Volume 75 • Number S4 • April 2023

Arthritis & Rheumatology

AN OFFICIAL JOURNAL OF THE AMERICAN COLLEGE OF RHEUMATOLOGY

ABSTRACT SUPPLEMENT

Pediatric Rheumatology Symposium 2023

March 29–April 1, 2023

New Orleans, LA

WILEY | American College of Rheumatology
Empowering Rheumatology Professionals

Table of Contents

Pediatric Rheumatology Symposium

Thursday, March 30

11:30 AM – 1:00 PM
Plenary Session I
(#001–004)

5:10 PM – 5:30 PM
Poster Breakout Sessions
(#011–015) Poster Breakout 1 - Lupus Genetics, Epigenetics, & Social Determinants
(#016–020) Poster Breakout 2 - JIA & Uveitis: Genetics, Clinical & Therapeutic Aspects
(#021–025) Poster Breakout 3 - Pediatric Rheumatology Potpourri

6:00 PM – 7:00 PM
Poster Session A
(#041–077) Posters: Clinical and Therapeutic I
(#078–081) Posters: Genetics and Pathogenesis I
(#082–089) Posters: Quality, Health Services, and Education I

Friday, March 31

2:30 PM – 3:00 PM
Plenary Session II
(#005–006)

4:30 PM – 5:00 PM
Poster Breakout Sessions
(#026–030) Poster Breakout 4 - JDM & Scleroderma: Clinical & Therapeutic Aspects
(#031–035) Poster Breakout 5 - Autoinflammatory/Vasculitis: STAT3, NLRP3, KD and MIS-C
(#036–040) Poster Breakout 6 - Systemic JIA: Genomics, Transcriptomics & Disease States

5:00 PM – 6:00 PM
Poster Session B
(#090–126) Posters: Clinical and Therapeutic II
(#127–130) Posters: Genetics and Pathogenesis II
(#131–137) Posters: Quality, Health Services, and Education II

Saturday, April 1

11:00 AM – 12:00 PM
Plenary Session III
(#007–010)

Abstract Embargo Policy

Accepted abstracts are made available to the public online in advance of the meeting and are published in a special online supplement of our scientific journal, *Arthritis & Rheumatology*. Information contained in those abstracts may not be released until the abstracts appear online. Academic institutions, private organizations, and companies with products whose value may be influenced by information contained in an abstract may issue a press release to coincide with the availability of an ACR abstract on the ACR Abstract website. However, the ACR continues to require that information that goes beyond that contained in the abstract (e.g., discussion of the abstract done as part a scientific presentation or presentation of additional new information that will be available at the time of the meeting) is under embargo until **March 29 at 4:30 PM CT.** Journalists with access to embargoed information also cannot release articles before this time.

Violation of this policy may result in the abstract being withdrawn from the meeting and other measures deemed appropriate. Authors are responsible for notifying colleagues, institutions, communications firms, and all other stakeholders related to the development or promotion of the abstract about this policy. If you have questions about the ACR abstract embargo policy, please contact ACR abstracts staff at abstracts@rheumatology.org.

Abstract Reprint (Reproduction) Policy

For the purposes of this statement, the term **ACR Abstracts** refers to all ACR PRSYM abstracts as published in *Arthritis & Rheumatology* and posted online, including the abstracts accepted for presentation during ARP sessions and the late-breaking category.

For the purposes of this statement, the term **ACR Posters** refers to the accepted abstract **POSTER PRESENTATIONS** as presented in the poster hall during PRSYM. **This does not include abstract text published in the online supplement of *Arthritis & Rheumatology*.** All **ACR Posters** are the property of the ACR and the presenting author and cannot be reproduced or distributed without written permission from the ACR and the presenting author.

- For the purposes of this statement, the term "reproduce" includes all forms of reproduction, including, but not limited to, print, electronic and photographed formats.
- For the purposes of this statement, the term "presenting author" refers to the author who is designated as the individual who will present the work during PRSYM, as identified through the abstract submission process.

Approval Process for ACR Abstracts

- Excerpts or the entirety of **ACR Abstracts** may not be reproduced without the prior written permission of the publisher.
- Permission requests for abstract content and other permission inquiries should be addressed to:

  Permissions Department
  c/o John Wiley & Sons, Inc.
  111 River Street
  Hoboken, NJ 07030
  Fax: 201-748-6008
  wiley.com/go/permissions
  - Commercial entities seeking permission to reprint must obtain all materials from the author and/or publisher John Wiley & Sons, Inc. The ACR cannot provide any materials.

Approval Process for ACR Posters

- Reprint requests for the actual poster abstract text published in the *Arthritis & Rheumatology* supplement are considered **ACR Abstracts** and must submitted to Wiley (see approval process above).
- Requests to reproduce individual ACR posters, figures from ACR posters, or booklets of poster presentations (e.g., two or more) must be submitted via e-mail to abstractreprints@rheumatology.org.
- Poster reproduction requests must include the following:
  – Abstract ID Number
  – Abstract Title
  – Presenting Author's Name
  – A copy of Presenting Author's written approval (Please Note: An e-mail approval from Presenting Author is acceptable)

Reproducing ACR Abstracts and Posters for Dissemination Prior to PRSYM 2023

- Requests to reproduce abstracts for dissemination prior to PRSYM 2023 *will not be approved*.
- Per the ACR Embargo Policy (see above), academic

institutions, private organizations, and companies with products whose value may be influenced by information contained in an abstract may issue a press release to coincide with the availability of an abstract online.
- Permission to issue a press release does not require ACR approval. However, it must comply with the ACR Embargo Policy; violation of this policy may result in the abstract being withdrawn from the meeting or other measures deemed appropriate.
- For more information regarding press releases, please contact the ACR public relations department at pr@rheumatology.org.

Reproducing ACR Abstracts and Posters for Dissemination During PRSYM 2023

Following approval (see approval process above), an exhibiting organization may:

- Following approval (see approval process above), an exhibiting organization may disseminate copies of individual ACR Abstracts from its exhibit space. Booklets of abstracts (e.g., two or more) may not be produced.
- Following approval, an exhibiting organization may disseminate information summaries (title/date/time/poster number) of ACR Abstracts from its exhibit space. Summaries may not reference company or product names. Requests for approval must be submitted in writing to abstractreprints@rheumatology.org.

Reproducing ACR Abstracts and ACR Posters for Dissemination After PRSYM 2023

ACR Abstracts

Following approval from Wiley (see approval process above), the ACR permits ACR Abstracts (i.e., all abstract content published in the online supplement) to be reprinted and disseminated following PRSYM 2023.

- Abstracts and booklets of abstracts (e.g., two or more) must include the following statement on the front of the abstract/booklet:
  Abstract(s) reprinted from ACR Convergence held [insert meeting dates]. The American College of Rheumatology does not guarantee, warrant, or endorse any commercial products or services. Reprinted by [insert name of supporting company].
- Booklets cannot contain corporate or product logos or any advertisements. No exceptions.

ACR Posters

Following approval from the presenting author and the ACR (see approval process above), copies of actual ACR poster presentations (i.e., the actual graphic file of the poster) may be reproduced.

1. Reprint requests for the actual poster abstract text published in the *Arthritis & Rheumatology* supplement are considered **ACR Abstracts** and must submitted to Wiley (see approval process above).

- **IMPORTANT: The ACR does not retain and cannot provide poster presentation images.**
- The following statement must be listed under each Poster reprint:
  Reprinted from the Pediatric Rheumatology Symposium held [insert meeting dates]. The American College of Rheumatology does not guarantee, warrant, or endorse any commercial products or services. Reprinted by [insert name of supporting company].

Use of the ACR Name

The names, insignias, logos and acronyms of the ACR, the ARP, and the Rheumatology Research Foundation are proprietary marks. Use of the names in any fashion, by any entity, for any purpose, is prohibited without the express written permission of the American College of Rheumatology.

Use of the ACR Scientific Program Content

- Information displayed or presented at all sessions during PRSYM 2023 is the property of the ACR or the presenter. Information may not be recorded, photographed, copied, photocopied, transferred to electronic format, reproduced or distributed without the prior written permission of the ACR and the presenter.
- Any use of the program content, which includes but is not limited to oral presentations, audiovisual materials used by speakers and program handouts, without the written consent of the ACR is prohibited.
- This policy applies before, during and after the meeting.
- The ACR will enforce its intellectual property rights and penalize those who infringe upon it.

Use of the ACR Disclosure Key

It is suggested when referencing disclosures in the reprints, that the ACR's disclosure key be added to provide adequate context for abstracts:

None: Has no relevant financial relationship to disclose.

1. Advisor or Review Panel member
2. Consultant
3. Employee
4. Officer or Board Member
5. Grant/Research Support
6. Speaker/Honoraria includes speakers bureau, symposia, and expert witness
7. Independent Contractor
8. Ownership Interest
9. Royalties
10. Intellectual Property / Patents
11. Stock options or bond holdings in a for-profit corporation or self-directed pension plan
12. Other Financial or Material Support

Arthritis & Rheumatology
Vol. 75, No. S4, April 2023, pp 1–243
DOI 10.1002/art.42509

American College of Rheumatology
Empowering Rheumatology Professionals

Abstract Number: 001

Serious Infections Among Children with Systemic Lupus Erythematosus in the Pediatric Health Information System

Jordan Roberts, Anna Faino, Mersine Bryan, Jonathan Cogen and Esi Morgan, Seattle Children's Hospital, Seattle, WA

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Plenary Abstracts Session I
Session Type: Plenary Session
Session Time: 2:30PM–3:30PM

Background/Purpose: High rates of serious infection have beenreported in pediatric systemic lupus erythematosus (pSLE), but modern care practices including pneumococcal vaccination, changing corticosteroid use, and B-cell-depletion may modify infectious risks. The current burden of infections in pSLE is unknown. We conducted a retrospective study using

Table 1: Characteristics of children with pSLE hospitalized for any indication and with serious infection from 2009-2021

Variable	All Lupus Hospitalizations (32,414 hospitalizations among 11,178 patients)	Lupus Hospitalizations with Serious Infection (3,839 hospitalizations among 2,485 patients)
Age at Admission (years [IQR])	15 [13 - 17]	15.8 [13.2 - 17.9]
	Number (% of hospitalizations)	*Number (% of hospitalizations)*
Female	26253 (81%)	3124 (81%)
Race/Ethnicity		
Asian	1680 (5%)	191 (5%)
Hispanic	8931 (28%)	1177 (31%)
Multiracial	326 (1%)	35 (1%)
Non-Hispanic Black	10561 (33%)	1348 (35%)
Non-Hispanic White	8360 (26%)	786 (20%)
Other	1837 (6%)	221 (6%)
Unknown	719 (2%)	81 (2%)
Insurance Type		
Private	11528 (36%)	1180 (31%)
Public (non-military)	18788 (58%)	2433 (63%)
Military	632 (2%)	76 (2%)
Self-Pay	569 (2%)	52 (1%)
Other	833 (3%)	94 (2%)
Uninsured	64 (0%)	4 (0%)
Household Income by Zip Code		
<$25,000/year	2633 (8%)	345 (9%)
$25-50,000/year	19358 (62%)	2304 (62%)
$50-75,000/year	7787 (25%)	904 (24%)
$75-100,000/year	1397 (4%)	157 (4%)
>$100,000/year	293 (1%)	27 (1%)
Disease Characteristics		
Lupus Nephritis	15398 (48%)	1943 (51%)
End Stage Renal Disease	1818 (6%)	247 (6%)
Hospitalization Outcomes		
ICU Admission	3347 (10%)	1050 (27%)
	Number (% of patients)	*Number (% of patients)*
In-hospital Mortality	132 (1%)	74 (3%)

the Pediatric Health Information System (PHIS) to determine number and type of serious infections in hospitalized children with pSLE, and to identify risk factors for poor outcomes.

Methods: We included patients 2-21 years of age with ICD-9 or ICD-10 code for SLE (710.0, M32*) during admission to a hospital participating in PHIS, an administrative database of 50 freestanding childrens hospitals in the United States, from 2009-2021. Serious infections were identified using ICD-9 and ICD-10 codes and antimicrobial medication use. Summary statistics were used to describe demographic and disease features of the entire PHIS pSLE cohort, and the subset hospitalized with serious infection. We calculated the frequency of admission, intensive care admission, and mortality by type of

Table 2: Hospitalization Outcome by Type of Infection and Disease Characteristics *Death during hospitalization was considered either with or without ICU admission.

Variable	**No ICU Stay (n=2789)**	**ICU Stay (n=1050)**	**Death during Hospitalization* (n=74)**
	Number (% of total hospitalization without ICU)	*Number (% of total hospitalization with ICU)*	*Number (% of total deaths)*
Type of Infection			
≥2 serious infection codes	283 (10%)	338 (32%)	40 (54%)
Bacterial			
Bacterial Pneumonia	763 (27%)	396 (38%)	16 (22%)
Cellulitis	694 (25%)	116 (11%)	11 (15%)
Urinary Tract Infection	393 (14%)	105 (10%)	2 (3%)
Sepsis	218 (8%)	475 (45%)	46 (62%)
Septic arthritis	55 (2%)	8 (1%)	0 (0%)
Endocarditis	14 (1%)	30 (3%)	5 (7%)
Osteomyelitis	10 (0%)	5 (0%)	1 (1%)
Bacterial encephalitis	8 (0%)	5 (0%)	0 (0%)
Bacterial meningitis	5 (0%)	12 (1%)	0 (0%)
Fungal			
Candidiasis	125 (4%)	86 (8%)	15 (20%)
Aspergillosis	34 (1%)	27 (3%)	9 (12%)
Non-tuberculosis or unspecified mycobacterial	21 (1%)	4 (0%)	0 (0%)
Other mycoses	18 (1%)	27 (3%)	8 (11%)
Histoplasmosis	12 (0%)	3 (0%)	0 (0%)
Coccidiomycosis	9 (0%)	3 (0%)	1 (1%)
Pneumocystis jirovecii pneumonia	4 (0%)	14 (1%)	4 (5%)
Cryptococcus	2 (0%)	4 (0%)	0 (0%)
Viral			
Herpes simplex	255 (9%)	51 (5%)	3 (4%)
Influenza	164 (6%)	28 (3%)	1 (1%)
Herpes zoster	138 (5%)	17 (2%)	2 (3%)
CMV	91 (3%)	51 (5%)	8 (11%)
COVID	73 (3%)	21 (2%)	2 (3%)
Adenovirus	15 (1%)	19 (2%)	4 (5%)
Patient Characteristics			
Disease Characteristics			
Lupus Nephritis	1377 (46%)	648 (56%)	58 (67%)
End Stage Renal Disease	154 (5%)	123 (11%)	13 (15%)

infection. Logistic regression using generalized estimating equations was used to identify risk factors for ICU admission and in-hospital mortality among children hospitalized with infection.

Results: We identified 11,178 unique patients with pSLE and at least one hospitalization in PHIS. Among this cohort, there were 32,414 hospitalizations, of which 3,839 (11.8%) had codes for serious infections. Patients hospitalized with serious infections were similar in demographic and disease characteristics to the entire PHIS pSLE cohort (Table 1). Specific infections and outcomes of hospitalization are reported in Table 2. The most common infections were bacterial pneumonia (n=1159), cellulitis (n=810), sepsis (n=693) and urinary tract infections (n=498). In-hospital mortality occurred in 1% of children with pSLE hospitalized for all indications and in 3% of hospitalizations with a code for serious infection. The highest in-hospital mortality rates occurred in hospitalizations with codes for pneumocystis jirovecii pneumonia (22%), aspergillosis (15%), and other fungal infections (18%). Approximately half of children who died had ≥2 serious infections during the hospitalization in which death occurred. Lupus nephritis and end-stage renal disease (ESRD) were significantly associated with increased odds of ICU admission (OR [95% CI] of 1.6 [1.3, 1.8] and 2.1 [1.6, 2.7], respectively) and in-hospital mortality (OR [95% CI] 2.3 [1.3, 4.0] and 2.1 [1.0, 4.2]) among children admitted for a serious infection (Figure 1).

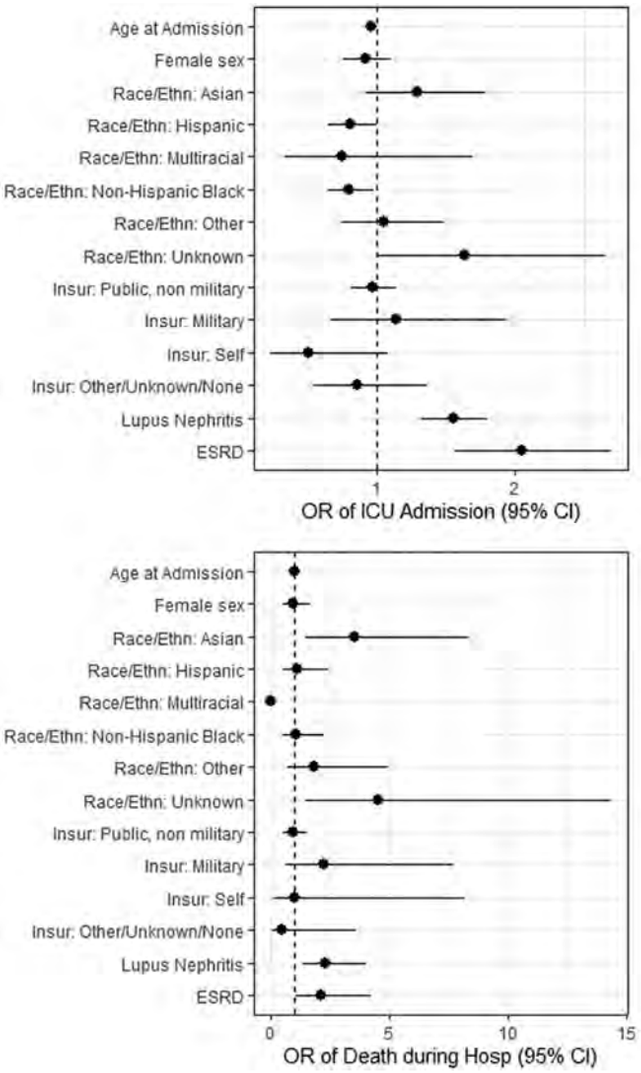


Figure 1: Odds of ICU admission and mortality among children with pSLE hospitalized with infection

Conclusion: Hospitalizations including codes for serious infection comprised a small proportion of pSLE admissions but accounted for the majority resulting in in-hospital mortality among patients with pSLE in the PHIS cohort. While bacterial infections were more common, mortality rates of hospitalizations with codes for fungal infections were high. Patients with pSLE and renal disease were at increased risk for poor outcomes. Further research is needed to understand modifiable risk factors for infection and severe outcomes, including immunosuppressant medication use.

Disclosure: J. Roberts: None; **A. Faino**: None; **M. Bryan**: None; **J. Cogen**: None; **E. Morgan**: None.

Abstract Number: 002

Re-analysis of the APPLE (Atherosclerosis Prevention in Paediatric Lupus Erythematosus) Trial Identifies Novel Determinants of Patient Heterogeneity and a Distinct Lipid Metabolomic Signature of Atherosclerosis Progression

Coziana Ciurtin[1], Junjie Peng[1], Pierre Donnes[1], Stacy Ardoin[2], Laura Schanberg[3], Laura Lewandowski[4], George A Robinson[1] and Elizabeth Jury[1], [1]University College London, London, United Kingdom, [2]Nationwide Children's Hospital, Columbus, OH, [3]Duke University Medical Center, Durham, NC, [4]NIAMS, NIH, Bethesda, MD

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Plenary Abstracts Session I
Session Type: Plenary Session
Session Time: 2:30PM–3:30PM

Background/Purpose: Juvenile-onset systemic lupus erythematosus (JSLE) is associated with chronic inflammation and increased risk of atherosclerosis. The APPLE trial was a randomised, placebo-controlled trial of atorvastatin for atherosclerosis progression in JSLE, using carotid intima-media thickness (CIMT) measurements as primary outcome.

Methods: Unsupervised clustering analysis was used to stratify JSLE patients by their baseline CIMT and identify patterns of CIMT progression over 36 months. An additional in-depth metabolomic analysis was performed to identify lipidomic signatures predictive of CIMT progression. Correlation and univariate regression analyses explored associations between patient and disease characteristics and serum biomarkers. Machine learning techniques and ROC analyses were used to identify and validate a serum metabolomic signature of high CIMT progression.

Results: Baseline CIMT measurements stratified 151 JSLE patients recruited to the APPLE trial into three groups with distinct CIMT progression trajectories irrespective of the treatment allocation (Figure). Two distinct CIMT progression rates (high vs. low), characterised by higher total and low-density lipoprotein (LDL) cholesterol levels (P=0.001 and P=0.002, respectively) were found in the placebo group, while patients treated with atorvastatin had three distinct CIMT trajectories (high, intermediate and low progression), not associated with any relevant biomarkers. A robust metabolomic signature predictive of high CIMT progression in the placebo arm was identified (AUC = 80.7%).

Conclusion: This complementary analysis of the APPLE trial provides new evidence for the significant heterogeneity of subclinical atherosclerosis in JSLE and its distinct progression trajectories irrespective of treatment allocation. Clinical trial patient stratification using the newly identified metabolomic signature predictive of increased natural atherosclerosis

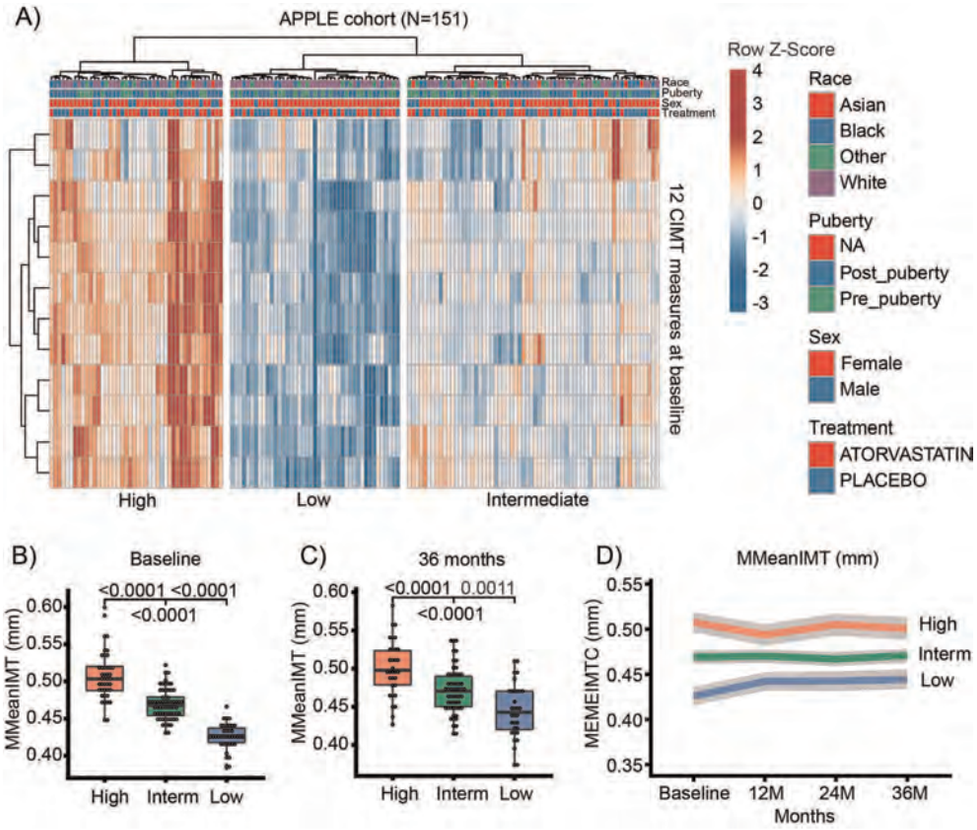


Figure 1: JSLE stratification (all APPLE patients with complete baseline data, N=151) by baseline CIMT (12 measures). A) Baseline CIMT measures of patients with juvenile-onset SLE were stratified using unsupervised hierarchical clustering. All 12 CIMT measures were standardised within each row by Z score and plotted as a heat map, representing the relationship to the mean of the group (red represents relatively high CIMT measures and blue represents relatively low CIMT measures). Each column represents a patient with JSLE. Three groups of patients with distinct baseline CIMT profiles were identified. B-C) Box and whisker plots show baseline and 36-month MMeanIMT measurements (APPLE primary outcome) in the identified high, intermediate and low baseline CIMT groups. Comparisons between groups were performed using Wilcoxon signed-rank test (* p<0.05; ** p<0.01; *** p<0.001). D) Distinct longitudinal MMeanIMT progression from baseline to 36 months of the high, intermediate and low CIMT progression groups (Mean, 95% CI), irrespective of treatment allocation. (Only JSLE patients with completed CIMT data at 36 months were included in the panel C-D, N=121). Legend: CIMT- carotid intima-media thickness; MMeanIMT - Mean-Mean IMT common carotid artery measurement.

progression rate may improve results. Despite being effective in lowering serum lipids, atorvastatin did not prevent the CIMT progression in many at risk JSLE patients, highlighting the need for personalised therapies to address various molecular mechanism driving atherosclerosis in JSLE.

Disclosure: C. Ciurtin: GlaxoSmithKlein(GSK), 5, UCB, 6; **J. Peng**: None; **P. Donnes**: GlaxoSmithKlein(GSK), 5; **S. Ardoin**: None; **L. Schanberg**: Bristol-Myers Squibb(BMS), 5, Sanofi, 12, DSMB member, UCB, 12, DSMB chair; **L. Lewandowski**: None; **G. Robinson**: None; **E. Jury**: GlaxoSmithKlein(GSK), 5.

Abstract Number: 003

Inflammation or Infection? Understanding Linguistic and Cultural Nuances Impacting the Care of Somali Children with Juvenile Idiopathic Arthritis

Emily Hause, Abdirazak Ali and Muna Sunni, University of Minnesota, Minneapolis, MN

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Plenary Abstracts Session I
Session Type: Plenary Session
Session Time: 2:30PM–3:30PM

Table 1. Focus group input from patient/family- and community-based focus groups organized by overarching theme.

Theme	Somali patient/family focus groups	Somali community member focus group
	Comments	
Juvenile arthritis knowledge	Patients don't share the diagnosis with others due to concern of being treated as "fragile"	Not familiar with JIA although familiar with rheumatism & arthritis in adults
	No commonly identified stigma around JIA	The concepts of autoimmunity & inflammation are vague and not well-understood
	Patients don't tell people they have arthritis since they don't want to be treated differently	A child with a swollen joint would be concerning for infection
		Concern for a causative agent (inadequate milk intake, exposure to vaccines or other environmental factors)
Medications & management	No stigma regarding medication use, chronicity, or injectable formulations although it is common to stop taking medications and to experience subsequent flare-ups	No stigma regarding obtaining treatment for JIA or around injectable medications
	Utilize adjunctive therapies (such as cupping, skin burning, massage, and prayer) before or alongside medications. Realigning joints by force is common	Home remedies for swollen joints e.g., burning the skin of the affected area, cupping, massage, stretching, herbal therapies, assistance from a religious figure/prayer, and drinking more milk
	Interest in changing of ambient temperature from a "cold" location such as Minnesota to a "warm" location to see if it helps symptoms	Sick children should return to their country of origin (Somalia) to look for a traditional remedy
	Want additional support from schools and guidance on physical activity limitations	
	Unlikely to utilize resources like "arthritis camp", but desire connection with other Somali families in the JIA community	
Translation	Should create a word or phrase to describe inflammation that can be used consistently in clinic and taught to interpreters	There is not a word in Somali that appropriately matches the English word "inflammation", thus difficult to understand inflammation vs infection
	Strong preference for in-person rather than video or telephone interpreters	Large preference for in-person rather than virtual interpreters
	There are many words in English that do not have exactly corollaries in Somali such as fingertips and wrists	Some translators speak Somali, but don't have the technical vocabulary necessary in specialized conditions
	Clinic materials should focus on visual aids rather than text	Picture-based educational materials are preferred over text
	The many dialects of Somali present a translation challenge	Frustration regarding not being able to utilize family members to translate as there are many dialects of Somali, which are not accounted for when using a translation service
	Prefer gender of physician match the gender of the patient, but gender concordance is less important in an interpreter	Female participants strongly preferred a female interpreter, especially if the topic was deemed sensitive

Background/Purpose: Data regarding the linguistic and cultural factors impacting the Somali patient experience and understanding with regards to juvenile idiopathic arthritis (JIA) are limited. We aimed to learn these factors from Somali patients, families, and the community to better inform future practices.

Methods: This cross-sectional, qualitative study abstracted common themes via open-ended questions from 2 Somali focus groups: 1) patients with JIA and their families, and 2) Somali community members.

Somali children with more than one visit to the University of Minnesota pediatric rheumatology clinic from 2020-2022, aged 18 months to 22 years, diagnosed with JIA or uveitis, were identified via Slicer-Dicer searches in the Epic electronic medical record and were recruited. Community members were recruited via mosques, social networks, and word-of-mouth. Focus groups were conducted during the summer at a local community center. Open-ended questions guided focus group discussions that were facilitated by a Somali interpreter. Major themes that resulted from these discussions were recorded.

Results: Patient/family focus groups: Six out of fourteen eligible Somali patients/families attended two group sessions. Participating patients ages were 17, 18, and 19 years old. Participating families included four mothers, one father, and one first-degree relative (cousin). The community-based focus group was comprised of seven women and six men aged >18 years.

In both types of focus groups, three themes emerged: JIA knowledge, medications & management, and translation. Particularly pertinent to the diagnosis of JIA was the realization that there is not a word or corresponding concept for inflammation and autoimmunity. All groups mentioned common home remedies and a desire for adjunctive therapies along with allopathic medications. They noted many linguistic hurdles with interpreters and varying Somali dialects. Specific concepts organized by theme are presented in Table 1.

There was a strong preference for in-person rather than virtual interpreters, a desire for picture-based education materials as opposed to text-based, and a need for a standardized word or phrase that can be used to describe inflammation. There was unanimous support for a proposed educational intervention of a 5-minute video to be shown at the time of JIA diagnosis that introduces important concepts without worrying about variability in translation capability.

Conclusion: This is the first study describing attitudes towards JIA in the Somali population. Data from this study provide valuable insights for pediatric rheumatologists to better understand cultural and linguistic factors impacting the care of Somali youth with JIA, with the potential to improve the delivery of culturally sensitive care to Somali JIA patients through appropriately targeted interventions.

Disclosure: E. Hause: None; **A. Ali**: None; **M. Sunni**: None.

Abstract Number: 004

Analysis of Patients with Juvenile Dermatomyositis Compared to Healthy Controls Using CITE-seq Identifies Differences in Cell Composition and Gene and Epitope Expression

Camilla Wibrand[1], Emily Flynn[2], Gabrielle Rabadam[2], George Hartoularos[2], Yang Sun[2], Chun Ye[2], Susan Kim[3], Marina Sirota[2] and Jessica Neely[1], [1]UCSF, San Francisco, CA, [2]University of California, San Francisco, San Francisco, CA, [3]UCSF Benioff Children's Hospital, San Francisco, CA

SESSION INFORMATION

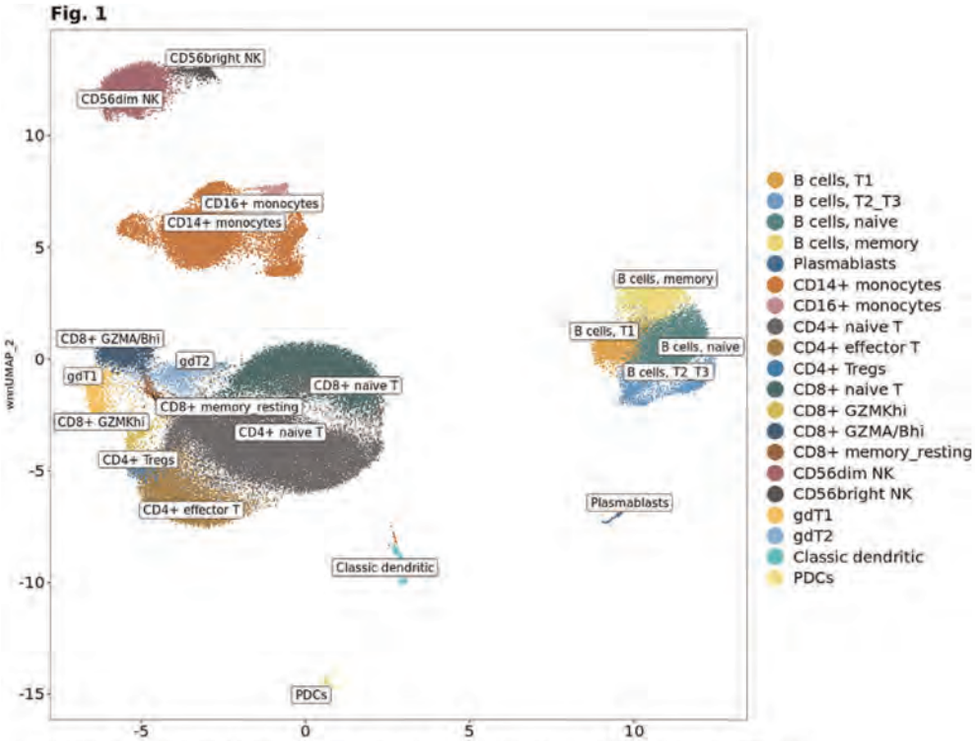
Session Date: Thursday, March 30, 2023
Session Title: Plenary Abstracts Session I
Session Type: Plenary Session
Session Time: 2:30PM–3:30PM

Background/Purpose: Juvenile dermatomyositis (JDM) is a rare and serious systemic autoimmune condition, and much remains unknown about the pathogenesis, the immune cell types and cell-specific disease associated pathways. Increasing knowledge of immune dysregulation in JDM with detailed immunophenotyping at the cellular level could lead to insight into disease mechanisms.

Methods: Multiplexed single cell RNA and protein sequencing was applied to 27 peripheral blood samples to simultaneously profile the gene expression and cell surface antibody (ADT) signatures associated with JDM (n=15, samples=22) compared to healthy pediatric controls (HC)(n=5). Data processing was performed using Demuxlet, SoupX, Harmony, DSB, and Seurat and included doublet removal, quality control, dimensionality reduction, clustering, and cell type annotation using canonical ADT and gene markers. To determine changes associated with JDM, we performed: 1) cell type proportion analysis between individual groups (treatment naïve (TN), inactive, flare, HC) and for different distributions of abundances pr. cell type, 2) correlation analysis between cell type proportion and disease activity measures for all JDM samples, 3) differential expression (DE) between TN JDM and HC pr. cell type using DESeq2 for genes and likelihood-ratios for ADTs (FDR < 0.05), and 4) gene overrepresentation analysis (GOA) with clusterProfiler.

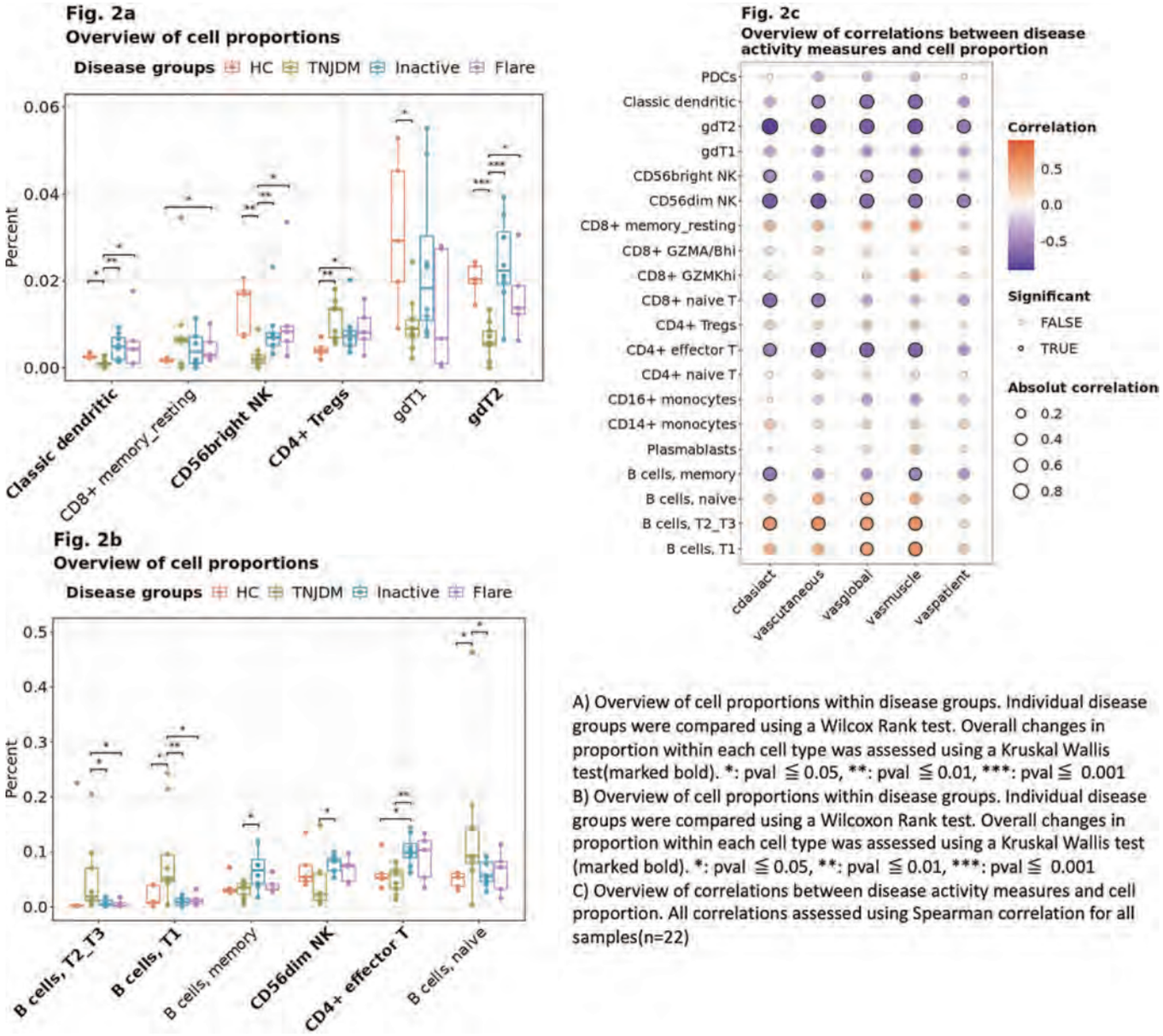
Results: ~110,000 immune cells were analyzed across 26,057 genes and 268 ADTs. Using unsupervised clustering, we identified 29 clusters, which compromised 20 unique immune cell populations (Fig 1). TN patients had statistically significantly higher levels of transitional and naïve B cells compared to inactive JDM and HC, and cell type proportion was significantly positively correlated with measures of disease activity (Fig 2, pval < 0.05). Patients with inactive disease had significantly higher proportions of memory B cells compared to TN patients, and the proportion of memory B cells was significantly negatively correlated with disease activity measures (Fig 2, pval < 0.05). TN patients had significantly fewer NK and gd T cells than both healthy controls and other JDM disease states, and cell type proportion was significantly negatively correlated with disease activity measures (Fig 2, pval < 0.05). Tregs were elevated in JDM irrespective of disease state

Fig. 1



Weighted-nearest neighbor UMAP embeddings and subset annotations of single cell CITE-seq dataset from patients with juvenile dermatomyositis (n=15, samples = 22) and healthy pediatric controls(n=5)

wnnUMAP

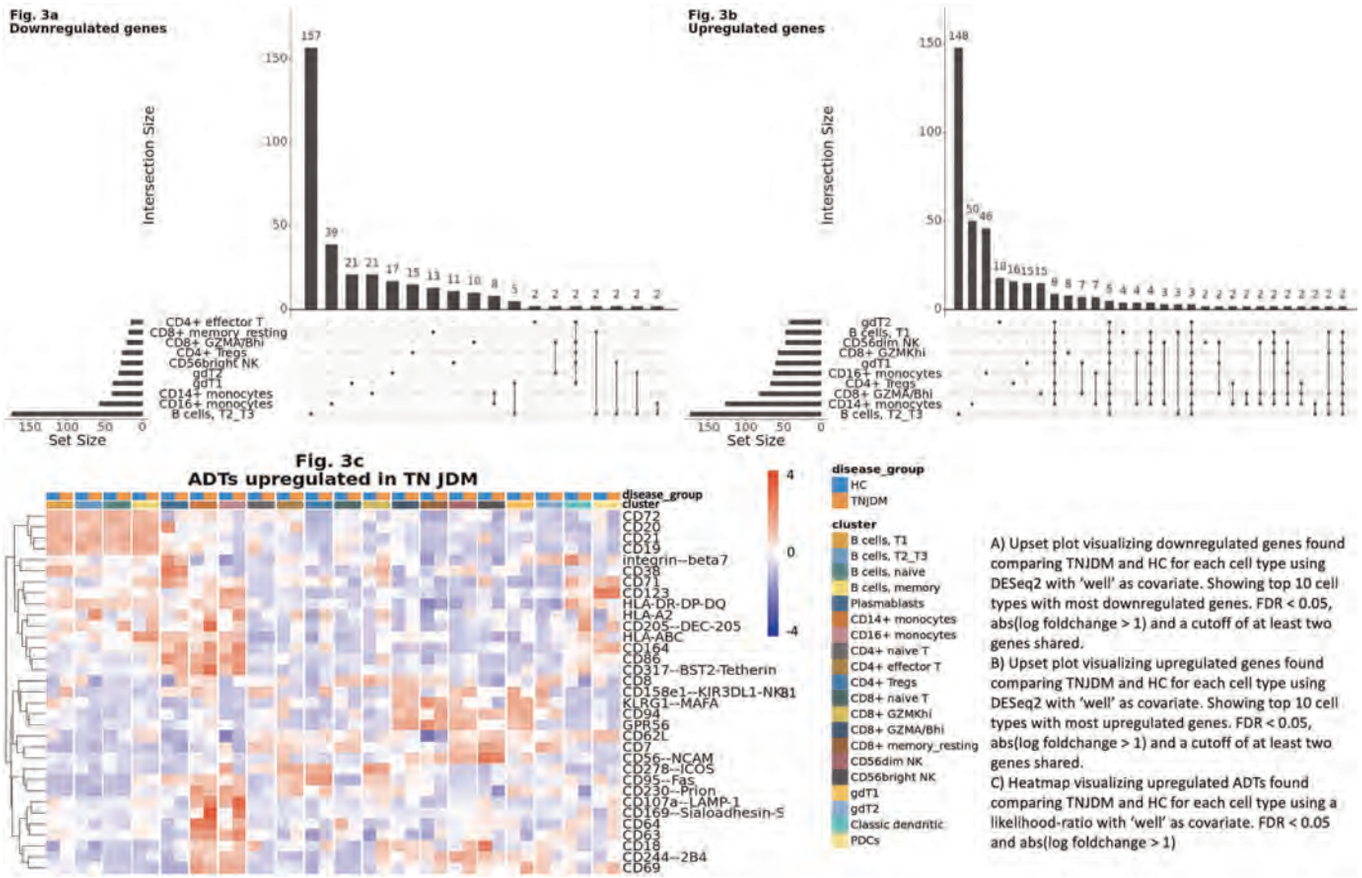


A) Overview of cell proportions within disease groups. Individual disease groups were compared using a Wilcox Rank test. Overall changes in proportion within each cell type was assessed using a Kruskal Wallis test(marked bold). *: pval ≦ 0.05, **: pval ≦ 0.01, ***: pval ≦ 0.001
B) Overview of cell proportions within disease groups. Individual disease groups were compared using a Wilcoxon Rank test. Overall changes in proportion within each cell type was assessed using a Kruskal Wallis test (marked bold). *: pval ≦ 0.05, **: pval ≦ 0.01, ***: pval ≦ 0.001
C) Overview of correlations between disease activity measures and cell proportion. All correlations assessed using Spearman correlation for all samples(n=22)

Cell proportion and correlation

compared to HC (Fig 2, pval < 0.05). Most DE genes were found within transitional B cells, monocytes, and Tregs (Fig 3). However, many DE genes were shared among cell types, and GOA revealed overrepresentation in genes involved in type 1 interferon and interferon gamma pathways for most cell types. Differential protein analysis revealed upregulation of SIGLEC-1 and LAMP-1 on the surface of both CD14+ and CD16+ monocytes (Fig 3).

Conclusion: Alterations in immune cell composition within the B, $CD4^+$ T, gd T, and NK cell compartments are associated with disease activity in JDM. Monocytes and transitional B cells show major gene expression changes during disease, but all cell types are heavily influenced by interferon signaling. Future directions include characterization of JDM-associated immunophenotypes at the transcriptomic and proteomic levels as well as network analysis.



Differential expression

Disclosure: C. Wibrand: AMBU, 11, GenMab, 11, Lundbeck, 11, NovoNordisk, 11; **E. Flynn**: None; **G. Rabadam**: None; **G. Hartoularos**: None; **Y. Sun**: None; **C. Ye**: Chan Zuckerberg Biohub, 5, Chan Zuckerberg Initiative, 5, Genentech, 5, ImmunAI, 1, 11, Maze Therapeutics, 2, 11, Related Sciences, 1, 11, TReX Bio, 2; **S. Kim**: None; **M. Sirota**: Exxagen, 1; **J. Neely**: None.

Abstract Number: 005

Real-World Application of the Pediatric Glucocorticoid Toxicity Index in Children with Lupus Nephritis: A Feasibility and Initial Validation Study

Emily Zhang[1], Gabrielle Alonzi[1], Madeline Hlobik[1], Esra Meidan[1], Mindy Lo[1], Olha Halyabar[2], Melissa Hazen[1], Ezra Cohen[3], Lauren Henderson[1], Siobhan Case[4], Margaret Chang[1], Camille Frank[1], Ankana Daga[1], Jonathan Hausmann[5], Ahmad Bakhsh[1], Liyoung Kim[1], Daniel Ibanez[1], Holly Wobma[1], Mia Chandler[6], Fatma Dedeoglu[1], Robert Sundel[1], Peter Nigrovic[1], Karen Costenbader[7], Mary Beth Son[1] and Joyce Chang[1], [1]Boston Children's Hospital, Boston, MA, [2]Children's Hospital/Boston Medical Center, Boston, MA, [3]Boston Medical Center, Boston, MA, [4]Brigham and Women's Hospital, Boston Children's Hospital, Boston, MA, [5]Boston Children's Hospital / Massachusetts General Hospital, Boston, MA, [6]Boston Children's Hospital; Brigham and Women's Hospital, Boston, MA, [7]Brigham and Women's Hospital/ Harvard Medical School, Boston, MA

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Plenary Abstracts Session II

Session Type: Plenary Session

Session Time: 2:30PM–3:00PM

Background/Purpose: The morbidity of chronic glucocorticoid (GC) use is rarely captured as a standardized clinical outcome in pediatric rheumatic conditions. The newly developed pediatric glucocorticoid toxicity index (pGTI) (*Brogan et al. 2022*) presents a novel opportunity to operationalize measurement of GC toxicity for research and clinical care. We tested the feasibility and construct validity of the pGTI in our pediatric lupus nephritis (pLN) cohort.

Methods: We conducted a retrospective cohort study of patients with pLN in the Boston Childrens Hospital Lupus Registry with rheumatology visits since January 2016 and ^{3}6 months of systemic GC treatment. The index visit was the visit date at which GC were initiated. We scored steroid toxicity items in 12/15 domains at visits occurring every 6 months (+/-2) for up to 3 years, with positive scores indicating new/worsening toxicity and negative scores indicating improvement. Scores were collected prospectively for skin and neuropsychiatric toxicity domains as of November 2022; the remainder were abstracted via manual chart review. We calculated total pGTI for each visit interval, Cumulative Worsening Score (CWS) and Aggregate Improvement Score (AIS). Pearsons correlation coefficients and mixed effects linear regression for repeated measures were used to evaluate relationships between daily, average, or cumulative prednisone dose and respective pGTI indices.

Table 1. Characteristics of patients in the pediatric lupus nephritis cohort (N = 45)

Demographic Characteristics	
Age at SLE diagnosis (years), mean (SD)	13.4 (3.3)
Disease duration at index visit (months), median [IQR]	0 [0-1]
Female, n (%)	35 (78%)
Race	
Asian	8 (18%)
Black	10 (22%)
White	13 (29%)
Unknown	5 (11%)
Other	9 (20%)
Hispanic ethnicity	13 (29%)
Insurance status	
Public	20 (44%)
Private	24 (53%)
Uninsured	1 (2%)
BMI at index visit	
Normal	31 (69%)
≥ 85 and <95%ile	9 (20%)
≥ 95ile	5 (11%)
SLEDAI-2K score at index visit, median [IQR]	20.5 [17-24]
Calendar year of index visit	
Before 2016	16 (36%)
2016-2022	29 (64%)
Disease Features	
Nephritis Class	
III/IV	35 (78%)
Mixed Class III/IV and V	6 (13%)
V	3 (7%)
Unavailable	1 (2%)
Serositis	14 (31%)
Dialysis	4 (10%)
Neurologic manifestations	11 (24%)
anti-dsDNA antibody positive	41 (93%)
Immunosuppressive Medication Usage	
Initial Cyclophosphamide (up to 30 days after index visit date)	12 (27%)
Initial Mycophenolate	23 (51%)
Initial Other DMARD	5 (11%)
Initial Belimumab	0 (0%)
Initial Rituximab	6 (13%)
Cyclophosphamide ever	21 (47%)
Belimumab ever	11 (24%)
Rituximab ever	19 (42%)

BMI = body mass index; SLEDAI-2K = Systemic Lupus Erythematosus Disease Activity Index 2000; DMARD = Disease-modifying antirheumatic drug

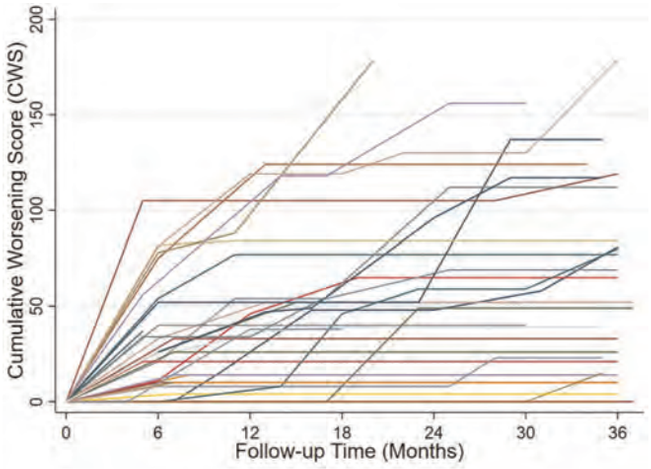


Figure 1. Cumulative Worsening Score (CWS) trajectories over three years of follow-up. CWS is a continuous summation of any toxicity that has been accrued after the index visit. Each line represents the score trajectory of an individual patient. Following discontinuation of GCs, if there was no glucocorticoid use during subsequent visit intervals, then further changes in toxicity domains could not be reliably attributed to glucocorticoid use and therefore did not contribute to the CWS.

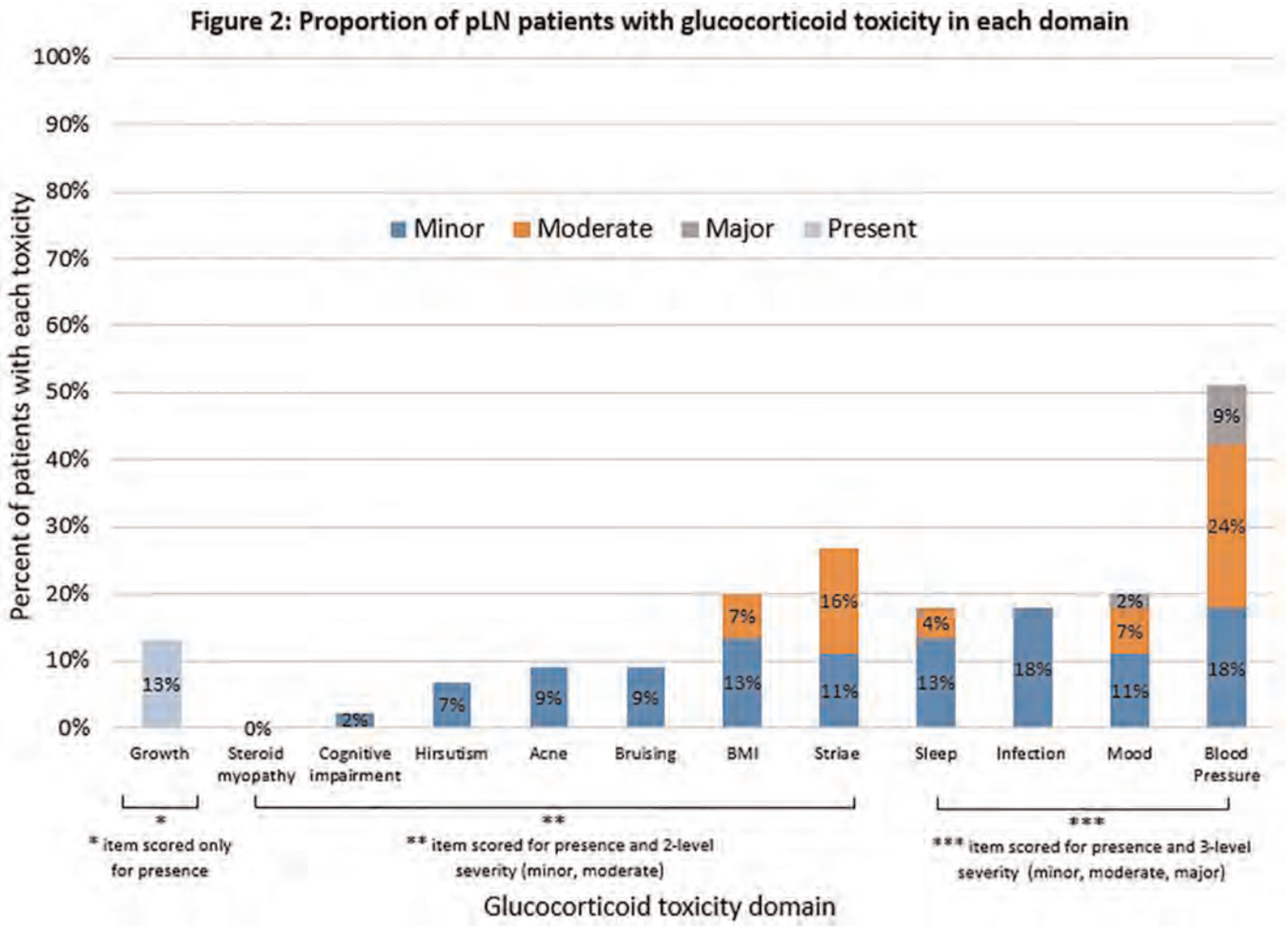


Figure 2. Proportion of pediatric lupus nephritis (pLN) patients (N=45) with new or worsening toxicity for each pediatric Glucocorticoid Toxicity item at any time during follow-up. Neuropsychiatric and skin toxicity items were captured prospectively for 4/227 (2%) of visits and otherwise abstracted from medical records.

Results: We included 227 visits by 45 patients (median of 5 visits/patient [IQR 2-4]) (Table 1). Glucose metabolism (hemoglobin A1c), hyperlipidemia, and bone mineral density domains were dropped due to infrequent clinical assessment (7 observations).

The largest increases in toxicity occurred in the first 6 months, but 33% of patients accrued new/worsening toxicity beyond 12 months (Fig 1). Median CWS was 14 (IQR [0-33]; N = 45) at month 6, 18 ([0-52], N = 40) at month 12, and 30 (IQR [0-53], N = 38) at month 24. The most common toxicity (51% of patients) was worsening blood pressure, including hypertensive emergency (11%) and posterior reversible encephalopathy syndrome (9%). Other common toxicities were striae, increasing body mass index (BMI), and mood disturbance (Fig 2).

pGTI score correlated modestly with current (r^2 0.25, p 0.01) and interval-averaged (r^2 0.27, p 0.01) prednisone dose (mg/day). There was modest correlation between cumulative dose (mg) and CWS (r^2 0.41, p 0.01). In repeated measures analysis adjusted for time, cumulative dose associated with greater CWS (7 unit increase in CWS for every 100 mg higher cumulative prednisone dose, 95% CI [3.8 – 10.9]), but there was large between-subject variance.

Conclusion: Patients with pLN exhibit significant GC toxicity, particularly in the blood pressure, skin, mood and BMI domains. Calculation of the pGTI using real-world data was generally feasible, but required adaptations for missing clinical data. Modest correlation between GC dose and pGTI supports construct validity of this tool but also highlights between-subject variability in toxicity. As a standardized, pediatric-specific tool for assessing GC toxicity, the pGTI can be considered

Disclosure: E. Zhang: None; **G. Alonzi**: None; **M. Hlobik**: None; **E. Meidan**: None; **M. Lo**: None; **O. Halyabar**: None; **M. Hazen**: Aditum Bio, 2, Sobi, 2; **E. Cohen**: None; **L. Henderson**: Adaptive Biotechnologies, 2, BMS, 5, Pfizer, 2, Sobi, 2, 12, Site PI for Sobi sponsored clinical trial; **S. Case**: None; **M. Chang**: None; **C. Frank**: None; **A. Daga**: None; **J. Hausmann**: Fresenius Kabi, 2, Novartis, 2; **A. Bakhsh**: None; **L. Kim**: None; **D. Ibanez**: None; **H. Wobma**: None; **M. Chandler**: None; **F. Dedeoglu**: UptoDate, 9; **R. Sundel**: None; **P. Nigrovic**: None; **K. Costenbader**: Amgen, 2, 5, AstraZeneca Pharmaceuticals LP, 2, 5, Eli Lilly, 5, Glaxo Smith Kline, 2, 5, Merck, 5, Neutrolis, 2; **M. Son**: None; **J. Chang**: GlaxoSmithKlein(GSK), 5.

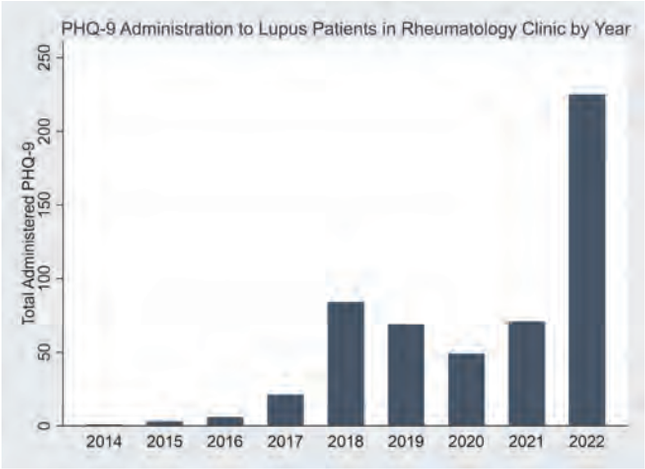
Abstract Number: 006

Implementation of Automated Depression Screening in Patients with Lupus in a Tertiary Pediatric Rheumatology Clinic

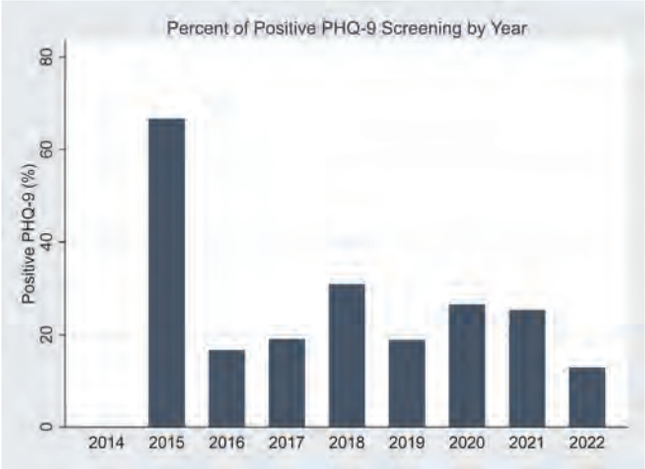
Lauren Harper[1], Alana Goldstein-Leever[1], James Gallup[1], Vidya Sivaraman[2], Stacy Ardoin[1], Kyla Driest[1], Evan Mulvhihill[3] and Alysha Taxter[4], [1]Nationwide Children's Hospital, Columbus, OH, [2]Nationwide Children's Hospital/ The Ohio State University, Columbus, OH, [3]Nemours Children's Hospital, [4]Nationwide Children's Hospital, Columbus, OH

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Plenary Abstracts Session II
Session Type: Plenary Session
Session Time: 2:30PM–3:00PM

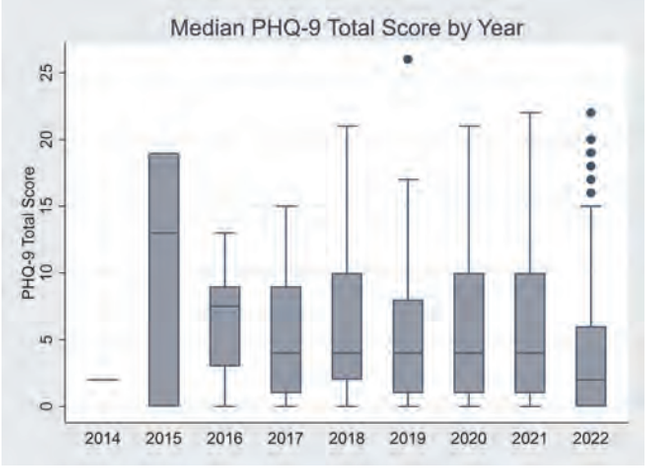
Background/Purpose: Patients with chronic rheumatic conditions, particularly lupus, have higher rates of depression, which significantly impacts their lives and can lead to poor medication compliance and increased disease activity. Regular mental health screening is essential to optimize the care for these patients. This study aims to evaluate an automated electronic depression screening process and its integration into routine clinical care.



Legend: PHQ-9 = Patient Health Questionnaire-9



Legend: PHQ-9 = Patient Health Questionnaire-9



Legend: PHQ-9 = Patient Health Questionnaire-9

Methods: Patients ≥12 years old with a diagnosis of lupus who were evaluated at a tertiary pediatric rheumatology clinic between 2014 and 2022 were included. Depression screening using the Patient Health Questionnaire-9 (PHQ-9) was completed by paper screenings annually, transcribed into the electronic health record (EHR), and later automated in 2022 to be completed at every visit via i-Pad tablet. Positive scores were defined as score ≥10, or if the patient endorsed suicidality. During the automated phase of this study, an intrusive alert was displayed for positive screens during a providers workflow. Data were extracted from the EHR and providers were surveyed about the new workflows.

Results: During the study period, 149 patients completed 529 screenings. PHQ-9 administration increased from 1 patient in 2014 to 21 in 2017; after automation, screenings increased to 225 ($p < 0.01$) (Figure 1). Percent positive screening increased from 0% in 2014 to 25-30% in 2018-2021 and decreased to 12% in 2022 ($p < 0.01$) (Figure 2). The incidence of positive screens during the study period was 20%, whereas the prevalence of positive PHQ-9 screen for a single patient was 38%. The median PHQ-9 score was 3 [0, 6], and scores decreased as screening increased (Figure 3). Ten automated alerts were triggered; 9 (90%) met with psychology or social work, 9 (90%) completed suicide risk assessment, which included the Ask Suicide-Screening Questions & the Columbia Suicide Severity Rating Scale. Providers expressed satisfaction with screening and did not have objections to intrusive alerts.

Conclusion: Results demonstrate that automated screening procedures maximize the number of patients screened for depression and minimize demand on support staff. Screening at each visit provides more opportunities to identify and treat depression. Such procedures may be emulated at parallel institutions to streamline mental health screening efforts while minimizing demands.

Disclosure: L. Harper: None; **A. Goldstein-Leever**: None; **J. Gallup**: None; **V. Sivaraman**: None; **S. Ardoin**: None; **K. Driest**: None; **E. Mulvhihill**: None; **A. Taxter**: None.

Abstract Number: 007

Single Cell RNA Sequencing Analysis of the Skin to Evaluate the Effect of Autologous Stem Cell Transplant on Fibroblast Populations in Juvenile Onset Systemic Sclerosis

Claire Cheng, Giffin Werner, Anwesha Sanyal and **Kathryn Torok**, University of Pittsburgh, Pittsburgh, PA

SESSION INFORMATION
Session Date: Saturday, April 1, 2023
Session Title: Plenary Abstracts Session III
Session Type: Plenary Session
Session Time: 11:00AM–12:00PM

Background/Purpose: Juvenile systemic sclerosis (JSSc) is a rare autoimmune disease associated with high morbidity. Inflammatory driven multi-organ fibrosis is similar to adult-onset SSc, with 40% of JSSc patients recently identified in an international cohort to have interstitial lung disease. There are a group of JSSc patients that have severe, refractory disease to standard immunosuppression regimens. For these patients, autologous stem cell therapy (ASCT) provides a possibility to reset the immune system and provide clinical benefit with much lower transplant-related mortality. Our center reported the clinical improvement of the first 3 JSSc patients with a CD-34 selected ASCT protocol at the American College of Rheumatology 2021 Convergence (Abstract 0778). The current abstract describes the single-cell RNA sequencing (scRNA-seq) profile of these 3 JSSc patients skin at baseline (pre-ASCT), 6-mos and 12-mos post-ASCT to define the shift in fibroblast populations and transcriptome expression.

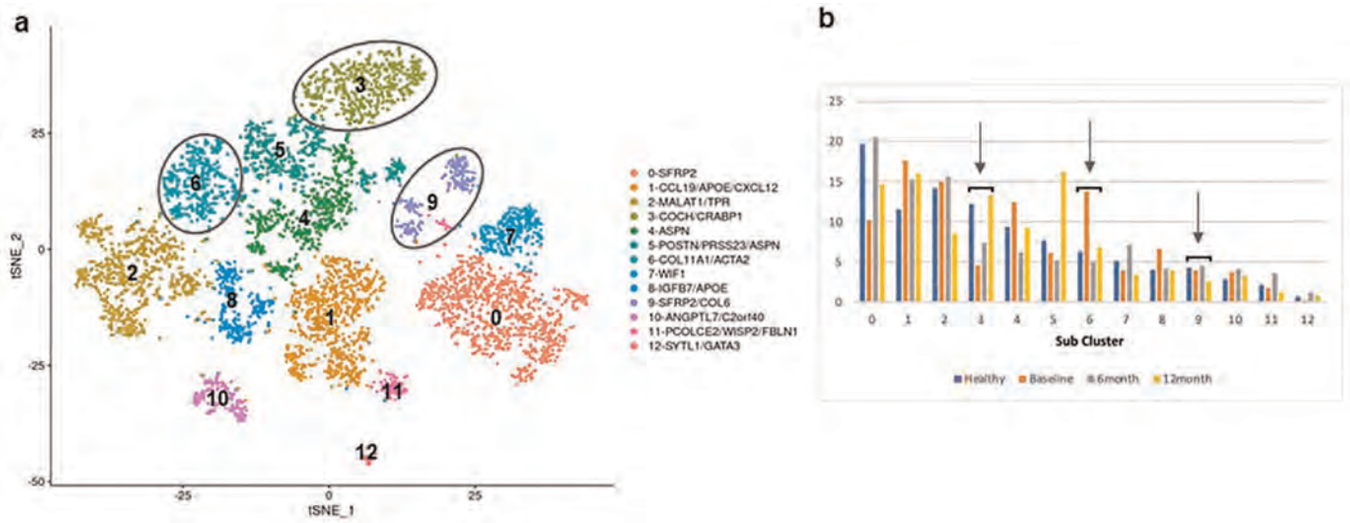


Figure 1 a & b: (a) tSNE analysis of fibroblast transcriptomes (total fibroblast cell count = 6216) from 3 JSSc patients and 3 healthy controls. (b) Relative cell proportion difference per fibroblast subcluster compared between healthy controls to baseline, 6-months and 12-months post-ASCT in JSSc.

Methods: scRNA-seq was performed on the skin of 3 JSSc patients at the baseline, 6-mos and 12-mos post-ASCT, along with 3 healthy aged-matched controls. Library preparation with a 10X Genomics® Chromium instrument and sequencing on Illumina NextSeq instrument was performed. Transcripts were mapped to reference human genome GRCh38 and assigned

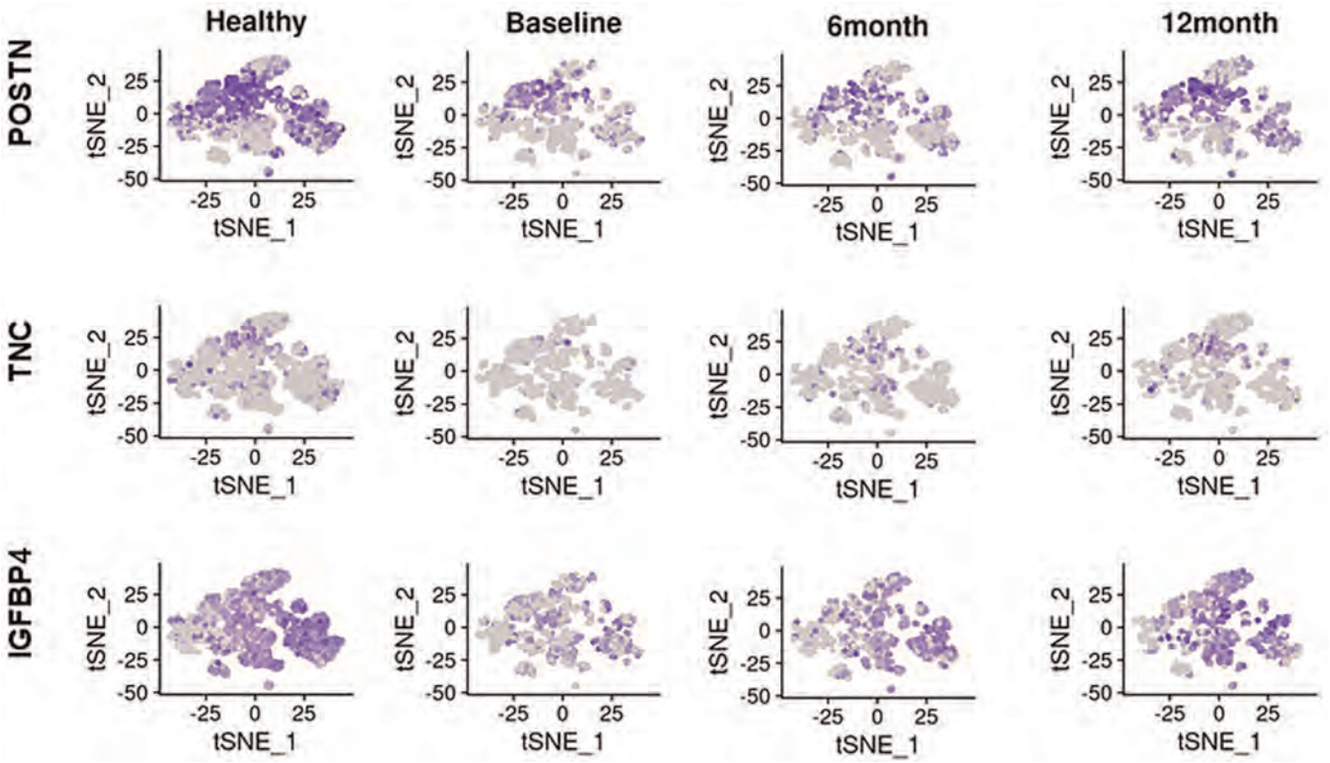


Figure 2a: Feature plots of fibroblasts demonstrating increased gene expression of POSTN, TNC and IGFB4 in JSSc after ASCT with matching expression to healthy controls at 12-months post-ASCT. Supporting wound healing genes increased expression.

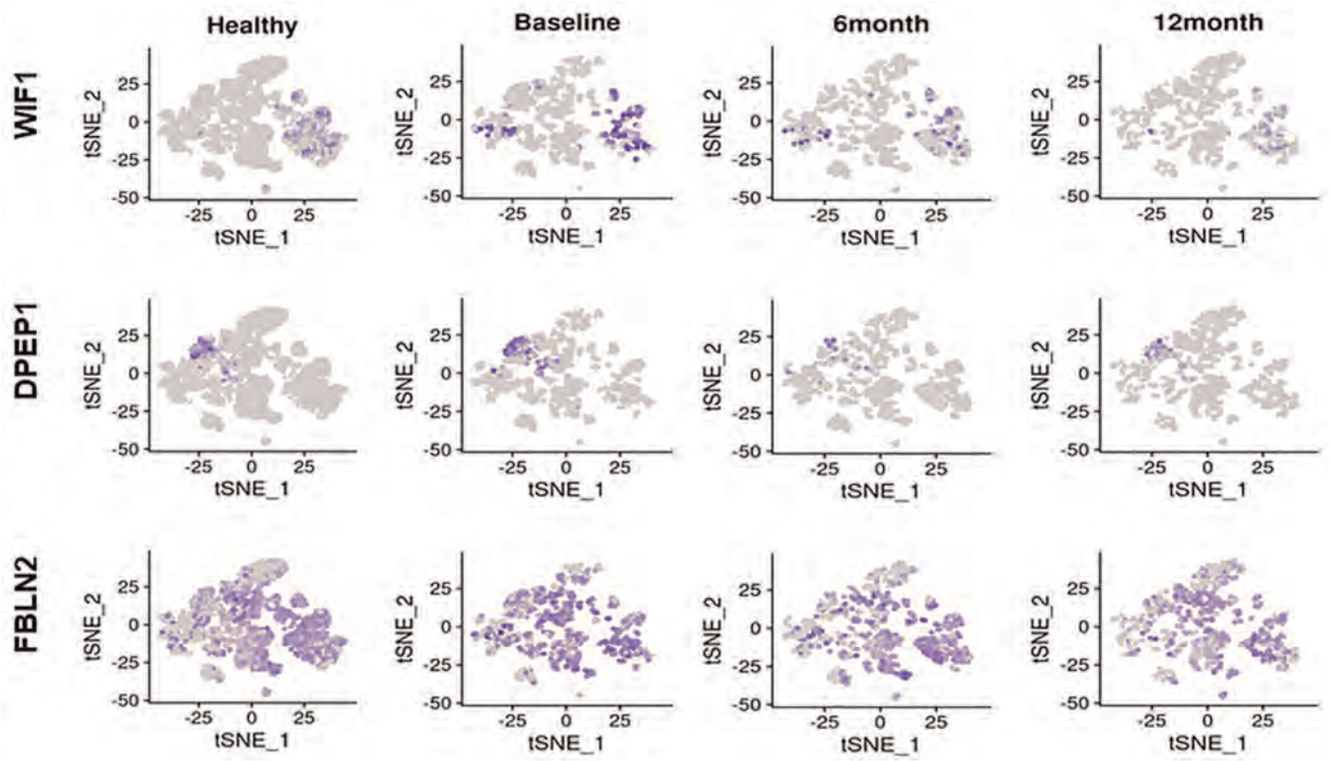


Figure 2b: Decreased fibrotic genes, WIFI, DPEP1 and FBLN2, across fibroblast populations after ASCT, with expression decreased to level of healthy controls at 12 months post-ASCT. Reflecting decreased fibrotic signal, and mirroring decreased skin thickness.

to cells of origin using the Cell Ranger pipeline (10X Genomics®). R-language analyses using Seurat and Harmony identified and visualized distinct cell populations by clustering methodologies.

Results: JSSc and healthy control fibroblasts clustered into 13 unique subclusters (Fig 1a). One fibroblast subcluster, defined by COCH/CRABP1(3) expression, increased after ASCT and at 12-mos post-ASCT it matched the frequency found in healthy controls (Fig 1b). Two fibroblast subpopulations, defined by COL11A1/ACTA2 (6) and SFRP2/COL6 (9) expression, decreased by half after ASCT, both mirroring healthy control frequency by 12 mos post-ASCT. In addition to the shift of these fibroblast subclusters towards a healthy distribution after ASCT, differential expression of gene (DEG) analyses found significant changes in pro-fibrotic genes: WIF1, DPEP1, and FBLN2, with decreased expression after ASCT (Figure 2a), and changes in wound-healing genes: POSTN, TNC, and IGFBP4, with increased expression after ASCT (Figure 2b). All 3 patients experienced dramatic improvement in skin score and other global outcome measures by 6 mos and sustained this at 12 mos (data not shown here; ACR 2021 Abstract 0778).

Conclusion: Dynamic changes were noted in JSSc fibroblast subclusters, overall shifting toward a healthy skin fibroblast distribution by 12-mos post-ASCT. The COL11A1/ACTA2 cluster represents contractile follicle-related cells containing myofibroblasts, and the SFRP2/COL6 cluster represent reticular and papillary dermal cells, both decreasing after ASCT and possibly pathogenic, and will be further studied. DEG analysis showed an upregulation of wound-healing associated genes: POSTN, TNC, and a downregulation of fibrosis-related genes: WIF1, DPEP1, and FBLN2, reflecting shift in fibroblast phenotype toward decreased fibrosis and increased elasticity.

Disclosure: C. Cheng: None; **G. Werner**: None; **A. Sanyal**: None; **K. Torok**: None.

Abstract Number: 008

Treatment Response and Outcomes of 63 Cases of Juvenile Dermatomyositis-Associated Calcinosis

Belina Yi[1], Dawn Wahezi[2], Lauren Covert[3], Kaveh Ardalan[4], Joyce Hui-Yuen[5], Natalia Vasquez Canizares[2], Doaa Mosad Mosa[6], Madison Jones[7], Colleen Correll[8], Alexis Begezda[9], Susan Shenoi[10], Eveline Wu[11], Leonard Kovalick[12], William Lapin[13], Stacey Tarvin[14], Melissa Oliver[15], Martha Rodriguez[14], Itay Marmor[16], Kevin Baszis[17], Alysha Taxter[18], Andrew Hanson[19], Cynthia Crowson[19] and Amir Orandi[19], [1]Children's Hospital Los Angeles, Los Angeles, CA, [2]Children's Hospital at Montefiore, New York, NY, [3]Duke University, Durham, NC, [4]Duke University School of Medicine, Durham, NC, [5]Cohen Children's Medical Center, Northwell Health, Lake Success, New York; Center for Autoimmune, Musculoskeletal, and Hematopoietic Diseases Research, Feinstein Institutes for Medical Research, Northwell Health, Manhasset, NY, [6]Mansoura University Hospitals, Mansoura University Faculty of Medicine, El Mansoura, Egypt, [7]Keck School of Medicine at University of Southern California, Los Angeles, CCA, [8]University of Minnesota, Minneapolis, MN, [9]Penn State, State College, PA, [10]Seattle Children's Hospital, Seattle, WA, [11]UNC Chapel Hill, Chapel Hill, NC, [12]UNC Health Care, Durham, NC, [13]Connecticut Children's, Hartford, CT, [14]Riley Hospital for Children at Indiana University Health, Indianapolis, IN, [15]Indiana University, Indianapolis, IN, [16]Dana-Dwek Children's Hospital, Hod Hasharon, Israel, [17]Washington University School of Medicine, St Louis, MO, [18]Nationwide Children's Hospital, Columbus, OH, [19]Mayo Clinic, Rochester, MN

SESSION INFORMATION

Session Date: Saturday, April 1, 2023

Session Title: Plenary Abstracts Session III

Session Type: Plenary Session

Session Time: 11:00AM–12:00PM

Background/Purpose: Calcinosis is a poorly understood and morbid complication of juvenile dermatomyositis (JDM). As there is no consensus treatment approach for calcinosis, and limited knowledge of outcomes, we seek to inform future treatment guidance for this significant complication of JDM by performing this multi-institutional retrospective review of treated cases of JDM calcinosis to assess outcomes as they relate to JDM severity, initial treatment, and calcinosis-directed treatment.

Methods: Collaborators of the Childhood Arthritis and Rheumatology Research Alliance (CARRA) submitted retrospectively reviewed cases with data collected at timepoints of JDM and calcinosis diagnoses, respectively, as well as the determined outcome of each specific calcinosis treatment(s). All cases were sourced from electronic health record searches of clinical terms or billing codes. Only cases diagnosed and treated from 2003 to 2019 were included to capture the contemporary era of JDM treatment. Cases were required to have probable/definitive JDM by Bohan and Peter criteria and calcinosis. Outcome was assessed in univariable and multivariable analyses as time to any improvement by clinician judgement utilizing all available follow-up. Proportion with any improvement along with 95% confidence intervals was estimated using Kaplan-Meier analysis. Multivariable Cox models were used to assess the association between patient characteristics and time to any improvement beginning at calcinosis diagnosis.

Results: Data for 63 patients were collected from 11 institutions. Median age was 7.8 years at JDM diagnosis with symptom duration occurring for median of 6 months prior, and 9.4 years at calcinosis diagnosis with calcinosis symptoms 2.4 months prior to diagnosis. Calcinosis was present at JDM diagnosis in 32%, and JDM was considered active in 76% of cases at the time of calcinosis diagnosis. Fifty percent of patients had a positive myositis antibody, while 71% had a chronic continuous disease course (Table 1). Seventy nine percent of patients ultimately showed improvement and 22% reached complete resolution. Patients received multiple treatment regimens including immunomodulating agents with or without other calcium modifying treatments (Table 1 footnote). IVIG was associated with greater probability of calcinosis improvement (adjusted HR 1.95; 95% CI 1.10 to 3.45; p=0.02) (Table 2) compared to treatment without IVIG. Patients who received immunomodulatory treatment with other calcium modifying agents tended to show improvement more quickly (Figure 1).

	Overall (N=63)
Age at calcinosis diagnosis, years	9.4 [5.7, 13.3]
Age at JDM diagnosis, years	7.8 [4.1, 11.1]
Duration of calcinosis symptoms prior to calcinosis diagnosis, years	0.2 [0.1, 0.3]
Duration of JDM symptoms prior to JDM diagnosis, years	0.5 [0.2, 1.0]
Male sex, n (%)	18 (28.6)
Race, n (%)	
White	25 (39.7)
Hispanic, Latino or Spanish origin	17 (27.0)
Black, African American, African or Afro Caribbean	12 (19.0)
Middle Eastern or North African	6 (9.5)
Asian	1 (1.6)
Unknown	2 (3.2)
Disease course, n (%)	
Monocyclic	2 (3.2)
Polycyclic	10 (15.9)
Chronic	44 (69.8)
Not applicable	7 (11.1)
Any myositis antibody (n=50), n (%)	25 (50.0)
Characteristics at JDM diagnosis, n (%)	
Calcinosis present	20 (31.7)
Aggressive treatment of JDM†	38 (60.3)
Characteristics at calcinosis diagnosis, n (%)	
Active JDM (n=62)	47 (75.8)
Moderate to severe JDM	33 (52.4)
Active muscle involvement	31 (49.2)
Active skin involvement	51 (81.0)
Background medication(s)‡	41 (65.1)
Treatment, n (%)	
None	11 (17.5)
Immunomodulatory agent(s)§	28 (44.4)
Immunomodulatory agent(s) plus Ca-modifying treatment(s)	19 (30.2)
Calcium modifying treatment(s)‖	5 (7.9)
Immunosuppressant(s)	30 (47.6)
Bisphosphonates	13 (20.6)
IVIG	20 (31.7)
Biologics	4 (6.3)
Surgical excision resulting from referral	7 (11.1)

* Continuous variables are summarized as median [interquartile range]. Categorical variables are summarized as frequency (percentage). Observations with complete information are presented when fewer than 63.

† Within the first 3 months of JDM diagnosis, receiving IV methylprednisolone ≥ 2 mg/kg/day continued for more than 1 week, and/or cyclophosphamide, and/or IVIG and/or rituximab

‡ Includes glucocorticoids, immunomodulatory drugs, IVIG, and biologics.

§ Includes glucocorticoids, immunosuppressants (Methotrexate, Leflunomide, Azathioprine, Mycophenolate mofetil, Mycophenolic acid, Sirolimus, Tacrolimus, Thalidomide, Lenalidomide, Cyclosporine, Hydroxychloroquine, Sulfasalazine, Cyclophosphamide, Colchicine), non-biologic DMARDs (tofacitinib, baricitinib, ruxolitinib), IVIG, Biologic DMARDs (Rituximab, Etanercept, Infliximab, Adalimumab, Certolizumab, Golimumab, Anakinra, Canakinumab, Rilonacept, Abatacept, Tocilizumab).

‖ Includes agents that effect calcium or phosphorus (Bisphosphonates, Vitamin D, Vitamin C, Calcium-channel blocker, Sodium thiosulfate, Aluminum hydroxide, Warfarin, Minocycline, Probenecid).

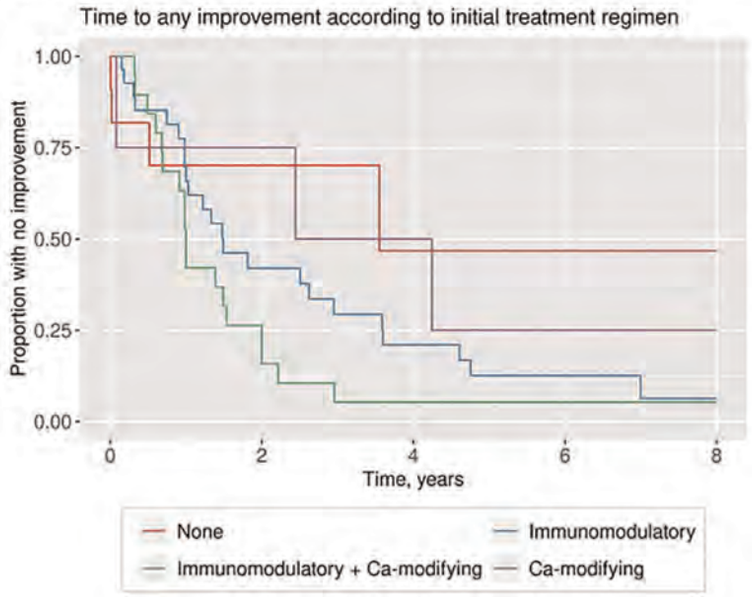
Conclusion: Our study showed that patients with JDM calcinosis received multiple treatment regimens including both immunomodulating therapies and calcium modifying agents, and reassuringly, most patients showed improvement over time. Among the immunomodulating therapies, IVIG was statistically significantly associated with improvement when compared to treatment without IVIG. We plan to study other factors associated with calcinosis outcomes, including analysis of patients with complete resolution. Improved knowledge of treatment choices and outcomes can support future prospective studies.

Table 2. Summary of Adjusted Univariable Analyses of Time-dependent Treatment Regimens*

Treatment Regimen	*HR (95% CI, adjusted)*	*P*
None	Ref.	
Immunomodulatory Agent(s)	2.57 (0.74, 8.96)	0.139
Calcium modifying treatment(s)	1.79 (0.38, 8.32)	0.458
Immunomodulatory Agent(s) + Ca-modifying treatment(s)	3.08 (0.90, 10.55)	0.074
Types of Immunomodulatory Agent		
Immunosuppressant(s)	1.40 (0.78, 2.49)	0.259
IVIG	1.95 (1.10, 3.45)	0.022
Biologics	1.40 (0.66, 3.00)	0.382
Bisphosphonates	1.23 (0.67, 2.24)	0.506
Surgical Excision	0.82 (0.24, 2.79)	0.751

*Results are from Cox proportional hazards models adjusted for age, sex, and time between initial JDM diagnosis and calcinosis diagnosis. Association between treatment and improvement was assessed using time-dependent indicators for the given treatments. Hazard ratios represent the multiplicative increase in hazard for improvement associated with the given treatments.

Figure 1. Time to Improvement of Calcinosis According to Treatment Regimen



Disclosure: B. Yi: None; **D. Wahezi**: None; **L. Covert**: None; **K. Ardalan**: None; **J. Hui-Yuen**: None; **N. Vasquez Canizares**: None; **D. Mosad Mosa**: None; **M. Jones**: None; **C. Correll**: None; **A. Begezda**: None; **S. Shenoi**: Novartis, 2, Pfizer, 1; **E. Wu**: Enzyvant, 2, Pharming Healthcare, Inc, 2; **L. Kovalick**: None; **W. Lapin**: None; **S. Tarvin**: None; **M. Oliver**: None; **M. Rodriguez**: None; **I. Marmor**: None; **K. Baszis**: None; **A. Taxter**: None; **A. Hanson**: None; **C. Crowson**: None; **A. Orandi**: None.

Abstract Number: 009

The Bridge to Adult Care from Childhood for Young Adults with Rheumatic Disease (BACC YARD) Program, a Pediatric-to-Adult Rheumatology Transition Program

John Bridges[1], Livie Huie[2], Amanda Alexander[2], Randy Cron[2], Maria I. (\"Maio\") Danila[3], Victoria Gennaro[4], Laura Hughes[2], Bailey Lipham[5], Linda McAllister[4], Matthew Mullen[2], Annelle Reed[6], Daniel Reiff[2], Carolyn Smith[6], Emily Smitherman[2], Matthew Stoll[2], Peter Weiser[2] and Melissa Mannion[2], [1]University of Alabama at Birmingham/Children's of Alabama, Birmingham, AL, [2]University of Alabama at Birmingham, Birmingham, AL, [3]University of Alabama at Birmingham (UAB), Birmingham VA Medical Center, Birmingham, AL, [4]Children's of Alabama, Birmingham, AL, [5]University of Alabama Medical School, Birmingham, AL, [6]Children's of Alabama, Birmingham, AL

SESSION INFORMATION
Session Date: Saturday, April 1, 2023
Session Title: Plenary Abstracts Session III
Session Type: Plenary Session
Session Time: 11:00AM–12:00PM

Background/Purpose: Children with chronic rheumatic conditions age and require transfer to adult rheumatologists for continued care. The transition period from pediatric to adult-oriented care is a high-risk time for disease flare and poor outcomes. Our objective was to design a structured transition program and determine the impact on time to first adult rheumatology appointment compared to historical controls.

Methods: In 2020 we initiated a specialized transition program structure utilizing a dual adult/pediatric rheumatology clinician with the goal to improve transition outcomes, the **B**ridge to **A**dult **C**are from **C**hildhood for **Y**oung **A**dults with **R**heumatic **D**isease (**BACC YARD**) Program. Young adults who received the BACC YARD intervention were compiled into an observational registry. In this first phase of the program, an adult/pediatric rheumatologist joined the care team for the final pediatric rheumatology visit, and then saw the patient with an adult rheumatology attending for the first visit in the adult rheumatology clinic. Requested timing for first adult rheumatology visit was determined by joint decision between the adult/pediatric rheumatologist and the pediatric attending at time of pre-transfer visit. The historical control cohort was obtained from the electronic records of Childrens of Alabama and University of Alabama at Birmingham (UAB) between March 1, 2018 and March 1, 2020.

Table 1: Characteristics of BACC YARD Program Cohort, 7/2020-5/2022 (N=86)

Characteristic		N (%)
Sex	Female	63 (73)
Age [Median (Range)]		20 (17-26)
Race		
	Black	29 (34)
	White	56 (65)
	Asian	1 (1)
Ethnicity		
	Hispanic	6 (7)
Rheumatic Disease of Childhood		
	JIA	50 (58)
	SLE and related CTDs	26 (30)
	Vasculitis	3 (3)
	Uveitis	2 (2)
	Inflammatory Myopathy	2 (2)
	RPC	1 (1)
	Morphea	1 (1)
	SURF	1 (1)

BACC YARD: Bridge to Adult Care from Childhood for Young Adults with Rheumatic Disease; **JIA**: juvenile idiopathic arthritis; **SLE**: systemic lupus erythematosus; **CTD**: connective tissue disease; **RPC**: relapsing polychondritis; **SURF**: syndrome of undifferentiated recurrent fever

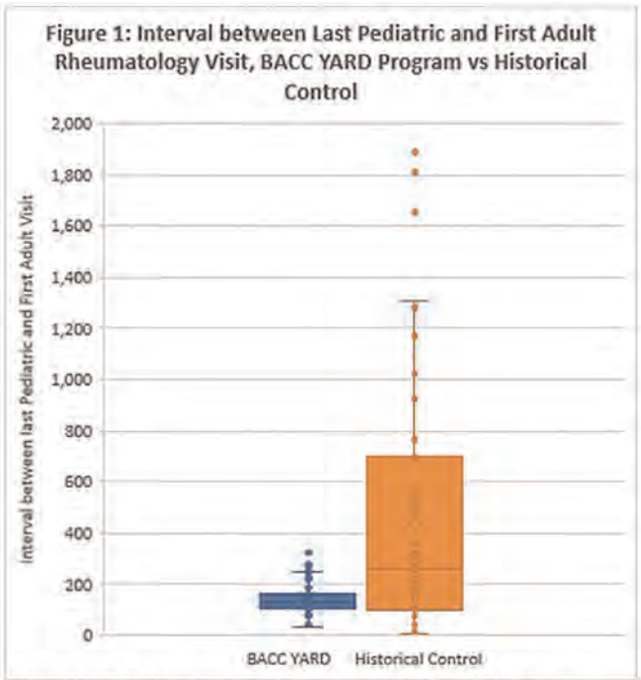
All historical patients were identified who had at least two pediatric rheumatology visits prior to transfer and were successfully transferred to UAB rheumatology. Data harvest was supplemented by individual chart review to ensure that there was no documentation of visits with a rheumatologist outside of the Childrens of Alabama/UAB system in the peri-transfer period.

Results: The BACC YARD program included 86 participants from 7/2020-5/2022. 8% of participants were lost to follow-up at Childrens of Alabama prior to the BACC YARD program but successfully re-established care at UAB through the BACC YARD program. 3.5% of participants were seen at other childrens hospitals for a childhood-onset rheumatic disease but

Table 2: Median Transfer Intervals in Days, BACC YARD Program Cohort vs Historical Control Cohort

	Median (Range)	p value
BACC YARD from Children's of Alabama Transferred to UAB (7/2020-4/2022, N=56)		
	JIA (N=35)	
Requested	128 (43-207)	
Actual	129 (43-342)	
	SLE (N=14)	
Requested	100 (43-157)	
Actual	119 (36-249)	
	Other (N=7)	
Requested	119 (99-149)	
Actual	149 (127-323)	
	Total (N=56)	
Requested	119 (43-207)	
Actual	141 (43-342)	<0.05
Historical Control Cohort from Children's of Alabama Transferred to UAB (3/2018-2/2020, N=45)		
Actual	261 (8-1888)	<0.001

BACC YARD: Bridge to Adult Care from Childhood for Young Adults with Rheumatic Disease; **JIA**: juvenile idiopathic arthritis; **SLE**: systemic lupus erythematosus



BACC YARD: Bridge to Adult Care from Childhood for Young Adults with Rheumatic Disease

established adult care at UAB through the BACC YARD program. 19% of participants had upcoming appointments scheduled at UAB but not completed by time of this analysis. 65% of participants successfully completed a pre-transfer visit at Childrens of Alabama and a post-transfer visit at UAB completed by 5/2022. 58% of the participants had a diagnosis of juvenile idiopathic arthritis and 30% had a diagnosis of systemic lupus erythematosus or similar connective tissue disease (**Table 1**). Actual median transfer intervals were 129 days for JIA, 119 days for SLE, and 141 days for other diagnoses (**Table 2**). There was a statistically significant shorter median interval between last pediatric and first adult visit in BACC YARD compared to the historical control cohort (141 days vs 261 days, respectively, p 0.001; **Figure 1**).

Conclusion: The BACC YARD Program cohort had a significantly reduced median time to first adult visit compared to historical controls. After further study, the BACC YARD intervention could be disseminated to other clinics for young adults with childhood onset rheumatic disease.

Disclosure: J. Bridges: None; **L. Huie**: None; **A. Alexander**: None; **R. Cron**: None; **M. Danila**: None; **V. Gennaro**: None; **L. Hughes**: None; **B. Lipham**: None; **L. McAllister**: None; **M. Mullen**: None; **A. Reed**: None; **D. Reiff**: None; **C. Smith**: None; **E. Smitherman**: None; **M. Stoll**: None; **P. Weiser**: None; **M. Mannion**: None.

Abstract Number: 010

Remotely Delivered Psychological Intervention May Be Beneficial to Youth with Childhood-Onset Lupus: A Preliminary Investigation

Natoshia Cunningham[1], Michelle Adler[2], Ashley Danguecan[3], Mallet Reid[2], Samantha Ely[4], Mathew Reeves[2], Lawrence Ng[3], Paris Moaf[3], Sarah Mossad[3], Tala El Tal[3], Luana Flores Pereira[3], Deborah Levy[3], Linda Hiraki[3], Jennifer Stinson[3], Sarah Ahola Kohut[3], Khalid Abulaban[5], Elizabeth Kessler[5], Stacy Allen[5], Tamar Rubinstein[6], Evin Rothschild[6], Natalie Rosenwasser[7], Kabita Nanda[7], Susan Canny[7], Emily Smitherman[8], Livie Huie[8], James Birmingham[9], Allison Thompson[10], Janel Thompson[10], Miranda Moyer[10], Emily Nguyen[10], Angela Chapson[10] and **Andrea Knight**[3], [1]Michigan State University, Grand Rapids, MI, [2]Michigan State University, East Lansing, [3]The Hospital for Sick Children, Toronto, ON, Canada, [4]Wayne State University, Michigan State University, Detroit, MI, [5]Helen DeVos Children's Hospital, Grand Rapids, MI, [6]Children's Hospital at Montefiore, New York, NY, [7]Seattle Children's Hospital, Seattle, WA, [8]University of Alabama at Birmingham, Birmingham, AL, [9]Metro Health, Cleveland, [10]Patient Co-Investigative Team

SESSION INFORMATION
Session Date: Saturday, April 1, 2023
Session Title: Plenary Abstracts Session III
Session Type: Plenary Session
Session Time: 11:00AM–12:00PM

Background/Purpose: Childhood-onset systemic lupus erythematosus (cSLE) is associated with fatigue, pain, and depressive symptoms that adversely impact health-related quality of life. The Treatment and Education Approach for Childhood-Onset Lupus (TEACH), a 6-week cognitive behavioral intervention, has been shown to reduce fatigue and depressive symptoms in pilot testing. Therefore, we worked with a patient/caregiver co-investigative team to adapt TEACH into a remote intervention. This ongoing multisite trial examines whether the remotely delivered TEACH program is feasible, and explores the effect of TEACH on fatigue, depressive symptoms, and pain compared to standard medical treatment after eight and twenty weeks. The present study examines interim results from participants who have completed the TEACH program.

Methods: We conducted a randomized controlled trial of TEACH. Youth ages 12-22 years meeting the ACR classification criteria for cSLE were referred to the study from 7 rheumatology sites across the U.S. and Canada, with 60 patients anticipated to complete the study. Referred patients were screened and were eligible for the study if they had clinical elevations in ≥1 domain using validated measures for fatigue (PROMIS Fatigue T-Score ≥ 60), average pain intensity (visual analog

scale ≥ 3/10), and depressive symptoms (Childrens/Beck Depression Inventory T-Score ≥ 60, 90). Eligible participants were randomized to either 1) TEACH, delivered by a live provider over six weekly, one-hour zoom sessions and standard medical care; or 2) standard medical care alone over a period of six weeks. TEACH recruitment and retention rates were used to assess preliminary feasibility. Paired samples *t*-tests examined changes in symptoms for TEACH completers at post assessment 8 weeks and 20 weeks from baseline, respectively.

Results: Of the 188 patients referred and approached, 85 (45%) completed screening measures, with 63 (74%) qualifying and enrolling for the study. Retention of enrolled participants from baseline to post assessment was high (92%). At the 8-week post assessment, those who completed TEACH (current n = 26) showed significant reductions in fatigue (t (25) = 5.680, p 0.001, M_{pre} = 62.4 ± 7.4 M_{post} =53.5 ± 8.2) and improved mood symptoms (t(25) = 5.823, p 0.001, M_{pre} = 64.6 ± 11.2, M_{post} = 51.3 ± 9.5), and trending reductions in pain (t(25) = 1.805, p = 0.083, M_{pre} = 3.2 ± 2.0, M_{post} = 2.3 ± 1.8) at the post-assessment. At the 20-week follow-up assessment, TEACH completers (current n = 17), maintained significantly improved fatigue (t(16) = 2.757, p = 0.013, M_{pre} = 62.7 ± 8.2, $M_{follow\text{-}up}$ = 56.4 ± 10.1) and mood (t(16) = 3.402, p = 0.003, M_{pre} = 61.5 ± 11.4, $M_{follow\text{-}up}$ = 53.3 ± 11.7) symptoms from baseline.

Conclusion: To date, remotely delivered TEACH appears to be feasible as demonstrated by high retention rates. Preliminary analyses suggest TEACH is beneficial regarding symptom reduction which is maintained for 20 weeks for youth with cSLE. Future analyses upon the trials completion will determine whether TEACH is more beneficial than standard medical care alone.

Disclosure: N. Cunningham: None; **M. Adler**: None; **A. Danguecan**: None; **M. Reid**: None; **S. Ely**: None; **M. Reeves**: None; **L. Ng**: None; **P. Moaf**: None; **S. Mossad**: None; **T. El Tal**: None; **L. Flores Pereira**: None; **D. Levy**: Janssen, 1, Roche, 5, Sobi, 1, 5; **L. Hiraki**: None; **J. Stinson**: None; **S. Ahola Kohut**: None; **K. Abulaban**: None; **E. Kessler**: None; **S. Allen**: None; **T. Rubinstein**: None; **E. Rothschild**: None; **N. Rosenwasser**: None; **K. Nanda**: None; **S. Canny**: None; **E. Smitherman**: None; **L. Huie**: None; **J. Birmingham**: None; **A. Thompson**: None; **J. Thompson**: None; **M. Moyer**: None; **E. Nguyen**: None; **A. Chapson**: None; **A. Knight**: None.

Abstract Number: 011

The Impact of Social Inequities on Presentation of Childhood-Onset Systemic Lupus Erythematosus (cSLE) at a Large Tertiary Center

Emily Beil[1], Eyal Muscal[2], Danielle Guffey[2], Marietta Deguzman[1] and Erin Peckham-Gregory[2], [1]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [2]Baylor College of Medicine, Houston, TX

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 1 - Lupus: Genetics, Epigenetics, & Social Determinants
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Differences in prevalence rates of childhood-onset SLE (cSLE) among different racial/ethnic groups have been well described. Yet, the role of social determinants of health (SDoH) in severity of presentation is not well understood. We hypothesized that in an urban center with a large, diverse catchment area, SDoH may influence the severity of cSLE at initial diagnosis.

Methods: We completed an IRB-approved retrospective review of children diagnosed with cSLE between 1/1/18 - 5/31/22 at Texas Childrens Hospital in Houston, TX. Patients were excluded if diagnosis was made at an outside institution or diagnosed at 18 years. We abstracted demographics, clinical characteristics and SDoH data such as insurance status,

Table 1. Select Patient and Social Characteristics (N=136)

Patient Characteristics		Social Characteristics	
Age	13.4 years ± 3 SD	**Primary Language**	
Sex		English	92 (67.6%)
Female	Female 112 (82.4%)	Spanish	42 (30.9%)
Male	Male 24 (17.6%)	Other	2 (1.5%)
Race/Ethnicity		**PCP**	
Caucasian	8 (5.9%)	Yes	91 (66.9%)
African American	35 (25.7)	No	20 (14.7%)
Asian	11 (8.1%)	Unknown	25 (18.4%)
Biracial	10 (7.4%)	**Insurance**	
Hispanic	72 (52.9%)	Medicaid/CHIP	68 (50%)
Organ Involvement		Private	54 (39.7%)
Hematologic	122 (89.7%)	None	14 (10.3%)
Joint	78 (57.4%)	**Referral Source**	
Cutaneous	63 (46.3%)	PCP	36 (26.5%)
Renal	48 (35.3%)	ED/Hospital	67 (49.3%)
Mucosal	35 (26.5%)	Subspecialty	33 (24.3%)
Pulmonary	14 (10.3%)	**Diagnosis Location**	
Cardiac	12 (8.8%)	Outpatient	68 (50%)
Gastrointestinal	10 (7.4%)	Inpatient Floor	49 (36%)
CNS	8 (5.9%)	Inpatient ICU	19 (14%)
Muscle	5 (3.7%)	**Social Work Consulted**	109 (80.1%)
SLICC Criteria	7 ± 2.4 SD	Insurance	31 (28.4%)
ACR Criteria	4.5 ± 1.4 SD	Psychiatric	28 (25.7%)
SLEDAI Score	12.5 + 7.1 SD	Transportation	21 (19.3%)
Medications Used		Transition	19 (17.4%)
IV Steroids	82 (60.3%)	Financial	8 (7.3%)
PO Steroids	45 (33.1%)	Legal Assistance	5 (4.9%)
Hydroxychloroquine	132 (97.1%)	School Needs	5 (4.9%)
Azathioprine	23 (16.9%)	Death Support	4 (3.7%)
Methotrexate	16 (11.8%)	Food Insecurity	4 (3.7%)
Cyclophosphamide	14 (10.3%)	Medical Noncompliance	3 (2.8%)
Rituximab	43 (31.6%)	Unstable Employment	2 (1.8%)
MMF	50 (36.8%)	Other	3 (2.8%)
ASA	51 (37.5%)	No Needs Identified	27 (24.8%)
Enoxaparin	4 (2.9%)		
Plasmapheresis	3 (2.2%)		
Other	4 (2.9%)		
Deaths	3 (2.2%)		

social work consultation and access to PCP. Statistical analyses were performed using Stata v15.1 (StataCorp, College Station, TX, USA) and comparisons were made using ANOVA and Kruskal-Wallis tests.

Results: The mean age of the 136 patients who met inclusion criteria was 13.4 years ± 3 SD. The majority of patients were female (n=112, 82.4%) and ~80% were Hispanic White (n=72, 52.9%) or Black (n=35, 25.7%). Fourteen (10.3%) patients were uninsured and 50% (n=68) had Medicaid or Childrens Health Insurance Program (CHIP). Half of the patients (n=68) were diagnosed during an inpatient admission with 49 (36%) on the floor and 19 (14%) in the ICU. The average SLEDAI was 12.5 ± 7.1 SD with 48.5% (n=66) of patients presenting with SLEDAI 12 (severe disease). One third of patients preferred a language other than English (n=44, 32.4%). Only 66% (n=91) of patients had a documented PCP. Social work was consulted in most patients as per section protocol (n=109, 80.1%); specific reasons listed in Table 1.

The distribution of race/ethnicity showed varied disease activity (SLEDAI and number of ACR criteria), CNS involvement and CYC therapy at levels that reached statistical significance (Table 2). Black and Biracial populations had higher SLEDAI scores at presentation (P =0.01). Non-Hispanic White patients were less likely to have a social work consult compared to the other

Table 2. Summary Statistics by Race/Ethnicity

	Non-Hispanic White (N=8)	Black (N=35)	Asian (N=11)	Biracial (N=10)	Hispanic White (N=72)	*P*-value
	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	
Age at Dx	13.7 (2.5)	13.4 (3.4)	13 (3.0)	11.9 (2.4)	13.6 (2.8)	0.531
SLICC	6.4 (2.4)	7.5 (2.7)	6.8 (2.3)	7.8 (2.7)	6.8 (2.3)	0.424
SLEDAI	8.5 (6.1)	14.8 (7.7)	10.6 (5.9)	17.1 (6.7)	11.5 (6.6)	**0.010**
ACR	4.5 (1.5)	4.9 (1.4)	4.2 (1.3)	5.4 (1.4)	4.2 (1.3)	**0.021**
Dx Location	N (%)	N (%)	N (%)	N (%)	N (%)	0.222
Outpatient	5 (62.5)	12 (34.3)	9 (81.8)	4 (40)	38 (52.8)	
Inpatient Floor	3 (37.5)	16 (45.7)	1 (9.1)	4 (40)	25 (34.7)0	
Inpatient ICU	0	7 (20)	1 (9.1)	2 (20)	9 (12.5)	
Referral Source	N (%)	N (%)	N (%)	N (%)	N (%)	0.082
PCP	2 (25)	5 (14.3)	5 (45.5)	3 (30)	21 (29.2)	
ED/Hospital	3 (37.5)	25 (71.4)	2 (18.2)	5 (50)	32 (44.4)	
Subspecialty	3 (37.5)	5 (14.3)	4 (36.4)	2 (20)	19 (26.4)	
Social Work Consult	N (%)	N (%)	N (%)	N (%)	N (%)	**0.010**
Yes	3 (37.5)	30 (85.7)	7 (63.6)	10 (100)	59 (81.9)	
No	5 (62.5)	5 (14.3)	4 (36.4)	0	13 (18.1)	
Severe Disease Characteristics	N (%)	N (%)	N (%)	N (%)	N (%)	
Renal Disease	2 (25)	17 (48.6)	1 (9.1)	5 (50)	23 (31.9)	0.102
CNS Disease	0	6 (17.1)	1 (9.1)	1 (10)	0	**0.004**
CYC Use	0	9 (25.7)	1 (9.1)	2 (20)	2 (2.8)	**0.003**

race/ethnicities (*P* =0.01). CNS involvement was highest among Black children (*P*=0.004). CYC was most often used in Black and Biracial populations (*P*=0.003).

Several demographic and SDoH parameters influenced disease severity at levels that reached statistical significance, including insurance status, race/ethnicity, referral source, PCP availability, primary language, and transportation needs. When assessed by insurance status, uninsured patients were most likely to be diagnosed on an inpatient floor and those with Medicaid had the highest proportion of ICU admissions (*P*=0.034; Table 3). Uninsured patients and those on Medicaid had the highest percentage of social work consults (*P* =0.001).

Conclusion: In children drawn from a large urban catchment area, we observed an influence of race/ethnicity on disease severity, and also potential SDoH proxy measures (non-English primary language, insurance status and transportation barriers). Work is underway to target modifiable SDoH in our catchment area.

Table 3. Summary Statistics by Insurance Status

	None (N=14)	Medicaid/CHIP (N=68)	Private (N=54)	*P*-value
	Mean (SD)	Mean (SD)	Mean (SD)	
Age at Dx	14.1 (2.5)	13.2 (3.1)	13.3 (2.9)	0.613
SLICC Criteria	7.9 (2.1)	7.1 (2.2)	6.6 (2.6)	0.166
SLEDAI	15.1 (8.9)	13.1 (6.7)	11.1 (6.8)	0.100
ACR Criteria	4.8 (1.1)	4.6 (1.4)	4.4 (1.4)	0.472
Dx Location	N (%)	N (%)	N (%)	**0.034**
Outpatient	3 (21.4)	32 (47.1)	33 (61.1)	
Inpatient Floor	9 (64.3)	23 (33.8)	17 (31.5)	
Inpatient ICU	2 (14.3)	13 (19.1)	4 (7.4)	
Race/Ethnicity	N (%)	N (%)	N (%)	**0.007**
Non-Hispanic White	1 (7.1)	2 (2.9)	5 (9.3)	
Black	0	21 (30.9)	14 (25.9)	
Asian	0	3 (4.4)	8 (14.8)	
Biracial	0	4 (5.9)	6 (11.1)	
Hispanic White	13 (92.9)	38 (55.9)	21 (38.9)	
Referral Source	N (%)	N (%)	N (%)	**0.011**
PCP	1 (7.1)	19 (27.9)	16 (29.6)	
ED/Hospital	12 (85.7)	36 (52.9)	19 (35.2)	
Subspecialty	1 (7.1)	13 (19.1)	19 (35.2)	
Access to PCP	N (%)	N (%)	N(%)	**0.001**
Yes	5 (35.7)	45 (66.2)	41 (75.9)	
No	8 (57.1)	8 (11.8)	4 (7.4)	
Unknown	1 (7.1)	15 (22.1)	9 (16.7)	
Primary Language English	N (%)	N (%)	N (%)	**0.026**
Yes	5 (35.7)	47 (69.1)	40 (74.1)	
No	9 (64.3)	21 (30.9)	14 (25.9)	
Social Work Consulted	N (%)	N (%)	N (%)	**0.001**
Yes	14 (100)	60 (88.2)	35 (64.8)	
No	0	8 (11.8)	19 (35.2)	
Transportation Difficulty	N (%)	N (%)	N (%)	**0.001**
Yes	14 (100)	9 (13.2)	8 (14.8)	
No	0	59 (86.8)	46 (85.2)	

Disclosure: E. Beil: None; **E. Muscal**: sobi, 1; **D. Guffey**: None; **M. Deguzman**: None; **E. Peckham-Gregory**: None.

Abstract Number: 012

Thrombotic Microangiopathic Changes in Kidney Biopsies of Childhood-Onset Systemic Lupus Erythematous Patients with and Without Severe Hematologic Disturbances

Cathy Tsin[1], Megan Troxell[1], Vivek Charu[1], Rufei Lu[2] and Joyce Hsu[3], [1]Stanford University, Palo Alto, CA, [2]UCSF, San Francisco, CA, [3]Stanford Medicine, Children's Health, Palo Alto, CA

SESSION INFORMATION

Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 1 - Lupus: Genetics, Epigenetics, & Social Determinants
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Outcomes for pediatric patients with lupus nephritis (LN) remain suboptimal. LN may present with thrombotic microangiopathy (TMA) seen on kidney biopsy. Childhood-onset SLE patients with or without TMA often also presents with hematologic disturbances. In LN renal biopsies, subtle endothelial changes that are indicative of TMA may be visualized only via electron microscopy (EM), but ultimately are not classified as overt TMA on the final impression of the pathology report. We theorized that there is an intermediate group of LN biopsies with TMA-like changes only seen by EM, which we referred to as "TMA-EM.To date, there are no studies investigating the frequency, predictors or outcomes of TMA in LN in a pediatric population. This study examined the frequency of TMA and TMA EM in our pediatric LN cohort, as well as the association of hematologic disturbances in these patients. We then examined the association of TMA and TMA-EM on renal outcomes after 1 year of treatment of LN.

Methods: We conducted a single-center, retrospective study of all cases of class III, IV and/or V LN confirmed on kidney biopsy from all pediatric patients seen at Stanford Medicine Childrens Health (July 2003 – Feb 2021). Electron microscopies of LN patients were re-reviewed by a single renal pathologist and patients were placed into 3 distinct categories of renal pathology findings: TMA, TMA-EM or no TMA. The primary outcome variable was type of renal pathology finding in patients with severe hematologic disturbance, mild/moderate disturbance or no disturbance. The secondary outcome variable was renal response after 1 year of treatment in patients with TMA, TMA-EM and no TMA. Fishers exact test was used to assess association between type of hematologic disturbances and renal pathology category. This test was also used to compare renal pathology categories to renal response outcomes (complete renal response, partial response or no response).

Results: There were 88 patients studied: 5 patients (5.7%) with TMA on kidney biopsy, 31 (35.2%) with TMA-EM, and 52 (59.1%) without TMA. We found that the distribution of TMA changes varied with hematologic changes present at the time of biopsy ($p = 0.006$ [Fishers exact test]). Among patients with no hematologic disturbances, 0% had evidence of TMA on biopsy. Severe hematologic changes were seen in 60.0% of TMA patients, 16.1% of TMA-EM patients and 5.8% of the no TMA patients on biopsy. We found that the distribution of renal response did not vary by TMA status on biopsy ($p=0.897$). Among patients with no TMA on biopsy, 26.9% showed complete renal response, compared with 59.6% in TMA-EM and 20% in TMA.

Conclusion: Lupus nephritis patients with TMA are more likely to exhibit associated hematologic changes than patients without hematologic disturbances, suggesting the development of TMA is not independent of systemic hematologic changes. TMA-EM does not appear to stand out as a distinct histopathologic grouping resulting in clinical significance. LN patients with TMA and TMA-EM are likely to respond to therapy similarly to those without TMA and there was no significant difference in renal response between the TMA categories.

Disclosure: C. Tsin: None; **M. Troxell**: None; **V. Charu**: None; **R. Lu**: None; **J. Hsu**: None.

Abstract Number: 013

Applying Pathway Analysis to Whole Genome Data to Identify Pathophysiologic Pathways in Childhood-onset Systemic Lupus Erythematosus

Katie Heitzman[1], Luis Franco[2], Linda Hiraki[3], Earl Silverman[3], Christiaan Scott[4], Ana Barrera-Vargas[5], Zuoming Deng[2], Mariana Kaplan[2] and Laura Lewandowski[2], [1]NIH, Bethesda, MD, [2]NIAMS, NIH, Bethesda, MD, [3]The Hospital for Sick Children, Toronto, ON, Canada, [4]University of Cape Town, South Africa, [5]Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán, Ciudad de Mexico, Mexico

SESSION INFORMATION

Session Date: Thursday, March 30, 2023

Session Title: Poster Breakout 1 - Lupus: Genetics, Epigenetics, & Social Determinants

Session Type: Abstract Session

Session Time: 5:10PM–5:40PM

Background/Purpose: Systemic lupus erythematosus (SLE) is a chronic, systemic autoimmune disorder. The pathogenesis of SLE is not fully understood, but twin/sibling high concordance rate suggests a genetic component triggered by stochastic environmental events. Childhood-onset SLE (cSLE) patients have an extreme phenotype, with early age of diagnosis and more severe disease relative to adult-onset SLE. Therefore, cSLE are an ideal population in which to study genetic contributors to disease. Historically, rare variant analyses have focused on a single gene shared by multiple patients in unrelated families. This approach does not consider that rare variants in people may cluster in genes that participate in similar biological processes. Therefore we aim to identify rare, enriched variants in genes of the same biological pathways that may contribute to the pathogenesis of SLE by applying pathway analysis to whole genome sequencing data of a diverse cSLE cohort.

Methods: SLE subjects met at least 4 of 11 revised American College of Rheumatology classification SLE criteria with disease onset prior to age 18. Whole blood samples collected from 81 subjects and 111 parents underwent whole genome sequencing (Table 1). Rare (minor allele frequency cutoff of 0.01), nonsynonymous variants in coding exons present in cSLE patients were selected, resulting in 483 variants in 232 genes. We restricted to variants present in 2 or more cSLE patients and not present in unaffected parents, or variants that were present more often in the cSLE patients than parental controls, assessed by Fisher's exact test ($p < 0.05$). The resulting gene list was sorted into Gene Ontolgoy: Biological Processes (GO:BP) using Metascape. The Cytoscape Enrichment Map App (Cytoscape v. 3.7) was then used to generate a network of pathways enriched for genes in which cSLE patients harbored rare variants.

Results: Of the 483 unique rare exonic variants, 239 variants were sorted into 133 GO:BP genesets. (Table 2) Enrichment map analysis to clustered them into 18 biologic pathways including RNA processing and apoptosis. (Figure 1) Some pathways (e.g. nucleic acid regulation) are well-established causes of monogenic SLE, while others have not previously been described in the SLE.

Table 1. Demographic data for the cSLE cohort

	cSLE (n=81)
% female n	80% (65)
Age at diagnosis, years, median (range)	12 (5-17)
Ancestry	
% European	22 (18)
% African	11 (9)
% Amerindian	17 (14)
% East Asian	16 (13)
% South Asian	6 (5)
% Admixed	27(22)

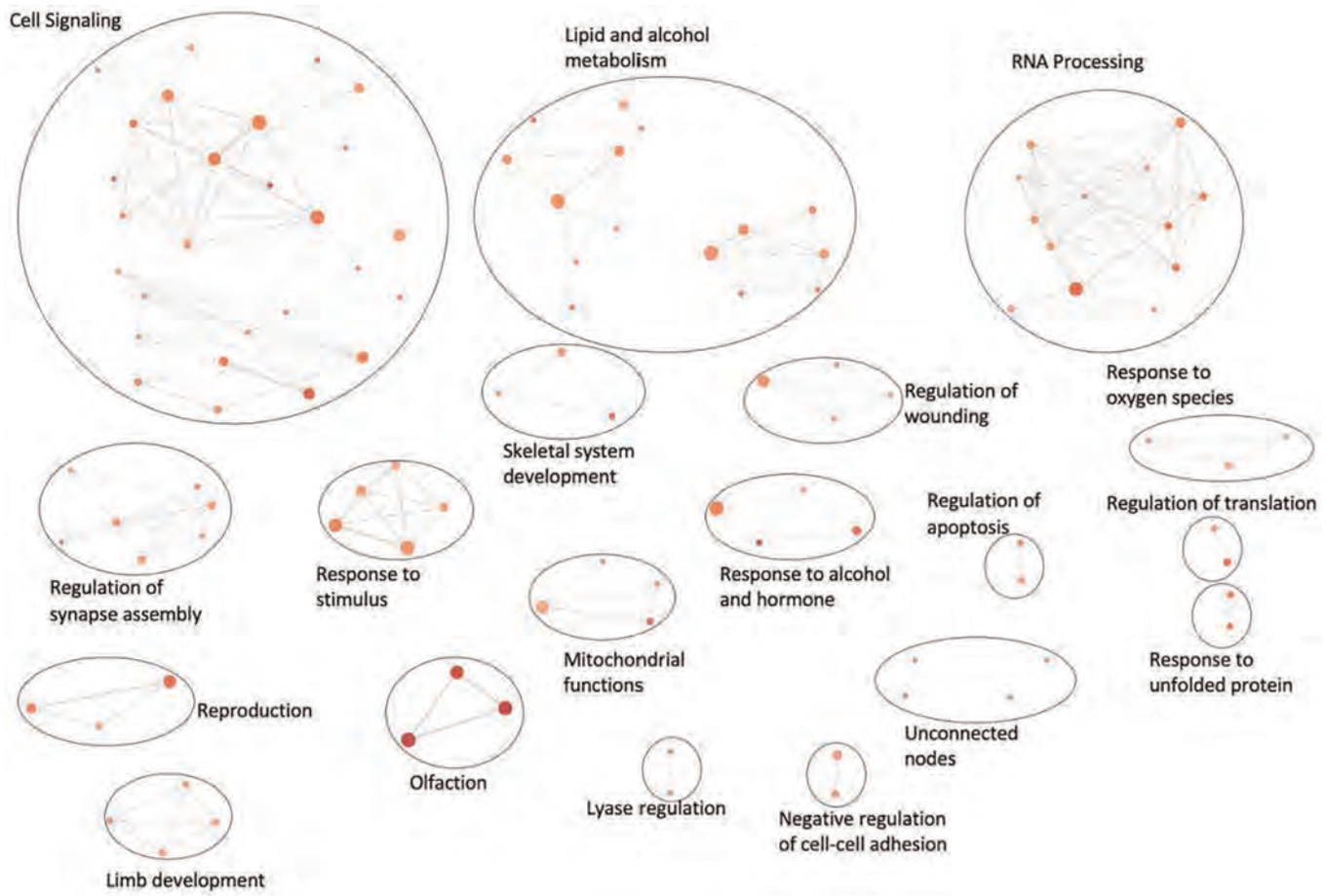


Figure 1. Biologic Pathways Enriched in Rare Coding Variants in cSLE patients. Each red circle is a geneset, represented as a node. The deeper the color of the node, the more the gene set is enriched in rare variants.

Table 2. Comparison of number rare variants in coding regions between cSLE patients and unaffected parents.

	cSLE Patients	Unaffected parents
Total number of individual variants	483	42
Number of variants which were enriched in GO:BP genesets	239	20

Conclusion: Rare, exotic variants were enriched in biologic pathways in cSLE patients compared to parental controls. Pathway-network analysis is a useful approach to identify biological pathways and specific genes that could contribute to SLE risk. Ongoing analysis of the specific variants identified in each pathway will allow us to prioritize key genes and pathways for further study. Additionally, assessing clinical genotype/phenotype correlations could contribute to advancing SLE past a broad clinical phenotype to a more precise molecular diagnosis.

Disclosure: K. Heitzman: None; **L. Franco**: None; **L. Hiraki**: None; **E. Silverman**: None; **C. Scott**: None; **A. Barrera-Vargas**: None; **Z. Deng**: None; **M. Kaplan**: None; **L. Lewandowski**: None.

Abstract Number: 014

Characterizing Lupus in African American Children in Southern United States

Anita Dhanrajani[1], Taylor Long[1], Spencer Hagwood[2], Leslie Johnson[1] and Cynthia Karlson[1], [1]University of Mississipi Medical Center, Jackson, MS, [2]University of Mississipi Medical Center School of Medicine, Jackson, MS

SESSION INFORMATION

Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 1 - Lupus: Genetics, Epigenetics, & Social Determinants
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: African-American (AA) ethnicity is a known predisposing factor for childhood onset systemic lupus erythematosus (cSLE) and a predictor of poor outcomes. In addition to race/ethnicity, income and geographical location are known drivers of health care disparities in cSLE. Despite this knowledge, there is lack of literature describing demographic and clinical features of cSLE in a predominantAA cohort. This study aims to characterize demographic and clinical features of cSLE patients in the Southeast United States compared to current literature describing predominantly Caucasian cohorts. The results from this studywill drive future work in comparing outcomes in cSLE and uncovering factors leading to disparities in other regions of the United States.

Methods: This is a cross sectional study of cSLE patients from 2 centers in Southeast United States – University of Mississippi Medical center (UMMC) and University of Alabama at Birmingham (UAB). Prevalent and incident patients with cSLE at UMMC were enrolled after obtaining consent. Demographic, social and clinical data was retrospectively collected by review of the medical charts. Similar data was obtained for the UAB cohort for patients with cSLE via the CARRA registry database. The data from both cohorts was combined and analyzed using SPSS statistical software. Descriptive statistics were used for demographic and clinical features. Unpaired t test and chi square test were used to compare outcomes in this cohort with those reported in the literature.

Results: Results are described for a total cohort of 36 patients, comprising 17 from UMMC and 19 from UAB. 31/36 (86.11%) were female, 28/36 (77.78%) were of AA ethnicity, and 23/36 (64%) had Medicaid insurance. Mean age at diagnosis (+/- SD) of SLE for the cohort was 13.7 years (+/- 2.9). Mean ACR score at diagnosis was 5.2 (+/- 1.29), SLICC score was 8.8 (+/- 2.2). Average baseline SLEDAI was 12.14 (+/- 9.6), SLEDAI at 6 months and 1 year respectively was 6.84 and 3.95. Average distance traveled to see a rheumatologist was 74.72 miles compared to a national average of 42.8 miles. 30/36 patients (83.33%) belonged to medium-high or high Social Vulnerability Index group based on zip code. Twenty five percent of patients had some renal involvement. Average SLEDAI compared to a multiethnic Canadian cohort with 10%

	Study cohort N=36 N (%)	**1000 faces of lupus cohort N=213 N (%)**	**Chi-square test results**
Female sex	31 (87)	176 (83)	0.26, p= 0.6
African American ethnicity	28 (78)	22 (11)	87.29, **p < 0.00001**
Renal involvement present	9 (25)	72 (36)	1.63, p = 0.20
Neurologic involvement present	7 (19)	26 (13)	1.05, p = 0.30
SDI > 0	15 (47)	32 (16)	24.09, **p < 0.00001**
SDI > 2	12 (33)	14 (7)	21.42, **p < 0.00001**
Social vulnerability index >= 2	30 (83)	NA	NA

	Study cohort (N=36) Mean (+/- SD)	1000 faces of lupus cohort (N= 199) Mean (+/- SD)	Unpaired t test results
Age at diagnosis (years)	13.7 (2.9)	12.6 (3)	**p=0.04**
ACR criteria	5.2 (1.29)	N/A	N/A
SLEDAI baseline	12.14 (9.6)	3.1 (2.1)	**p<0.0001**
SLEDAI at 6 months	6.84 (7.19)	N/A	N/A
SLEDAI at 1 year	3.95 (3.32)	N/A	N/A
Social vulnerability index score	0.71	N/A	N/A

black population, was significantly higher: 12.14 versus 3.1 ($p < 0.00001$). 15/36 had SDI > 0 (46.66%) versus 16% reported in the literature ($p < 0.0001$). 12/36 (33.3%) had SDI >/= 2 compared to 7% reported in the literature ($p < 0.0001$).

Conclusion: This study described the clinical and socioeconomic characteristics of a predominately AA cSLE population from 2 centers in the Southeastern United States. Compared to multiethnic cohorts of cSLE from Canada, this patient population has significantly higher disease activity and greater damage accrual. Social risk factors for this population include a higher social vulnerability index, longer distance from an academic pediatric rheumatology center, and having Medicaid insurance. The effect of these factors on disparity of disease outcomes needs to be further explored with larger cohorts.

Disclosure: A. Dhanrajani: None; **T. Long**: None; **S. Hagwood**: None; **L. Johnson**: None; **C. Karlson**: None.

Abstract Number: 015

Epigenetically-Distinct B Cell Profiles Pre- and Post-Induction Therapy in Pediatric Lupus

Joyce Hui-Yuen[1], Kaiyu Jiang[2], Susan malkiel[3], Betty Diamond[4] and James Jarvis[5], [1]Cohen Children's Medical Center, Northwell Health, Lake Success, New York; Center for Autoimmune, Musculoskeletal, and Hematopoietic Diseases Research, Feinstein Institutes for Medical Research, Northwell Health, Manhasset, NY, [2]University at Buffalo, Buffalo, NY, [3]Feinstein Institutes for Medical Research, Manhasset, NY, [4]The Feinstein Institutes for Medical Research, Manhasset, NY, [5]University at Buffalo Jacobs School of Medicine, Buffalo, NY

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 1 - Lupus: Genetics, Epigenetics, & Social Determinants
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Systemic lupus erythematosus (SLE) may be triggered by gene-environment interactions. Data are scarce on how epigenetic variance contributes to disease risk in pediatric SLE (pSLE). Our objective was to identify differences in chromatin accessibility in treatment-naïve pSLE compared to healthy children (HC) and pSLE patients post-induction therapy.

Methods: We used assays for transposase-accessible chromatin-sequencing (ATACseq) in 8 pSLE patients pre- and post-induction therapy and 5 HC to investigate whether regions of open chromatin unique to pSLE patients demonstrate enrichment for transcriptional regulators, using standard computational approaches and a false discovery rate of < 0.05.

Results: The mean age of onset was 13.75 (range 7-17) years in pSLE, and mean lupus activity index was 12.8 (range 6-24). There were 245 differentially accessible regions (DAR) around peaks unique to treatment-naïve pSLE patients; 64.3% were more accessible in pSLE than HC. Many of the DAR are located more than 100kb from the nearest transcription start site

(nTSS), implying transcription factors (TF) may be acting on distal enhancers to regulate transcription. pSLE DAR were enriched for the enhancer H3K4me3. In DAR encompassing TF binding sites, pSLE samples, but not HC, were enriched for disease-associated SNPs previously identified in lupus association studies. Variant calling within DAR found 3864 genes belonging to disease-relevant biologic processes such as cellular activation in immune response. In contrast, 86.7% of peaks unique to pSLE patients post-induction therapy were located distal to nTSS. Induction therapy for pSLE patients included corticosteroids in all patients, cyclophosphamide in 5, and mycophenolate in 3. DAR from the pSLE patients post-induction therapy were not enriched for enhancers or disease-associated SNPs.

Conclusion: We demonstrate an epigenetically-distinct profile in pSLE B cells when compared to HC, indicating pSLE B cells are predisposed for disease development. Pathways of significance analyses identified immunologic pathways important in the pro-inflammatory response in treatment-naïve pSLE patients, that were absent in analyses from the same patients post-induction therapy. Thus, increased chromatin accessibility in genomic regions controlling activation of inflammatory and immune responses suggest transcriptional dysregulation of key players in immune cell activation plays an important role in pathogenesis of SLE. Treatment with corticosteroids and immunosuppressives changes this epigenetic profile, making pathways responsible for inflammation and B cell activation less accessible.

Disclosure: J. Hui-Yuen: None; **K. Jiang**: None; **S. malkiel**: None; **B. Diamond**: None; **J. Jarvis**: None.

Abstract Number: 016

Gene Expression Changes in Polyarticular Juvenile Idiopathic Arthritis Following Tofacitinib Treatment

Esraa Eloseily[1], Alex Pickering[2], Sanjeev Dhakal[3], Alexei Grom[3], Hermine Brunner[3] and Sherry Thornton[4], [1]Division of Pediatric Rheumatology, Cincinnati Children's Hospital Medical Center, Cincinnti, OH, [2]Department of Biomedical Informatics, Harvard Medical School, Boston, MA, [3]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, [4]Cincinnati Children's Hospital, Cincinnati, OH

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 2 - JIA & Uveitis: Genetics, Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Despite advances in the understanding of juvenile idiopathic arthritis (JIA) pathophysiology, personalized treatments informed by gene transcriptomic profiles remain elusive. We aimed toexamine the relationship between changes in gene expression and treatment response in patients with JIA treated with tofacitinib.

Methods: Whole blood samples from patients with JIA were collected using Paxgene tubes at baseline (BL) and again at week (wk) 18 of treatment with tofacitinib as part of a clinical trial (NCT02592434) (1). Patients were classified as treatment responders (TR) if they achieved at least a JIA-American College of Rheumatology score of (JIA-ACR70) or above (N_{BL}=47; N_{WK18}=38); and non-responders (NR) improvement was no more than a JIA-ACR30 response (N_{BL}=20; N_{WK18}=8). Bulk RNA was isolated, subjected to globin/rRNA depletion, and used for generation of cDNA libraries for subsequent sequencing. RNA sequencing via Illumina Nova-seq generated 50 million reads per sample. Gene expression was compared in TR and NR (BL, wk18; change BL to wk18). Kallisto 0.48.0 was used for pseudo-quantification (GRCh38 release 94 index). Differential expression and Gene Ontology (GO) over-representation analyses were performed with dseqr 0.34.0.

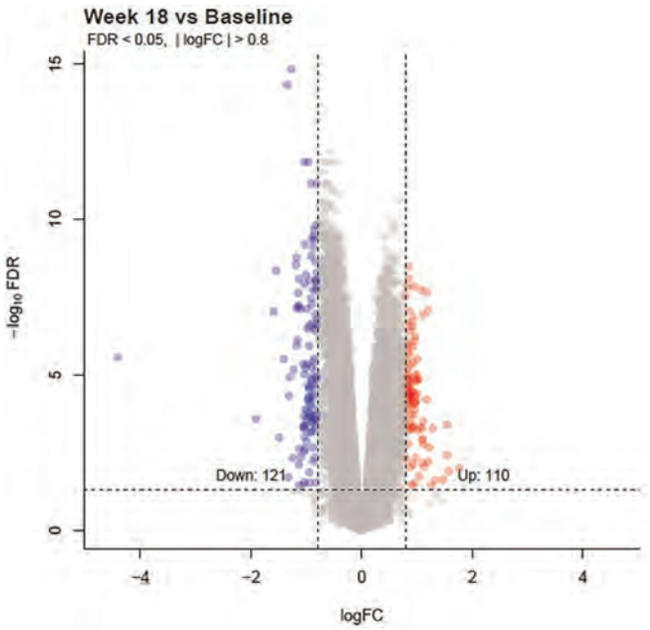


Figure 1: Volcano plot showing differential gene expression between BL and wk18 (FDR<0.05 and absolute logFC>0.8) n=231 (110 upregulated genes and 121 downregulated genes).

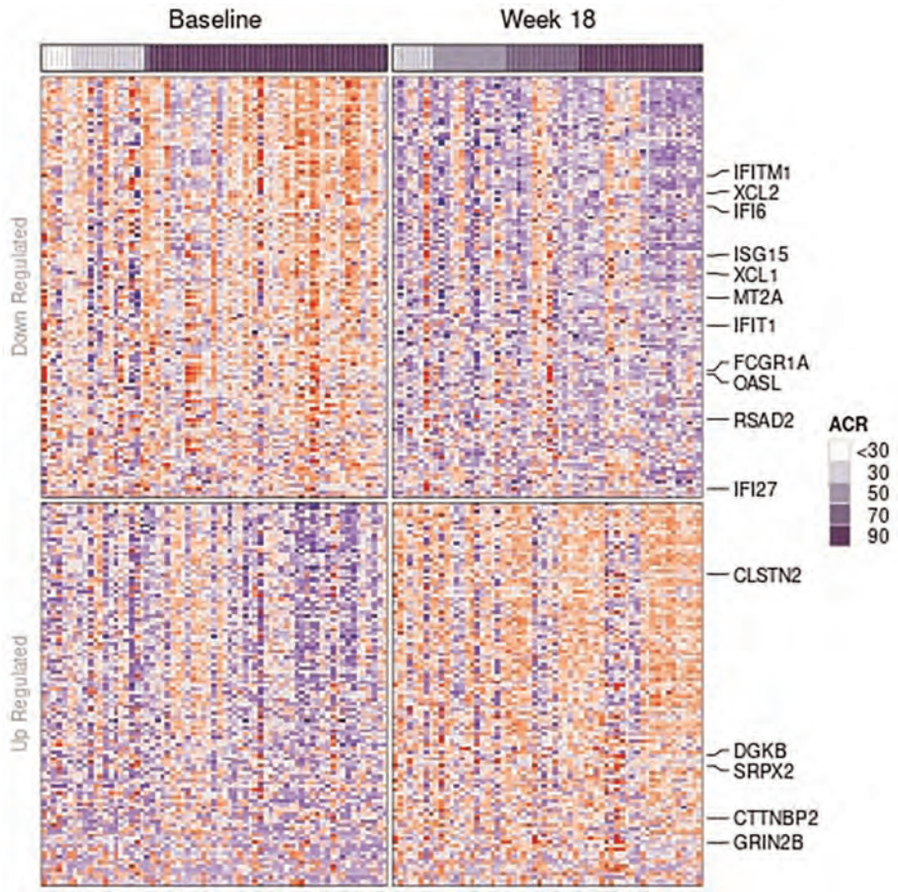


Figure 2: Heatmap showing differentially expressed genes (FDR < 0.05 and |logFC| > 1) at week 18 vs baseline in JIA patients receiving tofacitinib. Annotated genes are from ontologies related to type I and type II interferon activity (down-regulated), as well as from ontologies related to synapse organization (up-regulated).

Results: Significant differential expression (FDR< 0.05) was observed in 10,138 genes between BL and wk18. GO over-representation analysis was performed separately for up- and down-regulated genes with FDR< 0.05 and absolute logFC >0.8 (n=231) (Figure 1). For genes down-regulated with treatment (BL-wk18), ontologies related to negative regulation of gene expression, type I and type II interferon pathways were significantly over-represented (FDR< 0.05), while ontologies for synapse organization and activity were over-represented among up-regulated genes (Figure 2). The latter might reflect the normalization of the immunological synapse leading to improved cytotoxic NK and T cell functions, the impairment of which has been proposed to escalate the production of IFN-γ and other proinflammatory cytokines (2). There were no significantly differentially expressed genes (FDR < 0.05) between TR vs. NR at BL or wk18. Exploratory GO analysis in NR vs. TR at baseline (FDR< 0.13, n=76 genes) suggests up-regulation of genes in ontologies related to the activation of MAP kinase activity.

Conclusion: Tofacitinib treatment in JIA patients leads to widespread changes in whole blood transcriptional profiles. The most significant transcriptional changes observed included the down-regulation of genes associated with type I and type II interferon pathways, and the up-regulation of genes involved in synapse organization and activity. Potential association of MAP-kinase activation in NR at baseline needs to be further explored. MAP-kinase is one of the signalling pathways in inflammatory arthritis that can be targeted by small molecules other than JAK inhibitors. Thus, patients with a predominantly active MAP-kinase pathway could be less likely to respond to tofacitinib. If confirmed, these findings might be useful to personalize JIA treatment in the future.

Disclosure: E. Eloseily: None; **A. Pickering**: None; **S. Dhakal**: None; **A. Grom**: Novartis, 2, 5, Sobi, 2, 5; **H. Brunner**: GENENTECH, 12, provision of study drug for NIAMS funded study, Pfizer, 1, 2, 6; **S. Thornton**: None.

Abstract Number: 017

Potential Uveitic Biomarkers in Tears of Children with Juvenile Idiopathic Arthritis: A Pilot Study

Tiffany Nguyen[1], **Ilaria Maccora**[1], Mekibib Altaye[1], Wendy Haffey[2], Theresa Hennard[1], Alyssa Sproles[1], Sherry Thornton[1], Virginia Miraldi Utz[1], Ken Greis[2] and Sheila Angeles-Han[1], [1]Cincinnati Children's Hospital, Cincinnati, OH, [2]University of Cincinnati, Cincinnati, OH

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 2 - JIA & Uveitis: Genetics, Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Children diagnosed with oligoarticular juvenile idiopathic arthritis (JIA), the most commonly diagnosed subset of chronic arthritis, are at increased risk of developing uveitis. Known clinical risk factors such as JIA category, ANA positivity, ≤4 years JIA duration, and 6 years old at diagnosis are associated with increased risk but are not definitive. Regular ophthalmic screening is important since uveitis is asymptomatic. Tear fluid biomarkers are easily obtained specimens with great diagnostic potential. In this pilot study, we aim to identify potential tear-based inflammatory markers associated with uveitis development in children with JIA.

Methods: In this comparative cohort at Cincinnati Childrens with JIA-associated uveitis (JIA-U) and JIA without uveitis (JIA-no-U), participants were enrolled on the basis of high-risk factors: female, non-Hispanic, white and oligoarticular subtype. Medical charts were reviewed for demographics, uveitis characteristics, ophthalmic examination, and current treatment (Table 1). Tear fluid was collected in the left eye using Schirmer strips. We used advanced proteomic strategies (isobaric

tag for relative quantitation [iTRAQ] labeling and nanoLC-MS/MS) to quantify and compare proteins in JIA-U and JIA-no-U. A P-value 0.1, based on Wilcoxon rank sum exact test, was considered significant.

Results: Fifteen proteins exhibited differences in tears of 5 children with JIA-U and 5 with JIA-no-U (Table 2). Of these, 9 showed a difference at $p \leq 0.05$. These proteins have been associated with inflammatory arthritis and general inflammatory response pathways. However, Keratin type II cytoskeletal 2 epidermal (KRT2), cathepsin D (CTSD), Ras-related C3 botulinum toxin substrate 1 (RAC1), C-1-tetrahydrofolate synthase, cytoplasmic (MTHFD1), Golgi apparatus protein 1 (GLG1), Heat shock 70 (HSPA1B), Aldo-keto reductase family 1 member C1(AKR1C1), Immunoglobulin heavy variable 3-49 (IGHV3-49), Adseverin (SCIN) and Immunoglobulin lambda variable 3-10 (IGLV3-10) demonstrated notably higher expression in tears of those with JIA-U (Table 2). These 10 proteins are associated with the immune response, specifically at the level of retinal pigment epithelium (RPE) and blood retinal barrier (BRB).

Conclusion: Uveitis is a vision-threatening disease that warrants exploration of avenues for early detection and treatment. The eye is an immune-privileged organ, immunologically shielded by the RPE as part of the BRB.Wehypothesize that the immune response proteins identified within the tears of JIA-U patients are byproducts of inflammation-induced RPE and BRB breakdown and furthers the inflammatory cascade. Tears of children with JIA-U and JIA-no-U display differential expression of protein biomarkers that may be a useful clinical source of potential onset of uveitis for those diagnosed with oligoarticular JIA. Furthermore, differential expression of ocular versus joint inflammation may provide insight in the pathogenesis of both JIA and uveitis. Further study in larger cohorts is needed to verify these results.

Table 1. Demographics and clinical characteristics of children with JIA with and without uveitis

n (%) unless otherwise specified	All n =10 10 eyes	JIA-no-U n =5 5 eyes	JIA-U n =5 5 eyes
DEMOGRAPHICS			
Age at tear collection, years, median (IQR)	11 (10-14)	11 (10-13)	10 (9.5-15.5)
Female	10 (100)	5 (100)	5 (100)
White	10 (100)	5 (100)	5 (100)
Non-Hispanic	10 (100)	5 (100)	5 (100)
ANA positive	8 (80)	3 (60)	5 (100)
OLIGOARTICULAR JIA CHARACTERISTICS			
Age JIA diagnosis, years, median (IQR)	3.5 (2-5)	5 (2-7)	3 (1.5-4)
Active Arthritis at tear collection	3 (30)	1 (20)	2 (40)
Duration of JIA at time of evaluation, years, median (IQR)	6.5 (6-10)	6 (5.5-8.5)	8 (5.5-13.5)
UVEITIS CHARACTERISTICS			
Age Uveitis diagnosis, years, median (IQR)	3.5 (2.5-4)	N/A	3.5 (2.5-4)
Active Uveitis at tear collection[a]	0 (0)	N/A	0 (0)
Uveitis duration at time of evaluation, years, median (IQR)	7.5 (5.5-13)	N/A	7.5 (5.5-13)
Anterior location of Uveitis	5 (50)	N/A	5 (100)
Bilateral Disease	5 (50)	N/A	5 (100)
MEDICATIONS AT TIME OF TEAR COLLECTION			
Topical glucocorticoids	0 (0)	N/A	0 (0)
Methotrexate	4 (40)	2 (40)	2 (40)
Leflunomide	2 (20)	2 (40)	0 (0)
Infliximab	3 (30)	1 (20)	2 (40)
Tocilizumab	1 (10)	0 (0)	1 (20)
Adalimumab	1 (10)	0 (0)	1 (20)

[a]Values determined per SUN criteria, *JIA-U* JIA-associated uveitis, *JIA-no-U* JIA without uveitis

Table 2. Differentially expressed proteins in children with JIA-no-U and JIA-U, Mean (SD)

Protein	Protein function	JIA-no-U	JIA-U	p-value
Transitional endoplasmic reticulum ATPase	Protein degradation, intracellular membrane fusion, DNA repair and replication, regulation of cell cycle, activation of NF-kappa B pathway.	0.119 (0.06)	0.032 (0.02)	0.0159
Alpha-2-macroglobulin	Protease inhibitor, cytokine transporter. Inhibits proteases and inflammatory cytokines, disrupts inflammatory cascades.	0.075 (0.04)	-0.061 (0.09)	0.05
Keratin, type II cytoskeletal 2 epidermal	Keratinocyte activation, proliferation and keratinization. Maintenance of corneocytes and keratin filaments in suprabasal keratinocytes.	-0.18 (0.09)	-0.019 (0.1)	0.05
Cathepsin D	Enzyme exhibits pepsin-like activity, role in protein turnover and in proteolytic activation of hormones and growth factors.	-0.051 (0.11)	0.08 (0.07)	0.05
Proteasome subunit beta type-8	Subunit of proteasome, ATP-dependent proteolytic activity Antigen processing to generate class I binding peptides. Component of IFN gamma-induced sensitivity. May be involved in inflammatory response pathway.	0.142 (0.07)	0.033 (0.04)	0.05
Ras-related C3 botulinum toxin substrate 1	GTPase, belongs to RAS superfamily of small GTP-binding proteins. Regulates cellular events, control of cell growth, cytoskeletal reorganization, and activation of protein kinases. Regulates cellular responses	-0.075 (0.06)	0.008 (0.04)	0.05
Perilipin-3	Interacts with cytoplasmic domains of cation-indep.endent and cation-dependent MPRs. Required for endosome-to-Golgi transport. Protein binds to GTPase RAB9 (RAB9A), a member of RAS oncogene family.	0.03 (0.02)	-0.08 (0.08)	0.05
Fibrinogen alpha chain	Alpha subunit of the coagulation factor, fibrinogen.	0.096 (0.04)	-0.092 (0.11)	0.05
C-1-tetrahydrofolate synthase, cytoplasmic	Possesses 3 distinct enzymatic activities, dehydrogenase, cyclohydrolase and formyltetrahydrofolate synthetase. Each catalyzes 1 of 3 sequential reactions in interconversion of 1-carbon derivatives of tetrahydrofolate, which are substrates for methionine, thymidylate, and *de novo* purine syntheses.	-0.075 (0.06)	0.005 (0.04)	0.05
Golgi apparatus protein 1	Binds fibroblast growth factor and E selectin *Cell-adhesion lectin on endothelial cells mediating neutrophil binding	-0.125 (0.08)	0.045 (0.08)	0.0635
Heat shock 70	Molecular chaperone in cellular processes. Protection of proteome from stress, folding and transport of newly synthesized polypeptides, activation of proteolysis of misfolded proteins, formation and dissociation of protein complexes.	-0.016 (0.05)	0.051 (0.07)	0.0952
Aldo-keto reductase family 1 member C1	Reductase *in vivo* since the oxidase activity measured *in vitro* is inhibited by physiological concentrations of NADPH.	-0.055 (0.05)	0.08 (0.10)	0.0952
Immunoglobulin heavy variable 3-49	V region of variable domain of immunoglobulin heavy chains that participates in antigen recognition.	-0.159 (0.06)	0.071 (0.28)	0.0952
Adseverin	Regulatory function in exocytosis by affecting organization of microfilament network underlying plasma membrane. Signaling mediated by MAPK, p38 and JNK pathways.	-0.050 (0.07)	0.04 (0.07)	0.0952
Immunoglobulin lambda variable 3-10	V region of variable domain of immunoglobulin light chains that participates in antigen recognition.	-0.223 (0.06)	0.104 (0.09)	0.1

Comparison of mean values is indicated by color. Red and blue reflect higher and lower expression, respectively.

Disclosure: T. Nguyen: None; **I. Maccora**: None; **M. Altaye**: None; **W. Haffey**: None; **T. Hennard**: None; **A. Sproles**: None; **S. Thornton**: None; **V. Miraldi Utz**: None; **K. Greis**: None; **S. Angeles-Han**: None.

Abstract Number: 018

Using the Electronic Health Record to Identify Subjects with Rheumatic Disease

Alysha Taxter[1], Matthew Basiaga[2], Rajdeep Pooni[3], Caitlan Pinotti[4], Lisa Buckley[5] and CARRA Registry Investigators[6], [1]Nationwide Children's Hospital, Columbus, OH, [2]Mayo Clinic, Rochester, MN, [3]Stanford University, Palo Alto, CA, [4]Duke, Durham, NC, [5]Vanderbilt University Medical Center, Nashville, TN, [6]CARRA, Washington, DC

SESSION INFORMATION

Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 2 - JIA & Uveitis: Genetics, Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Research teams spend hours manually searching the electronic health records (EHRs) to identify potential candidates eligible for recruitment to the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry. There are multiple existing tools, including self-service reporting tools and more detailed data extraction methods, which may increase the efficiency of eligible subject identification. This study aims to build a predictive electronic algorithm to identify children with rheumatic disease using electronic health record data.

Methods: Five diverse clinical sites (Duke, Mayo, Nationwide Childrens, Stanford, Vanderbilt) evaluated currently available EHR reporting functionality and developed self-service queries. All sites used the same electronic health record vendor (Epic Systems Corporation). Available shared reporting tools were identified. Queries were created and evaluated for their sensitivity and positive predictive value of identifying children with juvenile arthritis (ICD10-CM M08.*, M35.7), lupus and associated conditions (M32.*, L93.*, H01.12, M35.*), and dermatomyositis (M33.*). The observation period was a convenience sample limited to two weeks duration; clinic schedules were reviewed to validate patients with these diagnosis, and were compared to the output of the queries. Descriptive statistics were used.

Results: Three automated extraction tools were identified. Clarity reports, which can pull nearly all EHR data with associated complex data linkages from Epics back-end database, requires custom programming and were not available without significant funding at most centers. Two other tools, Slicer-Dicer and Reporting Workbench, were available without cost, on-demand, and were readily available to be run by end-users. Slicer-Dicer was the easiest to configure and was further evaluated (Tables). The highest performing queries to identify patients with rheumatic disease included limiting queries by diagnosis, department, and encounter type. Although additional discrete data elements can be configured to report in Slicer-Dicer, these require access to specialized programming environments.

Table 1. Sensitivity and Positive Predictive Value of Identifying Juvenile Idiopathic Arthritis Through Electronic Health Record Reporting Tools

	Total Subjects Identified by Slicer-Dicer Query	Total Subjects with JIA seen in Clinic During Past 2 Weeks	True Positives	False Negatives	Sensitivity	Positive Predictive Value
Diagnosis	18712	198	184	14	0.93	<0.01
Diagnosis, age <18	4063	198	174	24	0.88	0.04
Diagnosis, age <18, department	456	198	177	21	0.89	0.39
Diagnosis, age <18, department	429	198	161	37	0.81	1.0
Diagnosis, department, encounter type	161	198	134	22	0.85	0.83
Diagnosis, department, encounter type, visit type, age <18	125	198	125	73	0.63	1.0
Diagnosis and Care Team	206	198	108	90	0.55	0.52

Legend: JIA= juvenile idiopathic arthritis

Conclusion: Although highly detailed reports can include additional variables and output, end-user usability was a key factor to configure and utilize on-demand and readily available reporting tools. Queries with high sensitivity and positive predictive value across all conditions included diagnosis, department, and encounter criteria. Enabling availability of additional discrete data elements could likely improve identification of children with rheumatic disease. Integration of reporting tools into research screening and recruitment workflows are needed.

Table 2. Sensitivity and Positive Predictive Value of Identifying Lupus and Associated Conditions Through Electronic Health Record Reporting Tools

	Total Subjects Identified by Slicer-Dicer Query	Total Subjects with SLE seen in Clinic During Past 2 Weeks	True Positives	False Negatives	Sensitivity	Positive Predictive Value
Diagnosis	57315	64	64	0	1.0	<0.01
Diagnosis, age <18	3748	64	60	4	0.94	0.02
Diagnosis, age <18, department	249	64	59	5	0.92	0.23
Diagnosis, age <18, department	233	64	55	9	0.86	0.23
Diagnosis, department, encounter type	97	64	58	6	0.91	0.60
Diagnosis, department, encounter type, visit type, age <18	82	64	37	27	0.58	0.45
Diagnosis and Care Team	131	64	31	33	0.48	0.23

Legend: SLE = systemic lupus erythematosus

Sensitivity and Positive Predictive Value of Identifying Lupus and Associated Conditions Through Electronic Health Record Reporting Tools

Table 3. Sensitivity and Positive Predictive Value of Identifying Juvenile Dermatomyositis Through Electronic Health Record Reporting Tools

	Total Subjects Identified by Slicer-Dicer Query	Total Subjects with JDM seen in Clinic During Past 2 Weeks	True Positives	False Negatives	Sensitivity	Positive Predictive Value
Diagnosis	4223	25	25	0	1.0	<0.01
Diagnosis, age <18	376	25	22	3	0.88	0.06
Diagnosis, age <18, department	48	25	20	1	0.95	0.42
Diagnosis, age <18, department	45	25	21	4	0.84	0.47
Diagnosis, department, encounter type	19	25	19	6	0.76	1.0
Diagnosis, department, encounter type, visit type, age <18	17	25	17	8	0.68	1.0
Diagnosis and Care Team	16	25	6	17	0.32	0.50

Legend: JDM = juvenile dermatomyositis

Sensitivity and Positive Predictive Value of Identifying Juvenile Dermatomyositis Through Electronic Health Record Reporting Tools

Disclosure: A. Taxter: None; **M. Basiaga**: None; **R. Pooni**: None; **C. Pinotti**: None; **L. Buckley**: None; **C. Investigators**: None.

Abstract Number: 019

Developing Standard Improvement Curves for Disease Activity, Pain and Quality of Life in Children with Newly Diagnosed Juvenile Idiopathic Arthritis: Results from the CAPRI Registry

Amieleena Chhabra[1], Matt Berkiwitz[2], Thomas Loughin[2], Lori Tucker[3], Giles Boire[4], Karine Toupin-April[5], Dax Rumsey[6], Michelle Batthish[7], Linda Li[8], Adam Huber[9], Brian Feldman[10], Jean-Phillippe Proulx-Gauthier[11], Ciaran Duffy[12], Paul Dancey[13], Gordon Soon[14], Heinrike Schmeling[6] and Jaime Guzman[15], [1]University of British Columbia, Vancouver, BC, Canada, [2]Simon Fraser University, Burnaby, BC, Canada, [3]BC Children's Hospital, Vancouver, BC, Canada, [4]Sherbrooke University, Sherbrooke, QC, Canada, [5]University of ottawa, Ottawa, ON, [6]University of Alberta, Edmonton, AB, Canada, [7]McMaster Children's Hospital, Hamilton, ON, Canada, [8]Arthritis research, Vancouver, BC, Canada, [9]University of Nova Scotia, Halifax, NS, Canada, [10]Hospital for Sick Children / University of Toronto, Toronto, ON, Canada, [11]Université Laval, Québec, QC, Canada, [12]Children's Hospital of Eastern Ontario (CHEO), Ottawa, ON, Canada, [13]Université de Montréal, Montreal, QC, Canada, [14]University of Toronto, Toronto, ON, [15]University of British Columbia, Vancouver, BC

SESSION INFORMATION

Session Date: Thursday, March 30, 2023

Session Title: Poster Breakout 2 - JIA & Uveitis: Genetics, Clinical & Therapeutic Aspects

Session Type: Abstract Session

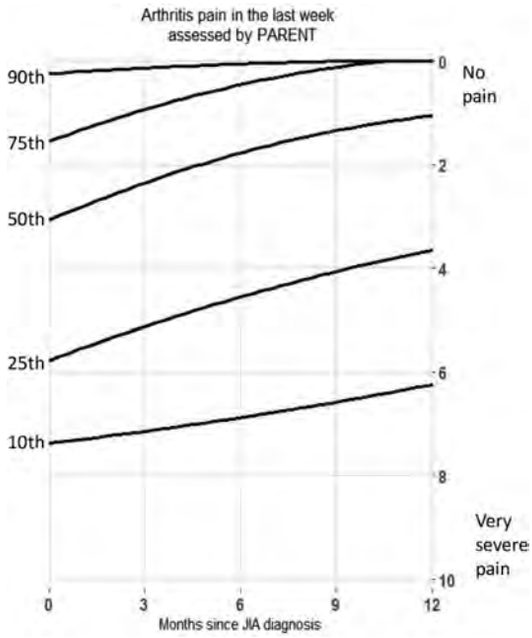
Session Time: 5:10PM–5:40PM

Background/Purpose: When a diagnosis of juvenile idiopathic arthritis (JIA) is made, many parents are shocked to find out that arthritis occurs in children and may have concerns that arthritis is an incurable disease leading to inexorable deformities and disability. We know from research in the last 20 years that this is not the case for most, as the prognosis of JIA has improved substantially. But how do we convey this to families in an understandable and helpful way? We propose to use the analogy of growth charts and developed standard improvement curves of expected treatment-related improvements during the first year on pain, quality of life, and disease activity in children with JIA.

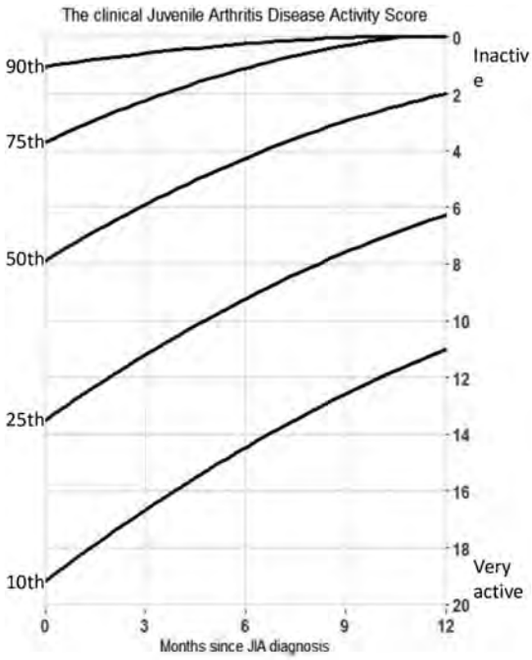
Table 1: Baseline characteristics of included patients

Characteristic	Value
Number of patients	721
Age at diagnosis, years (25th, 75th centiles)	9.4 (4.1, 13.2)
Female	61%
Weeks from diagnosis to enrolment	2 (0, 8)
Disease duration, weeks (25th, 75th centiles)	25 (13, 52)
JIA category, n (%)	
Oligoarthritis	321 (44.5)
Polyarthritis RF-negative	131 (18.2)
Enthesitis related arthritis	116 (16.1)
Psoriatic	46 (6.4)
Systemic	31 (4.3)
Polyarthritis RF-positive	26 (3.6)
Undifferentiated	50 (6.9)
Active joint count	2 (1, 4)
Physician global assessment of disease activity	3 (1.5, 5)
Parent global assessment of wellbeing	2 (0.5, 5)
cJADAS10	8 (4.5, 13)
QoML score at baseline (n=450) *	7.5 (5, 9)
Medications used in the first year, cumulative incidence, % and (95% CI)	
NSAID	83 (79, 86)
Joint Injections	33 (29, 37)
Prednisone	14 (11, 17)
Conventional DMARD	54 (50, 59)
Biological DMARD	20 (17, 24)
Clinical Inactive Disease in the first year	73 (69, 76)

Methods: Data from the Canadian Alliance of Pediatric Rheumatology Investigators JIA Registry was used. Patients newly diagnosed with JIA were recruited from February 2017 to December 2021 (N=721) and followed as clinically indicated. At each clinic visit, self-reported pain intensity, Quality of My Life (QoML), and physician and parent global assessments were recorded in numerical rating scales from 0 to 10. The clinical Juvenile Arthritis Disease Activity Score 10 (cJADAS10) was the sum of physician and parent global, and the number of active joints up to 10.



Standard improvement curves for the 10th, 25th, 75th and 90th centiles in pain intensity during the first year after JIA diagnosis



Standard improvement curves for the 10th, 25th, 50th, 75th and 90th centiles in CJADAS10 scores during the first year after JIA diagnosis.

Results: After assessing several parametric and non-parametric methods, Quantile Random Forests and LOWESS smoothing were used to chart the course of the 10th, 25th, 50th, 75th and 90th centiles for each measure during the first year after diagnosis and treatment initiation. A total of 721 children were recruited a median of 2 weeks after diagnosis, 61% were female and 44.5% had oligoarthritis (Table 1). During the first two years after diagnosis, there were a total of 3,671 visits that were used for plotting the charts. Using two years of data to produce curves for the first year improved their accuracy. During the first year, the 50th centiles changed from 3.5 to 1 for pain, from 3.5 to 1 for QoML, from 3 to 1 for the active joint count, and from 8 to 2 for the cJADAS10. The Figures provide examples of the resulting Charts.

Conclusion: Using real clinic data from Canadian children diagnosed with JIA in 2017-2021, we have produced standard curves of expected improvements during the first year after diagnosis that can be used to track individual childs progress and display for families how their child is doing in comparison to other children with JIA. These JIA Improvement Charts may assist families and physicians in making treatment decisions for the child. They may also help detect when early treatment adjustments are needed if improvement is faltering, in order to prevent long term damage.

Disclosure: A. Chhabra: None; **M. Berkiwitz**: None; **T. Loughin**: None; **L. Tucker**: None; **G. Boire**: None; **K. Toupin-April**: None; **D. Rumsey**: None; **M. Batthish**: AbbVie/Abbott, 5, Novartis, 6, Viatris, 12, AdBoard; **L. Li**: None; **A. Huber**: None; **B. Feldman**: None; **J. Proulx-Gauthier**: None; **C. Duffy**: None; **P. Dancey**: None; **G. Soon**: None; **H. Schmeling**: None; **J. Guzman**: None.

Abstract Number: 020

Generation of Human Resident Memory T Cells in 3D Synovial Organoid Model

Margaret Chang[1], Maryrose Hahn[1], Brian Wauford[1], Rachel Blaustein[2], Kevin Wei[2] and Peter Nigrovic[1], [1]Boston Children's Hospital, Boston, MA, [2]Brigham and Women's Hospital, Boston, MA

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 2 - JIA & Uveitis: Genetics, Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Most rheumatoid arthritis (RA) and juvenile idiopathic arthritis (JIA) patients experience episodic arthritis flares and require life-long medications to control their disease. We observed that arthritis tends to flare repeatedly in the same joints, even after decades of remission, displaying joint-specific memory. We have identified tissue resident memory T cells (TRM) in arthritic joints and demonstrated in murine models that these long-lived T_{RM} remain in synovium during remission and act as key mediators of arthritis flares. However, strategies to model synovial TRM development and function in a human context are lacking.

Methods: To recapitulate the synovial stromal structure in a 3D organoid, we encapsulated fibroblast-like synoviocytes isolated from synovial biopsies from human RA donors with human umbilical vein endothelial cells in Matrigel. These cells were seeded on a polyHEMA-coated low-attachment plate and cultured in a 1:1 mixture of complete RPMI media and human endothelial cell growth media. CD8 memory T cells from healthy donors were isolated from peripheral blood and co-cultured with the synovial organoid for 2-3 weeks. Where indicated, TNF was added to the culture media for the first 3-5 days to simulate inflammation. Cells were dissociated from the organoids and assayed for TRM surface phenotype by flow cytometry. T cells were evaluated for their ability to take up free fatty acids compared to other T cell populations, reflective of a shift toward fatty acid metabolism. To assess the propensity of TRM to remain in tissues,

the capacity of organoid-derived T cells to migrate across a transwell membrane in response to tissue egress signal CCL21 was also tested.

Results: Culturing CD8 memory T cells isolated from human blood in 3D synovial organoids, particularly with TNF, supported development of CD8 cells with T_{RM} surface phenotype (CD45RO+CD62L-CCR7-HLADR-CD25-CD103+CD49a+). These TRM-like cells have enhanced free fatty acid uptake compared to other T cell subtypes, consistent with a shift in metabolic activity seen in TRM. Unlike central memory T cells extracted from synovial organoids and naïve T cells isolated from blood, TRM differentiated in the synovial organoids did not migrate across the transwell membrane to CCL21, confirming this key sessile feature of TRM in tissues.

Conclusion: We developed a novel model for studying human TRM by differentiating TRM within 3D synovial organoids composed of stromal cells from human RA synovium. This model system will be valuable in gaining insights to synovial TRM biology including factors that may drive human synovial TRM development and function.

Disclosure: M. Chang: None; **M. Hahn**: None; **B. Wauford**: None; **R. Blaustein**: None; **K. Wei**: None; **P. Nigrovic**: None.

Abstract Number: 021

Distinguishing Childhood Sjogren's Disease in Patients Presenting with Recurrent Sialadenitis in a Multidisciplinary Clinic

Anna Holley[1] and **Sara Stern**[2], [1]University of Utah School of Medicine, Salt Lake City, UT, [2]University of Utah Department of Pediatrics, Salt Lake City, UT

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 3 - Pediatric Rheumatology Potpourri
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Sialadenitis is a common distinguishing manifestation of Childhood Sjogrens Disease (cSD). We have previously reported a surprisingly high rate of Childhood Sjogrens Disease (cSD) patients who presented with recurrent parotitis. For this reason, we created a pediatric multidisciplinary otolaryngology/rheumatology clinic (PMORC) which was founded in December 2019 at the University of Utah. Currently little is known about cSD and there is no widely accepted classification criteria. The aim of the study was to analyze clinical characteristics of patients with cSD and evaluate how many patients currently met 3 proposed but not widely accepted or validated classification criteria for children in this clinic.

Methods: Retrospective analysis of patients seen in this PMORC between December 2019 and July 2022 presenting with chronic sialadenitis was performed. Patients were included in the cSD group if diagnosed with cSD by a pediatric rheumatologist. Systematic evaluation of demographic information, clinical symptoms, and lab/biopsy/imaging results were collected. Patients were evaluated to see if they met the 2016 ACR-EULAR criteria, 1999 Japanese criteria, and proposed Bartunkova criteria for cSD. The framework of this PMORC included a new patient evaluation, a minor salivary gland biopsy and therapeutic sialendoscopy with triamcinolone acetonide injection, and follow up.

Results: 27 patients presented to the PMORC and 14 (51.9%) were diagnosed with cSD. 7 (50%) patients were female with a median age of symptom onset of 6.5 (1-15) years. 14 (100%) patients had parotid swelling, 4 (28.6%) had joint pain, 9 (64.3%) had dry eyes symptoms, 10 (71.4%) had dry mouth symptoms, 2 (14.3%) had fevers. 13 (92.9%) of the patients showed histologic proof of focal lymphocytic infiltration. 5 (35.7%) patients were positive for anti-SSA, anti-SSB and/or ANA autoantibodies.

2016 ACR-EULAR	1999 Japanese	1999 Bartunkova
Weighted sum of 5 items with a total score of ≥4:	Requires at least 2 of 4 items:	Requires at least 4 of 12 items: Exclusion of all other autoimmune diseases
1. anti-SSA/Ro antibody positivity (scoring 3) 2. Focal lymphocytic sialadenitis with a focus score of ≥1 foci/4 mm² (scoring 3) 3. Abnormal ocular staining score ≥5 or van Bijsterveld score of ≥4 (scoring 1) 4. Schirmer's test result of ≤ 5 mm/5 minutes (scoring 1) 5. Unstimulated salivary flow rate of ≤ 0.1 ml/minute (scoring 1)	1. Schirmer test ≤ 5 mm/5 min AND van Bijsterveld score >3 or positive fluorescein staining test 2. Abnormal sialography ≥Stage I OR Decreased Salivary secretion (≤ 10 ml/10min chewing gum test or (≤ 2 g/2min Saxon test) and decreased salivary function on scintigraphy 3. Anti-Ro/SSA and/or anti-La/SSB autoantibodies 4. Focal lymphocytic sialadenitis with focus score ≥1 lymphocytic focus per 4 mm² glandular tissue OR focal lymphocytic dacryoadenitis with focus score ≥1 lymphocytic focus per 3 mm² glandular tissue	1. Recurrent parotitis or enlargement of parotid gland 2. Recurrent conjunctivitis or keratoconjunctivitis sicca 3. Recurrent vaginitis 4. Systemic: fever of unknown origin, arthralgias, hypokalemic paralysis, abdominal pain 5. Positive anti-SSA, anti-SSB, high titer of ANA (speckled type), or RF 6. Elevated serum amylase 7. Leukopenia, high ESR 8.Hyperimmunoglobulinemia 9. Renal tubular acidosis 10. Histological proof of lymphocytic infiltration of salivary glands or other organs 11. Objective documentation of ocular dryness (Bengal red staining or Schirmer test) 12. Objective documentation of parotid gland affection (sialography)

Criteria for Classification of SD

Criteria met	Clinically dx with SD and met criteria (n=7)	Clinically dx with SD and did not meet criteria (n=7)	P-Value
Median number of criteria met	4 (4-7)	3 (2-3)	-
Parotitis and/or dry mouth	7	7	-
Focal lymphocytic infiltration	6	7	0.30
Systemic	**5**	**1**	**0.031**
Clinical Ocular	6	3	0.094
Objective ocular	2	0	0.13
Positive ANA, SSA and/or SSB	3	2	0.58
Amylase	1	0	0.30
ESR	1	0	0.30
Vaginal Dryness	1	0	0.30

Bartunkova Criteria - cSD Cohort

Criteria met	Met Bartunkova Criteria (n=8)	Did not meet Bartunkova Criteria (n=19)	P-Value
Parotitis and/or dry mouth	8	18	0.51
Focal lymphocytic infiltration	6	10	0.28
Systemic	6	4	0.17
Clinical Ocular	**7**	**4**	**0.0013**
Objective ocular	**3**	**0**	**0.0046**
Positive ANA, SSA and/or SSB	3	2	0.099
Amylase	1	2	0.88
ESR	1	0	0.12
Vaginal Dryness	1	0	0.12

Bartunkova Criteria - Entire Cohort

13 cSD patients and 3 non-cSD patients had focal lymphocytic infiltrate on biopsy (p=0.00023), 13 cSD and 8 non-cSD patients had abnormal ultrasounds (p=0.050), 9 cSD and 2 non-cSD patients had symptoms of dry eyes (p=0.0098), and 6 cSD and 0 non-cSD patients received a focus score of greater than one on biopsy (p=0.0074).

2 patients (14.32%) met the 2016 ACR-EULAR criteria and 2 (14.32%) met the Japanese criteria based on a positive biopsy score and positive serology.7 (50%) cSD patients met the pediatric Bartunkova criteria. Additionally, 1 patient in the non-cSD cohort met the Bartunkova criteria.

Conclusion: Given the variable features of cSD compared to adult SD, there is a need for a validated pediatric criterion. The 2016 ACR-EULAR criteria and 1999 Japanese criteria were not sufficient in identifying cSD in our clinic. Additionally, Bartunkovas proposed cSD criteria only diagnosed 50% of our cohort that received a clinical diagnosis of cSD in our clinic. Therefore, providers should have a high index of suspicion of cSD for children presenting with recurrent sialadenitis.

Disclosure: A. Holley: None; **S. Stern**: None.

Abstract Number: 022

Narrative Medicine and Pediatric Rheumatology: Addressing Burnout and Bias

aviya lanis, Natalie Rosenwasser and Esi Morgan, Seattle Children's Hospital, Seattle, WA

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 3 - Pediatric Rheumatology Potpourri
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Burnout, a syndrome of emotional exhaustion and depersonalization, adversely impacts healthcare. This can result in mood disturbances, poor patient and provider satisfaction and high turnover. Studies have shown burnout also plays a critical role in physician bias, with increased bias among those with higher rates of burnout. The SARS-CoV-2

pandemic increased levels of burnout and exposed the high prevalence of racial bias in healthcare. There is a critical need for methods to address burnout and bias in healthcare. Narrative medicine (NM) incorporates the stories of human experience into the medical realm and has been proven to strengthen relationships and increase empathy. To date, limited data investigating narrative medicines impact on burnout and bias has been promising. This pilot study aims to investigate the use of NM as a tool to address burnout and explore bias amongst a pediatric rheumatology division.

Methods: Physicians, nurses, medical assistants and staff at a single center enrolled in a series of six monthly 1-hour video-based sessions lead by co-facilitators (AL and a NM-trained facilitator) after obtaining informed consent. Sessions utilized poetry, photography, paintings, and spoken word to inspire writing prompts around the medical experience. A Plan-Do-Study-Act approach was used to guide intervention adjustments based on participant post-session feedback.Demographics

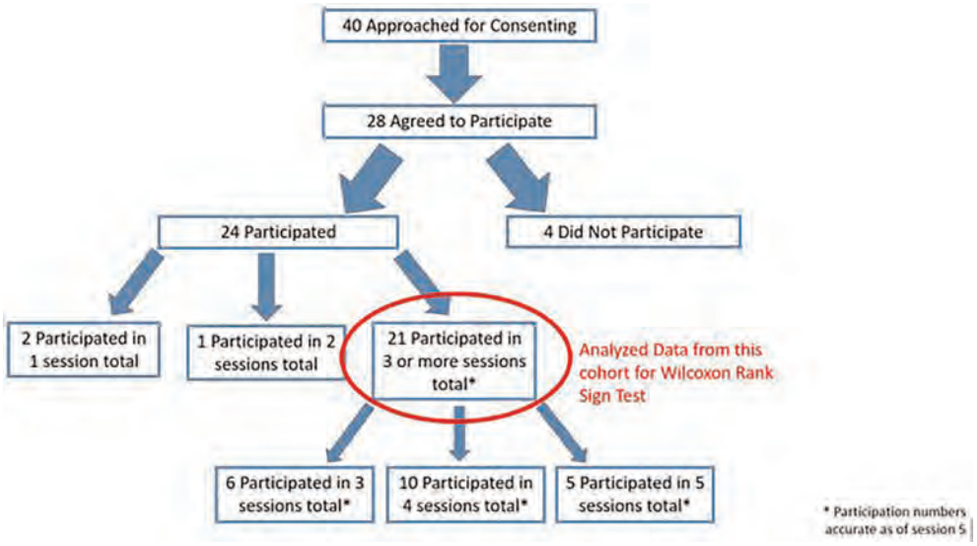


Figure 1: Breakdown of individuals consented compared to those who participated.

Table 1: Pre- and intra-participation questionnaire results including observations, mean, standard deviation (SD), median, range, and Wilcoxon signed-rank test p-values.

	Mini Z Burnout						Copenhagen Burnout Index		PHQ-9	
	Total Pre	Total Intra	Satisfaction Pre	Satisfaction Intra	Stress Pre	Stress Intra	Pre	Intra	Pre	Intra
Observations	21	20	21	20	21	20	21	19	21	18
Mean	29.2	29.5	12.5	12.9	13.7	13.5	42.8	38.4	6.0	4.6
SD	6.4	6.2	3.3	2.9	2.6	3.0	13.2	16.7	5.0	3.8
Median	30	30	14	13.5	14	14	39.5	34.2	5	3.5
Range	15-38	18-38	5-17	7-17	7-18	8-19	25-75	17.1-84.2	0-17	0-13
Wilcoxon signed rank test p-value	0.7754		0.43342		0.48933		0.25825		0.72723	

were collected, and standard surveys were administered at baseline, and after the third and sixth sessions, specifically: Mini Z Burnout, Copenhagen Burnout Inventory (CBI), Provider Health Questionnaire-9 (PHQ-9) as well as Implicit Association Testing (IAT) and explicit bias Feeling Thermometer (FT).Survey data were assessed for differences with a Wilcoxon signed rank test. Interim analysis of data from baseline and 3 months was analyzed; 6-month data will be analyzed once available.

Results: Twenty-four participants with 21 females, 2 males, and one person identifying as non-binary were divided amongst 6 narrative medicine groups for 6 sessions in total. Data analysis was completed for those who attended 3 or more sessions (n=21) (Figure 1). Self-identified race included 18 White, 4 Asian, 1 Black, and 1 who preferred not to answer. Participants included 8 attendings, 4 fellows, 2 nurses, 2 medical assistants, and 8 staff members. While no statistical significance was seen evaluating data from 0 to 3 months in the Mini Z Burnout, CBI, or PHQ-9, there was a trend toward improvement (median CBI improved from 39.47 to 34.21, and median PHQ-9 score improved from 5 to 3.5) (Table 1). IAT showed a statistically significant shift towards positive association with African Americans following participation (p-value= 0.012), while FT showed a non-statistically significant explicit bias shift away from positive association with European Americans (p=0.096).

Conclusion: Narrative medicine is a feasible intervention that warrants further study as a means to address bias. Additional long-term analysis is needed to understand if these changes are durable.

Disclosure: a. Ianis: None; **N. Rosenwasser**: None; **E. Morgan**: None.

Abstract Number: 023

Effect of Type 1 Interferons and JAK Inhibitors on Gene Expression in Bioengineered Pediatric Skeletal Muscle

Lauren Covert[1], Joseph Prinz[2], Hailee Patel[3], Jeffrey Dvergsten[4] and George Truskey[3], [1]Duke University, Durham, NC, [2]Duke University School of Medicine, Department of Biostatistics and Bioinformatics, Durham, NC, [3]Duke University, Department of Biomedical Engineering, Durham, NC, [4]Duke University Hospital, Durham, NC

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 3 - Pediatric Rheumatology Potpourri
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Genetic studies of new-onset juvenile dermatomyositis (JDM) exhibit elevation of Type 1 interferons (IFN 1) IFNα and IFNβ in blood, skin, and muscle. To understand the relationship of IFN 1 to muscle dysfunction we used an *in vitro* 3D biomimetic construct derived from healthy pediatric muscle (myobundles). IFN 1 reduces myobundle contractile force, upregulates MHC class 1 and induces donor-specific expression of myositis-specific antigens. Janus kinase inhibitors (JAKi) have therapeutic effect in Type 1 interferonopathies. In this study we evaluated gene expression of myobundles exposed to IFNα or β as well as after cotreatment with IFNβ and JAKi, tofacitinib or baricitinib.

Methods: Myoblasts isolated from 3 healthy pediatric donors were cultured and used to create donor-specific myobundles, exposed to 0 (control), 5, 10 or 20 ng/mL IFNα or β. In a separate protocol myobundles were exposed to 0 (control) or 20 ng/mL IFNβ for a total of 10 days then treated with tofacitinib 1 μM or baricitinib 500 nM from days 3-10 of IFNβ exposure. Myobundle mRNA was isolated for sequencing. Quality profiling and alignment of mRNA was performed using the fastp toolkit and splice aware STAR RNA-seq alignment tool, respectively. Differential expression was performed using the DESeq2 Bioconductor package. Independent filtering was used prior to calculating adjusted p-values, and moderated

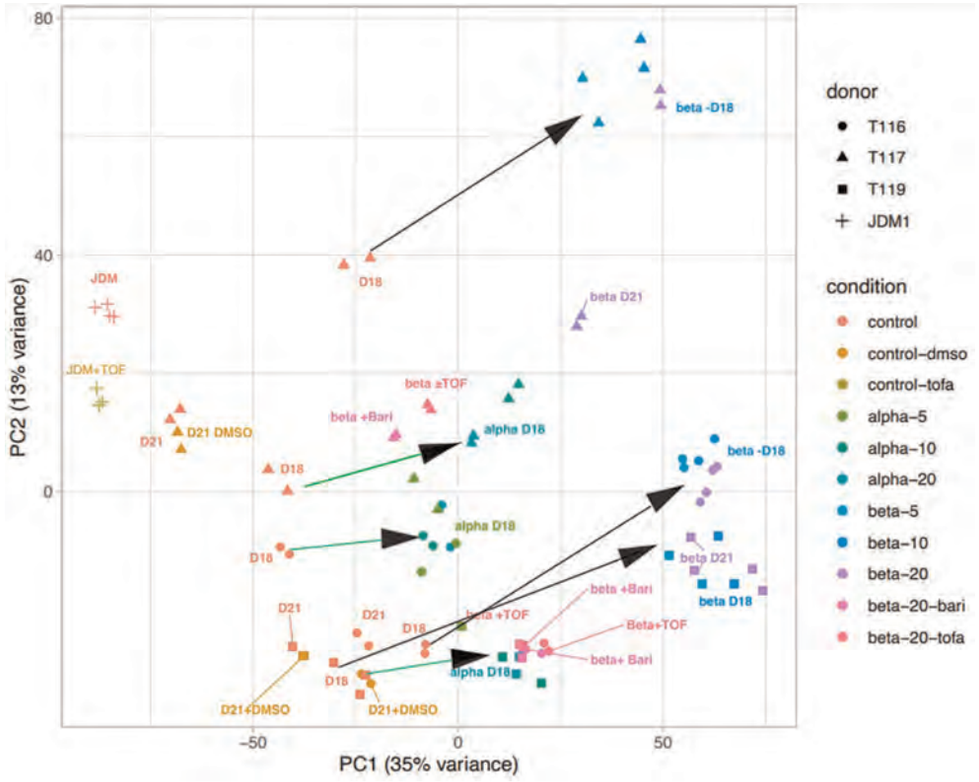


Figure 1. PCA plot of all myobundle samples. Green arrows show shift after IFNα; black arrows show greater shift in PC1 after IFNβ.

Table 1. Most overexpressed and downregulated genes after IFN 1 exposure in pediatric skeletal myobundles. lncRNA = long non-coding RNA; IRG = interferon response gene

5 most overexpressed genes in untreated vs. IFNα-exposed myobundles					
Name and rank by expression	Symbol	Biotype	Function	LogFC	Adjusted P
1. HLA complex P5	HCP5	lncRNA	Adaptive and innate immune response	4.540682616	6.70725E-30
2. Interferon alpha inducible protein 6	IFI6	protein coding	IRG; regulator of apoptosis	2.92726042	6.4896E-23
3. Interferon alpha inducible protein 27	IFI27	protein coding	IRG; cellular metabolism and apoptosis	2.703931143	1.45898E-10
4. Bone marrow stromal antigen 2	BST2	protein coding	May play a role in pre-B-cell growth	2.513927347	4.88315E-10
5. ISG15 ubiquitin like modifier	ISG15	protein coding	IRG; antiviral response, neutrophil recruitment	2.262263275	1.20655E-09
5 most downregulated genes in untreated vs. IFNα-exposed myobundles					
Name and rank by expression	Symbol	Biotype	Function	LogFC	Adjusted P
1. Long intergenic non-protein coding RNA 1790	LINC01790	lncRNA	Remodelling chromatin, RNA stabilization	-4.807994499	1.79077E-27
2. Deleted in malignant brain tumors 1	DMBT1	protein coding	Immune response to tumor cells	-2.544302048	9.21128E-16
3. Repetin	RPTN	protein coding	Calcium and metal ion binding activity	-2.088118024	4.90224E-07
4. Complement C1q like 4	C1QL4	protein coding	Regulator of cell differentiation, survival	-2.046888717	2.56891E-17
5. Ankyrin repeat domain 2	ANKRD2	protein coding	Muscle stress response, slow muscle function	-1.928906524	9.58798E-22

5 most overexpressed genes in untreated vs. IFNβ-exposed myobundles					
Name and rank by expression	Symbol	Biotype	Function	LogFC	Adjusted P
1. HLA complex P5	HCP5	lncRNA	Adaptive and innate immune response	6.54673393	2.5806E-68
2. C-X-C motif chemokine ligand 11	CXCL11	protein coding	Activated T-cell chemotactic	5.626802992	4.11483E-36
3. Phospholipase A and acyltransferase 2	PLAAT2	protein coding	Tumor suppressor	4.974492227	5.06341E-21
4. C-X-C motif chemokine ligand 10	CXCL10	protein coding	Antimicrobial response, immune cell recruiter	4.957058183	1.84442E-17
5. CD74 molecule	CD74	protein coding	Regulator of antigen presentation	4.906537725	1.45553E-36
5 most downregulated genes in untreated vs. IFNβ-exposed myobundles					
Name and rank by expression	Symbol	Biotype	Function	LogFC	Adjusted P
1. Long intergenic non-protein coding RNA 1790	LINC01790	lncRNA	Remodelling chromatin, RNA stabilization	-6.74468563	1.33099E-41
2. ENSG00000285671	n/a	lncRNA	Remodelling chromatin, RNA stabilization	-4.363826584	6.57012E-31
3. Mitochondrially encoded cytochrome c oxidase III	MT-CO3	protein coding	Respiratory chain complex IV function	-3.6612686	1.44311E-55
4. Stimulator of chondrogenesis 1	SCRG1	protein coding	Differentiation of mesenchymal stem cells	-3.61030522	3.20986E-30
5. Apolipoprotein L5	APOL5	protein coding	Lipid movement and binding	-3.583403961	1.65475E-37

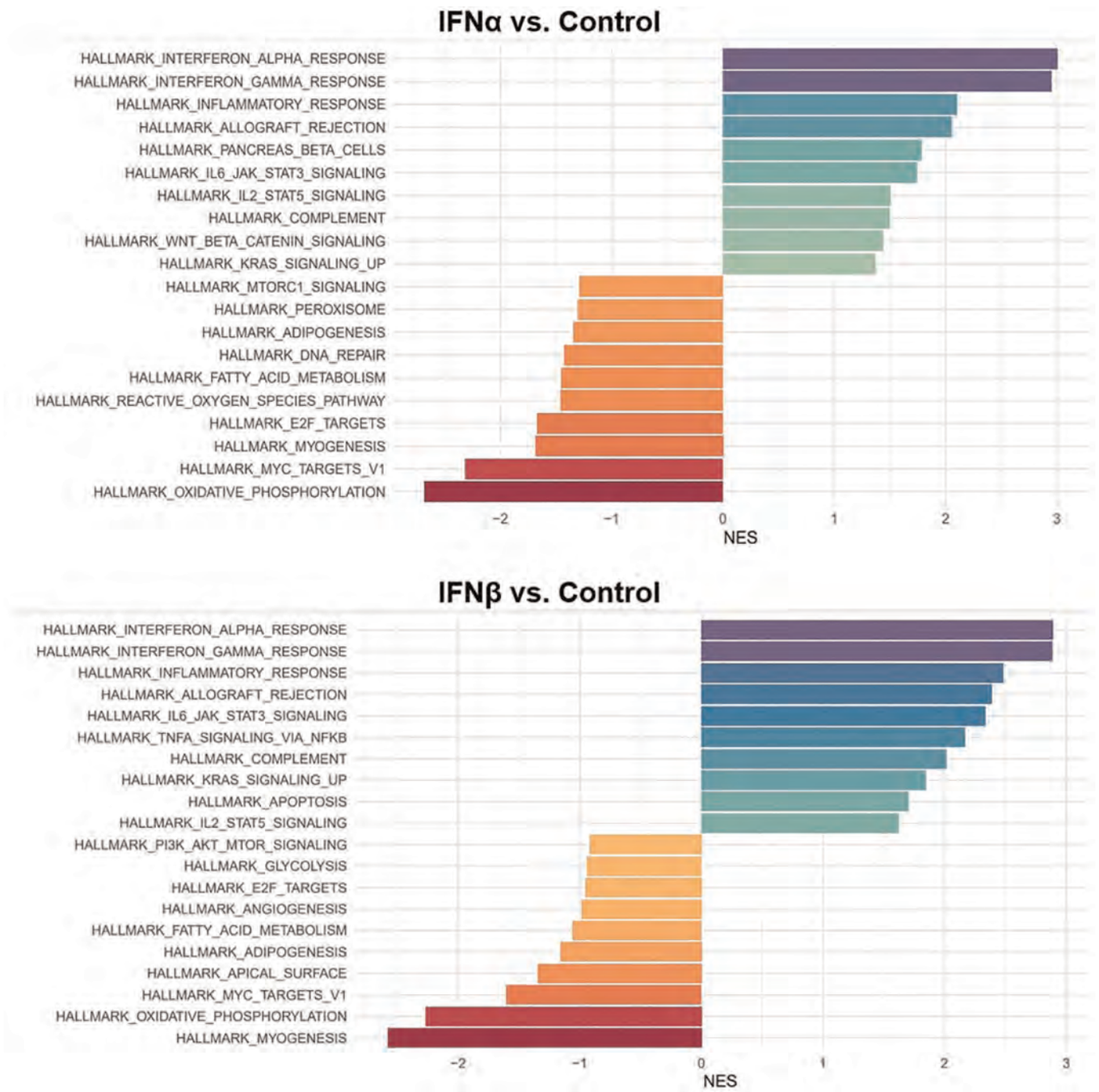


Figure 2. GSEA pathways after IFN I exposure

log2 fold-changes were derived using the ashr package. Gene set enrichment analysis was performed using hallmark pathways associated with altered gene expression for each comparison performed.

Results: Eighty-three myobundles from 3 healthy pediatric donors were analyzed. PCA analysis indicates distinct clusters of similar gene expression across IFNα-exposed and IFNβ-exposed myobundles vs. untreated controls, respectively (**Figure 1**). IFNβ-exposed myobundles treated with JAKi have gene profiles more similar to controls. The top 5 most overexpressed and downregulated genes after exposure to 20 ng/mL IFNα or IFNβ vs. control are listed in **Table 1** and included interferon response genes and proinflammatory genes. Gene Set Enrichment Analysis showed that IFNα induced enrichment of 29/50

pathways (58%) with IFNβ enrichment of 33/50 pathways (66%). Key enriched pathways included IFNα and γ response genes and inflammatory response genes. IFNα downregulated oxidative phosphorylation genes while IFNβ decreased expression of myogenesis genes (**Figure 2**). Treatment with IFNβ and JAKi had significantly less pathway enrichment (baricitinib with 13/50 and tofacitinib 16/50 enriched) with key enriched pathways involving skeletal muscle myogenesis.

Conclusion: IFNα and IFNβ have distinct effects on pediatric skeletal muscle gene expression, with upregulation of IFN and inflammatory response genes and downregulation of oxidative phosphorylation and myogenesis genes. Genetic profiles are significantly altered by JAKi, baricitinib and tofacitinib. Results demonstrate the potential of bioengineered pediatric skeletal muscle to advance our knowledge of JDM pathogenesis and provide a platform for investigating promising therapeutics.

Disclosure: L. Covert: None; **J. Prinz**: None; **H. Patel**: None; **J. Dvergsten**: None; **G. Truskey**: None.

Abstract Number: 024

Effects of Corticosteroids on Central Nervous System Microvascular Properties Assessed by Neuroimaging

Mark DiFrancesco, Ekemini Ogbu, Catherine Robben, Jennifer Huggins and **Hermine Brunner**, Cincinnati Children's Hospital Medical Center, Cincinnati, OH

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 3 - Pediatric Rheumatology Potpourri
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Prior research suggests that microvascular (MV) changes might be a biomarker for central nervous system (CNS) involvement of patients with systemic lupus erythematosus (SLE). Given that corticosteroids (CS) are widely used for the treatment of SLE and could also influence MV changes, CS effects need to be distinguished from MV changes secondary to CNS disease. We aimed to quantify and isolate the dose-dependent effects of oral CS on MV properties as assessed by diffusion-weighted imaging (DWI).

Methods: Using convenience sampling, 11 patients with chronic inflammatory conditions were enrolled and studied at two visits: (1) the time of high dose CS use (HD-CS; prednisone $\geq$20 mg/day) and (2) after tapering to low dose (LD-CS). At each study visit, cognitive ability was measured using the Pediatric Neuropsychological Assessment Metrics (PedANAM) and MRI was acquired at 3-Tesla, including T1 and T2 relaxometry, blood-brain-barrier permeability measurement by arterial spin labeling, and diffusion weighted imaging (DWI) at 19 weightings (b-values) ranging from 0 to 5000 s/mm^2. DWI signal was analyzed as a biexponential function of b-value under the intravoxel incoherent motion (IVIM) model. Besides bulk tissue diffusion (D), the IVIM model provides parameters describing microvasculature including pseudodiffusion (D*), dependent on flow through randomly-oriented microvessels, the volume fraction of blood (vbw), and their product, D* x vbw, proportional to blood perfusion. Voxel-wise change in each IVIM parameter was calculated between the HD-CS and LD-CS visits for each patient. Parameter changes were fitted across patients to a linear model including 2 terms 1) the mean parameter change and 2) the degree of CS dose reduction between HD-CS and LD-CS. Model results were expressed as effect size, represented by Cohens f^2, for the coefficients of the mean and dose reduction terms. with Cohens $f^2 > 0.35$ considered a large effect size.

Results: All patients completed the HD-CS visit and 9 the LD-CS visit. Table 1 summarizes patient characteristics. Regionally, there were mean changes in the 4 IVIM parameters between HD-CS and LD-CS with $f^2 > 0.35$. (Figure 1). Similarly, the dependence of parameter changes between HD-CS and LD-CS on the amount of dose reduction also had large effect size

regionally ($f^2 > 0.35$) (Figure 2). Vascular parameters were mostly greater at HD-CS, with increases in the blood perfusion-related product, D* x vbw, especially widespread. There was mostly positive dependence of vascular parameters with degree of dose reduction.

Conclusion: This pilot study provides evidence of at least moderate impact of oral CS on CNS MV as measured by DWI results in SLE. Further, CS impact on brain MV varies regionally across the brain. If confirmed in a larger cohort, the parameter estimates from this study can be used to isolate the brain MV effects from the inflammation associated with CNS lupus from those caused by CS treatment.

Table 1: Patient Characteristics

ID	Age @HD [yrs]	Sex	Race	Ethnicity	Diagnosis	Age at Diagnosis	Weeks between HD, LD	CS Dose HD:LD [mg/day]	CRP HD:LD [mg/dL]	BMI HD:LD	BP HD:LD
1	20	F	AA	NH	JIA	10	38.9	20:0	6.1:6.2	43.9:46.9	131/78:117/76
2	18	M	C	H	SLE	18	37.1	40:4	0:0.1	18.4:16.2	135/76:115/67
3	17	F	C	NH	Granulomatosis with polyangiitis	17	38.9	50:0	n/a:0.4	19.7:21.5	127/73:115/75
4	17	F	AA	NH	Sarcoidosis	17	41.6	20:0	0.3:0.4	27.0:28.9	123/73:126/59
5	18	F	A	NH	Autoimmune thrombocytopenia	18	14.1	20:0	n/a:n/a	22.3:20.8	113/67:107/59
6	20	M	C	NH	SLE	20	38.9	20:8.75	0.4:n/a	23.8:22.4	133/75:152/75
7	20	F	A	NH	SLE	19	38.9	30:0	0.3:n/a	19.8:18.3	117/77:107/70
8	20	F	AA	NH	SLE	19	38.9	20:0	0.8:n/a	55.5:53.7	114/78:120/78
9	15	F	C	NH	JIA	15	38.9	40:0	0.4:0.6	28.1:25.9	113/68:119/69

CS=corticosteroids, HD=high dose CS, LD=low dose CS, F=female, M=male, AA=African American, A=Asian, C=Caucasian, NH=non-Hispanic, H=Hispanic, JIA=juvenile idiopathic arthritis, SLE=systemic lupus erythematosus, CRP=C-reactive protein, BMI=body mass index, BP=blood pressure, n/a=not available.

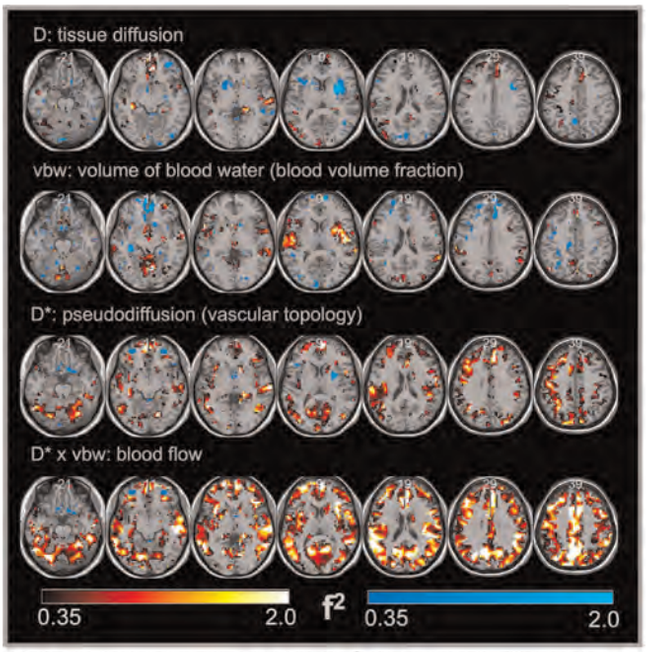


Figure 1: Effect size, expressed as Cohen's f^2, of mean pairwise difference in IVIM parameters between high dose and low dose CS. Effect sizes exceeding 0.35 are shown. Hot colors indicate where the mean difference is positive. Cool colors indicate where the mean difference is negative. Neurologic orientation convention used.

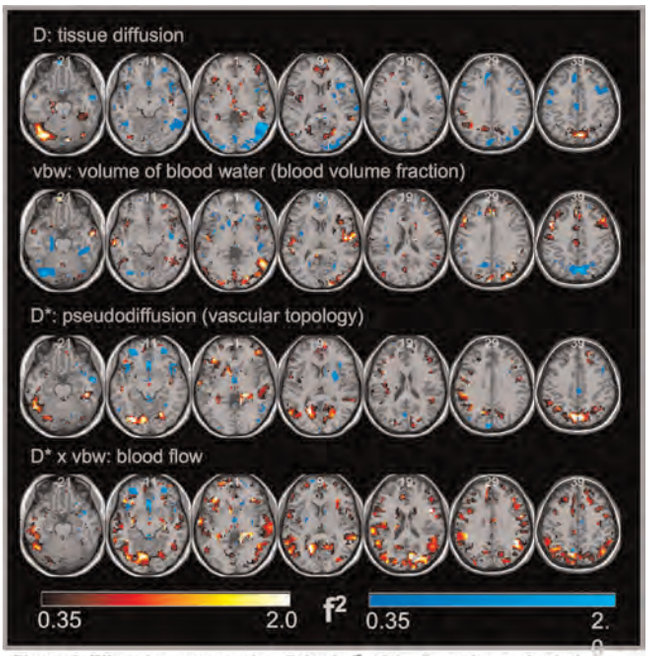


Figure 2: Effect size, expressed as Cohen's f^2, of the dependence of pairwise difference in IVIM parameters between high dose and low dose CS on dose reduction. Effect sizes exceeding 0.35 are shown. Hot colors indicate where the dependence is positive. Cool colors indicate where the dependence is negative. Neurologic orientation convention used.

Disclosure: M. DiFrancesco: None; **E. Ogbu**: None; **C. Robben**: None; **J. Huggins**: None; **H. Brunner**: GENENTECH, 12, provision of study drug for NIAMS funded study, Pfizer, 1, 2, 6.

Abstract Number: 025

Patient- and Center-level Risk Factors for Research Lost to Follow-up Using the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry

Monica Aswani, Livie Huie, Kristine Hearld, Melissa Mannion and **Emily Smitherman**, University of Alabama at Birmingham, Birmingham, AL

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Poster Breakout 3 - Pediatric Rheumatology Potpourri
Session Type: Abstract Session
Session Time: 5:10PM–5:40PM

Background/Purpose: Clinical registries are typically envisioned to be representative of a target patient population and reflective of health care delivery practices for said population. Variation in clinical practices by different sites and concerns related to recruitment/retention can have profound implications for the validity of data collected and extent to which it may be impacted by selection bias. Thus, the objective of this study is to assess the role of patient- and site-level factors associated with research attrition in the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry.

Table 1: Descriptive Statistics (N = 9730)

	Not Lost to Follow-up			Lost to Follow-up		
	Mean/Prop.	Freq.	SD	Mean/Prop.	Freq.	SD
LTFU	.000	9002		1.000	728	
Age	11.540		4.795	12.341		4.837
Female	.720	6483		.668	486	
% Missed Appointments	6.964		14.410	47.449		18.756
% Unscheduled Appointments	1.674		6.407	.755		3.699
Race/Ethnicity						
White	.711	6403		.720	524	
Black	.064	575		.088	64	
Asian	.046	410		.027	20	
Native American	.013	115		.008	6	
Middle Eastern	.006	56		.004	3	
Native Hawaiian or Pacific Islander	.006	54		.003	2	
Other	.016	140		.023	17	
Decline to Answer	.019	175		.011	8	
Hispanic	.119	1074		.115	84	
Insurance						
Private Insurance	.618	5559		.635	462	
Medicaid	.195	1755		.232	169	
Medicare	.021	186		.021	15	
Non-Medicaid State Insurance	.025	229		.029	21	
Military Insurance	.016	146		.014	10	
Indian Health Insurance	.012	108		.016	12	
No Insurance	.048	435		.003	2	
Non-US Insurance	.065	584		.051	37	
Other Insurance						
Income						
<25,000	.079	715		.092	67	
25,000-49,999	.117	1054		.143	104	
50,000-74,999	.117	1056		.137	100	
75,000-99,999	.106	957		.117	85	
100,000-150,000	.167	1499		.120	87	
>150,000	.151	1363		.106	77	
Unknown	.134	1209		.183	133	
Decline to Answer	.128	1149		.103	75	
Education						
Less than High School	.037	332		.018	13	
High School	.133	1193		.076	55	
College or Higher	.611	5503		.266	194	
Decline or Unknown	.219	1974		.640	466	
PR-CQIN Site	.414			.492		
Year Site Enrolled 1st Patient						
2015	.493	4438		.613	446	
2016	.462	4157		.368	268	
2017	.019	170		.003	2	
2018	.019	170		.015	11	
2019	.007	67		.001	1	

Methods: Patients enrolled in the CARRA Registry from 2015 to 2019 were eligible. The effects of patient (level 1) and site (level 2) factors on lost to follow-up were assessed using a 2-level logistic model, followed by empirical Bayes estimation to investigate cross-center variation. Sites classify patients as inactive if they are referred elsewhere, move to a non-CARRA site, transition to adult rheumatology, achieve remission, or are lost to follow-up. We defined our outcome of interest as patients lost to follow-up (= 1) versus all other patients (=0), who may be active or inactive due to other reasons. The empirical Bayes estimates are used to graphically illustrate variation in lost to follow-up rates between unadjusted and adjusted models (Figure 1).

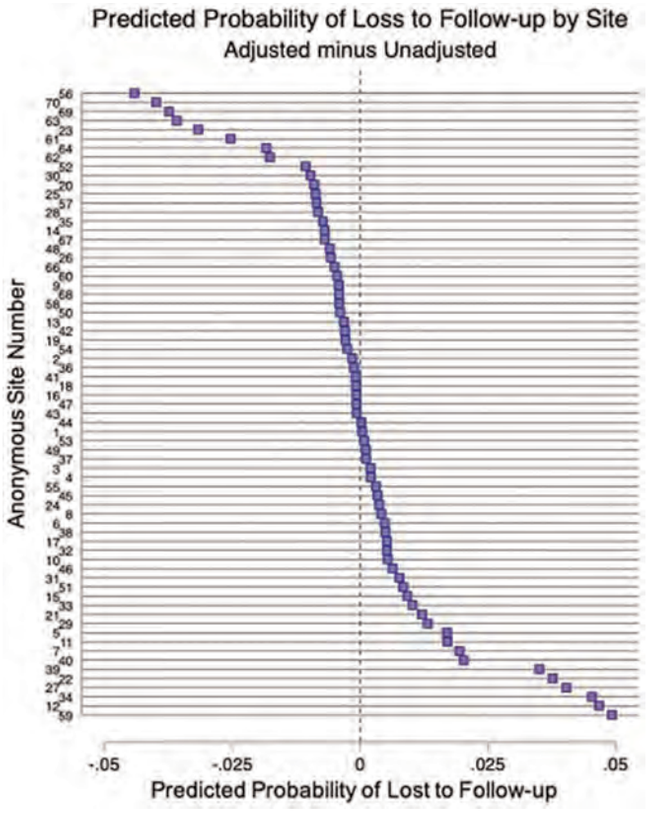


Figure 1: Site-level Variation. Sites with a negative difference (i.e., higher unadjusted predicted probability, to the left of zero) means controlling for patient- and site-level factors made their lost to follow-up look more favorable. Sites with a positive difference (i.e., higher adjusted predicted probability, to the right of zero) means controlling for patient- and site-level factors made their lost to follow-up look worse off.

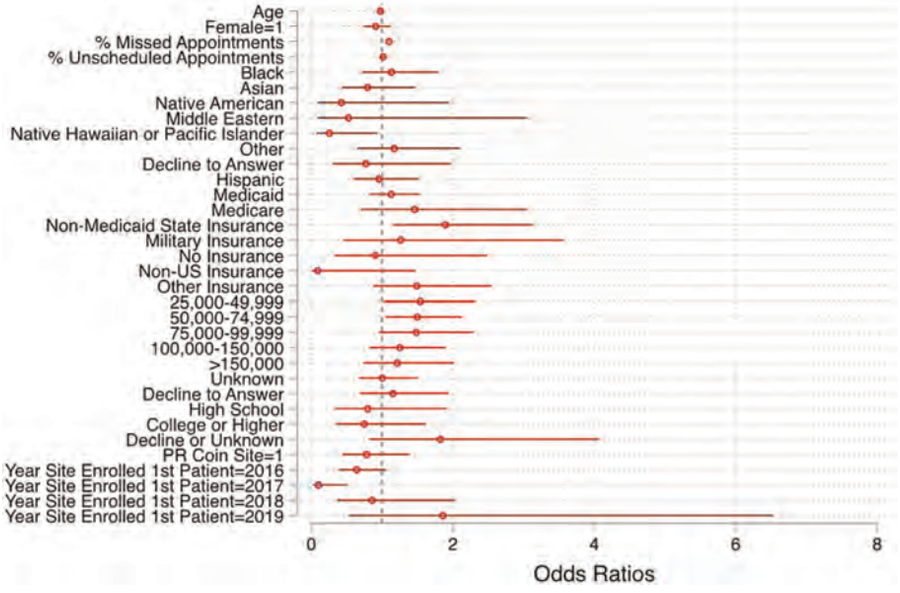


Figure 2: Multilevel Logistic Regression. Reference categories include: Male, White race, private insurance, less than high school, non-PR-COIN site, and site started CARRA enrollment in 2015.

Results: The final analytic sample contained 9,730 patients nested within 69 sites (Table 1). Most of the sample was female (n=6,481, 72%), White (n=6,403, 64%), and privately insured (n=5,559, 62%). Overall, 728 (7.5%) patients in the cohort were lost to follow-up. The intra-class correlation coefficient (ICC) from null model signified that 20.3% of the variance in lost to follow-up is at the registry site level. Percent of missed Registry visits (OR 1.09, 95% CI: 1.086, 1.12), non-Medicaid state insurance (OR 1.95, 95% CI: 1.15, 3.12), and an income between $25,000 and $49,999 (OR 1.52, 95% CI: 1.003, 2.31) were significantly associated with higher odds of lost to follow-up (Figure 2).

Conclusion: Significant variation in research participant lost to follow-up exists at the site level. Moreover, individual-level characteristics such as visit history, insurance status, and household socioeconomic status were associated with the likelihood of being lost to follow-up. This non-random attrition may also highlight concerns related to patient retention in clinical practice.

Disclosure: M. Aswani: None; **L. Huie**: None; **K. Hearld**: None; **M. Mannion**: None; **E. Smitherman**: None.

Abstract Number: 026

Performance of the UCLA Scleroderma Clinical Trials Consortium Gastrointestinal Tract 2.0 Instrument in a Juvenile Systemic Sclerosis Cohort

Sophie Stefancic, Amanda Robinson, Haley Havrilla, Samantha Branton, Vibha Sood and **Kathryn Torok**, University of Pittsburgh, Pittsburgh, PA

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 4 - JDM & Scleroderma: Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: Gastrointestinal (GI) manifestations in juvenile onset systemic sclerosis (jSSc) reflect adult disease with a range of involvement along the GI tract, including oropharyngeal dysphagia and intestinal dysmotility, which lead to malnutrition (15-56% jSSc) and increased mortality. Patient reported outcomes (PRO) to capture the impact of GI disease in children would be useful in detecting underlying GI problems and to establish outcome measures that capture change, optimizing understanding of therapeutic impact. The UCLA Scleroderma Clinical Trial Consortium Gastrointestinal Tract 2.0 (UCLA-GIT 2.0; GIT) is one of the most widely used and validated PRO for severity of GI involvement in adult-onset SSc (aSSc). To our knowledge the GIT 2.0s performance in jSSc has not been established.

Methods: All jSSc subjects enrolled in the National Registry for Childhood Onset Scleroderma, a prospective observational clinical research registry at a multi-disciplinary center, with a GIT collected at study visit were included. Demographic, clinical, and PRO data of interest were extracted. Summary statistics were applied to outcome measures. Spearman correlation coefficient was applied to analyze relationships between the GIT total and subscales to a traditional global GI involvement PRO, the intestinal visual analog scale of the Scleroderma Health Assessment Questionnaire (SHAQ-GI-VAS) for convergent construct validity (significance defined as p 0.05).

Results: Data for a total of 54 patients with jSSc were extracted. The average age of onset was 9.9 years old, and there was 3.3 years disease duration at time of initial GIT collection. Demographics, autoantibody, classification and clinical variables are summarized in Table 1. The median (IQR) of the GIT 2.0 and its subscales in our jSSc cohort are displayed in Table 2,

Table 1. Demographic and Clinical Data Summary n=54 Juvenile onset Systemic Sclerosis Patients.

	Juvenile Systemic Scleroderma Patients n=54
Demographics (median, IQR) or (n, %)	
Age at Disease Onset	9.9 (6.3 – 13.8)
Disease Duration at Visit	3.3 (1.3 – 4.3)
Female	39 (72.2)
Male	15 (27.8)
Race **White** **Black** **Asian** **Mixed** **Other**	 38 (70.4) 11 (20.4) 3 (5.6) 1 (1.9) 1 (1.9)
Subtype (n, %)	
Diffuse Cutaneous	24 (44.4)
Limited Cutaneous	5 (9.3)
Overlap SSc	18 (33.3)
Unclassified	7 (13.0)
Autoantibody Positivity (n , %)	
Scl-70	17 (31.5)
PM-SCL	10 (18.5)
U3-RNP	6 (11.1)
U1-RNP	4 (7.4)
Centromere	3 (5.6)
RO60	2 (3.7)
RNA pol III	1 (1.9)
Clinical Variables (median, IQR) or (n, %)	
Modified Rodnan Skin Score	9.5 (0 – 15.5)
Any Musculoskeletal Involvement **Myositis** **Muscle Weakness**	52 (96.3) 21 (38.9) 28 (51.9)
Any Gastrointestinal Involvement	37 (68.5)
Any Pulmonary Involvement **Abnormal PFTs** **Abnormal CT Chest**	46 (85.2) 24 (44.4) 21 (38.9)
Any Vascular Involvement **Digital Ulcer**	52 (96.3) 27 (50.0)
Other Dermatologic Involvement (not including skin thickness) **Calcinosis** **Gottron Papules**	47 (87.04) 5 (9.26) 11 (20.37)
CHAQ	0.64 (0.09 – 0.88)
Intestinal VAS (SHAQ GI-VAS)	0.08 (0 – 0.81)
UCLA-GIT 2.0 Total	0.22 (0.07 – 0.49)

The Childhood Health Assessment Questionnaire (CHAQ)
The Scleroderma Health Assessment Questionnaire Intestinal Visual Analogue Scale (SHAQ GI-VAS)
The University of California Los Angeles Scleroderma Clinical Trial Consortium gastrointestinal tract 2.0 (UCLA-GIT 2.0)

adjacent to a published adult comparison cohort. Most values are comparable to those found in adult SSc subjects with Distension and Reflux having the most impact. The GIT and its subscales correlated well with the SHAQ-GI-VAS scale, with the Total score having the strongest association, and notable moderate correlations with Social functioning and Emotional well-being (Table 3). Diarrhea was the only subscale not to correlate with SHAQ-GI-VAS. In addition, the Total score also correlated moderately to the SHAQ Global overall Disease impact (r_s 0.64, p 0.001).

Table 2. The UCLA Scleroderma Clinical Trials Consortium Gastrointestinal Tract 2.0 (GIT) Instrument Scores, Total and Subscores, in Juvenile systemic sclerosis (jSSc) (n=54) compared with *Adult SSc (n=346; all visits = 940 instances GIT)

UCLA GIT 2.0 subscales (0-3)[†]	Juvenile SSc Score n=54	*Adult SSc Score n=346
	Median (Q1-Q3) Range (min-max)	
Reflux Average GIT Score	**0.25** (0.00 – 0.50) R 0 – 2.13	**0.25** (0.00 – 0.63) R 0 – 3
Distension Average GIT Score	**0.50** (0.25 – 1.00) R 0 – 2.75	**0.50** (0.00 – 1.00) R 0 – 3
Soilage Average GIT Score	0.00 (0.00 – 0.00) R 0 – 2	0.00 (0.00 – 0.00) R 0 – 3
Diarrhea Average GIT Score	0.00 (0.00 – 0.00) R 0 – 2	0.00 (0.00 – 0.50) R 0 – 2
Social Functioning Average GIT Score	0.00 (0.00 – 0.33) R 0 – 2	0.00 (0.00 – 0.33) R 0 – 2.5
Emotional Well-being Average GIT Score	0.00 (0.00 – 0.00) R 0 – 1.22	0.00 (0.00 – 0.22) R 0 – 3
Constipation Average GIT Score	0.00 (0.00 – 0.69) R 0 – 2.5	0.00 (0.00 – 0.50) R 0 – 2.5
Total score of UCLA GIT 2.0	**0.16 (0.05 – 0.42) R 0 – 1.93**	**0.22 (0.07 – 0.49) R 0 – 2.5**

***Adult comparison SSc data obtained from:** Zampatti, N., Garaiman, A., Jordan, S. *et al.* Performance of the UCLA Scleroderma Clinical Trials Consortium Gastrointestinal Tract 2.0 instrument as a clinical decision aid in the routine clinical care of patients with systemic sclerosis. *Arthritis Res Ther* 23, 125 (2021). https://doi.org/10.1186/s13075-021-02506-x

The UCLA SCTC 2.0[†] includes 34 items and seven multi-item scales (reflux, distension/bloating, diarrhea, fecal soilage, constipation, emotional well-being and social functioning) and a total GIT score to assess health-related quality of life (HRQOL) and severity of GIT symptoms in SSc. All scales are scored from 0 (better HRQOL) to 3 (worse HRQOL), except the diarrhea and constipation(range from 0 to 2 and from 0 to 2.5, respectively), and this provides a total GIT severity scale. The GIT 2.0 has feasibility, reliability (test-retest and internal consistency) and validity in multiple studies. Minimally important differences have been estimated for all scales.
- Furst DE, Braun-Moscovic Y, Khanna D. Points to consider for clinical trials of the gastrointestinal tract in systemic sclerosis. *Rheumatology* (Oxford). 2017 Sep 1;56(suppl_5):v4-v11. doi: 10.1093/rheumatology/kex195. PMID: 28992166; PMCID: PMC5850471.

Conclusion: Results from a single center jSSc cohort demonstrate the GIT 2.0 is likely a useful tool in pediatric scleroderma, despite its development in adult disease. The literature finds the median Total score and subscale scores in adults to be similar to those reported here in jSSc, with greatest impact of GI issues to be from Distention and Reflux domains. Targeting supportive and pharmacological therapy in these areas should be pursued. This study also supports the social and emotional impact of GI disease using a PRO in pediatrics, which is important as patients (teens especially) are reluctant to fully answer GI related questions directly from the physician. The next step is to evaluate the GITs sensitivity to change in jSSc via longitudinal study visits in relation to GI and other outcomes.

	T Statistic	*Spearman's Coefficient (r_s)	P Value
SHAQ GI VAS Scale correlation			
GIT Reflux	5.09	**0.58**	**< 0.001**
GIT Distension	5.54	**0.61**	**< 0.001**
GIT Soilage	2.94	**0.38**	**0.005**
GIT Diarrhea	2.29	0.31	0.03
GIT Social Functioning	6.20	**0.66**	**< 0.001**
GIT Emotional Wellbeing	5.54	**0.61**	**< 0.001**
GIT Constipation	3.13	**0.40**	**0.003**
GIT Total	7.30	**0.71**	**< 0.001**
SHAQ Global Disease VAS Scale correlation			
GIT Total	5.91	**0.64**	**< 0.001**

***Correlation coefficient value scale**
< 0.4 Weak Correlation
0.4 – 0.7 Moderate Correlation
> 0.7 Strong Correlation

Spearman's Correlation Coefficient comparison between the intestinal visual analog scale of the Scleroderma Health Assessment Questionnaire (SHAQ-GI-VAS) and the UCLA Scleroderma Clinical Trials Consortium Gastrointestinal Tract 2.0 Instrument (GIT) Total and Subscores Demonstrate convergent construct validity. N=53 Juvenile systemic sclerosis (jSSc) patients (51 Degrees of Freedom). Significant ($p<0.05$) values are bolded.

Disclosure: S. Stefancic: None; **A. Robinson**: None; **H. Havrilla**: None; **S. Branton**: None; **V. Sood**: None; **K. Torok**: None.

Abstract Number: 027

Identifying and Understanding JDM in Africa: A Survey of Rheumatology Care Providers from Africa

Jessica Perfetto[1], Laura Lewandowski[2], Dawn Wahezi[1], Christiaan Scott[3] and Angela Migowa[4], [1]Children's Hospital at Montefiore, New York, NY, [2]NIAMS, NIH, Bethesda, MD, [3]Red Cross War Memorial Children's Hospital, [4]Aga Khan University, Nairobi, Kenya

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 4 - JDM & Scleroderma: Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: There is a paucity of data on pediatric rheumatic disease (PRD) in low and middle-income countries (LMIC), creating a false perception of low prevalence when this gap is driven by limited diagnostic capacities, scarcity of pediatric rheumatologists, and limited awareness of PRD. Juvenile dermatomyositis (JDM) can cause significant morbidity

and irreversible damage when inadequately treated, resulting in medical, social, and economic burden on patients, families, and healthcare systems, particularly in LMIC. The few studies of JDM in Africa suggest high rates of severe manifestations, including calcinosis and pulmonary involvement. The objective of this survey is to better understand the scope of JDM burden in Africa by obtaining an overview of JDM and comparing this to data in previously published work.

Figure 1: Country of origin of partial or complete responses (n=20)

Table 1: Clinical outcomes in children with juvenile dermatomyositis in Africa among survey respondents (n=20)

Country (Number of Respondents)	Total Patients (n=216)*	Clinically Inactive Disease†	Remission‡	Calcinosis	Interstitial Lung Disease	Deaths
Northern Africa (12)						
Egypt (6)	68*	28	13	36	5	2
Libya (3)	55*	18*	39*	12	5*	3
Morocco (1)	19	6 (31.6%)	4 (21.1%)	7 (36.8%)	6 (31.6%)	0 (0%)
Tunisia (2)	3*	6	6	0	4	0*
Eastern Africa (3)						
Kenya (2)	42	15 (35.7%)	12 (28.6%)	13 (31.0%)	8 (19.0%)	6 (14.3%)
Zambia (1)	7	5 (71.4%)	0 (0%)	3 (42.9%)	1 (14.3%)	2 (28.6%)
Central Africa (3)						
Cameroon (2)**	11	6 (54.5%)	5 (45.5%)	3 (27.3%)	2 (18.2%)	0 (0%)
Democratic Republic of the Congo (1)**	3	3 (100%)	1 (33.3%)	3 (100%)	0 (0%)	+
Western Africa (1)						
Nigeria (1)	4	2 (50.0%)	1 (25.0%)	0 (0%)	1 (25.0%)	0 (0%)
Southern Africa (1)						
South Africa (1)	4	1 (25.0%)	1 (25.0%)	2 (50.0%)	0 (0%)	0 (0%)

Number of patients reported as n; (%) also reported when total patients known
† Defined as lack of evidence of myositis disease activity as assessed by global and extra-muscular assessments, stable muscle strength and function, and normal muscle enzyme levels, per the International Myositis Assessment & Clinical Studies (IMACS) criteria for lack of evidence of active myositis
‡ Defined according to the IMACS 2005 definition: clinically inactive disease while not receiving any drug therapy for a 6-month continuous period
*At least 1 respondent unsure of total number of patients, therefore number provided is an underestimation and percentages were unable to be calculated; Egypt=3 of 6 respondents unsure about prior patients, Libya=1 of 3 respondents unsure about total current and total prior patients, Tunisia=2 of 2 respondents unsure about total current patients and 1 of 2 unsure about prior patients
**Respondents answered questions only about current, not prior, patients (Cameroon=1 respondent, Democratic Republic of Congo=1 respondent)
+ Response missing from 1 respondent

Table 2: Resources available and accessible for diagnosing and managing JDM in Africa (n=17*)

	Resources available, not considering access and/or cost	Resources typically used after considering access and/or cost	Countries+
General Diagnostic Tools	**n=18**	**n=17**	
None of the above	*n/a*	0 (0%)	
Inflammatory markers	18 (100%)	15 (88.2%)	
Muscle enzymes	18 (100%)	17 (100%)	
vWF antigen	4 (22.2%)	1 (5.9%)	**Egypt**
ANA	15 (83.3%)	9 (52.9%)	
MSA and/or MAA**	10 (55.6%)	5 (29.4%)	**Egypt, Tunisia**, South Africa
Muscle biopsy	11 (61.1%)	1 (5.9%)	**Tunisia**
EMG	13 (72.2%)	7 (41.1%)	**Egypt, Libya, Tunisia**
Disease assessment tools***	13 (72.2%)	9 (52.9%)	
Other†	2 (11.1%)	2 (11.8%)	
Diagnostic Tools for Calcinosis	**n=17**	**n=17**	
None of the above	*n/a*	1 (5.9%)	
Physical exam	14 (82.4%)	13 (76.5%)	
X-rays	15 (88.2%)	12 (70.6%)	
MRI	9 (52.9%)	4 (23.5%)	**Egypt, Libya**
Ultrasound	12 (70.6%)	7 (41.2%)	**Egypt, Tunisia**, Zambia, Cameroon
Other‡	1 (5.9%)	0 (0%)	
Diagnostic Tools for ILD	**n=17**	**n=17**	
None of the above	*n/a*	0 (0%)	
X-rays	10 (58.8%)	9 (52.9%)	
CT	15 (88.2%)	12 (70.6%)	
PFTs	14 (82.4%)	13 (76.5%)	
Other	0 (0%)	0 (0%)	
Medications	**n=17**	**n=16**	
None of the above	*n/a*	1 (6.3%)	
Steroids			
Prednisone or prednisolone	17 (100%)	16 (100%)	
Intravenous Methylprednisolone Pulse Dosing (15-30mg/kg)	16 (94.1%)	14 (87.5%)	
Conventional DMARDs			
Hydroxychloroquine or chloroquine	14 (82.4%)	8 (50%)	**Egypt, Tunisia, Morocco**, Zambia
Methotrexate	17 (100%)	16 (100%)	
Mycophenolate mofetil or mycophenolic acid	14 (82.4%)	11 (68.8%)	
Azathioprine	14 (82.4%)	10 (62.5%)	
Tacrolimus	7 (41.2%)	0 (0%)	
Cyclosporine or ciclosporin	10 (58.8%)	3 (18.8%)	**Egypt, Morocco**
Cyclophosphamide	12 (70.6%)	8 (50%)	**Egypt, Tunisia, Morocco,** South Africa, Kenya
Biologic DMARDs			
Rituximab	13 (76.5%)	8 (50%)	**Egypt, Tunisia, Morocco, Libya,** South Africa, Kenya
TNF inhibitors	9 (52.9%)	4 (25%)	**Egypt,** South Africa
Abatacept	2 (11.8%)	1 (6.3%)	**Egypt**
Other			
IVIG	14 (82.4%)	9 (56.3%)	
Janus kinase inhibitors	6 (35.2%)	2 (12.5%)	**Egypt**
Other	0 (0%)	0 (0%)	

JDM: juvenile dermatomyositis, vWF: von Willebrand factor, ANA: antinuclear antibody, MSA: myositis-specific antibody, MAA: myositis-associated antibody, EMG: electromyography, MRI: magnetic resonance imaging, ILD: interstitial lung disease, CT: computed tomography, PFTs: pulmonary function tests, DMARD: disease-modifying antirheumatic drugs, TNF: tumor necrosis factor, IVIG: intravenous immune globulin

* 17 respondents except where otherwise noted

+ Country listed when a resource was available and/or accessible to ≤ 50% of respondents; countries in bold represent Northern African countries

** MSA include: Mi-2, MDA5 (CADM140), NXP-2 (MJ), TIF1 (p155/140), SRP, anti-synthetase (Jo-1, PL-7, PL-12, EJ, OJ, KS, Zo, Ha, YRS); MAA include: Pm-Scl, U1RNP, U1/U2RNP, U3RNP, Ro, La, Ku

*** Including any of the following: Childhood Myositis Assessment Scale (CMAS), Childhood Health Assessment Questionnaire (CHAQ), Manual Muscle Testing (MMT), Physician Global Activity Visual Analogue Scale (VAS), Patient/Parent Global Activity VAS, Myositis Disease Activity Assessment Tool (MDAAT), Myositis Disease Damage Index (MDI), Physician Global Assessment of Disease Damage, Patient/Parent Global Assessment of Disease Damage

† Other = MRI (1), ultrasound (1)

‡ Other = diagnosis made clinically but all tools available (1)

Methods: A survey was distributed to members of the African League of Associations for Rheumatology (AFLAR; n=233) and Paediatric Society of the African League Against Rheumatism (PAFLAR; n=130) via WhatsApp groups. Respondents were queried about the total number of JDM patients seen currently or within the last 10 years; morbidities and clinical outcomes, including calcinosis, interstitial lung disease (ILD), clinically inactive disease, remission, and death; and availability and accessibility of diagnostic tools and medications.

Results: A total of 43 (12%) individuals started the survey, with 4 (9%) partially and 16 (37%) fully completing it, describing 216 JDM patients. One general pediatrician participated; the remainder were adult (n=10; 50%), pediatric (n=7; 35%), or adult/pediatric (n=2; 10%) rheumatologists. Respondents represented the 5 regions of Africa, primarily Northern Africa (n=12; 60%) (Figure 1). Mortality was reported at 14% and 29% in Kenya and Zambia, respectively (Table 1). Many respondents identified limited availability of medications (56%), unfamiliarity with JDM by caregivers (69%) and other medical providers (44%), and delayed presentation to care (75%). Access and/or cost limited use of diagnostic tools and medications (Table 2). 13 diagnostic tools and medications were typically used by 50% or fewer of respondents, the majority (69%) of which are accessible in only Northern or Southern African countries. Despite the lack of cost or other apparent access barriers, disease assessment tools are used by only 53% of respondents.

Conclusion: This is the first study to explore JDM on a broad scale across the African continent. We identified 216 African children with JDM within the past 10 years, exceeding the previously published reports of 196 JDM patients seen in the last 25 years and likely still underestimating prevalence. Our results suggest substantially higher rates of severe disease, with mortality rates in some countries vastly different from the 1-3% reported in high-income countries. Many respondents were limited in their capacity to diagnose and treat JDM, but there was a wide range across African regions. This survey highlighted important possible differences in disease manifestations and outcomes that warrant further study. Future collaboration with our African colleagues is critical to raise awareness of JDM to ultimately improve disease severity and outcomes.

Disclosure: J. Perfetto: None; **L. Lewandowski**: None; **D. Wahezi**: None; **C. Scott**: None; **A. Migowa**: None.

Abstract Number: 028

Achieving Medication-Free Remission in Juvenile Dermatomyositis

Harneet Ghumman[1], Ilaria Maccora[2], Hermine Brunner[1], Amy Cassedy[3], Mekibib Altaye[2], Asra Firdous[1], Alexei Grom[1], Daniel Lovell[1], Angela Merritt[1], Megan Quinlan-Waters[1] and **Sheila Angeles-Han**[2], [1]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, [2]Cincinnati Children's Hospital, Cincinnati, OH, [3]Cincinnati Children's Hospital Medical Center, Cincinnati, OH

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 4 - JDM & Scleroderma: Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: Juvenile dermatomyositis (JDM) is characterized by symmetric proximal muscle weakness, distinct rash, and a risk for calcinosis. Systemic immunosuppression is needed. Evidence is limited on the factors associated with disease remission off of medication. The aims of this study are to describe the demographic, clinical, laboratory, imaging characteristics in our JDM cohort, and to identify variables associated with medication-free remission.

Methods: This is a cross-sectional retrospective study of a convenience sample of children diagnosed with JDM at ≤18 years of age, with a minimum clinical follow-up of 2 years at Cincinnati Children's Rheumatology Clinic. Medication-free remission was defined as inactive JDM off all systemic medications for at least 6 months. Medical records were reviewed. We compared the following variables in children who achieved remission to those who did not: demographics, clinical features, muscle enzymes, ANA, myositis-specific auto-antibodies, Childhood Myositis Assessment Scale (CMAS) scores, and physician, parent and patient global assessment scores. Group differences were tested using Fisher's Exact tests and Wilcoxon Rank Sum tests. A $p < 0.05$ was considered statistically significant. Analysis was conducted using SAS 9.3©.

Results: Of 70 participants, 47 (67%) achieved medication-free remission while 23 (33%) were still on medication at last follow-up. Overall, most were Non-Hispanic White (87%), females (64%), had a median age of JDM onset 5 years (range 3-11), and median age of diagnosis of JDM of 5 years (4-11). Those who achieved medication-free remission required treatment for 43 (interquartile [IQR] 28-77) months before discontinuation. They were of a younger median age at JDM onset (5 [IQR 3-6] vs 9 [5-12] years) and JDM diagnosis (5 [3-7] v. 9 [5-12] years) compared to those who did not achieve remission. Males were less likely to achieve remission (28% [n=13] in remission vs. 52% [12], p=0.044). Both groups had similar presentation of rash, muscle weakness, elevated muscle enzyme levels, ANA positivity, abnormal muscle and skin biopsy, and MRI findings (in those who had them done). Time to treatment initiation was similar, and prednisone (94%), methotrexate (87%) and IVIG (56%) were most commonly prescribed. Children unable to achieve medication-free remission were more likely to be on IVIG (74% [17/39], p=0.032), tofacitinib (100% [3/3], p=0.032), and abatacept (22% [5/7], p=0.030). At last follow up, they also had worse patient/parent scores global assessment scores (0.0 [0-1.5] vs 1.0 [0-5.0], p=0.012), but similar CMAS and physician global scores (Table 3).

Table 1. Characteristics of Children with Juvenile Dermatomyositis

n (%), unless otherwise specified	Total n = 70	Children Who Achieved Medication-Free Remission n = 47	Children Who Did Not Achieve Medication-Free Remission n = 23	χ^2/Z Score	p-Value
Demographics					
Ethnicity				0.26	0.451
Non-Hispanic	35 (97.2)	29 (100.0)	6 (85.7)		
Race				3.10	0.055*
White	60 (87.0)	42 (89.4)	18 (81.8)		
Asian	2 (2.9)	2 (4.3)	0 (0.0)		
African-American	7 (10.1)	3 (6.4)	4 (18.2)		
Gender				4.04	0.044*
Female	45 (64.3)	34 (72.3)	11 (47.8)		
Disease Characteristics					
Season Diagnosed				1.11	0.774
Fall	19 (27.1)	13 (27.7)	5 (26.1)		
Winter	13 (18.6)	10 (21.3)	3 (13.0)		
Spring	17 (24.3)	10 (21.3)	7 (30.4)		
Summer	21 (30.0)	14 (29.8)	7 (30.4)		
Age at Baseline, Median (IQR)	9.5 (5.0 – 16.0)	7.0 (4.0 – 15.0)	13.0 (8.0 – 19.0)	2.36	0.018*
Age at Onset, Median (IQR)	5.0 (3.0 – 11.0)	5.0 (3.0 – 7.0)	9.0 (4.0 – 12.0)	2.44	0.015*
Age at Diagnosis, Median (IQR)	5.0 (4.0 – 11.0)	5.0 (3.0 – 7.0)	9.0 (5.0 – 12.0)	2.53	0.011*
Days to Initiation of Treatment, Median (IQR)	0.0 (0.0 – 0.5)	0.0 (0.0 – 0.5)	0.1 (0.0 – 0.9)	1.06	0.291
Medication Duration in Months, Median (IQR)	41.1 (28.6 – 73.1)	43.2 (28.6 – 76.7)	39.3 (24.2 – 56.4)	-0.894	0.371
Follow Up Duration in Months, Median (IQR)	43.2 (29.5 – 74.2)	43.2 (28.6 – 81.3)	39.6 (29.8 – 71.3)	-0.100	0.460
Clinical Features					
Heliotrope Rash	41 (58.6)	28 (59.6)	13 (56.5)	.06	0.808
Gottron's Papules	50 (71.4)	36 (76.)	14 (60.9)	1.87	0.171
Proximal Muscle Weakness	54 (77.1)	35 (74.5)	19 (82.6)	0.58	0.446
Nailfold Capillary Changes	43 (61.4)	31 (66.0)	12 (52.2)	1.24	0.266
Elevated Muscle Enzymes^, n = those with results					
ALT, (n= 62)	24 (34.2)	14 (34.2)	10 (47.6)	1.06	0.303
AST, (n= 62)	37 (59.7)	25 (61.0)	12 (57.1)	0.08	0.771
Aldolase, (n=61)	44 (72.1)	28 (68.3)	16 (80.0)	0.92	0.338
CK, (n=63)	29 (46.0)	20 (47.6)	9 (42.9)	0.13	0.721
LDH, (n=54)	49 (90.7)	31 (91.2)	18 (90.0)	0.02	0.359
Other Abnormal Test Findings, n = those with results					
Muscle Biopsy, (n=25)	23 (92.0)	13 (86.7)	10 (100.0)	1.45	0.229
MRI, (n=53)	47 (88.7)	32 (88.9)	15 (88.2)	0.01	0.944
Skin Biopsy, (n=31)	8 (25.8)	5 (23.8)	3 (30.0)	0.14	0.713
Positive Myositis Auto-antibodies, n = those with results					
ANA, (n=46)	34 (73.9)	23 (76.7)	11 (68.8)	0.34	0.560
Anti-Jo-1, (n= 38)	2 (5.3)	0 (0.0)	2 (16.7)	4.57	0.094
Anti-Ro/SSA, (n=40)	2 (5.0)	0 (0.0)	2 (14.3)	3.91	0.117
Anti-La/SSB, (n=39)	0 (0.0)	0 (0.0)	0 (0.0)	NA	NA
Anti-RNP, (n=39)	1 92.6)	0 (0.0)	1 (7.7)	2.05	0.333
Anti-Sm (n= 40)	1 (2.5)	0 (0.0)	1 (7.1)	1.90	0.350
Anti-dsDNA, (n=34)	1 (2.9)	0 (0.0)	1 (8.3)	1.89	0.353
RF, (n=11)	0 (0.0)	0 (0.0)	0 (0.0)	N/A	N/A
Anti-Mi-2, (n=1)	2 (50.0)	0 (0.0)	1 (50.0)	N/A	N/A

* p<0.05 was considered statistically significant, ^Normal lab values: AST : ≤ 47, ALT: ≤49, Aldolase: ≤ 7.6, CK: ≤ 192, LDH: ≤ 246

Table 2. Medication Administration in Management of Juvenile Dermatomyositis

	Total n=70	Children Who Achieved Medication-Free Remission n = 47	Children Who Did Not Achieve Medication-Free Remission n = 23	χ^2/Z Score	p-Value
Medication Administration, n (%)					
Abatacept	7 (10.0)	2 (4.3)	5 (21.7)	5.24	**0.030***
Adalimumab	1 (1.4)	0 (0.0)	1 (4.4)	2.07	0.329
Azathioprine	5 (7.1)	4 (8.5)	1 (4.4)	0.40	0.339
Cyclophosphamide	1 (1.4)	1 (2.1)	0 (0.0)	0.50	0.671
Hydroxychloroquine	44 (62.9)	27 (57.5)	17 (73.9)	1.79	0.181
IVIG	39 (55.7)	22 (46.8)	17 (73.9)	.60	**0.032***
Leflunomide	3 (4.3)	1 (2.1)	2 (8.7)	1.62	0.217
Methotrexate Total	61 (87.1)	42 (89.4)	19 (82.6)	0.63	0.209*
Methotrexate PO[1]	28 (40.0)	19 (40.4)	9 (39.1)	0.01	0.917
Methotrexate SQ[1]	54 (77.1)	37 (8.7)	17 (73.9)	0.20	0.652
Mycophenolate Mofetil,	14 (20.0)	8 (17.0)	6 (26.1)	0.79	0.164
Nirmatrelvir/Ritonavir	1 (1.4)	0 (0.0)	1 (4.)	2.07	0.329
Prednisone	66 (94.3)	44 (93.6)	22 (95.6)	0.12	0.407
Rituximab	4 (5.7)	2 (4.3)	2 (8.7)	0.56	0.298
Tacrolimus	17 (24.3)	14 (29.8)	3 (13.0	2.35	0.077
Tofacitinib	3 (4.3)	0 (0.0)	3 (13.0)	6.40	**0.032***
Triamcinolone	1 1.4)	1 (2.1)	0 (0.0)	0.50	0.671
>2 Systemic Medications	68 (97.1)	46 (97.9)	22 (95.6)	0.27	0.448

NOTE: *Fisher's Exact Test; [1]Not mutually exclusive categories

Table 3. Study Outcomes by Treatment Group of 70 Patients with Juvenile Dermatomyositis

	Children Who Achieved Medication-Free Remission n = 47		Children Who Did Not Achieve Medication-Free Remission n =23	
	Baseline	Last Follow-up	Baseline	Last Follow-up
Outcome Scores, median (IQR)				
CMAS Score[b]	42.5 (34.0 -46.5)	51.0 (49.0 – 52.0)	35.5 (36.0 – 49.0)	51.0 (46.0 – 52.0)
Parent/Patient Global Overall Well-Being Score[ac]	4.0 (1.0 – 6.0)	0.0 (0.0 – 1.5)	2.0 (0.0 – 6.5)	1.0 (0.0 – 5.0)
Physician Global Assessment[d]	1.8 (1.0 – 3.0)	0.5 (0.0 - 0.5)	2.5 (1.5 – 4.0)	1.3 (0.3 – 2.3)

NOTE: [a]Z-Score=-2.30, p 0.021, [b]CMAS Score: higher score = better strength/endurance, Median (IQR), Range 0-51, [c]Parent/Patient Global Assessment Score: Overall Well-being Score higher score = worse: Median (IQR), Range 0-10, [d]Physician Global Assessment Score: higher score = worse: Median (IQR), Range 0-10

Conclusion: Although 2/3 of patients achieved medication-free remission, 1/3 of our cohort remained on treatment. Sex and age may be important factors as more children who were males and older at JDM onset/ diagnosis were less likely to achieve medication-free remission. These children were also found to be on tofactitinib, IVIG, and abatacept.

Disclosure: H. Ghumman: None; **I. Maccora**: None; **H. Brunner**: GENENTECH, 12, provision of study drug for NIAMS funded study, Pfizer, 1, 2, 6; **A. Cassedy**: None; **M. Altaye**: None; **A. Firdous**: None; **A. Grom**: Novartis, 2, 5, Sobi, 2, 5; **D. Lovell**: AstraZeneca, 2, Boehringer-Ingelheim, 2, Bristol-Myers Squibb(BMS), 12, Prinicipal Investigator, GlaxoSmithKlein(GSK), 2, Janssen, 2, NIH/NIAMS, 12, Co-Investigator, NIH/NICHD, 12, Co-Investigator, Novartis, 2, Pfizer, 12, Prinicipal Investigator, Roche, 2, 5, UCB, 2; **A. Merritt**: None; **M. Quinlan-Waters**: None; **S. Angeles-Han**: None.

Abstract Number: 029

Cardiac Magnetic Resonance Imaging in Children with Systemic Lupus Erythematosus and Scleroderma Spectrum Disorders: A Single Center Experience

Meredith Rae, Tam Doan and Eyal Muscal, Baylor College of Medicine, Houston, TX

SESSION INFORMATION

Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 4 - JDM & Scleroderma: Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: Cardiac disease is a major cause of morbidity and mortality in children with SLE and SSc. Various studies have demonstrated an association between duration of disease and abnormalities on cardiac magnetic resonance imaging (CMR) in pediatric rheumatic conditions. CMR may be complementary to echocardiogram as it assesses edema and fibrosis, as well as right ventricular ejection fraction (RV EF). We assessed additional cardiac inflammation/damage data gleaned from children with SLE and SSc during times of cardiac symptomatology.

Methods: We performed an IRB-approved query of our CMR database (patients 2014-2022) and abstracted data on 11 children (16 CMRs) with SLE-spectrum or SSc. We recorded echocardiographic findings from the most recent echo prior to CMRs. Echo parameters included tricuspid regurgitation (TR; jet >2.5 m/sec), ventricular dysfunction (LV and RV), and

Table 1. Patient characteristics at time of CMR

Patient	Diagnosis	Age (years)	Sex	Ethnicity	SLEDAI score	SSc manifestations	Disease duration (years)
1	SSc with myositis	15	F	Hispanic		Cardiac, GI, ILD	0
1b		15				Cardiac, GI, ILD	0.5
2	SSc/JDM overlap	10	F	African American		Cardiac, GI, PH	2
3	SSc/JDM overlap	13	F	African American		Cardiac, GI, PH	0
4	SSc	18	F	Hispanic		Cardiac, GI, ILD	5
5	SSc/SLE overlap	13	F	Hispanic	7	Cardiac, ILD, PH	0
6	JDM/SLE overlap	17	F	Hispanic	14		10.5
7	SLE	18	M	Hispanic	1		4
8	SLE	18	F	Hispanic	10		9
9a	SLE	13	F	Hispanic	8		1
9b		14			4		2
9c		15			4		3
9d		16			4		4
9e		17			22		5
10	MCTD	17	F	Hispanic	4		1.5
11	MCTD with APS	16	F	Hispanic	6		1

SSc=systemic sclerosis, JDM=juvenile dermatomyositis, SLE=systemic lupus erythematosus, APS=antiphospholipid syndrome, MCTD=mixed connective tissue disease, GI=gastrointestinal, ILD=interstitial lung disease, PH=pulmonary hypertension

pericardial effusion (PCE). CMR evaluated LV and RV function, myocardial fibrosis, myo/pericarditis, and PCE. We used STIR and T2 map sequences to determine edema indicative of myo/pericarditis and delayed enhancement sequence for fibrosis assessment. SLEDAI score was used to assess patient disease activity at the time of CMR.

Results: Patients median age at time of CMR (n=16 scans) was 16 years (range=10-18), and median disease duration was 1.5 years (range=0-10). 82% were female, 82% were Hispanic/Latino, and 18% were Black/African American. Median SLEDAI score of patients with SLE-spectrum disease at time of CMR was 7 (range=1-14). Two of the seven patients with SLE-spectrum disease (28.5%) had renal manifestations at time of CMR, and none had CNS disease. Among the five patients with SSc, three (60%) had interstitial lung disease (ILD), three (60%) had pulmonary hypertension (PH), and four (80%) had GI involvement (Table 1). Thirteen (81%) of the 16 echocardiograms done prior to CMR demonstrated significant abnormality (Table 2). CMRs were performed in three other patients with normal echos, one of whom who was receiving PH therapy, one with cardiac symptoms and elevated cardiac enzyme, and one with abnormal T waves concerning for myocarditis. Additional cardiac abnormalities were identified on CMR in seven (44%) of the scans: one (6%) with edema, four (25%) with fibrosis, one (6%) with LV/RV dysfunction not seen on echo, and one (6%) with RV dysfunction not seen on echo. LV function was worse on CMR as compared to echo in 11 (79%) of the cases (Table 2). CMRs led to escalation in immunomodulatory therapy in all three patients with edema or fibrosis on CMR. Of the patients with abnormalities on CMR, one showed resolution of fibrosis on subsequent imaging, and the others had stable depressed function on repeat echo or CMR.

Table 2. Echo findings vs. CMR findings

Patient	Echo findings				CMR findings				
	LV EF (normal ≥55%)	RV function	TR jet > 2.5 m/s	PCE	LV EF (normal ≥55%)	RV EF (normal ≥48%)	Edema	Fibrosis	PCE
1	Moderately to severely depressed	Moderately depressed	No	None	31%	35%	No	Yes	Trivial
1b	Severely depressed	Moderately depressed	No	None	30%	33%	No	Yes	None
2	70%	Mildly depressed	Yes	None	70%	55%	No	Equivocal	None
3	47%	Moderately depressed	Yes	Small	62%	43%	No	No	Small
4	70%	Normal	Yes	None	64%	70%	No	No	None
5	59%	Normal	No	None	47%	45%	No	No	Moderate
6	44%	Normal	No	Small	35%	58%	No	Yes	Small
7	67%	Normal	No	None	61%	58%	No	No	None
8	57%	Low normal	Yes	Small	48%	49%	No	No	None
9a	60%	Normal	Yes	Small to moderate	65%	59%	Yes	No	None
9b	65%	Normal	Yes	None	58%	62%	No	Yes	None
9c	57%	Normal	No	None	54%	51%	No	No	None
9d	59%	Normal	Yes	None	54%	51%	No	No	None
9e	64%	Normal	Yes	None	54%	51%	No	No	None
10	66%	Normal	No	None	55%	53%	No	No	None
11	69%	Normal	No	None	64%	61%	No	No	None

LV EF= left ventricular ejection fraction, RV EF=right ventricular ejection fraction, TR=tricuspid regurgitation, PCE=pericardial effusion

Conclusion: CMR performed in children with SLE and SSc revealed cardiac edema and fibrosis as well as lower LV/RV function than detected on echocardiography. Findings of cardiac fibrosis and/or edema led to a change in care in a third of patients. Future directions may include assessing CMR as an early screening tool, especially in patients with SSc who may have rapidly progressive and at times fatal cardiac manifestations.

Disclosure: M. Rae: None; **T. Doan**: None; **E. Muscal**: sobi, 1.

Abstract Number: 030

International Validation of the Total Morbidity Score for Juvenile Localized Scleroderma: 2023 Update

Christina ZIgler[1], Debra Henke[2], Clare Pain[3], Hanna Lythgoe[3], Kaveh Ardalan[2], Kathryn Torok[4] and Suzanne Li[5], [1]Duke, Durham, NC, [2]Duke University School of Medicine, Durham, NC, [3]Alder Hey Children's NHS Foundation Trust, Liverpool, United Kingdom, [4]University of Pittsburgh, Pittsburgh, PA, [5]Hackensack Meridian School of Medicine, Joseph M. Sanzari Children's Hospital, Hackensack, NJ

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 4 - JDM & Scleroderma: Clinical & Therapeutic Aspects
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: Juvenile localized scleroderma (jLS) is a rare condition causing inflammation and fibrosis that may impair health-related quality of life (HRQoL). Recent studies demonstrate extracutaneous manifestations (ECMs) are common and associated with poorer HRQoL, but current measures fail to capture this complexity. The Total Morbidity Score (TMS) is a weighted measure that scores both cutaneous damage and ECMs, providing a total score for disease morbidity.[1] Before this score can be used in clinical settings, there is a critical need for coordinated, international partnerships to obtain consensus on the weighting scheme within certain modules.

Methods: A virtual, modified Delphi process is being utilized to refine the weighting scheme for the Head and Neurological (HN) and the other organ (OO) modules of the TMS, building upon prior work.[1] Experienced jLS rheumatologists and dermatologists were recruited from CARRA, PReS, and PRINTO, and two of 3 planned survey rounds administered in REDCap. In the first round, clinicians were asked to rank the items within each module in terms of severity (5 – extremely severe to 1 – not severe). In the second round, clinicians were presented with the results from round 1 and asked to confirm the severity classifications for items within the modules. Consensus was defined a priori as >70% clinician agreement.

Results: Eighteen clinicians (12 female) representing 13 countries completed the first survey with 15 also completing round 2. All items reached consensus on severity rankings except for constipation (OO module; with 5/15 disagreement with classification as 'moderate). Reasons provided by clinicians for disagreement on this item included the high frequency of constipation in the non-jLS population, the challenge of attributing this symptom specifically to jLS, and the variation in severity. Also in round 2, participants were asked what additional information would help them determine the severity level (severe, mild, moderate) for the following items: seizure (history of, intractable), brain imaging abnormality in white/grey matter associated with symptoms, vision changes, and headaches/migraines. These responses will be used in round 3 to determine if greater clarity regarding ECM descriptions is needed when scoring/weighing these items on the TMS.

Table 1. Frequency and percent agreement for severity rankings by 15 clinicians responding to second round of Delphi survey.

Table 1. Frequency and percent agreement for severity rankings by 15 clinicians responding to second round of Delphi survey.

Head & Neurological Module – Items Ranked Severe	Agree *n* (%)
1. Brain imaging abnormality in white/grey matter associated with symptoms	15 (100)
2. Severe hemifacial atrophy, facial distortion (ex: obvious underdevelopment or atrophy of the hemi-face with deviation of face towards affected side for nose, mouth, asymmetry of eyes)	15 (100)
3. Brain imaging vascular abnormality associated with symptoms	15 (100)
4. Electroencephalogram (EEG) abnormality with associated seizure symptoms	15 (100)
5. Seizure (History of seizure, Intractable seizure)	15 (100)
6. Movement disorders, myotonia	15(100)
7. Peripheral neuropathy: pain, spasm, palsy	15 (100)
8. Moderate hemifacial atrophy, facial distortion (between mild and severe)	13 (86.7)
9. Neuropsychiatric changes: Cognitive, personality, mood/behavior	12 (80)
10. Vision changes	14 (93.3)
Head & Neurological Module – Items Ranked Moderate	
11. Eye inflammation (Episcleritis, Keratitis, Uveitis)	12 (80)
12. Malocclusion of the jaw	12 (80)
13. Enophthalmos	13 (86.7)
14. Tooth root abnormality, Missing tooth/teeth, Atrophic gums	13 (86.7)
15. Temporomandibular joint (TMJ) pain, arthralgia, arthritis	14 (93.3)
16. Mild hemifacial atrophy, facial distortion (ex: narrowing/ underdevelopment of affected side of face (globally or region forehead to jaw line), but no difference in teeth/jaw alignment, eye or nose position)	14 (93.3)
17. Headaches/migraines	11 (73.3)
18. Eye dryness, lacrimal, duct issues, Keratoconjunctivitis sicca	13 (86.7)
Head & Neurological Module – Items Ranked Mild	
19. Hair loss at the scalp	12 (80)
20. Hair loss on the face (eyebrow, eyelashes)	13 (86.7)
21. Electroencephalogram (EEG) WITHOUT associated seizure symptoms	13 (86.7)
22. Brain imaging vascular abnormality NOT associated with symptoms	13 (86.7)
23. Brain imaging abnormality in white/grey matter NOT associated seizure symptoms	11 (73.3)
24. Tongue hemiatrophy	13 (86.7)
25. Dry mouth	14 (93.3)
Other Organ Module – Items Ranked Severe	
1. Abnormal pulmonary function tests: (Forced Vital Capacity (FVC) </= 80%, Diffusing Capacity of the Lungs for Carbon Monoxide (DLCO) </= 80%)	14 (93.3)
2. Dyspnea	15 (100)
3. Digital ulcer	15 (100)
4. Dysphagia	15 (100)
Other Organ Module – Items Ranked Moderate	
5. Vasculitic rash, Livedo reticularis	14 (93.3)
6. Gastroesophageal reflux disease (GERD)	13 (86.7)
7. Raynaud's phenomena	13 (86.7)
8. Diarrhea	12 (80)
9. Constipation	10 (66.7)

Conclusion: In survey round 2, all items but one reached pre-specified thresholds for consensus on severity rankings. A third, final round of the modified Delphi process is currently underway to operationalize the TMS scoring approach, including item weightings and additional categories for certain items (e.g. seizures). Refined weighting of TMS HN and OO domain subscores will enhance the utility of this measure for clinicians, researchers, and international collaborations.

References

1. Li S, Patel A, Pope E, … Laxer R. Capturing the Range of Disease Involvement in Localized Scleroderma: The Total Morbidity Score [abstract]. *Arthritis Rheumatol*. 2020;72.

Disclosure: C. Zigler: None; **D. Henke**: None; **C. Pain**: None; **H. Lythgoe**: None; **K. Ardalan**: None; **K. Torok**: None; **S. Li**: Merck/MSD, 11.

Abstract Number: 031

Multisystem Inflammatory Syndrome in Children Phenotypes Vary Between SARS-CoV-2 Variants

Greta Mastrangelo[1], Ellen Go[2], Paul Tsoukas[2], Hua Lu[3], Amy Xu[2], Arthur Hoi Hin Cheng[2] and Rae Yeung[4], [1]The Hospital of Sick Children,Department of Paediatrics, University of Toronto, Division of Rheumatology, Toronto, ON, Canada, [2]The Hospital of Sick Children, Department of Paediatrics, University of Toronto, Division of Rheumatology; Cell Biology Program, The Hospital for Sick Children Research Institute, Toronto, ON, Canada, [3]The Hospital for Sick Children, Toronto, ON, Canada, [4]The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Poster Breakout 5 - Autoinflammatory/Vasculitis: STAT3, NLRP3, KD and MIS-C

Session Type: Abstract Session

Session Time: 4:30PM–5:00PM

Background/Purpose: Multisystem Inflammatory Syndrome in Children (MIS-C) is a serious complication associated with COVID-19, presenting as a hyperinflammatory disorder characterized by fever and multiorgan dysfunction. Whether the MIS-C phenotype varies accordingly to the SARS-CoV-2 variants is still unclear. We aim to compare MIS-C clinical features,treatments, and outcomes across the various waves of COVID-19 dividing the patient population into three cohorts according to the pre-Delta, Delta, and Omicron MIS-C waves. Our secondary objective is to evaluate if the clinical phenotype (shock, Kawasaki Disease (KD), fever with hyperinflammation) varies across the three cohorts.

Methods: Prospective cohort study of 252 patients with MIS-C, at a tertiary care pediatric center from March 2020 to March 2022. Clinical and laboratory features, complications,outcomes, and treatments were evaluated. The association with SARS-CoV-2 variants and MIS-C cohorts was assumed based on local epidemiology and sequencing data, representing the predominant strain across the three-time periods. The starting date of each MIS-C wave for study purposes was set at two weeks after the first case of COVID-19 from that respective variant in the community, as the actual time lag for developing MIS-C iswithin 2-6 weeks after the acute infection. Descriptive statistics were performed to assess differences between the 3 MIS-C cohorts and clinical phenotypes.

Results: Of the 252 patients (150 with pre-Delta variants, 59 with Delta, 43 with Omicron), the median age was 5.2 years, 58.7% were male, and 50.0 % had SARS-CoV-2 exposure. The 3 cohorts showed a significant difference in MIS-C phenotypes distribution (p=0.003). Fever and hyperinflammation was the predominant phenotype (20%) in the pre-Delta cohort; shock represented the majority (39%) in the Delta cohort; and the KD phenotype was prevalent (67%) in the Omicron cohort. Cardiac and gastrointestinal involvements were the most common features in all the cohorts, whereas, neurological involvement was the least prevalent. The Omicron cohort had more mucocutaneous involvement compared to the others. The main difference between the variant waves was reflected in measures of complications and outcome with the MIS-C cohort associated with the Delta variant capturing the most severe phenotype with a higher incidence of shock (39%), MAS (22%), andPICU admission (34%). The proportion of children developing coronary artery lesions was similar in all groups. Among all the 3 MIS-C cohorts, the majority of patients received either IVIG alone or together with upfront steroids. Pulsed high-dose steroids and anticoagulation therapy were more commonly used among children in the Delta MIS-C cohort, findings in keeping with the prevalence of the MIS-C shock phenotype in this group.

Conclusion: The MIS-C phenotype varies accordingly to the SARS-CoV-2 variants, and patients with the Delta variant have a more severe phenotype with a greater proportion of complications. These findings provide new insights into disease phenotype and SARS-CoV-2 variants and may have important implications for diagnosis and management.

Disclosure: G. Mastrangelo: None; **E. Go**: None; **P. Tsoukas**: None; **H. Lu**: None; **A. Xu**: None; **A. Hoi Hin Cheng**: None; **R. Yeung**: None.

Abstract Number: 032

From Bedside to Bench and Back: Discovery of a Novel Missense Variant in NLRP3 Causing Atypical Cryopyrin-Associated Periodic Syndromes with Hearing Loss as the Primary Presentation, Responsive to Anti-IL1 Therapy

Merav Birk-Bachar[1], Hadar Cohen[2], Yoel Levinsky[3], rotem tal[4], Gil Amarilyo[5], Meirav Sokolov[6], Efrat Sofrin-Drucker[7], Naama Orenstein[7], Gabriel Lidzbarsky[7], Liora Kornreich[8], Eyal Raveh[6], Nesya Kropach-Gilad[7], Motti Gerlic[2] and Liora Harel[9], [1]Pediatric Rheumatology Unit, Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [2]Sackler Faculty of Medicine, Immunology Department, Tel Aviv University, Tel Aviv, Israel, [3]Schneider Children's Medical Center of Israel, Tel Aviv University, Petach Tikva, Israel, [4]Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [5]Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [6]Pediatric Ear Nose and Throat Unit Unit, Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [7]Recanati Genetic Institute, Rabin Medical Center-Beilinson Hospital, Petach Tikva, Israel, [8]Pediatric Imaging Unit, Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [9]Scheiders Children Medical Center of Israel, Petah-Tiqva, Israel

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Poster Breakout 5 - Autoinflammatory/Vasculitis: STAT3, NLRP3, KD and MIS-C

Session Type: Abstract Session

Session Time: 4:30PM–5:00PM

Background/Purpose: Cryopyrin-associated periodic syndromes (CAPS) are a spectrum of rare autoinflammatory diseases caused by gain-of-function mutations inthe NLRP3 gene. These mutations cause inflammasome hyperactivity and subsequent uncontrolled release of Interleukin-1β (IL-1β). CAPS patients may develop progressive sensorineural hearing loss (SNHL) due to cochlear autoinflammation, initially affecting the high-ultrahigh frequency ranges. In rare instances this may be the sole presenting symptom. A Jewish Ashkenazi family presented with autosomal dominant SNHL, a novel missense variant of unknown significance in the NLRP3 gene 001079821:c.1790G >A p.Ser597Asn, however they did not fulfill CAPS classification criteria. In order to establish the suspected diagnosis of atypical CAPS causing SNHL, and enable initiation of Anti-IL1 therapy, we performed functional studies of the inflammasome to evaluate whether the variant leads to inflammasome hyperactivity.

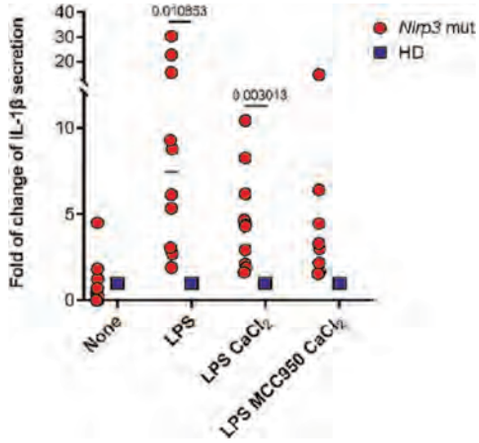


Figure 1. Ex Vivo NLRP3 Inflammasome Functional Assessment

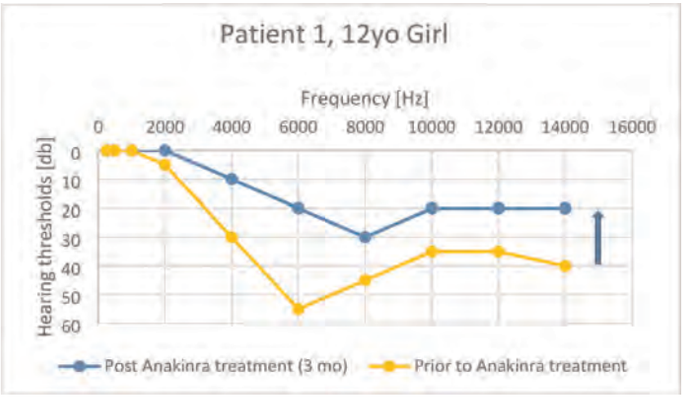


Figure 2. Pure-tone audiometry Pre and Post Anakinra Treatment

Methods: Our prospective study included 15 family members (10 known carriers, 5 age-group matched non-carriers who served as controls). We conducted clinical and hearing assessments along with ex-vivo functional studies of inflammasome activity. Secreted levels of IL-1β from peripheral-blood mononuclear cells (PBMCs) in carriers versus controls, were measured under 3 main conditions: basal state, in response to priming signal alone (LPS- lipopolysaccharide or LPS+$CaCl_2$), and in response to priming signal along with specific inflammasome inhibitor (MCC950).

Results: Of the 10 known carriers spanning 3 generations, 7 suffered from SNHL. Although most presented progressive deterioration classically associated with CAPS, some suffered from sudden episodes of unilateral SNHL as well. The severity of hearing impairment ranged from moderate, involving the high-ultrahigh frequency, to severe hearing loss requiring cochlear implant. Functional assessment of the inflammasome in carriers vs. non carriers (controls) revealed that although basal levels of secreted IL-1β were not significantly different, PBMCs isolated from NLRP3 variant carriers primed with either LPS or LPS+CaCl2 showed significantly higher secretion of IL-1β than healthy subjects. Inflammasome inhibition with MCC950 prior to addition of LPS+CaCl2 resulted in relative suppression of IL-1β secretion (Fig. 1). This evidence of NLRP3 inflammasome hyperactivity confirmed the suspected diagnosis of atypical CAPS. Indeed, administration of Anakinra therapy resulted in substantial clinical improvement, particularly among pediatric patients who exhibited near resolution of hearing impairment within 1-3 months of treatment (Fig. 2).

Conclusion: Our study revealed a novel pathogenic variant in NLRP3, causing atypical CAPS with a primary presentation of SNHL- responsive to Anti IL1 therapy. These findings highlight the crucial role of early diagnosis and subsequent treatment in reversing cochlear damage. Auditory assessment of the high and ultrahigh frequency range is important and can detect early subclinical SNHL. Incorporating functional inflammasome assessment as part of the clinical evaluation could establish the diagnosis in inconclusive cases.

Disclosure: M. Birk-Bachar: None; **H. Cohen**: None; **Y. Levinsky**: None; **r. tal**: None; **G. Amarilyo**: None; **M. Sokolov**: None; **E. Sofrin-Drucker**: None; **N. Orenstein**: None; **G. Lidzbarsky**: None; **L. Kornreich**: None; **E. Raveh**: None; **N. Kropach-Gilad**: None; **M. Gerlic**: None; **L. Harel**: None.

Abstract Number: 033

Kawasaki Disease (KD) Criteria Fulfillment and Associated Outcomes in Multisystem Inflammatory Syndrome in Children (MIS-C)

Lyndsey Cole, Marsha Anderson, Heather Heizer, Michelle Hite, Christina Osborne, Samuel Dominguez and Pei-Ni Jone, University of Colorado School of Medicine, Denver, CO

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 5 - Autoinflammatory/Vasculitis: STAT3, NLRP3, KD and MIS-C
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: Multisystem inflammatory syndrome in children (MIS-C) is a hyperinflammatory illness associated with SARS-CoV-2 infection and has overlapping features with Kawasaki Disease (KD). The objective was to describe fulfillment of complete and incomplete KD criteria among patients with MIS-C, and to compare features at admission and clinical outcomes among patients with MIS-C who did or did not meet KD criteria.

Methods: Single pediatric hospital retrospective chart review of all patients who met U.S. Centers for Disease Control and Prevention (CDC) MIS-C criteria and received treatment between April 2020 to February 2021.

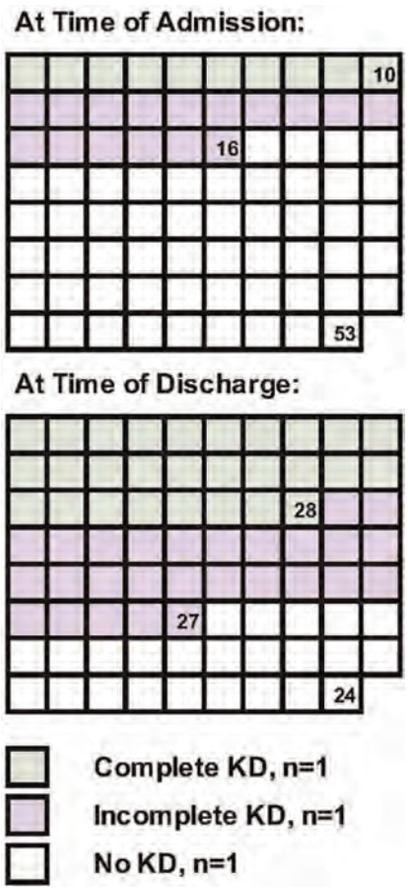


Figure 1: Patients with MIS-C Meeting KD Criteria

Table 1: Characteristics and Outcomes Among Patients with MIS-C Based on KD Criteria

	Comparison by Fulfillment of KD Criteria at Admission			Comparison by Later Fulfillment of KD Criteria Among Patients Without KD at Admission		
	Criteria met at Admission (n=26)	Criteria not met at Admission (n=53)	P-Value	Criteria met later in hospital stay (n=29)	Criteria never met (n=24)	P-value
Features at Admission, No. (%)						
Age (mean in years)	8.5	9	0.61	8.5	9.5	0.55
Day of illness (mean)	7	5	<0.001	4	5	0.45
SARS-CoV-2 PCR positive	11 (42%)	19 (36%)	0.58	9 (31%)	10 (42%)	0.42
SARS-CoV-2 immunoglobulin G positive	24 (92%)	46 (87%)	0.24	25 (86%)	21 (88%)	>1
KD Clinical Criteria						
Fever ≥ 5 days	26 (100%)	24 (45%)	<0.001	13 (45%)	11 (46%)	0.94
Rash	22 (85%)	30 (57%)	0.01	20 (69%)	10 (42%)	0.046
Mucous membrane changes	19 (73%)	21 (40%)	0.005	14 (48%)	7 (29%)	0.16
Conjunctivitis	26 (100%)	30 (57%)	<0.001	18 (62%)	12 (50%)	0.38
Extremity changes	16 (62%)	16 (30%)	0.008	10 (34%)	6 (25%)	0.45
Cervical lymph node ≥ 1.5 cm	1 (4%)	1 (2%)	0.89	1 (3%)	0 (0%)	>1
KD Lab Criteria						
Anemia for age	11 (42%)	13 (25%)	0.11	7 (24%)	6 (25%)	0.94
Albumin ≤ 3.0 g/dL	20 (77%)	14 (27%); n=51	<0.001	12 (43%); n=28	2 (9%); n=23	0.007
Elevated alanine transaminase (ALT)	20 (77%)	30 (59%); n=51	0.12	23 (82%); n=28	7 (30%); n=23	<0.001
White Blood Cells ≥ 15,000/mm^3	2 (8%)	6 (11%)	0.47	4 (14%)	2 (8%)	0.86
Platelets ≥ 450,000 after fever 7 days	1 (7%); n=15	0 (0%); n=0	>1	0 (0%); n=2	0 (0%); n=3	>1
Pyuria	9 (47%); n=19	7 (18%); n=40	>1	4 (20%); n=20	3 (15%); n=20	0.80
Outcomes, No. (%)						
LAD and/or RCA Z Score 2.0-2.49	3 (12%)	10 (19%); n=52	0.30	7 (24%)	3 (13%); n=23	0.92
LAD and/or RCA Z Score ≥ 2.5	8 (31%)	11 (21%); n=52	0.35	8 (28%)	3 (13%); n=23	0.95
Left ventricular dysfunction (EF <55%)	16 (62%)	24 (46%); n=52	0.20	16 (55%)	8 (35%); n=23	0.14
Need for intensive care	19 (73%)	29 (55%)	0.12	22 (76%)	7 (29%)	<0.001
Vasoactive medication requirement	12 (46%)	19 (36%)	0.38	18 (62%)	1 (4%)	<0.001
Need for second-line therapy	9 (35%)	24 (45%)	0.37	19 (66%)	5 (21%)	0.001
Need for third-line therapy	1 (4%)	8 (15%)	0.13	8 (28%)	0 (0%)	0.005
Respiratory failure	8 (31%)	15 (28%)	0.82	12 (41%)	3 (13%)	0.02

ABBREVIATIONS: PCR, polymerase chain reaction; LAD, left anterior descending coronary artery; RCA, right coronary artery; EF, ejection fraction

Results: 79 patients with MIS-C were identified. 55/79 patients (70%) met criteria for KD at admission (n=26) or later in the hospitalization (n=29). Of those, 28/55 (51%) met criteria for complete KD and 27/55 (49%) for incomplete KD. Patients meeting KD criteria at admission presented later (mean day 7 vs. 5, P< 0.001) and were more likely to have rash, mucous membrane changes, conjunctivitis, extremity changes, and hypoalbuminemia at admission compared to those who did not (P=0.01, 0.005, < 0.001, 0.008, and < 0.001, respectively). There were no differences in coronary artery lesions (CAL), left ventricular (LV) dysfunction, respiratory failure, or needs for intensive care, vasoactive medications, or second- or third-line MIS-C therapy. Patients who did not meet KD criteria at admission but did later in the hospitalization were more likely to have rash, hypoalbuminemia, and elevated alanine transaminase at admission (P=0.05, 0.007, and < 0.001, respectively), respiratory failure (P=0.02) and had greater need for intensive care, vasoactive medications, and second- and third-line therapy than patients never meeting KD criteria (P< 0.001, < 0.001, 0.001, and 0.005, respectively). There were no differences in CAL or LV dysfunction.

Conclusion: Most patients with MIS-C met criteria for KD by time of hospital discharge. Fulfillment of KD criteria at admission was not associated with changes in MIS-C patient outcomes, though fulfillment of KD criteria later during hospitalization was associated with greater need for intensive care, vasoactive medications, and second- and third-line MIS-C therapy. KD criteria fulfillment in patients with MIS-C at any timepoint was not associated with increased risk of CAL or LV dysfunction. Further investigation is needed to understand how pathophysiology of MIS-C compares to KD and if presence of KD features can help predict outcomes in MIS-C.

Disclosure: L. Cole: None; **M. Anderson**: None; **H. Heizer**: None; **M. Hite**: None; **C. Osborne**: Biofire, 5; **S. Dominguez**: Biofire, 2, 5, Karius, 2, Pfizer, 5; **P. Jone**: None.

Abstract Number: 034

STAT3 Gain-of-function Syndrome Mutations Are Susceptible to JAK Inhibition Despite a Spectrum of Potency

Herda Ona[1], Justin Branch[2], Isabella Osuna[1], Priscilla vasquez[2], Anaid Reyes[1], Phillip Baker[1], Michael Clowers[1], Stephanie Wood[1] and Tiphanie Vogel[1], [1]Baylor College of Medicine, Houston, TX, [2]Baylor College of Medicine, Texas Children's Hospital, Houston, TX

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 5 - Autoinflammatory/Vasculitis: STAT3, NLRP3, KD and MIS-C
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: STAT3 gain-of-function (GOF) syndrome is a rare inborn error of immunity that leads to early-onset lymphoproliferation, autoimmune cytopenias, multi-solid organ autoimmunity, hypogammaglobulinemia, and short stature. Approximately one-fifth of individuals with STAT3 GOF develop inflammatory arthritis. STAT3 GOF syndrome results from heterozygous variants in*STAT3*, an important transcription factor in numerous immune and non-immune cytokine signaling pathways. Investigating naturally occurring variants in *STAT3* can shed light on numerous cellular processes given the critical roles for STAT3 in development, differentiation and regulation.

Methods: STAT3 GOF was determined using a standardized luciferase assay. Inhibition was assessed in patient-derived cell lines by reduction of*SOCS3*transcript, as determined by quantitative real time PCR, in the presence of an inhibitor following stimulation with interleukin-21.

Results: There are 211 patients with STAT3 GOF syndrome caused by 84 unique mutations spanning the 6 domains of the molecule. There are 70 patients with 37 different mutations in the DNA binding domain, although the 3 most common mutations are in the coiled-coil (R152W) and transactivation (P715L and T716M) domains. The average relative potency of STAT3 GOF mutations at baseline is 28.5-fold above the transcriptional capacity of wild-type (WT) STAT3; however the range is 1.1- to 99.7-fold higher. Mutations that are not GOF at baseline, are GOF with cytokine stimulation. Unlike STAT3 mutations associated with the primary immunodeficiency autosomal-dominant hyper-IgE syndrome, STAT3 GOF mutations are not dominant to WT. Intriguingly, some, but not all, STAT3 GOF mutations do not require phosphorylation at the canonical tyrosine 705 amino acid to drive transcription at higher than WT rates. Other STAT3 GOF mutations appear to be tempered by serine 727 phosphorylation. All tested STAT3 GOF mutations were effectively inhibited by the JAK inhibitors ruxolitinib, tofacitinib, and/or baricitinib. The direct STAT3 inhibitor S3I-201 reduced*SOCS3* transcription by approximately 50% in control cells, and also in several of the STAT3 GOF cell lines tested.

Conclusion: We determined the relative potency of all known disease-associated STAT3 GOF variants. Fortunately, JAK inhibitors are effective against all tested STAT3 GOF variants and should be considered for targeted treatment of patients with STAT3 GOF syndrome. Work to elucidate the cellular mechanisms of STAT3 GOF variants is ongoing.

Disclosure: H. Ona: None; **J. Branch**: None; **I. Osuna**: None; **P. vasquez**: None; **A. Reyes**: None; **P. Baker**: None; **M. Clowers**: None; **S. Wood**: None; **T. Vogel**: Moderna, 2, Novartis, 2, Pfizer, 2, SOBI, 2.

Abstract Number: 035

SARS-CoV-2 Vaccination of Children with a History of Multisystem Inflammatory Syndrome

Mariana Sanchez Villa[1], Matthew Wisniewski[1], Jessica Nguyen[1], Eyal Muscal[1], Marietta Deguzman[2], Sara Kristen Sexson Tejtel[1], Sridevi Devaraj[1], Flor Munoz-Rivas[1], Leila Sahni[1] and Tiphanie Vogel[1], [1]Baylor College of Medicine, Houston, TX, [2]Baylor College of Medicine, Texas Children's Hospital, Houston, TX

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 5 - Autoinflammatory/Vasculitis: STAT3, NLRP3, KD and MIS-C
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: Most children who contract SARS-CoV-2 are asymptomatic or mildly symptomatic, but a subset subsequently develop the hyperinflammatory condition called multisystem inflammatory syndrome in children (MIS-C). There has been hesitation to vaccinate children with a history of MIS-C against SARS-CoV-2 over concerns that hyperinflammation may recur. As part of our post-MIS-C follow-up care, we have advocated for COVID-19 vaccination. We aimed to determine the frequency of COVID-19 vaccination among the cohort of patients with a history of MIS-C diagnosed at our institution.

Methods: Patients who presented May 2020-October 2022 with an acute febrile illness that fulfilled the 2020 Centers for Disease Control and Prevention MIS-C case definition were included. During outpatient subspecialty care following hospitalization, patients with a history of MIS-C were counseled on and encouraged to receive COVID-19 vaccination after 90 days from discharge. Charts were retrospectively reviewed to identify patients vaccinated against SARS-CoV-2. COVID-19 vaccine findings were confirmed using a state immunization registry.

Results: Vaccines were reviewed for 294 of 295 patients diagnosed with MIS-C: one patient died prior to discharge. 99 of 294 patients (34%) received at least one dose of COVID-19 vaccine after MIS-C diagnosis. Vaccinated patients were 58% male, and initiated vaccination at a mean of 10.8 years of age (range 3-19 years) and at 8.8 months post-MIS-C hospitalization (range 20 days-24.4 months). 90 of 99 patients are partially vaccinated: 13 received one dose of vaccine, 60 received 2 doses, and 17 received 3 or more doses of monovalent COVID-19 vaccine. 9 are fully vaccinated, including one 3-year-old who completed a 3-dose primary series and 3 patients who had 3 doses of monovalent vaccine prior to a bivalent booster.All patients received mRNA vaccine; 99% of doses (193 of 194) were BNT162b2. No patients have re-presented with a recurrence of MIS-C or any other hyperinflammatory condition over a mean of 11.3 months of follow-up since last vaccination (range 18 days-23.1 months). In our cohort, 15 of 295 patients (5%) received a COVID-19 vaccination at a time prior to the onset of MIS-C; all but one had evidence of prior or current SARS-CoV-2 infection at MIS-C presentation, either by nucleic acid amplification and/or nucleocapsid serology. 7 of these 15 patients have received subsequent doses of COVID-19 vaccine, and none have reported recurrence of hyperinflammation (mean 8.2 months since last dose, range 2.8-13.8 months).

Conclusion: SARS-CoV-2 vaccination is well-tolerated by children with a history of MIS-C. This is reassuring as SARS-CoV-2 becomes endemic and annual vaccination against SARS-CoV-2 is considered. Work is in progress to prospectively monitor patients with a history of MIS-C for vaccine reactogenicity and immune activation following SARS-CoV-2 vaccination.

Disclosure: M. Sanchez Villa: None; **M. Wisniewski**: None; **J. Nguyen**: None; **E. Muscal**: sobi, 1; **M. Deguzman**: None; **S. Sexson Tejtel**: sobi, 2; **S. Devaraj**: None; **F. Munoz-Rivas**: Moderna, 1, Pfizer, 1, 5; **L. Sahni**: None; **T. Vogel**: Moderna, 2, Novartis, 2, Pfizer, 2, sobi, 2.

Abstract Number: 036

Predictive Factors of Long-lasting Remission Following Anakinra Withdrawal in Patients with Systemic Juvenile Idiopathic Arthritis After Achievement of Clinical Inactive Disease

Germana Nardini[1], **Claudia Bracaglia**[2], Denise Pires Marafon[1], Emanuela Sacco[3], Arianna De Matteis[1], Ivan Caiello[1], Giusi Prencipe[1], Fabrizio De Benedetti[2] and Manuela Pardeo[2], [1]Division of Rheumatology, IRCCS Ospedale Pediatrico Bambino Gesu', Roma, Italy, [2]Division of Rheumatology, IRCCS Ospedale Pediatrico Bambino Gesù, Roma, Italy, [3]Fondazione Casa Sollievo della Sofferenza, Pediatria, San Giovanni Rotondo, Italy

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Poster Breakout 6 - Systemic JIA: Genomics, Transcriptomics & Disease States

Session Type: Abstract Session

Session Time: 4:30PM–5:00PM

Background/Purpose: Systemic juvenile idiopathic arthritis (sJIA) is a rare autoinflammatory disease of unknown etiology. Several uncontrolled studies showed that early treatment with anakinra is associated with a better outcome, according to the "window of opportunity" hypothesis. However, little evidence is available on withdrawal strategy. Anakinra withdrawal modalities are still heterogeneous among the different rheumatology centres. The aim of this study was to describe our strategy for anakinra withdrawal in sJIA patients following achievement of clinical inactive disease (CID) and to evaluate the association between clinical variables at baseline, including disease duration, and persistence of remission.

Methods: We retrospectively analysed data of 39 sJIA patients followed in our centre who withdrew anakinra after reaching CID off glucocorticoids for at least 6 months before withdrawal. Eight patients withdrew anakinra abruptly while 31 patients did it after tapering throughout one or two steps (alternate-day and/or once every three days regimens) of anakinra administration. All patients underwent a 24 months follow up after withdrawal. They were subsequently divided into two groups according to presence or absence of disease flare during the follow up. Demographic, clinical and laboratory data were evaluated in univariate and multivariate analysis as predictors of flare.

Results: During the follow up, 10/39 patients (25.6%) flared after a median time from anakinra withdrawal of 7.9 months. In univariate analysis the variable most strongly related with the absence of flare was the disease duration from the onset anakinra initiation (p = 0.0046), with an optimal cut-off of 3 months. Patients who started anakinra ≥ 3 months after disease onset had an 8-fold higher risk of flare after anakinra discontinuation (CI 95 2.0-32.4; p = 0.0003) (Figure 1). Furthermore,

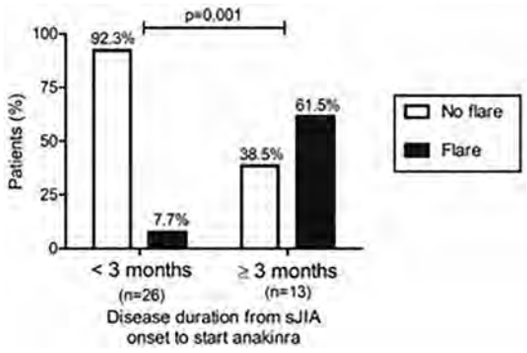


Figure 1. Relationship between disease duration from disease onset to anakinra initiation and rate of flare within the 24 months follow up after anakinra withdrawal.

we observed that a higher dose of anakinra (≥ 2 mg/kg/day) was associated with a lower risk of flare (p = 0.065). We also demonstrated that patients who tapered therapy before withdrawal were significantly associated with a lower percentage of flare compared to the ones who withdrew anakinra abruptly, with a 4-fold lower risk of flare (CI 95 1.47-10.1; p = 0.0164). In a model analysing at the same time disease duration, anakinra dose at baseline and anakinra tapering, disease duration ≥ 3 months at baseline resulted to be the only variable significantly associated with flare after anakinra withdrawal with an Odds Ratio of 15.16 (CI 95 1.7-131.9; p = 0.014).

Conclusion: Our results strengthen the evidence on IL-1 inhibition in sJIA treatment and provide new ones supporting the "window of opportunity" hypothesis. Early anakinra treatment may predict a good short-term outcome and also potentially prevent the development of a persistent disease course. These observations have a strong clinical relevance, an early diagnosis and a targeted treatment could control the disease and could also modify its natural history.

References:

- Nigrovic PA. Review. Arthritis Rheumatol 2014; 66:1405-13.
- Pardeo M, et al. Arthritis Rheumatol 2021; 73:1053-1061.

Disclosure: G. Nardini: None; **C. Bracaglia**: Sobi, 2, 6; **D. Pires Marafon**: None; **E. Sacco**: None; **A. De Matteis**: None; **I. Caiello**: None; **G. Prencipe**: None; **F. De Benedetti**: AbbVie/Abbott, 2, Novartis, 2, Novimmune, 2, Pfizer, 2, Roche, 2, Sobi, 2; **M. Pardeo**: SOBI, 2, 6.

Abstract Number: 037

Transcriptional Analysis of CD14+ Monocytes During Macrophage Activation Syndrome Highlights Role for Interferons and RNA Sensing in Monocytes

Susan Canny[1], Hannah DeBerg[2], Griffin Gessay[2], Ailing Lu[3], Mary Eckert[1], Andrea La Bella[4], Susan Shenoi[5], Joyce Hui-Yuen[6], Betsy Barnes[7] and Jessica Hamerman[2], [1]Seattle Children's Hospital, Seattle, WA, [2]Benaroya Research Institute, Seattle, WA, [3]Feinstein Institutes for Medical Research, Manhasset, NY, [4]Northwell Health, Cohen Children's Medical Center, New Hyde Park, NY, [5]Seattle Children's Hospital, Seattle, WA, [6]Cohen Children's Medical Center, Northwell Health, Lake Success, New York; Center for Autoimmune, Musculoskeletal, and Hematopoietic Diseases Research, Feinstein Institutes for Medical Research, Northwell Health, Manhasset, NY, [7]Center for Autoimmune, Musculoskeletal, and Hematopoietic Diseases Research, Feinstein Institutes for Medical Research, Northwell Health, Manhasset, NY

SESSION INFORMATION

Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 6 - Systemic JIA: Genomics, Transcriptomics & Disease States
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: Macrophage activation syndrome (MAS), a form of secondary hemophagocytic lymphohistiocytosis (HLH), is a potentially fatal complication of rheumatic diseases. MAS is a dysfunctional hyperinflammatory response characterized by abnormal activation of lymphocytes and phagocytes, leading to an overproduction of inflammatory cytokines and damage to host tissues. Circulating monocytes are highly responsive to their surrounding environment, are known to exhibit phenotypic and functional changes during inflammation, and can give rise of macrophages that phagocytose immune cells. However, monocytes and macrophages have not been well-studied in MAS. MAS is most commonly associated with systemic juvenile idiopathic arthritis (sJIA). At least 10% of sJIA patients will experience an overt episode of MAS with up to 50% exhibiting signs of subclinical inflammation.

Methods: We analyzed classical CD14+ monocytes from children with active MAS (6 subjects) compared to individuals with sJIA without MAS (4 subjects) and age/sex/race matched healthy children (8 subjects) by flow cytometry and RNA sequencing (RNA-Seq). Two MAS subjects and two age/sex/race matched healthy controls were analyzed by single cell RNA sequencing (scRNA-Seq). Subjects with MAS were defined based on the 2016 classification criteria by Ravelli and colleagues as well as the ratio of ferritin to ESR. Differentially expressed genes (DEGs) were defined as those with at least 2 fold change and false discovery rate less than 0.1.

Results: We found significant upregulation of CD16 surface expression during active MAS, which rapidly reversed after treatment with systemic steroids. Our RNA-Seq data show broad transcriptional changes in CD14+ monocytes from children with active MAS, including upregulation of RNase 2 (involved in processing RNAs for the innate immune sensor TLR8) and SLAMF7 (associated with monocyte/macrophage hyperinflammation in response to interferon gamma). scRNA-Seq analyses of myeloid cells from two subjects with active MAS revealed a strong interferon signature in MAS monocytes, including enrichment for STAT1, IRF1, IFITM3, ISG15, and GBPs, and upregulation of alarmins, including S100A8, S100A9, and S100A12. We identified hemoglobin transcripts specifically in the cells from MAS subjects by scRNA-Seq, suggesting the detection of hemophagocytes in circulation. We are currently analyzing myeloid cells of additional MAS subjects using scRNA-Seq.

Conclusion: These data confirm an important role for cytokines, specifically interferons, in driving gene expression in monocytes during MAS and suggest potential targets for future therapies. Together, our data show that CD14+ monocytes have a unique transcriptional signature in MAS and support a role for interferon signaling and enhanced RNA sensing in active disease.

Disclosure: S. Canny: None; **H. DeBerg**: None; **G. Gessay**: None; **A. Lu**: None; **M. Eckert**: None; **A. La Bella**: None; **S. Shenoi**: Novartis, 2, Pfizer, 1; **J. Hui-Yuen**: None; **B. Barnes**: None; **J. Hamerman**: None.

Abstract Number: 038

HLA-DRB1*15 Alleles in Systemic Juvenile Idiopathic Arthritis with Lung Disease and Macrophage Activation Syndrome in Italy

Claudia Bracaglia[1], Manuela Pardeo[1], Maria Troiano[2], Giuseppe Testa[2], Ivan Caiello[1], Arianna De Matteis[3], Matteo Trevisan[1], Franco Locatelli[4], Marco Andreani[2] and Fabrizio De Benedetti[1], [1]Division of Rheumatology, IRCCS Ospedale Pediatrico Bambino Gesù, Roma, Italy, [2]Laboratory of Transplant Immunogenetics, Department of Haematology/Oncology, Cell and Gene, IRCCS Ospedale Pediatrico Bambino Gesù, Roma, Lazio, Italy, [3]Division of Rheumatology, IRCCS Ospedale Pediatrico Bambino Gesu', Roma, Italy, [4]Ospedale Pediatrico Bambino Gesù, Department of Hematology/Oncology, Cell and Gene Therapy, Roma, Italy

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Poster Breakout 6 - Systemic JIA: Genomics, Transcriptomics & Disease States
Session Type: Abstract Session
Session Time: 4:30PM–5:00PM

Background/Purpose: Systemic juvenile idiopathic arthritis (sJIA) is characterized by unique clinical features and it is considered as a polygenic autoinflammatory disease. Macrophage activation syndrome (MAS) and lung disease (LD) are two life threatening complications of sJIA. An association of HLA-DRB1*15 alleles with sJIA complicated by LD has been recently reported.

Methods: DNA was extracted by automatic system Qiagen EZ1 Advanced XL from blood samples collected from sJIA patients followed in a single Pediatric Rheumatology tertiary center. The AllType NGS 11-Loci Amplification Kit (Thermo Fisher, One Lambda, Canoga Park, CA) was used to amplify target DNA regions. The PCR cycling conditions were followed as suggested by the manufacturer. The kit uses one multiplexed PCR amplifying the full HLA-A/B/C/DQA1/DPA1 genes and exon 2 to 3UTR for HLA-DRB1/3/4/5/DQB/ DPB1 genes. Reads were analyzed with the HLA TypeStream VisualTM Software (Thermo Fisher, One Lambda, Canoga Park, CA), version 2.0.0. The IPD-IMGT/HLA Database release used was the 3.41. The strength of association between HLA alleles was estimated by 99% confidence intervals.

Results: Samples were collected from 98 sJIA patients, 52 females, all White Caucasian, except of 2 of African origin (1 from North-Africa and 1 from Sub-Saharan Africa) and 1 of mixed origin (Caribbean and Caucasian), with median age at disease onset of 6.9 years. The HLA-DRB1*15 allele was found in 20 (20%) out of 98 sJIA patients with a median age at disease onset of 5.4 years. These results were compared with a reference group of 1017 healthy Italian individuals, previously typed in our laboratory and representative of HLA allele frequency distribution in Italy. Eighteen, out of the 196 different HLA haplotypes observed in the patients studied, were positive for HLA-DRB1*15:01 (9.1%) compared to 94, out of 2034 (4.6%) in the healthy group with a p value of 0.006. However, using the Bonferronis correction for multiple tests, the significant association was lost (p=0.14), probably due to the small sample size.

Of those 20 HLA-DRB1*15 positive patients, 7 (35%) had refractory sJIA, as defined by Erkens (1), compared to 14 (17%) of the HLA-DRB1*15 negative patients. Five out of 20 (25%) HLA-DRB1*15 positive patients had LD compared to 2 (3%) of negative patients. Furthermore, 12 out 20 (60%) HLA-DRB1*15 positive patients had one or more MAS episode compared to 41 (52%) negative patients. Only 3 patients of the entire cohort experienced a drug adverse reaction to tocilizumab; all 3 had LD and carried the HLA-DRB1*15 allele. Notably, the lung involvement in these 3 patients developed before the drug reaction.

Conclusion: The HLA-DRB1*15 allele seems to be more frequently carried by sJIA patients compared to healthy Italian population and therefore might be identified as a potential marker of susceptibility to the disease. This allele appears to be more frequent in a subgroup of sJIA patients with early disease onset, refractory course and with lung involvement. These data are of course limited to a small population and need to be confirmed in a larger international and multiracial cohort.

Reference.

1. Erkens R. et al, Rheum Dis Clin North Am. 2021

This project has been funded by the systemic JIA Foundation.

Disclosure: C. Bracaglia: Sobi, 2, 6; **M. Pardeo**: SOBI, 2, 6; **M. Troiano**: None; **G. Testa**: None; **I. Caiello**: None; **A. De Matteis**: None; **M. Trevisan**: None; **F. Locatelli**: Sobi, 2; **M. Andreani**: None; **F. De Benedetti**: AbbVie/Abbott, 2, Novartis, 2, Novimmune, 2, Pfizer, 2, Roche, 2, Sobi, 2.

Abstract Number: 039

Towards the Definition of Cutoff Values for Disease Activity States in Systemic JIA Using the Systemic Juvenile Arthritis Disease Activity Score

Ana Isabel Rebollo-Giménez[1], Yulia Vyzhga[2], Luca Carlini[3], **Silvia Rosina**[4], Elisa Patrone[1], Maria Katsikas[5], Claudia Magalhaes[6], Dalia El-Ghoneimy[7], Yasser El Miedany[8], Raju Khubchandani[9], Priyankar Pal[10], Gabriele Simonini[11], Giovanni Filocamo[12], Maurizio Gattinara[13], Fabrizio De Benedetti[14], Davide Montin[15], Adele Civino[16], Muatasem Alsuweiti[17], Valda Stanevicha[18], Vyacheslav Chasnyk[19], Ekaterina Alexeeva[20], Sulaiman M Al-Mayouf[21], Soamarat Vilaiyuk[22] and Angelo Ravelli[23], [1]IRCCS Istituto Giannina Gaslini, UOC Reumatologia e Malattie Infiammatorie, Genova, Italy, [2]IRCCS Istituto Giannina Gaslini, UOC Reumatologia e Malattia Infiammatorie, Genova, Italy, [3]IRCCS Istituto Giannina Gaslini, UOC Reumatologia e Malattie Autoinfiammatorie, Genova, Italy, [4]IRCCS Istituto Giannina Gaslini, Genova, Italy, [5]Hospital de Pediatria Juan P. Garrahan, Department of Immunology/Rheumatology, Buenos Aires, Argentina, [6]São Paulo State University, Pediatric Rheumatology Division, Botucatu, Brazil, [7]Children's Hospital, Ain Shams University, Pediatric Allergy, Immunology and Rheumatology Unit, Cairo, Egypt, [8]Ain Shams University, Department of Rheumatology and Rehabilitation, Cairo, Egypt, [9]Jaslok Hospital and Research Centre, Department of Paediatrics, Mumbai, India, [10]Institute of Child Health, Pediatric medicine, Kolkata, India, Kolkata, India, [11]IRCCS Meyer Children's Hospital, Rheumatology Unit, Florence, Italy, [12]Fondazione IRCCS Ca' Granda-Ospedale Maggiore Policlinico, Pediatric Rheumatology, Milano, Italy, [13]Istituto Gaetano Pini, Rheumatology Unit, Milano, Italy, Genova, [14]Division of Rheumatology, IRCCS Ospedale Pediatrico Bambino Gesù, Roma, Italy, [15]Regina Margherita Children Hospital, Immunology and Rheumatology Unit, Turin, Italy, [16]Ospedale Vito Fazzi, Paediatric Immunology/Rheumatology Service, Lecce, Italy, [17]King Hussein Medical Center, Department of Pediatrics - Pediatric Allergy, Immunology & Rheumatology Clinic, Amman, Jordan, [18]University Children Hospital, Department of Pediatrics, Riga, Latvia, [19]Saint-Petersburg State Pediatric Medical University, Department of Hospital Pediatrics, St. Petersburg, Russia, [20]Federal State Autonomous Institution "National Medical Research Center of Children's Health", Ministry of Health of the Russian Federation, Moscow, Russia, [21]King Faisal Specialist Hospital & Research Center, Alfaisal University, Department of Pediatric Rheumatology, Riyadh, Saudi Arabia, [22]Mahidol University Faculty of Medicine, Ramathibodi Hospital Department of Pediatrics, Bangkok, Thailand, [23]IRRCS Istituto Giannina Gaslini and Università degli Studi di Genova, Genova, Italy

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Poster Breakout 6 - Systemic JIA: Genomics, Transcriptomics & Disease States

Session Type: Abstract Session

Session Time: 4:30PM–5:00PM

Background/Purpose: Systemic juvenile idiopathic arthritis (sJIA) accounts up to 15% of all patients with JIA and is distinctfrom the other disease categories due to the association of articular and extra-articular manifestations. The systemic Juvenile Arthritis Disease Activity Score (sJADAS) is a composite disease activity score validated specifically for use in sJIA that includes, beside the 4 components of the original JADAS [Tibaldi J, Pistorio A, Aldera E, et al. Development and initial validation of a composite disease activity score for systemic juvenile idiopathic arthritis. Rheumatology (Oxford). 2020;59(11):3505-3514], a fifth item aimed to quantify the burden of systemic features. The interpretation of scores of sJADAS requires criteria that identify the states of disease activity. These criteria can be used to monitor the disease course over time and to define therapeutic targets. Our purpose was to compare the clinical and laboratory data of each disease activity state in patients enrolled in the multinational study aimed to define the sJADAS cutoffs.

Methods: Data were extracted from a multinational cross-sectional dataset that included patients diagnosed with sJIA by ILAR criteria, recruited between February 2022 and November 2022. At study visit, each patient was categorized subjectively by the caring physician into one of the following disease activity states: inactive disease (ID), low (or minimal) disease activity (LDA), moderate disease activity (MDA), or high disease activity (HDA). Study data was collected through a standard case report form and entered into an electronic database.

	ID	LDA	MDA	HDA
Number of patients (%)	87 (37.7)	39 (16.9)	46 (19.9)	59 (25.5)
Age at onset, years (mean, SD)	6.17 (4.24)	5.44 (3.06)	5.11 (3.42)	5.36 (4.06)
MD global VAS (mean, SD)	1.08 (2.64)	2.5 (2.59)	5.03 (2.59)	6.61 (2.91)
MD systemic VAS (mean, SD)	0.06 (0.23)	0.88 (1.13)	3.08 (2.69)	7.52 (2.17)
Systemic features (%)	1 (1.1)	7 (17.9)	22 (47.8)	56 (94.9)
• Fever	0 (0)	0 (0)	19 (41.3)	55 (93.2)
• Rash	0 (0)	4 (10.3)	9 (19.6)	29 (49.2)
• Hepatomegaly	0 (0	0 (0)	2 (4.3)	21 (35.6)
• Splenomegaly	0 (0)	1 (2.6)	4 (8.7)	14 (23.7)
• Lymphadenopathy	1 (1.1)	2 (5.1)	5 (10.9)	20 (33.9)
• Serositis	0 (0)	0 (0)	4 (8.7)	9 (15.3)
ESR, mm/h (mean, SD)	8.46 (7.63)	16.26 (14.43)	46.11 (34.53)	66.85 (32.4)
CRP, mg/dl (mean, SD)	0.39 (0.85)	0.88 (1.93)	5.17 (7.03)	7.89 (6.06)
NAJ > 1 (%)	1 (1.1)	13 (33.3)	34 (73.9)	53 (89.8)

Comparison of clinical and laboratory features across disease activity states (n = 231). MD global VAS=physician global assessment of disease activity; MD systemic VAS= physician global assessment of systemic disease activity; ESR= erythrocyte sedimentation rate; CRP= C-reactive protein; NAJ=Number of active joints; ID=inactive disease; LDA= low (or minimal) disease activity; MDA=moderate disease activity; HDA=high disease activity.

Results: A total of 231 patients were enrolled in 29 centers in 12 countries. The mean age at diagnosis was 5.63 years. 87 patients (37.7%) were judged as having ID, 39 (16.9%) LDA, 46 (19.9%) MDA and 59 (25.5%) HDA. The comparison of the main clinical and laboratory features across patients with the 4 disease activity states is shown in the Table. Overall, the presence of extra-articular manifestations was more common in patients with MDA and HDA (p 0.00001), whereas fever, rash, hepatosplenomegaly, and lymphadenopathy were more frequent in HDA patients (p 0.00001). The count of active joints increased progressively from ID to HDA (p 0.00001). The mean values of physician global assessment of disease activity and systemic manifestations, as well as the mean values of acute phase reactants, were highest in patients with HDA, with gradual decrease from MDA to LDA to ID.

Conclusion: This preliminary analysis of the study data indicates that the subjective assessment of disease state by the caring physicians led to discriminate reliably patients with different level of disease activity. This evaluation will, then, serve well as reference for the subsequent analyses aimed to identify the cutoffs for the main disease activity states in sJIA using the sJADAS.

Disclosure: A. Rebollo-Giménez: None; **Y. Vyzhga**: None; **L. Carlini**: None; **S. Rosina**: None; **E. Patrone**: None; **M. Katsikas**: None; **C. Magalhaes**: None; **D. El-Ghoneimy**: None; **Y. El Miedany**: None; **R. Khubchandani**: None; **P. Pal**: None; **G. Simonini**: AbbVie/Abbott, 12, Educational Grant, Novartis, 12, Educational Grant, Sobi, 12, Educational Grant; **G. Filocamo**: Novartis, 2, Sobi, 2; **M. Gattinara**: None; **F. De Benedetti**: AbbVie/Abbott, 2, Novartis, 2, Novimmune, 2, Pfizer, 2, Roche, 2, Sobi, 2; **D. Montin**: None; **A. Civino**: None; **M. Alsuweiti**: None; **V. Stanevicha**: None; **V. Chasnyk**: None; **E. Alexeeva**: AbbVie/Abbott, 5, Amgen, 5, Bristol-Myers Squibb(BMS), 5, Centocor, 5, Eli Lilly, 5, Merck/MSD, 5, Novartis, 5, 6, Pfizer, 5, 6, Roche, 5, 6, Sanofi, 5; **S. Al-Mayouf**: Novartis, 6, Sobi, 6; **S. Vilaiyuk**: None; **A. Ravelli**: AbbVie/Abbott, 6, Alexion, 6, Angelini, 6, Bristol-Myers Squibb(BMS), 6, Novartis, 6, Pfizer, 6, Reckitt Benckiser, 6, Roche, 6, Sobi, 6.

Abstract Number: 040

HLA DRB1*15 and Eosinophilia Are Common Among Patients with Systemic Juvenile Idiopathic Arthritis

Alison Lerman[1], Shawn Mahmud[1], Zineb Alfath[2], Benjamin Langworthy[3], Patricia Hobday[1], Mona Riskalla[1] and Bryce Binstadt[1], [1]Division of Pediatric Rheumatology, Allergy & Immunology, Department of Pediatrics, University of Minnesota and M Health Fairview Masonic Children's Hospital, Minneapolis, MN, [2]University of Minnesota, Minneapolis, [3]Division of Biostatistics, School of Public Health and Biostatistical Design and Analysis Center, Clinical and Translational Science Institute, University of Minnesota, Minneapolis, MN

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Poster Breakout 6 - Systemic JIA: Genomics, Transcriptomics & Disease States

Session Type: Abstract Session

Session Time: 4:30PM–5:00PM

Background/Purpose: Over the last two decades, some children with systemic juvenile idiopathic arthritis (SJIA) have developed a severe form of interstitial lung disease (ILD) termed SJIA-associated lung disease (SJIA-LD). This has paralleled increased use of biologic agents targeting IL-1 and IL-6 to treat SJIA, leading to concern that these medications might predispose to SJIA-LD. In retrospective analyses, the majority of patients with SJIA-LD have had exposure to IL-1 and IL-6 inhibitors, but interpreting the relevance of this drug exposure history is confounded by the increasingly widespread use of these agents to treat SJIA over the same time period. Carriage of HLA DRB1*15 has been reported in previous publications as a risk factor for adverse drug reactions among patients with SJIA. We performed a retrospective chart review to evaluate these factors at our center.

Methods: Subjects were considered for inclusion if they received a billing diagnosis code for SJIA between 1996 and 2022 (ICD-10-CM codes: M08.20, M08.2A, M08.29) before the age of 17 and had at least six months of follow-up in the University of Minnesota pediatric rheumatology clinics. Cases were then manually adjudicated to confirm the diagnosis of SJIA. 83 eligible patients were included. HLA typing wasperformed in 23 of the subjects. We compared characteristics of patients with or without eosinophilia. Among patients with HLA typing, we compared clinical characteristics of subjects with or without DRB1*15 and with or without SJIA-LD.The terminology DRB1*15:XX denotes DRB1*15:01 and DRB1*15:03. Eosinophilia was defined as an absolute eosinophil count ^{3}700/microliter or a percent eosinophils ^{3}10% of the white blood cell count.

TABLE 1
Characteristics of 83 patients with SJIA, divided by those with eosinophilia or not.

	All patients with SJIA (n=83)	Eosinophilia (n=33)	No eosinophilia (n=50)	P-value (eosinophilia vs no eosinophilia)
Age at Diagnosis (years) (mean, range)	6.5, (1-16)	6.2, (1-16)	6.8, (1-16)	0.61
Male (number, %)	33 (40%)	13 (39%)	20 (40%)	0.96
Exposed to IL-1 or IL-6 blockade (number, %)	79 (95%)	32 (97%)	47 (94%)	0.54
MAS (number, %)	15 (18%)	10 (30%)	5 (10%)	0.02
Lung disease (number, %)	7 (8.4%)	5 (15%)	2 (4%)	0.11
Trisomy 21 (number, %)	3 (4%)	2 (6%)	1 (2%)	0.56
History of drug reaction (number, %)	18 (22%)	11 (33%)	7 (14%)	0.04

SJIA = systemic juvenile idiopathic arthritis; MAS = macrophage activation syndrome.

TABLE 2
Comparison of patients with SJIA with or without HLA DRB1*15:XX

	SJIA patients with HLA DR Typing (n=23)		
	DRB1*15:XX positive (n=17)	DRB1*15:XX negative (n=6)	P-value
Age at Diagnosis (y) (mean, range)	4.3, (1-16)	9.3, (5-16)	0.03
Male (number, %)	7 (41%)	2 (33%)	1
Exposed to IL-1 or IL-6 blockade (number, %)	16 (94%)	6 (100%)	1
MAS (number, %)	6 (35%)	1 (17%)	0.62
Eosinophilia (number, %)	10 (59%)	3 (50%)	1
Lung disease (number, %)	7 (41%)	0 (0%)	N/A
Trisomy 21 (number, %)	3 (18%)	0 (0%)	0.54
History of drug reaction (number, %)	9 (53%)	3 (50%)	1

We do not report a p-value for the difference in proportion of SJIA-LD between those positive and negative for DRB1*15:XX, because patients with SJIA-LD were overrepresented in the sample of patients in whom HLA typing was performed.
SJIA = systemic juvenile idiopathic arthritis; MAS = macrophage activation syndrome.

TABLE 3
Comparison of patients with SJIA with lung disease (SJIA-LD) to those without lung disease

	SJIA patients with HLA DR Typing (n=23)		
	SJIA-LD n=7	No Lung Disease n=16	P-value (lung disease vs no lung disease)
HLA DRB1*15:XX positive (number, %)	7 (100%)	10 (63%)	0.12
Age at Diagnosis (y) (mean, range)	1.7, (1-3)	7.4, (1-16)	0.01
Male (number, %)	3 (43%)	6 (38%)	1
Exposed to IL-1 or IL-6 blockade (number, %)	7 (100%)	15 (94%)	1
MAS (number, %)	5 (71%)	2 (13%)	0.01
Eosinophilia (number, %)	5 (71%)	8 (50%)	0.41
Trisomy 21 (number, %)	0 (0%)	3 (19%)	0.53
History of drug reaction (number, %)	6 (86%)	6 (38%)	0.07

SJIA = systemic juvenile idiopathic arthritis; MAS = macrophage activation syndrome.

Results: Exposure to IL-1 and IL-6 blockers was common, occurring in 95% of patients. Eosinophilia occurred in 40% of patients with SJIA. Eosinophilia was associated with adverse drug reactions and macrophage activation syndrome. Among the 23 patients with HLA typing, 74% carried DRB1*15, and 63% of patients without SJIA-LD carried DRB1*15. Seven subjects had SJIA-LD, all of whom carried DRB1*15 and 6 of whom had drug reactions. Patients with SJIA-LD were younger at the time of diagnosis and more likely to have had macrophage activation syndrome. There were no deaths.

Conclusion: The most striking finding in this cohort of patients with SJIA is the high rate of DRB1*15:XX carriage (74%) as compared to the general population carriage rate of 25%. Even among subjects without lung disease, the rate of DRB1*15:XX carriage was high (63%). In our cohort, all 7 patients with SJIA-LD expressed DRB1*15:XX. Eosinophilia was

also common, occurring in 40% of all patients with SJIA and often prior to IL-1 or IL-6 inhibitor therapy.Eosinophilia did not differ based on the presence or absence of HLA-DRB1*15.Eosinophilia was more common among patients with severe SJIA complicated by MAS. Many patients who carried HLA-DRB1*15 did not have eosinophilia, adverse drug reactions, or lung disease despite 95% of all patients being exposed to IL-1 or IL-6 inhibitors. In our cohort, all patients with SJIA-LD expressed HLA-DRB1*15 and were also significantly younger at age of diagnosis and more likely to have had a history of MAS.

Disclosure: A. Lerman: None; **S. Mahmud**: None; **Z. Alfath**: None; **B. Langworthy**: None; **P. Hobday**: None; **M. Riskalla**: None; **B. Binstadt**: Sobi, 5.

Abstract Number: 041

Cognitive Performance Score of the Pediatric Automated Neuropsychological Assessment Metrics Software in a Brazilian Cohort

Jaqueline De Amorim[1], Simone Kishimoto[1], Paula Fernandes[1], Roberto Marini[1], Lilian Costallat[1], Zahi Touma[2], Hermine Brunner[3] and **Simone Appenzelle**[4], [1]UNICAMP, São Paulo, Brazil, [2]University of Toronto, Toronto, ON, Canada, [3]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, [4]University of Campinas, Campinas, Sao Paulo, Brazil

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: The PedAnam (Pediatric Automated Neuropsychological Assessment Metrics) is an automatic software to evaluate cognitive performance that has recently been validated into Portugues (Brazil). The purpose of this study was to: (1) Explore and test Cognitive Performance Score (CPS) in systemic lupus erythematosus (SLE) and (2) Compare different CPS scores between SLE and controls and to determine if there is a difference according to patientsage.

Methods: Consecutive SLE and healthy controls completed to the PedAnam. We performed the calculation of four scores: (1) PedANAM-CPSUWA using unweighted averages of the accuracy score; (2) PedANAM-CPSPCA principal component analysis; (3) PedANAM-CPSlogit used logistic regression; (4)PedANAM-CPSmultiscore logistic models based on performance parameters with selected subtests. After these calculations, we observed the correlation between the CPS indices and compared the performance of patients and healthy controls. Cognitive impairment was determined by z-scores and considered present if ≤2 SD from healthy controls.

Results: We included a total of 201 consecutive SLE (183 [48.4%] women; median age = 28 years; age range = 9–76 years) and 177 healthy controls (124 [32.8 %] women; median age = 22; age range = 9-60 years). We observed a correlation between the PedANAM-CPSUWA and the PedANAM-CPSPCA (r = 0.99), PedANAM-CPSPCA and PedANAM-CPSmultiscore (r = -0.60) logit and PedANAM-CPSUWA (r = -0.56). We observed statistically significant differences between patients and healthy controls in the PedANAM-CPSUWA and PedANAM-CPSPCA scores (P = 0.001). The PedANAM-CPSlogit and PedANAM-CPSmultiscore indices did not show significant differences between groups. Furthermore, separating SLE according to current age (< 18 years and ≥18 years), no difference between the four indices was noted. Cognitive impairment was observed in 16.42% SLE patients and 4.52% healthy controls ($p < 0.001$) by the PedANAM-CPSUWA index and in 17.91% SLE patients and 5.64% healthy controls ($p < 0.001$) using the PedANAM-CPSPCA index.

Conclusion: After the translation and validation process, this is the first study using the CPS in Brazilian version of PedA-NAM. Two of the metrics were able to differentiate between SLE and healthy controls. No difference according to current age was observed.

Disclosure: J. De Amorim: None; **S. Kishimoto**: None; **P. Fernandes**: None; **R. Marini**: None; **L. Costallat**: None; **Z. Touma**: None; **H. Brunner**: GENENTECH, 12, provision of study drug for NIAMS funded study, Pfizer, 1, 2, 6; **S. Appenzelle**: None.

Abstract Number: 042

Prevalence of Chronic Pain in Childhood-onset Systemic Lupus Erythematosus and Juvenile Dermatomyositis

Sara Patrizi[1], Susmita Kashikar-Zuck[2], CARRA Registry Investigators[3], Mekibib Altaye[4] and Jennifer Weiss[5], [1]Stanford Medicine, Children's Health, Palo Alto, CA, [2]University of Cincinnati College of Medicine; Cincinnati Children's Hospital Medical Center, Cincinnati, OH, [3]CARRA, Washington, DC, [4]Cincinnati Children's Hospital, Cincinnati, OH, [5]Pediatric Rheumatology, Hackensack Meridian School of Medicine, Hackensack University Medical Center, Hackensack, New Jersey, Hackensack, NJ

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: The prevalence, severity and impact of chronic pain in pediatric patients with autoimmune diseases such as childhood onset SLE (cSLE) and juvenile dermatomyositis (JDM) has yet to be well studied. This study used the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry to 1) describe the prevalence and severity of pain in cSLE and JDM patients and 2) evaluate the impact of pain symptoms on their daily functioning.

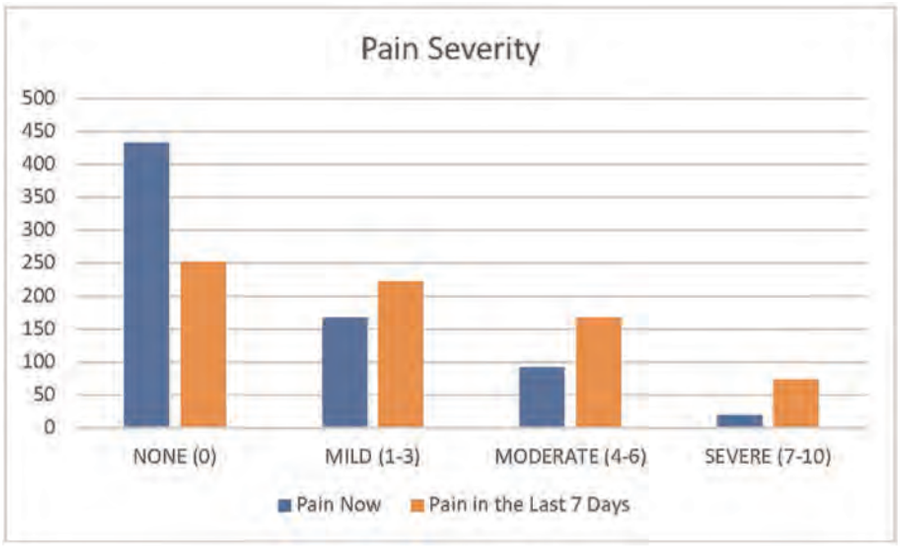


Figure 1: Pain Severity at Clinic Visit and in the Last Two Weeks Prior to Visit

Table 1: Results of Disease Activity and Pain Measures

	None/Mild Pain	Moderate/Severe Pain	P-value
PROMIS (N=498)			
T-Score *	49.40	59.63	<0.0001
SLEDAI (N=167)**			
Mean (Standard Deviation)	5.19 (6.27)	7.75 (8.27)	0.0407
Physician Global Assessment (N=196)			
Mean (Standard Deviation)	2.33 (2.19)	3.8 (2.43)	<0.001
Widespread Pain (N=200)			
Yes	4	9	
No	123	64	0.0112

*PROMIS T-Score: This is a standardized measurement of the summarized PROMIS score, with a mean of 50 and standard deviation of 10
**SLEDAI Range: 0-105

Methods: All patients 8 years and older diagnosed with lupus-spectrum diseases (SLE, MCTD, APLS, Sjogrens Syndrome and cutaneous lupus) as well as JDM in the CARRA Legacy registry enrolled between 2010 and 2014 were included. Data analyzed for this study included demographic information, disease severity, pain intensity scores and quality of life measures (PROMIS Pain Interference scale), Physician Global Assessment (PGA 0-10) and physician assessment of the presence of widespread pain (yes/no). Descriptive statistics were computed to examine pain and clinical characteristics of the cohort. Statistical comparisons between patients who reported no/mild pain versus those who reported moderate/severe pain were performed using the method of least mean squares (PROMIS Pain Interference), and to evaluate for differences in the disease activity between the none/mild pain cohort and the moderate/severe pain cohort, chi-squared test, and t-test or Wilcoxon test was performed as appropriate.

Results: CARRA registry data was obtained on all patients diagnosed with lupus-spectrum diseases and JDM, with an initial cohort of 788 patients. Out of the 717 patients with valid data, 476 (66.39%) patients reported no or mild pain, 239 (33.61%) patients reported moderate to severe pain in the last week. For the presence of current pain, 600 (84.39%) patients reported no or mild pain and 111 (15.61%) patients reported moderate or severe pain (Figure 1). Those who reported moderate/severe pain reported significantly greater (p 0.0001) scores on the PROMIS pain interference scale than those who reported no/mild pain in the past week. Those who reported moderate/severe pain in the last week had significantly higher (p=0.0407) mean SLEDAI scores, and statistically higher PGA (p 0.0001) than those who reported no/mild pain. Finally, those who reported moderate/severe pain in the last week had a significantly higher (p=0.0112) presence of widespread pain compared to those who reported no/mild pain (Figure 1).

Conclusion: This retrospective study shows that a subset of pediatric patients (~33%) diagnosed with lupus-spectrum diseases and JDM report having moderate/severe pain in the last 7 days and these patients experience significantly reduced quality of life due to pain interference compared to those who have no/mild pain. In addition, this study noted that there was frequent missing data in the evaluation for the presence of widespread pain in a cohort of pediatric patients with rheumatic diseases, highlighting the need for future prospective research to determine the prevalence of widespread pain, such as secondary juvenile fibromyalgia, in this cohort.

Disclosure: S. Patrizi: None; **S. Kashikar-Zuck**: None; **C. Investigators**: None; **M. Altaye**: None; **J. Weiss**: None.

Abstract Number: 043

Development and Usability Testing of Web-based Standardized Scoring Tool for Magnetic Resonance Images from Children with Chronic Nonbacterial Osteomyelitis (CNO)

Farzana Nuruzzaman[1], T. Shawn Sato[2], Andrew Carbert[3], Joel Paschke[4], Lauren Potts[5], Meinrad Beer[6], Ming Huang[7], Ramesh Iyer[8], Johanna Monsalve[9], Anh-Vo Ngo[8], Jennifer Stimec[10], Mahesh Thapa[8], Wei Hou[11], Walter P. Maksymowych[12], Polly Ferguson[13] and Yongdong (Dan) Zhao[8], [1]Stony Brook Children's Hospital, Stony Brook, NY, [2]University of Iowa, Iowa City, IA, [3]CARE Arthritis, Edmonton, AB, [4]CARE Arthritis, Edmonton, AB, Canada, [5]Long Beach, CA, [6]University Hospital, Ulm Germany, Ulm, Germany, [7]Mount Sinai Hospital, New York, NY, [8]University of Washington, Seattle, WA, [9]Stony Brook University Hospital, Stony Brook, NY, [10]The Hospital for Sick Children, Toronto, ON, Canada, [11]Stony Brook University, Stony Brook, NY, [12]University of Alberta, Edmonton, AB, Canada, [13]University of Iowa Carver College of Medicine, Iowa City, IA

SESSION INFORMATION

Session Date: Thursday, March 30, 2023

Session Title: Posters: Clinical and Therapeutic I

Session Type: Poster Session A

Session Time: 6:00PM–7:00PM

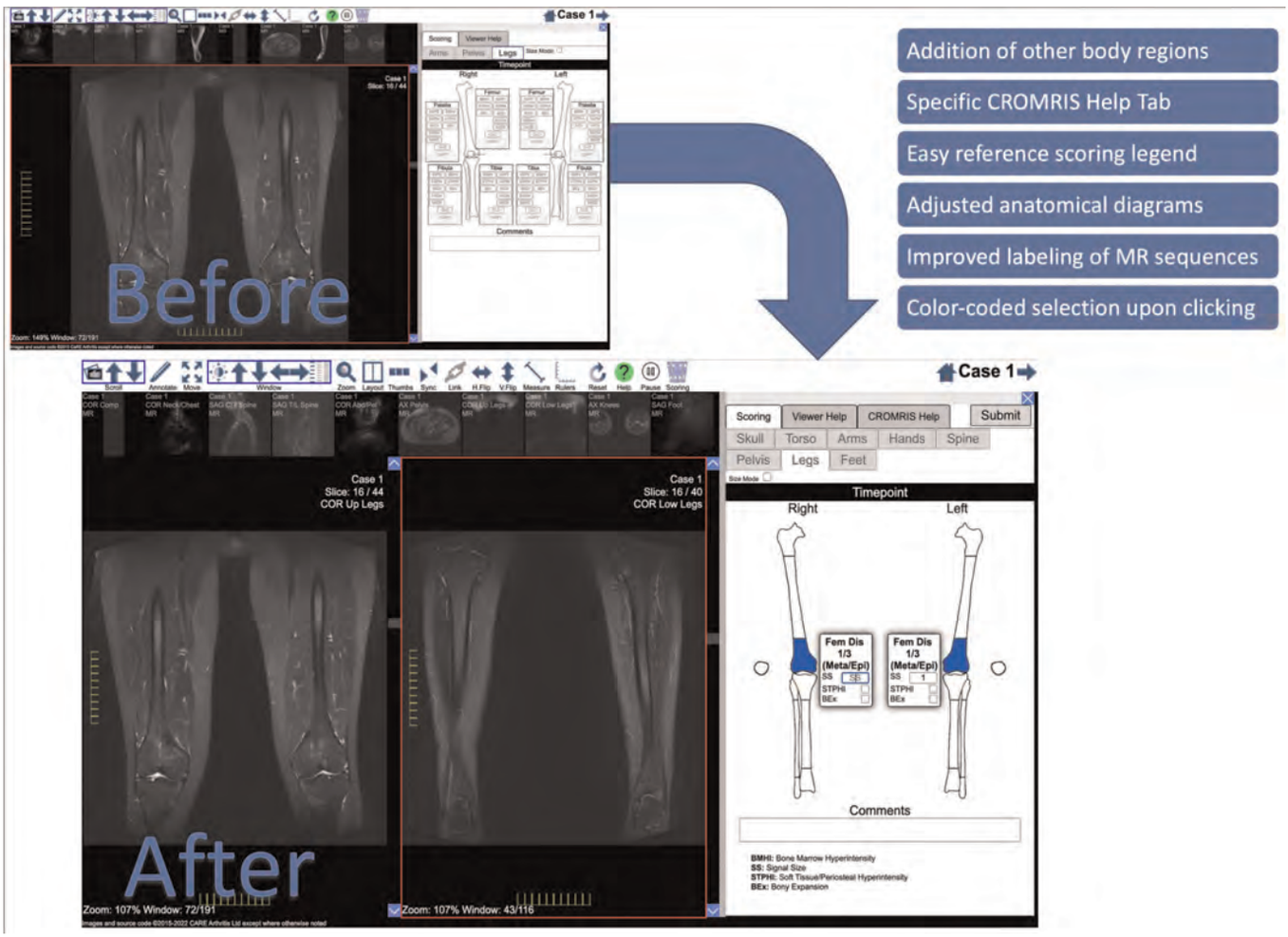


Figure 1. Example of Web-based CROMRIS Tool on Long Bones of Lower Extremity Before and After Feedback Sessions/Usability Testing

Background/Purpose: The ChRonic nonbacterial Osteomyelitis Magnetic Resonance Imaging Scoring (CROMRIS) tool was developed to assess specific characteristics of bone and soft tissue inflammation in MRI of patients with CNO, but was labor intensive to utilize. Primary objectives of this study are: 1) to adapt the CROMRIS tool to a web-based platform and 2) assess the usability of this web-based CROMRIS system among radiologists.

Methods: A prototype web-based CROMRIS tool limited to the arms and legs was developed by CARRAs CRMO Workgroup and CARE-Arthritis in 2019. Monthly meetings between software developers, rheumatologists, radiologists and an illustrator led to a beta version that includes the whole body. A purposive sample of radiologists (n=7) provided feedback on the beta version in a demo session on 4/11/22 via semi-structured surveys (Stony Brook University #IRB2021-00033). Usability was assessed in two phases (in 5/2022 and in 12/2022) using the System Usability Scale (SUS), a Likert scale in which respondents indicate their level of agreement or disagreement on a scale of 1 to 5 for 10 statements. Feedback was reported with descriptive content analysis, continuous variables as means and categorical variables as percentages.

Results: A clickable-schematic-based CROMRIS was developed to include all body regions: head (skull/mandible), spine, torso (clavicle, sternum, ribs), pelvis, hands, feet, arms and legs [Figure 1]. Notable features are the ability to immediately highlight a schematic region upon selection to directly input scores and the ability to zoom in on smaller areas. Suggested changes included flipping orientation of hands/feet drawings to match MRI presentation, labeling of individual spine and rib segments, insertion of scoring legend on each tab for reference and creation of a summary page with a composite diagram where one can visualize the location and size of lesions by color as well as a numerical CROMRIS activity index [-Figure 2]. A video tutorial and MRI Atlas is on the platform for training (https://www.carearthritis.com/mriportal/crmo/index/). Visual factors and anatomical diagrams were among the features "liked best" by survey respondents. Mean SUS scores increased from 64.5 (below average) to 75 (above average). All respondents agreed that the web-based CROMRIS was "easy to learn" and found that the "various functions of the web-based CROMRIS were well integrated."

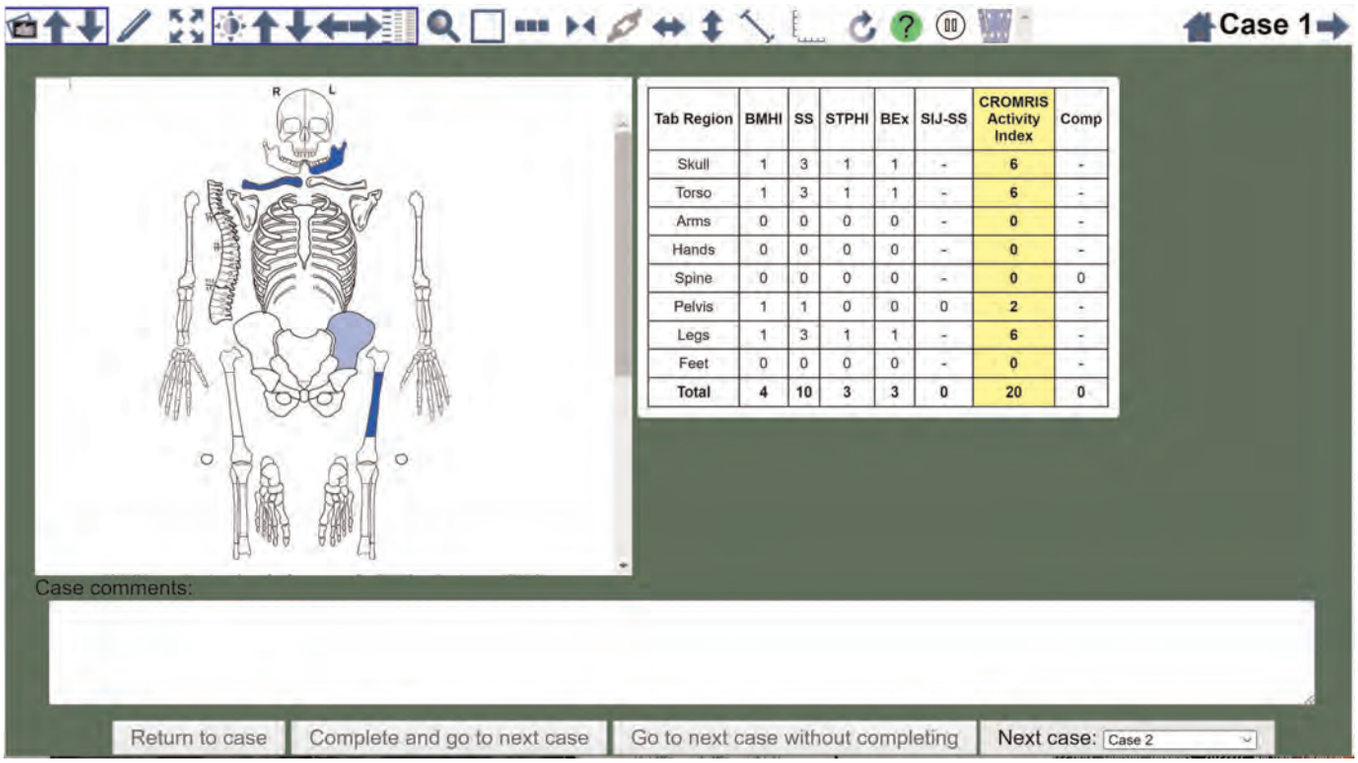


Figure 2. Screenshot of Summary Page for Final Web-based CROMRIS system

Conclusion: The web-based CROMRIS portal shows good usability amongst radiologists. Studies of inter-rater reliability among experienced pediatric radiologists are underway. Once validated, this tool can be used as a semi-quantitative MRI scoring tool to allow for standardization of reporting output of radiological interpretations of MRI in CNO.

Disclosure: F. Nuruzzaman: None; **T. Sato**: None; **A. Carbert**: None; **J. Paschke**: None; **L. Potts**: None; **M. Beer**: None; **M. Huang**: None; **R. Iyer**: None; **J. Monsalve**: None; **A. Ngo**: None; **J. Stimec**: None; **M. Thapa**: None; **W. Hou**: None; **W. Maksymowych**: Abbvie, 2, 5, 6, Bristol-Myers Squibb(BMS), 2, Celgene, 2, Eli Lilly, 2, 5, Galapagos, 2, 5, Janssen, 6, Novartis, 5, 6, Pfizer, 2, 5, 6, UCB, 2, 5, 6; **P. Ferguson**: None; **Y. Zhao**: Bristol-Myers Squibb(BMS), 5.

Abstract Number: 044

Validation of Newly Proposed Classification Criteria for Pediatric Chronic Nonbacterial Osteomyelitis: A Virginia Cohort

Kelley Lee[1], Ashley Kim[2] and Aarat Patel[3], [1]University of Richmond, Richmond, VA, [2]University of Virginia, Charlottesville, VA, [3]Bon Secours Mercy Health / University of Virginia, GLEN ALLEN, VA

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Chronic nonbacterial osteomyelitis (CNO) is an aseptic autoinflammatory bone disease of unknown etiology. This diagnosis can be delayed due to the non-specific nature of symptoms, referral patterns to non-rheumatology specialists and normal imaging studies. New classification criteria have been proposed for CNO and were presented at the American College of Rheumatology scientific meeting in 2022. This study seeks to validate these new classification criteria.

Methods: This is a retrospective case-series of pediatric patients diagnosed with CNO (n=34). They were diagnosed and managed between 2012 and 2022 at the University of Virginia in Charlottesville, Virginia and Bon Secours Mercy Health in Richmond, Virginia. The entry criteria of chronic bone pain, age 18 years, abnormal radiograph and/or MRI were used prior to applying the newly proposed classification criteria. Tendomains were evaluated to indicate whether a patient has CNO; those who scored 55 points or higher qualified (see table). The clinical manifestations, laboratory results, and imaging studies that lead to a diagnosis of CNO were reviewed in detail. We applied and validated the newly proposed classification criteria for CNO to this Virginia cohort of previously diagnosed pediatric CNO patients. We also reviewed the characteristics of those that did not fulfill this classification criteria.

Results: Upon application of the newly proposed classification criteria for CNO, 31 out of 34 patients (91%) were found to meet this classification criteria. Of those that did not fulfill the criteria but carry the diagnosis of CNO (n=3) none had "least specific sites" such as hands orneurocranium, all had elevated ESR and two out of three had an initial response to antibiotics. Their scores were 49, 51 and 53 with classification criteria being met with 55 or higher. The average age of diagnosis was 10 (range 2-17) years-old with the average time between onset of symptoms to confirmed radiographic evidence being 25 (range 1-120) months. The most common presenting symptoms were bone pain (100 %) and arthritis (52%) with fever (23%) and rash (3%) being less common. Elevated ESR (39%) and CRP (39%) were found at initial visit with rheumatology. Evidence of commonly seen radiographic abnormalities leading to CNO diagnosis was found with regional MRI (65%), bone scan (42%) with some requiring full body MRI (26%) to confirm other lesions. Some patients did have multiple imaging studies including X-ray, CT, bone scan, MRI and bone biopsy in their workup.

CNO Classification Criteria Domains

	Specific Bones Affected
0	Any least specific sites (hands, neurocranium) without any most specific
10	Any nonspecific sites without any least and most
18	Any of the most specific sites (mandible, clavicle)
	Lesion Distribution Pattern
0	Unifocal with whole body imaging
3	Unifocal without whole body imaging
8	Multifocal without any symmetrical pattern
16	Multifocal with any symmetrical pattern
	Response to antibiotics
0	Complete response to appropriate antibiotics monotherapy
13	NA
16	Partial or No response to antibiotics monotherapy
	Bone Pathology
0	Bone Bx no signs inflammation or fibrosis
6	No bone biopsy
10	Bone Bx - inflammation only
12	Bone Bx - signs of fibrosis only
13	Bone Bx - inflammation and fibrosis
	Age
0	<3
13	>=3
	Coexisting conditions (axial arthritis/IBD/PsO)
0	None or info is not available
4	Axial arthritis present without psoriasis/PPP/other rash or IBD
8	Psoriasis/PPP/other rash present without IBD
9	Presence of IBD
	Hemoglobin
0	<10 mg/dL
3	NA
5	>=10mg/dL
	Fever
0	Presence of fever
2	NA
4	Absence of fever
	ESR (normal -20mm/hr)
0	>=60 mm/hr
1	NA/not done
3	<60 mm/hr
	CRP (normal 0-10 mg/l)
0	>=30mg/l
1	NA
3	<30mg/l

*One score per domain. **Total of 55 points or higher qualify as having CNO.
CNO = chronic nonbacterial osteomyelitis; NA = not available; Bx = bone biopsy; IBD = inflammatory bowel disease; PsO = psoriasis; PPP = palmoplantar pustulosis; ESR = erythrocyte sedimentation rate; CRP = C-reactive protein

Conclusion: This retrospective chart review validates the newly proposed classification criteria for CNO. We conclude that these criteria are easy to apply even in the absence of a bone biopsy or advanced imaging studies. This will aid in the development of a homogeneous cohort for future studies in CNO.

Disclosure: K. Lee: None; **A. Kim**: None; **A. Patel**: AbbVie/Abbott, 1, 6, Amgen, 6, Eli Lilly, 6, GlaxoSmithKlein(GSK), 6.

Abstract Number: 045

Patient Reported Outcomes in Pediatric Vasculitis

Clare Peckenpaugh[1], Aimee Hersh[2], CJ Inman[1], Sara Stern[1], Erin Treemarcki[2], Peter Merkel[3] and Karen James[1], [1]University of Utah Department of Pediatrics, Salt Lake City, UT, [2]University of Utah, Salt Lake City, UT, [3]University of Pennsylvania, Philidelphia, PA

SESSION INFORMATION

Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Vasculitis is a group of multisystem, often relapsing diseases that can affect patients through disease activity, damage, and treatment toxicity. Vasculitis in adults often has significant effects on health-related quality of life (HRQoL). Vasculitis in children occurs during critical times in psychosocial and physical development yet little is known on how these diseases and treatments affect HRQoL in children. This study aimed to evaluate HRQoL in patients with pediatric-onset vasculitis.

Methods: This was a prospective study of patients with vasculitis seen by pediatric rheumatology and/or pediatric nephrology at a single center tertiary care facility. Enrolled patients were those diagnosed with a chronic type of vasculitis before their 18th birthday who agreed to participate. Subjects were given a series of patient-reported outcome (PRO) measures during clinic visits, including several pediatric Patient-Reported Outcome Measures Information Systems (PROMIS) short forms. The treating provider concurrently filled out disease activity and damage measures (Pediatric Vasculitis Activity Score (PVAS) and Pediatric Vasculitis Damage Index (PVDI). Patients were enrolled at any time point in their disease course. Descriptive statistics and Spearmans Rank correlation were used.

Results: 20 patients participated, the majority of whom had ANCA-associated vasculitis. Median age at enrollment and age at diagnosis were 15.5 and 13.7 years, respectively (**Table 1**). Median disease duration was 2.3 years. 7/20 patients had active disease, defined as a PVAS 0 (median12, IQR 7.2 among active patients). 19/20 had at least one damage item scored on the PVDI, median 2, IQR 1, 4.5, range 0-18. Physician and patient global assessments of disease activity correlated (spearmans rho=0.65, p=0.005). PROMIS scores are displayed in **Figure 1**. The median score in all 10 domains queried was worse than the reference population, most notably in sleep-related impairment, depressive symptoms, cognitive function, peer relationships, and mobility.

Conclusion: Patients with pediatric-onset vasculitis have substantially worse scores than reference populations in multiple domains of HRQoL, including physical and psychosocial domains, demonstrating the negative life-altering effect of pediatric-onset vasculitis on HRQoL. Patient and physician global assessments did not correlate, highlighting the need to capture patients perspectives when studying these diseases. Further work is needed to evaluate HRQoL in pediatric patients with vasculitis so that treatments can be targeted to both control active disease and improve HRQoL.

Table 1. Presenting characteristics of patients

	N(%) or Median(IQR)
Age at enrollment (years)	15.5 (13, 17.5)
Age at diagnosis (years)	13.7 (10.8, 15.9)
Median disease Duration (years)	2.3 (0.4, 3.6)
Race	
Asian	1 (5)
Caucasian	19 (95)
Ethnicity	
Hispanic	2 (10)
Diagnosis subtype	
ANCA-associated vasculitis	14 (70)
Behçet's disease	2 (10)
Polyarteritis nodosa	2 (10)
IgA vasculitis	1 (5)
Takayasu's arteritis	1 (5)
ANCA Type	
cANCA (N=16)	8 (50)
pANCA (N=16)	2 (13)
Anti-MPO (N=18)	2 (11)
Anti-PR3 (N=18)	11 (61)
Patient Global Assessment of Disease activity	2 (2,4)
Physician Global Assessment of Disease activity	0.5 (0,3)
Pediatric Vasculitis Activity Score	0 (0, 9.5)
Pediatric Vasculitis Damage Index Score	2 (1, 4.5)

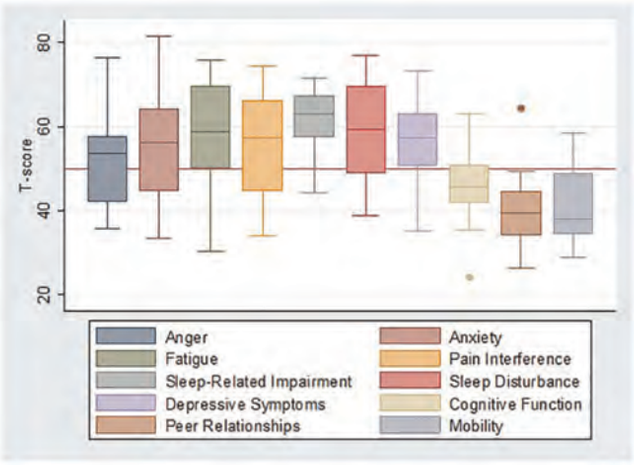


Figure 1. Boxplots of T scores from short forms of the *Patient Reported Outcome Measures Information System (PROMIS). T-scores for study subjects in each domain represents differences relative to the general population mean score that is set at a T score of 50. A higher T score represents more ore of the domain being measured, a lower T score represents less of the domain being measured.

Disclosure: C. Peckenpaugh: None; **A. Hersh**: None; **C. Inman**: None; **S. Stern**: None; **E. Treemarcki**: None; **P. Merkel**: AbbVie/Abbott, 2, 5, AstraZeneca, 2, 5, Boehringer-Ingelheim, 2, 5, Bristol-Myers Squibb(BMS), 2, 5, Caballetta, 2, Chemocentryx, 2, 5, CSL Behring, 2, Dynacure, 2, Eicos, 5, Electra, 5, EMDSerono, 2, Forbius, 2, 5, Genentech/Roche, 2, 5, Genzyme/Sanofi, 2, 5, GlaxoSmithKlein(GSK), 2, 5, Immagene, 2, InflaRx, 2, 5, Jannsen, 2, Jubilant, 2, Kiniksa, 2, Kyverna, 2, 11, Magenta, 2, MiroBio, 2, Mitsubishi, 2, Neutrolis, 2, 5, Novartis, 2, Pfizer, 2, Q32, 2, Regeneron, 2, Sanofi, 5, Sparrow, 2, Star, 5, Takeda, 2, 5, Uptodate, 9, Vistera, 2; **K. James**: None.

Abstract Number: 046

Colchicine Adherence Among Children and Young Adults with Familial Mediterranean Fever During Treatment with interleukin-1 Inhibitors

Yoel Levinsky[1], rotem tal[2], Liora Harel[3] and Gil Amarilyo[4], [1]Schneider Children's Medical Center of Israel, Tel Aviv University, Petach Tikva, Israel, [2]Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [3]Scheiders Children Medical Center of Israel, Petah-Tiqva, Israel, [4]Schneider Children's Medical Center of Israel, Petach Tikva, Israel

SESSION INFORMATION

Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: IL-1 inhibitor are approved for familial Mediterranean fever (FMF) patients who are resistant to colchicine. However, it is imperative to continue concomitant treatment with colchicine since it is the only drug proven to prevent secondary amyloidosis. We aimed to investigate the adherence to colchicine among colchicine resistant FMF patients treated with IL-1 inhibitors (crFMF) and compare them to colchicine sensitive FMF patients that were treated merely with colchicine (csFMF).

Table 1. Comparison of medication possession ratio (MPR) of colchicine between FMF patients treated with IL-1 inhibitors Versus patients without IL-1 inhibitors treatment

Parameter	With IL-1 inhibitors – "study group" (n=108)	Without IL-1 inhibitors – "matched group" (N=432)	P-value
Colchicine MPR – total	78.9±41.4	82.5±80.6	0.5
Age category, years			
≤6	74.9±24.9	75.8±33.7	0.9
7-14	90.2±53.6	75.6±70.3	0.09
15-29	76.5±47.9	91.1±123.3	0.3
Sex			
Male	75.7±49.9 (N=48)	79.2± 65.5 (N=192)	0.9
Female	81.4±33.2 (N=60)	85.1±91.0 (N=240)	0.03
Time of colchicine use			
< 8 years	102.8±46.2	100.9±99.5	0.2
8-15 years	66.6±35.8	56.7±36.5	0.2
> 15 years	61.9±24.8	60.4±25.8	0.7
Colchicine MPR category			0.1
< 20%	3 (2.8%)	46 (10.6%)	
20%-39%	16 (14.8%)	71 (16.4%)	
40%-59%	19 (17.6%)	61 (14.1%)	
60%-79%	19(17.6%)	73 (16.9%)	
≥80%	51 (47.2%)	181 (41.9%)	
Colchicine MPR for patients without Amyloidosis or CKD	85.8±44.3	84.9±83.8	0.8

Methods: The databases of Maccabi Health Services (MHS), a 2.6 million member state-mandated health provider in Israel was searched for patients 5-29y with FMF diagnosis. Medication possession ratio (MPR) was used as main outcome measure. crFMF were matched in a ratio of 1:4 to csFMF patients.

Results: The final cohort included 4526 patients. Among them, 108 crFMF patients, were matched to 432 csFMF patients. The total MPR in each groups was similar (78.9 ±41.4 versus 82.5 ± 80.6, P=0.5). No significant MPR differences between the groups were found for age and for time of colchicine use. However, the adherence to colchicine was insufficient (MPR < 80%) among more than 50% of the patients in both groups.

Conclusion: In contrast to initial concerns, adherence to colchicine in crFMF is similar to csFMF. However, in both groups, adherence to colchicine is poor and education of both caregivers and patients is essential in order to increase adherence.

Disclosure: Y. Levinsky: None; **r. tal**: None; **L. Harel**: None; **G. Amarilyo**: None.

Abstract Number: 047

Clinical Characteristics and Outcomes of North American Youth with Lupus Nephritis Requiring Dialysis Treated with Cyclophosphamide

Christine Wang[1], Rebecca Sadun[2], Wenru Zhou[3], Kristen Miller[3], Claire Palmer[3], Stacy Ardoin[4], Christine Bacha[5], Emily Hause[6], Joyce Hui-Yuen[7], Nicole Ling[8], Maria Pereira[9], Meredith Riebschleger[1], Kelly Rouster-Stevens[10], Aliese Sarkissian[11], Julia Shalen[12], William Soulsby[13], Marinka Twilt[14], Eveline Wu[15], Laura Lewandowski[16], Scott Wenderfer[17] and **Jennifer Cooper**[18], [1]University of Michigan, Ann Arbor, MI, [2]Duke University, Durham, NC, [3]University of Colorado, Denver, CO, [4]Nationwide Children's Hospital, Columbus, OH, [5]Akron Children's Hospital, Akron, OH, [6]University of Minnesota, Minneapolis, MN, [7]Cohen Children's Medical Center, Northwell Health, Lake Success, New York; Center for Autoimmune, Musculoskeletal, and Hematopoietic Diseases Research, Feinstein Institutes for Medical Research, Northwell Health, Manhasset, NY, [8]UCSF, San Francisco, CA, [9]Baylor College of Medicine, Houston, TX, [10]Emory University/Children's Healthcare of Atlanta, Atlanta, GA, [11]UNC- Chapel Hill, Durham, NC, [12]Johns Hopkins University, Baltimore, MD, [13]University of California, San Francisco, San Francisco, CA, [14]Alberta Children's Hospital, Calgary, AB, Canada, [15]UNC Chapel Hill, Chapel Hill, NC, [16]NIAMS, NIH, Bethesda, MD, [17]British Columbia Children's Hospital, Vancouver, BC, Canada, [18]University of Colorado/Children's Hospital Colorado, Denver, CO

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Few studies have evaluated the clinical characteristics and outcomes of youth with lupus nephritis (LN) treated with cyclophosphamide (CYC) who initially required kidney replacement therapy. CYC is partially renally cleared and the need for dose reduction with dialysis is controversial. Our study objectives were to summarize clinical characteristics of LN patients, describe treatment patterns, and compare the number of patients who required short-term dialysis versus those who developed end stage kidney disease (ESKD).

Methods: A multisite retrospective cohort study was conducted at 11 North American pediatric centers using electronic medical record data. Patients 6 to 22 years of age with active LN treated with either the lower dose EuroLupus (EL) or NIH CYC regimen from 7/1/2014 to 6/30/2021 were included. This subgroup analysis included patients who required dialysis at the time of CYC initiation (baseline). Patient data was collected at baseline, 3, 6, and 12 months. ESKD was defined as requiring dialysis therapy for >3 months. EL dosing was reviewed, and standard dosing was defined as 500 mg every 2 weeks for 6 doses, and any deviations from this dosing were recorded as modified. Baseline clinical characteristics and renal outcomes were summarized using descriptive statistics.

Results: Twenty patients met study inclusion criteria; of these, 15 (75%) received the EL regimen and 5 (25%) received the NIH regimen (**Table 1**). The average age at start of CYC during the study was 13.8 3.3 years. The cohort was predominantly female (75%), of diverse race/ethnicity, and publicly insured (60%). 80% of patients had new-onset LN while 20% had either relapsed or refractory disease. 95% of the cohort had proliferative LN and 30% had concurrent membranous disease. The mean cumulative dose of CYC was 5,823 mg 1,281 (NIH) and 2,681 mg 1,316 (EL). Of the 15 EL patients, 6 (40%) had modified dosing. Rituximab was given within 12 months of starting CYC to 40% of NIH regimen treated patients and 60% of EL treated patients. By 12 months, 3 patients (15%) had died, 4 (24%) met criteria for ESKD, and 13 (65%) had renal recovery off dialysis (**Table 2**). Of the patients with renal recovery, 11 of 13 (85%) were off before 3 months; 2 (15%) had an eGFR > 90 ml/min/1.73 m^2 at 12 months (**Table 2**). Infections requiring hospitalization during the CYC treatment period occurred in 1 (20%) patient who received the NIH regimen and 7 (47%) patients who received the EL regimen. 5 of 8 (63%) patients hospitalized due to infection also received rituximab.

Table 1: Baseline Demographics and Clinical Characteristics

	All (n=20)	NIH (n=5)	EL (n=15)
Age at diagnosis of SLE, years	13.1 ± 2.9	13.4 ± 3.2	13.0 ± 3.0
Age at diagnosis of LN, years	13.7 ± 3.4	14.0 ± 3.4	13.6 ± 3.6
Age at start of current CYC, years	13.8 ± 3.3	14.8 ± 3.5	13.4 ± 3.2
Female sex	15 (75%)	4 (80%)	11 (73%)
Race/Ethnicity			
Asian	2 (10%)	0 (0%)	2 (13%)
Black or African American	5 (25%)	2 (40%)	3 (20%)
Hispanic or Latino/a	7 (35%)	1 (20%)	6 (40%)
White	3 (15%)	1 (20%)	2 (13%)
Other[1]	3 (15%)	1 (20%)	2 (13%)
Insurance status			
Public	12 (60%)	3 (60%)	9 (60%)
Private	8 (40%)	2 (40%)	6 (40%)
Renal indication for CYC			
New-onset LN	16 (80%)	3 (60%)	13 (87%)
Relapsed or Refractory LN	4 (20%)	2 (40%)	2 (13%)
Complement 3 (C3) low	20 (100%)	5 (100%)	15 (100%)
Complement 4 (C4) low	18 (90%)	5 (100%)	13 (87%)
Sedimentation rate, mm/hr, n=17	60 (45 – 109)	54 (29 – 85)	67 (59 – 132)
Elevated blood pressure[2]	17 (85%)	4 (80%)	13 (87%)
ISN/RPS Lupus Nephritis class			
Class II	1 (5%)	0 (0%)	1 (7%)
Class III	3 (15%)	0 (0%)	3 (20%)
Class IV	16 (80%)	5 (100%)	11 (73%)
Class V, concurrent	6 (30%)	2 (40%)	4 (27%)
Crescents present n=19	10 (53%)	3 (60%)	7 (47%)
Glomerulosclerosis present n=19	7 (37%)	2 (40%)	5 (33%)
TMA present	2 (10%)	1 (20%)	1 (7%)
NIH activity score[3], n=15	9 (6 – 16)	12 (8 – 16)	7 (4 – 18)
NIH chronicity score[3], n=15	2 (1 – 6)	2 (1 – 2)	2 (1 – 6)

Results are presented as mean ± standard deviation for normally distributed continuous data and median with interquartile range for non-normally disturbed continuous data. Counts with percentages are summarized for categorical data.
[1]Includes Multi-Racial and Middle Eastern/North African patients
[2]Systolic blood pressure and/or diastolic blood pressure ≥ 95th percentile for age at baseline visit
[3]Modified NIH lupus nephritis activity and chronicity indices
Abbreviations: NIH = National Institutes of Health, EL = EuroLupus, SLE = Systemic Lupus Erythematosus, LN = Lupus Nephritis, CYC = Cyclophosphamide, ISN/RPS = International Society of Nephrology and the Renal Pathology Society. TMA = Thrombotic microangiopathy

Conclusion: A majority of youth with proliferative LN requiring dialysis at baseline had new-onset LN and were treated with a lower dose CYC regimen (standard or modified EL). Over half of the patients received concurrent rituximab. Although most patients were able to discontinue dialysis, almost a quarter developed ESKD, and 3 patients died. Limitations include small sample size, lack of data on dialysis indication or dialysis type, and a follow-up duration of only 12 months. There is also practice variation regarding CYC dosing modification for patients on dialysis and optimal dosing is unclear. Future larger prospective studies are needed to better understand outcomes for this vulnerable population.

Table 2: Outcomes of the Dialysis Cohort

	All (n=20)	NIH (n=5)	EL (n=15)
Three months			
Off dialysis, n (%)	11 (55%)	2 (40%)	9 (60%)
eGFR[1], ml/min/1.73 m^2	79 (63 – 111)	95 (n/a)[5]	68 (59 – 111)
Urine protein:creatinine ratio[2], n=10	1.4 (0.7 – 4.3)	3.6 (n/a)[5]	1.4 (0.4 – 3.4)
Death (cumulative)	2 (10%)	0 (0%)	2 (13%)
Six Months			
Off dialysis, n (%)	12 (60%)	3 (60%)	9 (60%)
eGFR[1], ml/min/1.73 m^2, n=12	84 (59 – 101)	103 (96 – 145)	63 (54 – 92)
Urine protein:creatinine ratio[2], n=11	0.6 (0.2 – 1.5)	1.3 (0.2 – 1.8)	0.6 (0.2 – 1.3)
End stage kidney disease, n=17	5/17 (29%)	2/5 (40%)	3/12 (25%)
Death (cumulative)	3 (15%)	0 (0%)	3 (20%)
Twelve Months			
Off dialysis, n (%)	13 (65%)	4 (80%)	9 (60%)
eGFR[1], ml/min/1.73 m^2, n=10	65 (57 – 100)	82 (57 – 146)	59 (57 – 85)
Urine protein:creatinine ratio[2], n=9	0.4 (0.2 – 1.3)	1.0 (0.2 – 1.9)	0.4 (0.1 – 0.7)
eGFR > 90 mL/min/1.73 m^2	2/13 (15%)	1/4 (25%)	1/9 (11%)
End stage kidney disease, n=17	4/17 (24%)	1/5 (20%)	3/12 (25%)
Death (cumulative)	3 (15%)	0 (0%)	3[4] (20%)
Infection requiring hospitalization during CYC therapy[3]	8 (40%)	1 (20%)	7 (47%)

Results are presented as mean ± standard deviation for normally distributed continuous data and median with interquartile range (IQR) for non-normally distributed continuous data. Counts with percentages are summarized for categorical data. Decreased "n" represent missing values.
[1]GFR estimated using the modified Schwartz equation
[2]Units in mg protein/mg creatinine
[3]Infections included Cytomegalovirus viremia (2), invasive Aspergillosis (2), Enterococcus urinary tract infection, Streptococcus viridians bacteremia, septic shock of presumed infectious origin, febrile viral illness
[4]Two deaths secondary to invasive Aspergillosis, one unclear etiology
[5]Sample size (n=2) insufficient to calculate IQR
Abbreviations: NIH = National Institutes of Health, EL = EuroLupus, eGFR = estimated glomerular filtration rate, CYC = cyclophosphamide

Disclosure: C. Wang: None; **R. Sadun**: None; **W. Zhou**: None; **K. Miller**: None; **C. Palmer**: None; **S. Ardoin**: None; **C. Bacha**: None; **E. Hause**: None; **J. Hui-Yuen**: None; **N. Ling**: None; **M. Pereira**: None; **M. Riebschleger**: None; **K. Rouster-Stevens**: Accordant, 2; **A. Sarkissian**: None; **J. Shalen**: None; **W. Soulsby**: None; **M. Twilt**: None; **E. Wu**: Enzyvant, 2, Pharming Healthcare, Inc, 2; **L. Lewandowski**: None; **S. Wenderfer**: Alnylam, 1, Bristol-Myers Squibb, 2, Novartis, 1; **J. Cooper**: None.

Abstract Number: 048

Comparative Efficacy and Safety of Ibuprofen and Naproxen in the Treatment of Oligoarticular Juvenile Idiopathic Arthritis (oJIA): Bi-national Cohort Study

Orly Ohana[1], Itay Marmor[2], Liora Harel[3], Shiri Rubin[4], rotem tal[5], Yoel Levinsky[6], Orit Peled[7] and **Gil Amarilyo**[3], [1]Pediatric C ward, Schneider Children's Medical Centre, Petach Tikva, Israel, [2]Dana-Dwek Children's Hospital, Hod Hasharon, Israel, [3]Pediatric rheumatology clinic, Schneider children's medical center of Israel, Petach Tikva, Israel, [4]Pediatric Ear Nose and Throat Unit Unit, Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [5]Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [6]Schneider Children's Medical Center of Israel, Tel Aviv University, Petach Tikva, Israel, [7]Department of Pharmacy, Schneider Children's Medical Center of Israel, Petach Tikva, Israel

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: JIA is the most common childhood rheumatic disease. NSAIDs and intraarticular corticosteroid injections (IACI) are first-line therapy for oJIA. NSAIDs Adverse events (AEs) include gastrointestinal ulcers/bleeding and impaired renal function.

The most prescribed NSAIDs for oJIA are ibuprofen (IBU) and naproxen (NAP). However, direct comparison between these drugs is lacking. We aimed to compare the efficacy and safety of IBU vs. NAP for oJIA.

Methods: This is bi-national retrospective study of oJIA patients from Schneider Childrens Medical Center (SCMCI) and in St. Louis Children's Hospital (STL) treated with either IBU or NAP as first-line therapy. Efficacy was defined as patients that achieved complete response (CR=no evidence for arthritis). Safety was assessed by the occurrence of AEs during follow-up.

Results: Of 171 patients, 104 were treated in SCMCI and 67 in STL. The study population had a mean age of 4.48 years, with female/male ratio ∼2.5:1 (71.9% vs. 28.1%). 110 patients (64%) were treated with NAP and 61 (36%) with IBU. No significant difference was found in drug efficacy [CR 10/61 (16.4%) IBU vs. 19/110 (17.3%) NAP (P=0.83)]. Treatment duration 28 days was associated with significantly higher odds for CR (p=0.017). For safety, 13 AEs were associated with NAP, whereas only one for IBU (p=0.02). Treatment was discontinued in all AEs cases.

Conclusion: IBU and NAP showed similar albeit low efficacy which emphasizes their role as bridging therapy until IACI is achieved. However, IBU showed better safety profile NAP and therefore should be considered as first-line therapy.

Disclosure: O. Ohana: None; **I. Marmor**: None; **L. Harel**: None; **S. Rubin**: None; **r. tal**: None; **Y. Levinsky**: None; **O. Peled**: None; **G. Amarilyo**: None.

Abstract Number: 049

What Happens After Juvenile Myositis Patients Screen Positive for Mental Health Comorbidities? Update from a Multicenter Juvenile Myositis Mental Health Screening Pilot Study

Kaveh Ardalan[1], Rebecca Fillipo[1], Christina ZIgler[2], Audrey Ward[1], Jeffrey Dvergsten[3], Ann Reed[1], Alison Manning[1], Gary Maslow[1], Brian Feldman[4], Ashley Danguecan[5], Sarah Mossad[5], Luana Flores Pereira[5], Susan Shenoi[6], Stacey Haynes[7], Joanna Patten[7] and Andrea Knight[5], [1]Duke University School of Medicine, Durham, NC, [2]Duke, Durham, NC, [3]Duke University Hospital, Durham, NC, [4]Hospital for Sick Children / University of Toronto, Toronto, ON, Canada, [5]The Hospital for Sick Children, Toronto, ON, Canada, [6]Seattle Children's Hospital, Seattle, WA, [7]Seattle Childrens Hospital and Research Center / University of Washington, Seattle, WA

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Juvenile myositis (JM) patients report high rates of emotional distress but qualitative studies suggest challenges accessing high quality mental health care. We present survey data on mental health specialist follow-up for JM patients who screened positive in a multicenter North American JM mental health screening pilot study.

Methods: JM patients/parents (5-21 yo) completed patient/parent-proxy mental health screeners (i.e. Pediatric Symptom Checklist-17 [PSC-17], Patient Health Questionnaire-9 [PHQ9], Screen for Child Anxiety Related Disorders [SCARED]). Demographic/clinical data and Patient-Reported Outcomes Measurement Information System (PROMIS) pediatric/parent-proxy mental health symptom intensity measures were collected. Patients with positive screening, defined as any total/domain scores above established clinical cutoffs, were referred for mental health specialist evaluation. Four weeks after referral, parents (or 18yo+ patients) were emailed surveys evaluating: 1) if they had seen a mental health specialist, 2) barriers to mental health evaluation, 3) facilitators for addressing mental health, and 4) satisfaction with mental health care. Barriers/facilitators were adapted from a prior Patients, Advocates, and Rheumatology Teams Network for Research and Service (PARTNERS) study and respondents could indicate presence/absence of as many as they liked. Descriptive statistics were calculated.

Results: Seventy-two participants enrolled, with 49 (68%) screening positive and 20 completing follow-up surveys (41% response rate). Most (n = 19) respondents were parents and survey responses were more often received for female patients with higher PROMIS physical function and lower PROMIS emotional distress scores, though responders/nonresponders

Table 1: Participant Descriptive Statistics

	Full Cohort (n)	n (%); median (IQR)	Screen Positive (n)	n (%); median (IQR)	Survey Responders (n)	n (%); median (IQR)	Survey Non-responders (n)	n (%); median (IQR)
Diagnosis	72		49		20		29	
Juvenile Dermatomyositis		66 (91.7)		44 (89.9)		19 (95)		25 (86.2)
Overlap Myositis		4 (5.8)		4 (8.2)		1 (5)		3 (10.34)
Immune-Mediated Necrotizing Myopathy		1 (1.4)		1 (2.0)		0 (0)		1 (3.45)
Other Myositis		1 (1.4)		0 (0)		0 (0)		0 (0)
Gender (Female)	72	49 (68.1)	49	35 (71.4)	20	17 (85)	29	18 (62)
Race/Ethnicity (non-Hispanic white)	72	42 (58.3)	49	27 (55.1)	20	11 (55)	29	16 (55.2)
Age at Study Visit (years)	72	11.5 (9-16)	49	12 (10-16)	20	12.3 (10.5-15)	29	12.9 (10-17)
Global Assessments								
Physician's Global Assessment of Total Disease Activity (PGA)	72	1 (0-2.5)	49	1 (0-2.5)	20	2.0 (0-3.25)	29	0 (0-2)
Patient Global Assessment of Total Disease Activity (Pt-GA)	54	1 (0.4-3.9)	41	1 (0.4-3.9)	18	1.65 (0-3.9)	23	0.9 (0.2-5)
Parent Global Assessment of Total Disease Activity (Par-GA)	62	1.55 (0.1-4.1)	41	2.0 (0.1-5)	19	1.6 (0-4.5)	22	2.65 (0.2-5)
PROMIS Physical Function Measures								
PROMIS - Mobility (patient self-report)^	56	54.5 (46.4-57.9)	43	52.5 (45.3-57.9)	18	54.9 (48.7 -61.7)	25	48.1 (45.0-57.9)
PROMIS - Mobility (parent-proxy report)^	67	51.7 (40.4-54.0)	44	48.7 (38.2-54)	19	51.7 (40.6-60.2)	25	41.5 (38.0-52.5)
PROMIS - Upper Extremity (patient self-report)	56	57.3 (45.7-57.3)	43	50.9 (45.4-57.3)	18	54.1 (49.3-57.3)	25	50.5 (45.4-57.3)
PROMIS - Upper Extremity (parent-proxy report)^	67	47.1 (38.6-55.7)	44	47.1 (38-55.7)	19	55.7 (39.9-55.7)	25	46.0 (36.6-55.7)
PROMIS Mental Health Measures								
PROMIS - Depressive Symptoms (patient self-report)^	56	51 (40.1-58.1)	43	53.4 (47.4-60.7)	18	49.6 (44.2-60.7)	25	54.9 (47.4-59.5)
PROMIS - Depressive Symptoms (parent-proxy report)	66	47.1 (40.6-55.8)	43	53.3 (40.6- 59.2)	19	53.7 (39.2-57.6)	24	52.8 (41.9-59.3)
PROMIS - Anxiety (patient self-report)	55	49.9 (36.6-57.9)	42	53.6 (47.1-59.6)	18	53.0 (47.1 -61.2)	24	54.4 (46.2-58.1)
PROMIS – Anxiety (parent-proxy report)^	65	44.4 (33.7-54.8)	43	47.8 (33.7-58.4)	19	44.4 (33.7-56.9)	24	51.3 (39.8-59.3)

*Patient self-report/parent-proxy report measures were respectively administered as follows: PSC-17 (12yo+ self; 5yo+ parent); PHQ9 (12yo+ self only); SCARED (8yo+ both self and parent report); PROMIS computerized adaptive testing item banks (8yo+ self; 5yo+ parent); Pt-GA (8yo+); Par-GA (5yo+)

^PROMIS score difference between responder/nonresponder groups exceeds minimal clinically important difference estimate in JM (Reference: Wolfe M, et al. Estimation of Clinically Important Differences in Patient-Reported Outcomes Measurement Information System (PROMIS) Measures in Juvenile Myositis [abstract]. *Arthritis Rheumatol.* 2020; 72 (suppl 10))

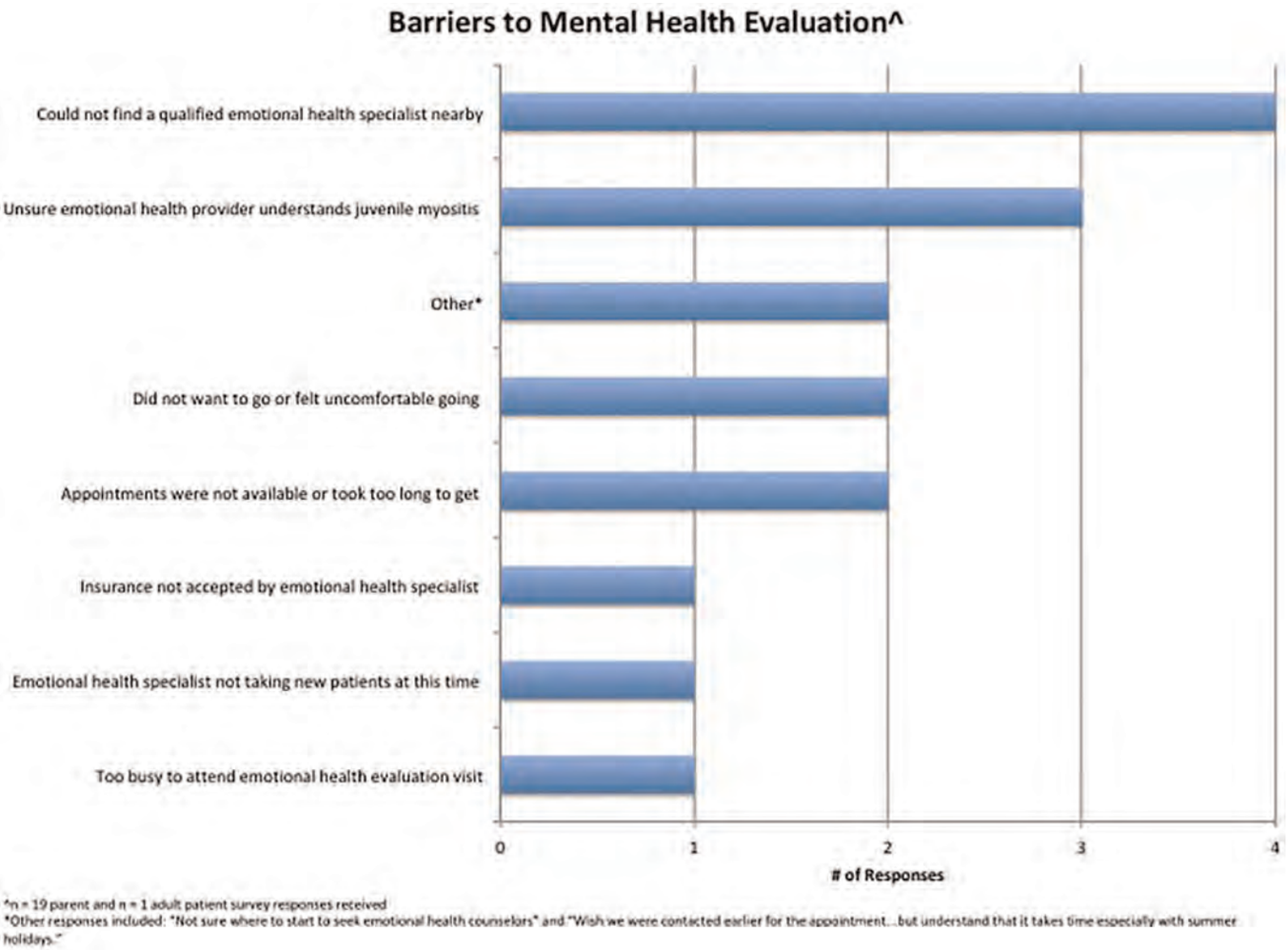


Figure 1: Barriers to Mental Health Evaluation

were otherwise comparable (Table 1). Of the patients screening positive, most patients (n = 15, 75%) had not yet scheduled mental health specialist evaluation, 1 (5%) had scheduled but not attended, and 4 (20%) had visited a specialist with only 1 of these 4 reporting initiation of psychotropic medication. Barriers included difficulty finding nearby specialists who understand JM patients experiences (Figure 1). Facilitators, such as pediatric rheumatologists discussing mental health needs, are shown (Figure 2). A plurality felt mental health needs were being 'very well met (n = 9, 45%), but the majority stated mental health needs were met only 'somewhat (n = 5, 25%) or 'a little bit/not at all (n = 6, 30%).

Conclusion: JM patients with positive mental health screening results did not often access timely mental health specialist follow-up. Barriers and facilitators, such as lack of mental health providers with experience with patients with JM and the role of pediatric rheumatologists in facilitating referral, were identified and replicate prior qualitative study findings. Given that respondents often endorsed unmet mental health needs and difficulty accessing mental health care, novel mental health care delivery approaches (e.g. integrated care) warrant further study to ensure appropriate mental health evaluation and treatment for JM patients.

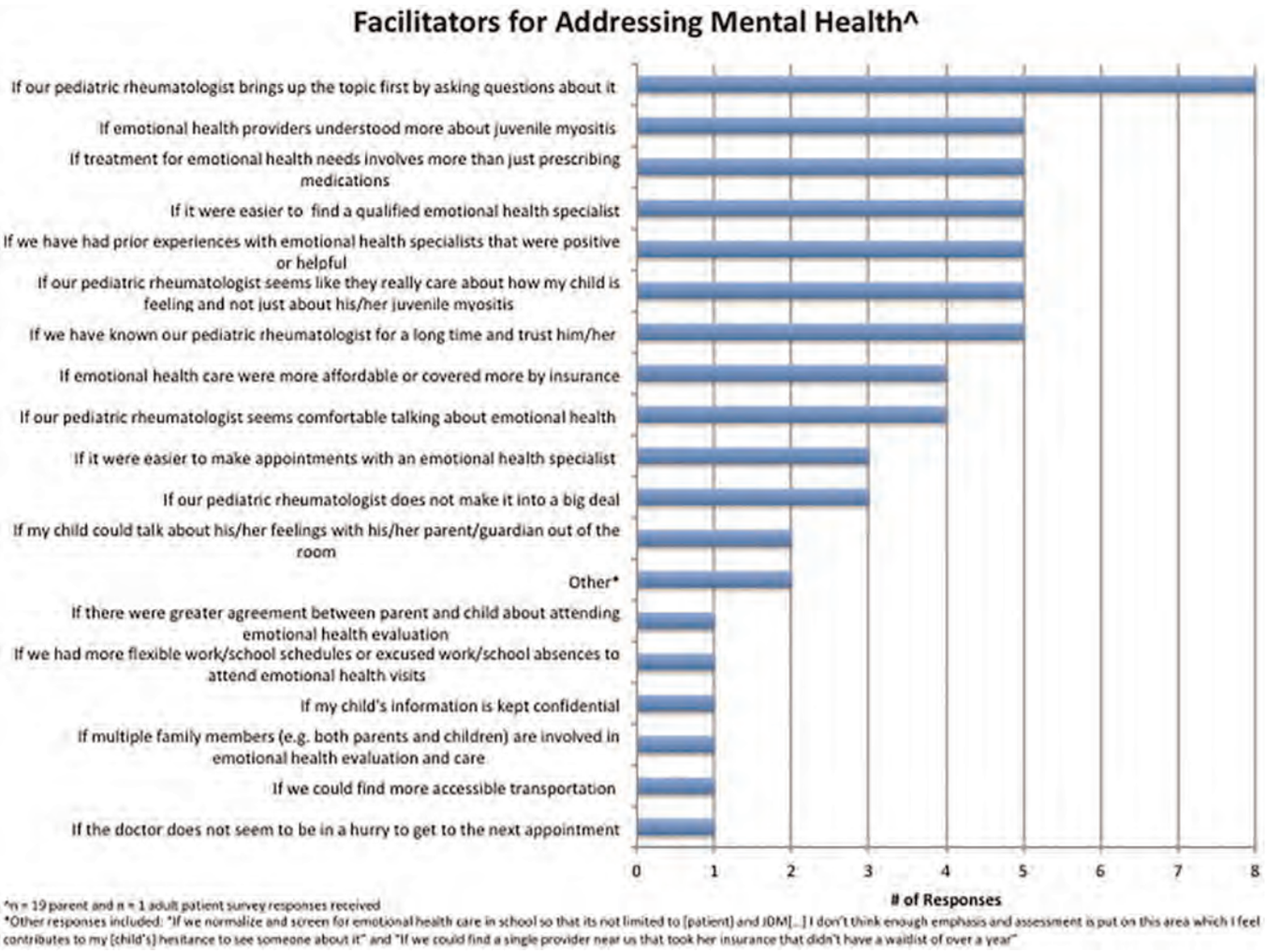


Figure 2: Facilitators for Addressing Mental Health

Disclosure: K. Ardalan: None; **R. Fillipo**: None; **C. Zigler**: None; **A. Ward**: None; **J. Dvergsten**: None; **A. Reed**: None; **A. Manning**: None; **G. Maslow**: None; **B. Feldman**: None; **A. Danguecan**: None; **S. Mossad**: None; **L. Flores Pereira**: None; **S. Shenoi**: Novartis, 2, Pfizer, 1; **S. Haynes**: None; **J. Patten**: None; **A. Knight**: None.

Abstract Number: 050

Predictive Value of the 2019 EULAR/ACR SLE Criteria's Extra-Renal Domains to Renal Response One Year After Treatment in a Pediatric Lupus Nephritis Cohort

Sara Patrizi[1], Megha Tandel[2], Derek Boothroyd[2] and Joyce Hsu[1], [1]Stanford Medicine, Children's Health, Palo Alto, CA, [2]Quantitative Sciences Unit, Department of Medicine, Stanford University School of Medicine, Palo Alto, CA

SESSION INFORMATION

Session Date: Thursday, March 30, 2023

Session Title: Posters: Clinical and Therapeutic I

Session Type: Poster Session A

Session Time: 6:00PM–7:00PM

Background/Purpose: In 2019, new classification criteria for SLE were developed by the EULAR/ACR. Prior research in adult lupus cohorts found a positive correlation between high summary score at diagnosis and SLE disease activity at 5 years, but it is unknown whether a higher summary score indicates more severe disease in a pediatric cohort. Using the new lupus criteria, this pediatric study evaluates the predictive value of higher summary scores and number of extra-renal domains at diagnosis to renal outcomes after treatment.

Methods: This is a retrospective, single center cohort study of all patients seen at Stanford Childrens Health diagnosed with-SLEwith kidney involvement prior to their 18th birthday from 2010- 2022. For each patient, the EULAR/ACR summary score was calculated based on the clinical and laboratory criteria. Patients were dichotomized into two subgroups, those with low/moderate number (1-5) of extra-renal domains at diagnosis vs those with a high number (6-9). Patient baseline information was collected at diagnosis and follow up data was collected after 1 year. The Childhood Arthritis and Rheumatology Research Alliance (CARRA) 2012 definition for renal response was utilized (no, partial [mild/moderate] or complete response) after 1 year of treatment as our outcome measure. Descriptive statistics were reported by group and an ordinal logistic regression was conducted to calculate adjusted odds ratio for renal response to determine if a higher summary score (30) and high number of extra-renal domains predicted poor renal response.

Table 1. Patient Demographics, Clinical Characteristics and EULAR/ACR Classification Domains (N=74)

	Total N=74 % (n)	Low/Moderate (1-5 Domains) n=37 % (n)	High (6-9 Domains) n=37 % (n)	SMD (*d*)#
Age (years), mean (SD)	12.8 (2.8)	13.2 (3.1)	12.3 (2.5)	-0.33
Gender				0.39
Female	76 (56)	68 (25)	84 (31)	
Male	24 (18)	32 (12)	16 (6)	
Race/Ethnicity				0.72
Asian	42 (31)	32 (12)	51 (19)	
Black	4 (3)	9 (3)	0 (0)	
Hispanic	34 (25)	32 (12)	36 (13)	
White	19 (14)	27 (10)	11 (4)	
Other	1 (1)	0 (0)	2 (1)	
Insurance				0.30
Private	54 (40)	59 (22)	49 (18)	
Public	46 (34)	41 (15)	51 (19)	
Duration of Disease Prior to Diagnosis (months), median (IQR)	2 (1, 3)	2 (1, 6)	1 (1, 3)	-0.32
Treatment				0.09
Cellcept	61 (45)	59 (22)	62 (23)	
Cytoxan	27 (20)	27 (10)	27 (10)	
Other	12 (9)	14 (5)	11 (4)	
Blood Pressure Medication at Diagnosis^	76 (56)	84 (31)	68 (25)	0.38
SLEDAI Score at Diagnosis, median (IQR)*	21 (17, 25)	18 (14, 22)	24 (19, 29)	1.07
SLEDAI Score at Follow-up, median (IQR)*	3.5 (0, 7)	4 (2, 6)	2 (0, 8)	0.14
ANA Titer Levels at Diagnosis				
1:80	5 (4)	8 (3)	3 (1)	0.24
1:160	10 (7)	14 (5)	5 (2)	0.28
1:320	12 (9)	11 (4)	14 (5)	0.08
1:640	7 (5)	5 (2)	8 (3)	0.11
1:1280	19 (14)	14 (5)	24 (9)	0.28
≥1:1280	41 (31)	43 (16)	41 (15)	0.05
EULAR/ACR Classification Domains at Diagnosis				
Constitutional	57 (42)	30 (11)	84 (31)	1.30
Hematologic	70 (52)	51 (19)	89 (33)	0.91
Neuropsychiatric	11 (8)	5 (2)	16 (6)	0.35
Mucocutaneous	69 (51)	46 (17)	92 (34)	1.14
Serosal	42 (31)	43 (16)	41 (15)	-0.05
Musculoskeletal	42 (31)	22 (8)	62 (23)	0.90
APL antibodies	58 (43)	41 (15)	76 (28)	0.76
Complements	93 (69)	89 (33)	97 (36)	0.33
SLE-specific antibodies	92 (68)	86 (32)	97 (36)	0.40

Footnotes:
^Patients could have had one or more of the following medications at diagnosis: Clonidine, Amlodipine, Chlorthalidone, Lasix or Lisinopril
*SLEDAI ranges from 0 to 105
#Standard Mean Difference (SMD): (d) ≤0.2 low, 0.5 medium, ≥0.8 large difference

Results: Patient characteristics of our final cohort of 74 children are depicted in Table 1. Patients with a high number of extra-renal domains have 1.47 (95% CI: 0.55 – 2.91, p=0.44) times the odds of having a complete renal response than patients with a low/moderate number of domains (Table 2).

There were no statistically significant differences between the summary scores and the three renal response groups (Figure 1), although the no renal response group were comprised of more patients with a summary score30. Patients with a summary score 30 have 1.31 (95% CI: 0.50-3.44, p=0.59) times the odds of having a complete renal response than patients with a summary score 30 (Table 2).

Conclusion: In this study, we evaluated the predictive potential of the extra-renal domains and the summary score from the 2019 EULAR/ACR classification criteria in a pediatric cohort with SLE with renal involvement. We found that the number of extra-renal domains at diagnosis did not have a significant impact on renal response at one year. While there was no significant difference in the overall EULAR/ACR summary score at diagnosis and renal response after 1 year of treatment, patients with a summary score 30 had an increased trend toward having a complete renal response. This suggests that the overall summary score (a more quantitative, weighted score), rather than the number of domains, may predict renal outcomes in a pediatric lupus nephritis cohort. Future studies on larger cohorts are needed.

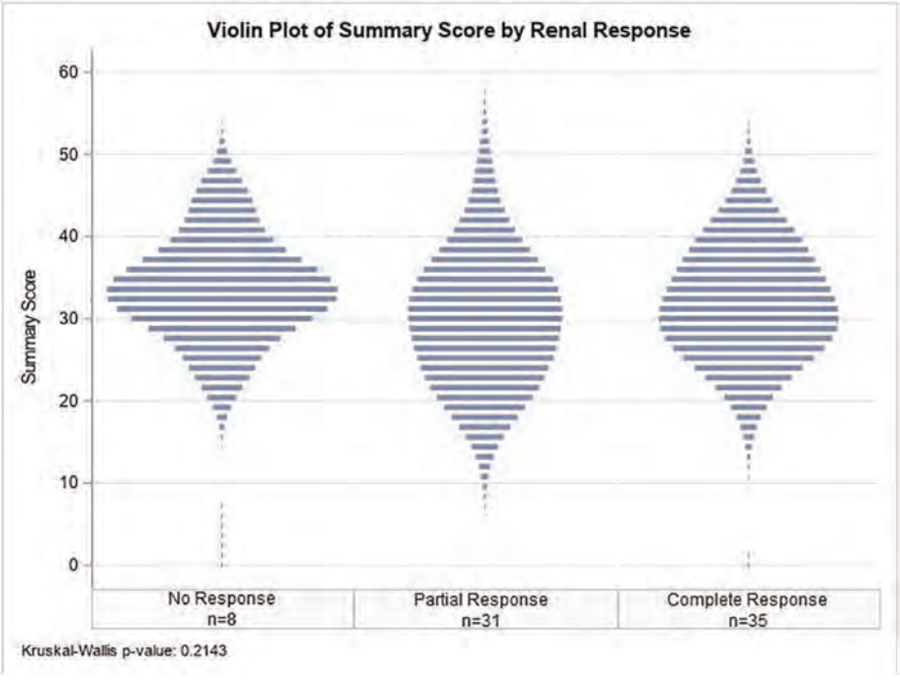


Figure 1. Overall Summary Score by Renal Response (N=74)

Table 2. Ordinal Logistic Regressions for Complete Renal Response (N=74)

	EULAR/ACR Extra-Renal Manifestation Domains		EULAR/ACR Overall Classification Summary Scores	
	OR (95% CI)	p-value	OR (95% CI)	p-value
Domain (referent: Low/Moderate)	1.47 (0.55 – 2.91)	0.4402		
Summary Score (referent: ≥30)			1.31 (0.50 – 3.44)	0.5880
Gender (referent: Female)	2.79 (0.90 – 8.81)	0.0786	2.43 (0.80 – 7.36)	0.1173
Race/Ethnicity (referent: White)	0.51 (0.15 – 1.75)	0.2834	0.57 (0.17 – 1.92)	0.3630
Insurance (referent: Public)	1.69 (0.67 – 4.27)	0.2644	1.78 (0.70 – 4.57)	0.2272
Duration of symptoms prior to diagnosis	1.02 (0.89 – 1.17)	0.9975	0.99 (0.87 – 1.14)	0.9337

Disclosure: S. Patrizi: None; **M. Tandel**: None; **D. Boothroyd**: None; **J. Hsu**: None.

Abstract Number: 051

Juvenile Relapsing Polychondritis: A Single Center Cohort

Angela Chun[1], Sarah Molina[1], Marietta De Guzman[2] and Maria Pereira Palacios[1], [1]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [2]Baylor College of Medicine, Texas Children's Hospital, Houston, TX

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Relapsing polychondritis (RPC) is a rare multisystemic autoimmune disease associated with recurrent, progressive inflammation in cartilaginous structures. In children, the most common manifestations are auricular chondritis, arthritis, and respiratory tract involvement, with some studies reporting increased severity of laryngotracheal complications compared to adults. Given the paucity of literature describing RPC in pediatrics, this retrospective chart review aims to provide additional data on five patients regarding clinical presentation, management, and outcomes.

Methods: With approval by our institutional review board, we retrospectively reviewed the clinical presentation, management, and outcomes of patients clinically diagnosed with RPC before 18 years of age at Texas Childrens Hospital between January 2004 and December 2022. The diagnosis was defined according to the modified McAdams criteria. We reported the Relapsing Polychondritis Disease Activity Index (RPDAI) scores at the initial and final clinic visits. Remission was defined as having less than 2 exacerbations per year.

Results: Of the five patients diagnosed with RPC, all were male, with a mean age of 9.6 years (range: 5-15 years) at presentation. The mean time to diagnosis was 18.4 months (range: 5-48 months). All patients had auricular chondritis, which was the presenting complaint for 4. Inflammatory arthritis was the next most common symptom in 3 patients, followed by nasal and bronchial chondritis in 2 and 1 with ocular involvement. The clinical presentations and initial labs/imaging are summarized in Table 1.

Two patients with tracheal chondritis presented in acute respiratory failure requiring emergent tracheostomy. One had dense calcific deposits on CT with resultant bronchial stenosis, and the other had evidence of bronchomalacia. A patient who reported vertigo during episodes of auricular chondritis underwent a CT temporal bone that revealed a soft tissue mass projecting into the external auditory canal. Two others developed conductive and sensorineural hearing loss. None developed a saddle nose deformity. Evaluation of cardiac manifestations noted one patient with mild aortic sinotubular junction dilation and another with tricuspid regurgitation. The mean RDPAI index on presentation was 27.6 (range: 9-61) and decreased by 48% from the first to the last visit.

Of the four patients who received treatment, all were given prednisone and methotrexate. Three patients received additional immune modulation, including anti-TNF agents and cyclophosphamide. Three achieved clinical remission. One patient required no therapy and remains in remission after five years. No secondary systemic autoimmune diseases were identified, or deaths reported.

Conclusion: Relapsing polychondritis is a rare autoimmune disease in children with variable severity, clinical course, and response to treatment. In our cohort, none developed a concomitant systemic autoimmune disease. Treatment with immunosuppression resulted in a decrease in RPDAI scores and clinical remission in 60%. Additional prospective studies are needed to determine the optimal treatment course and the validity of RPDAI for use in clinical practice.

Table 1

Gender, Age of Diagnosis	Time to Diagnosis (months)	Modified McAdam Criteria Met	Additional Presenting Features	Labs	Imaging	Complications	Current Treatment	Clinical Remission	Past Therapies	RDPAI Score at Presentation -> Last Visit	Mean Duration of Follow-Up (months)
M, 10	5	Bilateral auricular chondritis Nasal chondritis Respiratory tract chondritis	None	WBC 14.84 Hg/Hct 11.2/33.2 CRP 8.3 ESR 48 ANA Positive, 1:80	CT Neck/Chest: Severe subglottic tracheal stenosis, moderate-severe left and mild right bronchomalacia	Subglottic stenosis with respiratory failure requiring tracheostomy	Infliximab 5 mg/kg every 8 weeks	Yes	Prednisone Methotrexate	34 -> 9	87
M, 11	18	Bilateral auricular chondritis Nasal chondritis Nonerosive seronegative inflammatory arthritis Cochlear and/or vestibular dysfunction	Rash	WBC 7.1 Hg/Hct 12.5/36.7 CRP <0.5 ESR 5 ANA Negative Collagen Ab Positive, 21	CT Temporal Bones: Small soft tissue mass projecting into the superior anterior aspect of external auditory canal	Inner ear involvement	Adalimumab 40 mg every 2 weeks	No	Prednisone Methotrexate	22 -> 22	14
M, 5	11	Bilateral auricular chondritis Nonerosive seronegative inflammatory arthritis + Steroid responsive	Fever	WBC 6.9 Hg/Hct 11.6/34.1 CRP <0.5 ESR 9 ANA Negative	MRI whole body: Small amounts of fluid in shoulder, knee, ankle joint Echo: Aortic root at upper limits of normal, sinotubular junction mildly dilated, normal biventricular systolic function	Dilated sinotubular junction	Methotrexate 16 mg/m2 weekly	Yes	Prednisone	12 -> 0	34
M, 7	48	Bilateral auricular chondritis Cochlear and/or vestibular dysfunction	None	WBC 11.1 Hg/Hct 11.9/35.7	XRay Neck: Normal XRay Chest: Normal PFTs: Normal Echo: Normal	None	None	Yes	None	9 -> 0	60
M, 15	10	Bilateral auricular chondritis Respiratory tract chondritis Nonerosive seronegative inflammatory arthritis Ocular inflammation Cochlear and/or vestibular dysfunction	None	WBC 15.9 Hg/Hct 7.4/25 CRP 25.7 ESR 116 ANA Negative Collegen Ab Positive, 42.1	MRI Knee: Synovial thickening, enhancement, and suprapatellar effusion CT Chest/Abdomen/Pelvis: Diffuse mild thickening of wall of the trachea and major bronchi sparing the posterior membraneous wall of trachea. Multiple foci of dense calcific deposits within the wall of the major airways and laryngeal cartilages. Concentric stenosis of the mid portion of the right mainstem bronchus with reduction of luminal caliber by ~50%. CT Neck: Abnormal calcification of thyroid cartilage, cricoid cartilage, and region of the arytenoid cartilage. Abnormal incomplete ring-like calcification around the trachea with thickening of tracheal wall. No destructive lesion of nasal septum.	Subglottic stenosis with respiratory failure requiring tracheostomy Sensorineural hearing loss Ocular inflammation leading to impaired vision Trivial tricuspid regurgitation	Prednisone 20 mg daily Methotrexate 25 mg weekly Adalimumab 40 mg weekly	No	Prednisone Cyclophosphamide Etanercept Dapsone	61 -> 36	61

WBC = White Blood Cell in x10^3 /µL, Hg/Hct = Hemoglobin/Hematocrit in g/dL, CRP = C-reactive protein in mg/L, ESR = Erythrocyte sedimentation rate in mm/h

Disclosure: A. Chun: None; **S. Molina**: None; **M. De Guzman**: None; **M. Pereira Palacios**: None.

Abstract Number: 052

Extreme Periodic Fever, Aphthous Stomatitis, Pharyngitis, Adenitis (PFAPA): A Discrete Group of Patients

Yoel Levinsky[1], Rotem Tal[2], Liora Harel[2], Shoval Shoham[3], Sabreen Abu Ahmad[4], Yonatan Butbul Aviel[5], Gil Amarilyo[2] and **Mor Broide**[3], [1]Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [2]Pediatric rheumatology clinic, Schneider children's medical center of Israel, Petach Tikva, Israel, [3]Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [4]Ruth Rappaport Children's Hospital, Rambam Health Care, Haifa, Israel, [5]Rambam Medical center, Haifa, Israel

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Periodic fever, aphthous stomatitis, pharyngitis, and cervical adenitis (PFAPA) syndrome is the most common periodic fever syndrome in children; by definition, episodes occur every 2 to 8 weeks. However, in a subset of our patients, we noticed a higher frequency of attacks, of less than 2 weeks, which we refer to as extreme PFAPA (ePFAPA). This group consisted of patients who were extreme upon presentation of PFAPA, and those who became extreme after initiation of abortive corticosteroid treatment.

We aimed to characterize demographic and clinical features of these two groups and to compare them to patients with non-extreme PFAPA (nPFAPA).

Methods: The medical records of 365 patients with PFAPA who attended Schneider Children's Medical Center of Israel from March 2014 to April 2021 were reviewed. Patients with concomitant familial Mediterranean fever were excluded. Characteristics of the ePFAPA (including subgroups) and nPFAPA groups were compared using Wilcoxon rank sum, Pearson's chi-squared, and Fisher's exact tests.

Results: Forty-seven patients (12.9%) were identified as having ePFAPA.Among patients with ePFAPA, compared to patients with nPFAPA, disease onset (median (interquartile range)) was earlier: 1.5 years (0.7-2.5) vs 2.5 years (1.5-4.0), P< 0.001; and diagnosis was younger: 2.6 years (2.0-3.6) vs 4.5 years (3.0-6.2), P< 0.001. A higher proportion of patients with ePFAPA than nPFAPA were treated with colchicine prophylaxis (53% vs 19%, P< 0.001), but symptoms and signs during flares did not differ significantly between these groups. Demographic and clinical characteristics were similar between patients with ePFAPA from presentation of PFAPA (22, 47% of those with ePFAPA) and with ePFAPA from after corticosteroid treatment.

Conclusion: About half the patients categorized with ePFAPAsyndrome met the criteria upon presentation with PFAPA. Patients with ePFAPA compared to nPFAPA presented and were diagnosed at an earlier age

Disclosure: Y. Levinsky: None; **R. Tal**: None; **L. Harel**: None; **S. Shoham**: None; **S. Abu Ahmad**: None; **Y. Butbul Aviel**: None; **G. Amarilyo**: None; **M. Broide**: None.

Abstract Number: 053

Can Children with Colchicine Resistant FMF Be Treated with on Demand Canakinumab Regimen?– a Multicenter Study

Katy shehadeh[1], Yoel Levinsky[2], rotem tal[3], Neta Hana Aviran[3], Yonatan Butbul Aviel[4], Irit Tirosh[5], Shelly Kagan[6], Tarek Zoabi[3], Shiri Spielman[7], Adi Miller-Barmak[4], Rotem Semo Oz[8], Liora Harel[9], Gabriel Chodick[10] and **Gil Amarilyo**[6], [1]Tel Aviv University, Tel Aviv, Israel, [2]Schneider Children's Medical Center of Israel, Tel Aviv University, Petach Tikva, Israel, [3]Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [4]Rambam Medical center, Haifa, Israel, [5]Sheba Medical Center, Savyon, Israel, [6]Schneider Children's Medical Center of Israel, Petach Tikva, Israel, [7]Sheba Tel-HaShomer Medical Center, Givataim, Israel, [8]Sheba medical center, Herzelyia, Israel, [9]Scheiders Children Medical Center of Israel, Petah-Tiqva, Israel, [10]Maccabitech institute for research and innovation, Maccabi healthcare services, Tel Aviv, Israel

SESSION INFORMATION

Session Date: Thursday, March 30, 2023

Session Title: Posters: Clinical and Therapeutic I

Session Type: Poster Session A

Session Time: 6:00PM–7:00PM

Background/Purpose: Familial Mediterranean fever (FMF) is the most common autoinflammatory disease. Without therapy, it may lead to the development of secondary amyloidosis. Treatment with colchicine leads to long-term remission in ~70% of patients. 5% are resistant to colchicine therapy (crFMF) and may be treated with monthly dose of canakinumab (anti IL-1beta). However, colchicine, the only drug proved to prevent secondary amyloidosis. Canakinumab is immunosuppressive as well as expensive. Therefore, we aimed to compare on demand canakinumab (COD) dosage policy vs. canakinumab fixed frequency (CFF) policy.

Methods: Data from 3 Israeli pediatric rheumatology centers (Schneider Children's Medical Center of Israel, Sheba Tel-HaShomer Medical Center, Rambam Health Care Campus) were collected regarding crFMF patients treated with canakinumab. crFMF patients treated according to the COD policy were given 1 dose of sc-canakinumab injection 4mg/kg (max 150mg), with subsequent doses administered only after an additional attack. CFF patients were given fixed monthly doses according to the manufacturer instructions.

Results: Overall, 51 crFMF (25 COD vs. 26 FCC) with mean follow-up of 22.6 months were included. There were no significant demographic, clinical or genetic differences between the groups. The COD group received significantly lower cumulative canakinumab dosage during the follow-up period (15.688.95mg/kg vs.32.58.05mg/kg; P< 0.001). There were no differences between groups in mean FMF attacks nor in mean CRP levels at the end of follow-up period. None the less, the COD group necessitated higher colchicine doses (0.050.01mg/kg vs. 0.030.01mg/kg; P< 0.001). Overall, 51 crFMF (25 COD vs. 26 FCC) with mean follow-up of 22.6 months were included. There were no significant demographic, clinical or genetic differences between the groups. The COD group received significantly lower cumulative canakinumab dosage during the follow-up period (15.688.95mg/kg vs.32.58.05mg/kg; P< 0.001). There were no differences between groups in mean FMF attacks nor in mean CRP levels at the end of follow-up period. None the less, the COD group necessitated higher colchicine doses (0.050.01mg/kg vs. 0.030.01mg/kg; P< 0.001).

Conclusion: COD treatment in crFMF patients is as effective as CFF treatment. Using COD can reduce drug expenses and decrease immunosuppression exposure without negatively influencing the disease control.

Disclosure: K. shehadeh: None; **Y. Levinsky**: None; **r. tal**: None; **N. Aviran**: None; **Y. Butbul Aviel**: None; **I. Tirosh**: None; **S. Kagan**: None; **T. Zoabi**: None; **S. Spielman**: None; **A. Miller-Barmak**: None; **R. Semo Oz**: None; **L. Harel**: None; **G. Chodick**: None; **G. Amarilyo**: None.

Abstract Number: 054

Craniofacial Localized Scleroderma: A Single Center Retrospective Cohort

Leigh Stubbs, Ammar Hashemi, Raegan Hunt and Renata Maricevich, Baylor College of Medicine, Houston, TX

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Craniofacial localized scleroderma (LS) can lead to disfigurement and severe extracutaneous manifestations (ECMs). There is an ongoing need to standardize multidisciplinary evaluation and care. In this retrospective cohort, we report the multidisciplinary assessment and treatment over ten years at one center for 77 patients.

Methods: A retrospective Institutional Review Board-approved chart review was performed on 393 patients diagnosed with LS at Texas Childrens Hospital from January 1, 2012, to October 26, 2022. Patient inclusion criteria were as follows: 1) age < 18 at clinical diagnosis; 2) LS diagnosis using the Padua classification criteria; 3) confirmation of diagnosis by a pediatric rheumatologist, dermatologist, or plastic surgeon; 4) LS lesion involving the craniofacial area. Patients were excluded if any of the above criteria were not met as well as miscoding or insufficient medical records (Figure 1).

Results: Demographic and clinical information was collected for 77 patients who met the inclusion criteria. The mean age of diagnosis was 9 (range: 2-17) years. Most patients were female (64%) and Hispanic (55%). Diagnoses included linear scleroderma of the face/scalp (39%), Parry Romberg syndrome (21%), both (18%), mixed morphea (17%), and circumscribed morphea (5%). In most cases, a pediatric dermatologist (66.2%) made the diagnosis; 28 patients (36%) had a skin biopsy to confirm the clinical diagnosis. In 23% of patient charts, providers noted that the lesions would "burn out" or appear "burnt out." 27% of patients had a recurrence. Patients had a documented evaluation by the following pediatric specialists:

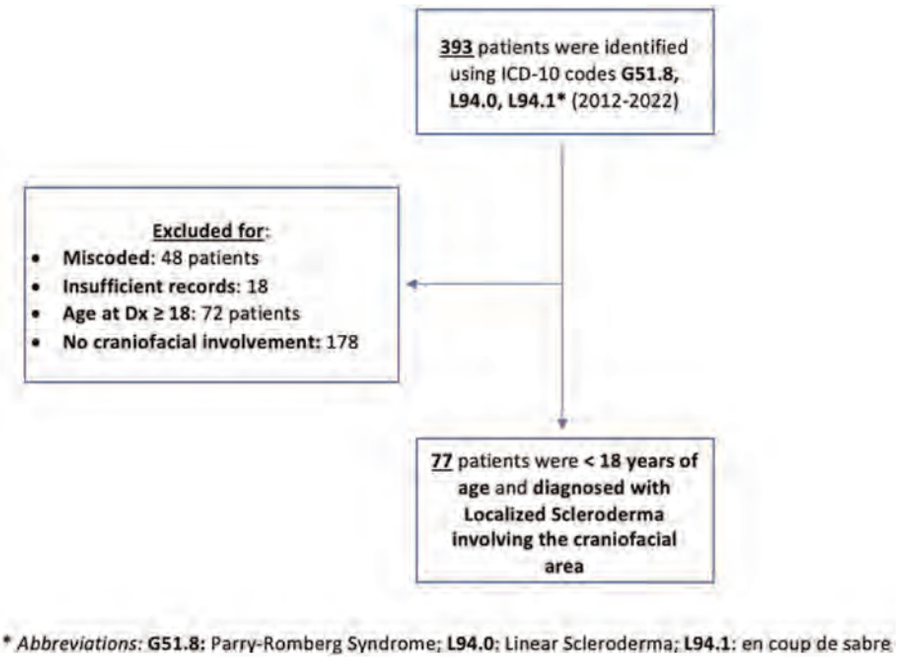


Figure 1. Retrospective cohort study design and flow chart.

Table 1. Extra-cutaneous manifestations in craniofacial localized scleroderma patients.

Extra-Cutaneous Manifestation	Number of Patients with Specific ECM	% of Total Cohort n = 77	% from Patients within ECM category
Neurological	26	33.8%	-
Headache	21	27.3%	80.8%
Cranial nerve palsy	5	6.5%	19.2%
Seizure	4	5.2%	15.4%
Other[a]	3	3.9%	11.5%
Psychological	26	33.8%	-
School issues (bullying, learning issues)	16	20.8%	61.5%
Mood disorder (depression, anxiety)	14	18.2%	53.8%
Referral to psychology/psychiatrist	12	15.6%	46.2%
Other[b]	10	13.0%	38.5%
Dental[c]	22	28.6%	-
Ophthalmological	15	19.5%	-
Dry Eye	10	13.0%	66.7%
Enophthalmos	8	10.4%	53.3%
Lagophthalmos	4	5.2%	26.7%
Other[d]	2	2.6%	13.3%
Uveitis	1	1.3%	6.7%
Musculoskeletal	11	14.3%	-
Arthritis	6	7.8%	54.5%
Arthralgia	4	5.2%	36.4%
Contractures	3	3.4%	27.3%
Limb Length Difference	3	3.4%	27.3%
Other functional limitations[e]	1	1.3%	9.1%

[a]Dizziness, Rasmussen's encephalitis, trigeminal neuralgia
[b]Attention-deficit/hyperactivity disorder, schizophrenia
[c]Shorten dental roots, missing secondary teeth, alveolar resorption, mandibular issues, gingival recession, atrophy of tongue
[d]Epiphora, penetrating keratoplasty
[e]Myositis

dermatology (84%), rheumatology (78%), ophthalmology (65%), dental (56%), and plastic surgery (47%). Of the 58 patients (75%) with an MRI brain or face, 41 had abnormal findings. ECMs included neurological (34%), psychological (34%), dental (29%), ophthalmological (20%), and musculoskeletal (14%) (Table 1). There were 55 patients (71%) on systemic medications including corticosteroids (76.4%), methotrexate (96.4%), mycophenolate mofetil (16.4%), biologics (7.3%), and other medications (hydroxychloroquine or intravenous immunoglobulin, 36.4%). Twenty-four patients (31%) have undergone surgery at a mean age of 13 (range: 5-18). The most common surgical procedure was an autologous fat graft (n =22) followed by rhinoplasty (n=2), eye repair (n=1), flap (n=1), hyaluronic filler (n=1), implant (n=1), and osteotomy (n=1). The mean number of fat grafts per patient was 2 (range: 1-7). The only surgical complications noted were a poor cosmetic outcome for the flap procedure and chronic infection of the malar implant leading to removal. Death occurred in one patient secondary to status epilepticus.

Conclusion: Despite published evaluation algorithms, there are still gaps in comprehensive multidisciplinary care for craniofacial LS. There needs to be improved collaboration with ophthalmology, dental, plastic surgery, and psychology. Topical treatment alone is inadequate, so referral to pediatric rheumatology for systemic immunosuppressant treatment is critical. Given the risk of recurrence, ongoing monitoring is essential.

Disclosure: L. Stubbs: None; **A. Hashemi**: None; **R. Hunt**: None; **R. Maricevich**: None.

Abstract Number: 055

COVID-19 Vaccination in Children with Rheumatic Diseases: Results of a CARRA-wide Survey

Beth Rutstein[1], Merav Heshin Bekenstein[2], Maria Schletzbaum[3], Nora Singer[4], Rebecca Sadun[5], Melanie Kohlheim[6], Vincent Del Gaizo[7], Kelly Wise[8], Melica Nikahd[9], Guy Brock[9], Monica Ardura[8], Vidya Sivaraman[10] and For the CARRA Investigators[11], [1]Children's Hospital of Philadelphia, Philadelphia, PA, [2]Tel Aviv Medical Center, Tel Aviv, Israel, [3]Medical College of Wisconsin, Milwaukee, WI, [4]Metro Health, Cleveland, OH, [5]Duke University, Durham, NC, [6]None, Columbus, OH, [7]CARRA, Washington, DC, [8]Nationwide Children's Hospital, Columbus, OH, [9]The Ohio State University, Columbus, OH, [10]Nationwide Children's Hospital/ The Ohio State University, Columbus, OH, [11]Duke University Medical Center, Durham, NC

SESSION INFORMATION

Session Date: Thursday, March 30, 2023

Session Title: Posters: Clinical and Therapeutic I

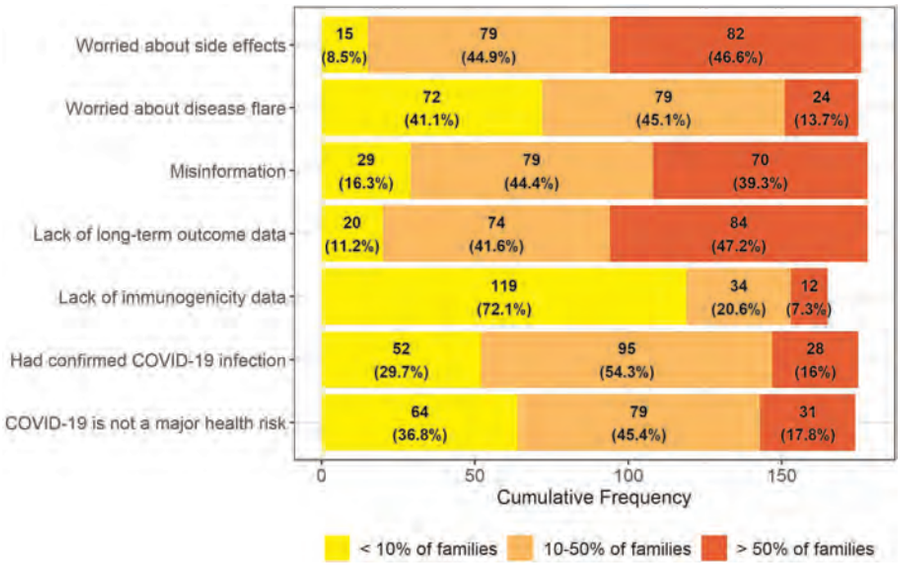
Session Type: Poster Session A

Session Time: 6:00PM–7:00PM

Background/Purpose: Children receiving immunosuppressive therapies (IST) have a higher risk of hospitalization from COVID-19. COVID-19 vaccines significantly reduce the likelihood of severe disease or death. Early studies demonstrate safety and immunogenicity of COVID-19 vaccines in children with rheumatic diseases. Yet, as of December 2022, 63% of children 6 months to 17 years of age remain unvaccinated against COVID-19. The CARRA Vaccination Working Group (WG) surveyed pediatric rheumatologists to evaluate current COVID-19 vaccination practices, vaccine hesitancy and barriers.

Methods: The CARRA Vaccination WG developed and distributed a survey to CARRA member healthcare providers from March-May 2022. The survey included questions about COVID-19 vaccination practices and provider report of parent perceptions about COVID-19 vaccination for their children. Results were collected via RedCap and responses were analyzed.

Results: The survey was completed by 219 members, with 74% pediatric rheumatologists and 21% fellows. The majority (98%) opinion was that disease flares after COVID-19 vaccination would be mild and/or rare. Provider concerns about vaccine-associated adverse events (AEs) included risk of myocarditis (76%), new autoimmune conditions (29%), and



Reasons for patients/ families declining COVID-19 vaccination per rheumatology providers

thrombosis (22%). These AEs were ranked as low risk, with 98% of providers recommending COVID-19 vaccines for their patients. Concerns of decreased vaccine efficacy were reported by 59%, particularly among patients receiving the following IST: rituximab (100%), systemic corticosteroids (86%), mycophenolate mofetil (59%), and JAK-inhibitors (46%). IST was temporarily modified for vaccination by 88% of providers, mostly based on ACR guidelines. Most providers (82%) did not routinely check post-vaccination serology and the remainder did so primarily for research purposes. Notably, 98% of providers reported encountering parents declining COVID-19 vaccination, with 75% reporting hesitancy in >10% of patients. In contrast, for routine vaccines, only 25% reported hesitancy in >10% of patients. Reported reasons for parent COVID-19 vaccine hesitancy were concerns about side effects, lack of long-term safety data, prior COVID-19 infection, and vaccine misinformation such as risk of infertility, genetic changes, or vaccination leading to COVID-19 infection.

Conclusion: Our survey showed discordance between provider and perceived parent opinions regarding COVID-19 vaccines for children with rheumatic diseases. Concerns about vaccine efficacy and AEs, including vaccine-associated myocarditis, did not reduce provider recommendations for COVID-19 vaccination. In contrast, parent concern of vaccine side effects, lack of long-term safety data, and misinformation reduced the likelihood of parents agreeing to vaccinate their child. These results highlight discrepancies between providers and parents of children with rheumatic disease in balancing the risks and benefits of COVID-19 vaccination. Limitations of the study are that providers rather than parents provided the reasons for vaccine hesitancy. Our study underscores the need to survey parents directly and include parents in the design phase of COVID-19 vaccination studies for children.

Disclosure: B. Rutstein: None; **M. Heshin Bekenstein**: None; **M. Schletzbaum**: None; **N. Singer**: None; **R. Sadun**: None; **M. Kohlheim**: None; **V. Del Gaizo**: None; **K. Wise**: None; **M. Nikahd**: None; **G. Brock**: None; **M. Ardura**: None; **V. Sivaraman**: None; **F. CARRA Investigators**: None.

Abstract Number: 056

An Advanced Physiotherapist Practitioner Model of Care Is Ideally Suited to Address Workforce Concerns in Pediatric Rheumatology: A Retrospective Chart Review

Julie Herrington[1], KAREN BEATTIE[1] and Michelle Batthish[2], [1]McMaster University, Hamilton, ON, Canada, [2]McMaster Children's Hospital, Hamilton, ON, Canada

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: An Advanced Physiotherapist Practitioner (APP) role was created in September 2020 at McMaster Children's Hospital, Ontario, Canada to support the growing demand for service within Pediatric Rheumatology. The APP assesses musculoskeletal focused referrals, orders and interprets investigations and labs, and provides management of these cases under medical directives. The purpose of this study is to evaluate the characteristics and outcomes of patients assessed by the APP as well as to investigate access to care for patients referred to Pediatric Rheumatology.

Methods: A retrospective chart review of initial patient assessments by the APP was performed on patients referred to pediatric rheumatology and triaged to the APP from September 2020 to December 2021. Extracted data included demographic characteristics of patients, wait times, clinical outcomes, and frequency of investigations performed requiring a medical directive.

Results: Initial triage (urgent, semi-urgent or non-urgent) by a Pediatric Rheumatologist assigned 118 patients to the APP. Of these, 70% were female, 61% were 10-15 years old and 50% were referred by primary care providers. The most common reason for referral was joint pain (87%). The APP saw cases from all triage categories including 17 (14%) urgent, 76 (65%) semi-urgent, 25 (21%) non-urgent. Of 17 cases deemed urgent, 14 (82%) were seen within the national benchmark of 4 weeks. After assessment, 25 (21%) had a confirmed rheumatic disease and 93 (79%) were considered non-rheumatic and discharged. A physiotherapy diagnosis was provided in 76 (64%) cases and physiotherapy interventions were provided in 95 (80%) cases. All assigned medical directives were used with imaging most frequent at 67 requisitions.

Conclusion: The base knowledge of a physiotherapist is an asset in this advanced practice role, however medical directives are necessary for the APP to fully perform. Most patients seen by the APP did not have a rheumatic disease and were managed with minimal involvement from a pediatric rheumatologist, potentially decreasing their burden of care. Access to care was improved as wait time benchmarks were met in most cases and the patient journey was shortened by providing immediate physiotherapy interventions. An APP model of care is ideally suited to address workforce shortages in pediatric rheumatology.

Disclosure: J. Herrington: None; **K. BEATTIE**: None; **M. Batthish**: AbbVie/Abbott, 5, Novartis, 6, Viatris, 12, AdBoard.

Abstract Number: 057

A "High-Risk" Depression/Fatigue Profile May Be Associated with Stronger Response to a Psychological Treatment for Childhood-Onset Systemic Lupus Erythematosus (cSLE)

Elizabeth Ross[1], khalid abulaban[2], Elizabeth Kessler[2], Andrea Knight[3] and Natoshia Cunningham[4], [1]Michigan State University College of Human Medicine, Grand Rapids, MI, [2]Helen DeVos Children's Hospital, Grand Rapids, MI, [3]The Hospital for Sick Children, Toronto, ON, Canada, [4]Michigan State University, Grand Rapids, MI

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Children with childhood-onset systemic lupus erythematosus (cSLE) experience more severe disease than their adult counterparts, in addition to high rates of clinical depressive symptoms (30%) and fatigue (65%) (1). A "high- risk" profile categorized by clinically elevated depressive symptoms and fatigue has been shown to be related to reduced health-related quality of life and poorer patient outcomes in cSLE patients (2). The **T**reatment and **E**ducation **A**pproach for **CH**ildhood-onset lupus (TEACH) is a tailored cognitive behavioral therapy (CBT) for youth with cSLE that may address these symptoms. (3). TEACH is currently being tested in an RCT with promising results after 6 weeks. However, it is unknown how the "high- risk" group may respond to this program. This study aims to assess the impact of risk status on the outcomes of cSLE patients undergoing CBT.

Methods: This study uses data from an ongoing multisite clinical trial of youth (average age of 17 years, 100% female, 70% identifying as from a minority group, n=46/60 currently completed evaluation for the study). All of these patients met the ACR criteria for cSLE. 16 of these youth have thus far been randomized to and completed TEACH. They were grouped into low- or high-risk categories based on baseline depression and fatigue t-scores (>70; >2SD from mean) (3). Repeated measures ANOVAs were conducted to compare the low- and high-risk groups pre and post 6 week TEACH protocol for main study

outcomes (e.g. depressive symptoms (CDI-II), fatigue (PROMIS), and pain (VAS)) and additional exploratory outcomes (anxiety (SCARED), disease severity (SLEDAI), and health-related quality of life (Peds-QL)). Independent samples t-test was used to determine if baseline outcome measures were significantly different between low- and high-risk groups.

Results: Out of the 16 patients who have completed the TEACH protocol, 6 (37.5%) met criteria for inclusion in the high-risk group. There was no significant difference between risk groups based on age, disease duration, or self-reported race. There was a significant difference between groups in baseline depression scores only (p= 0.005). At week 6 after completion of TEACH, depression scores decreased by 26% in the high- risk group, while the low- risk group only decreased 6% ($p < 0.001$; Table 1). Fatigue, anxiety, health-related quality of life, and disease severity followed the same trends (see Figure 1). Despite these trends, there were no significant differences found between the low- and high-risk groups in any outcomes other than depressive symptoms.

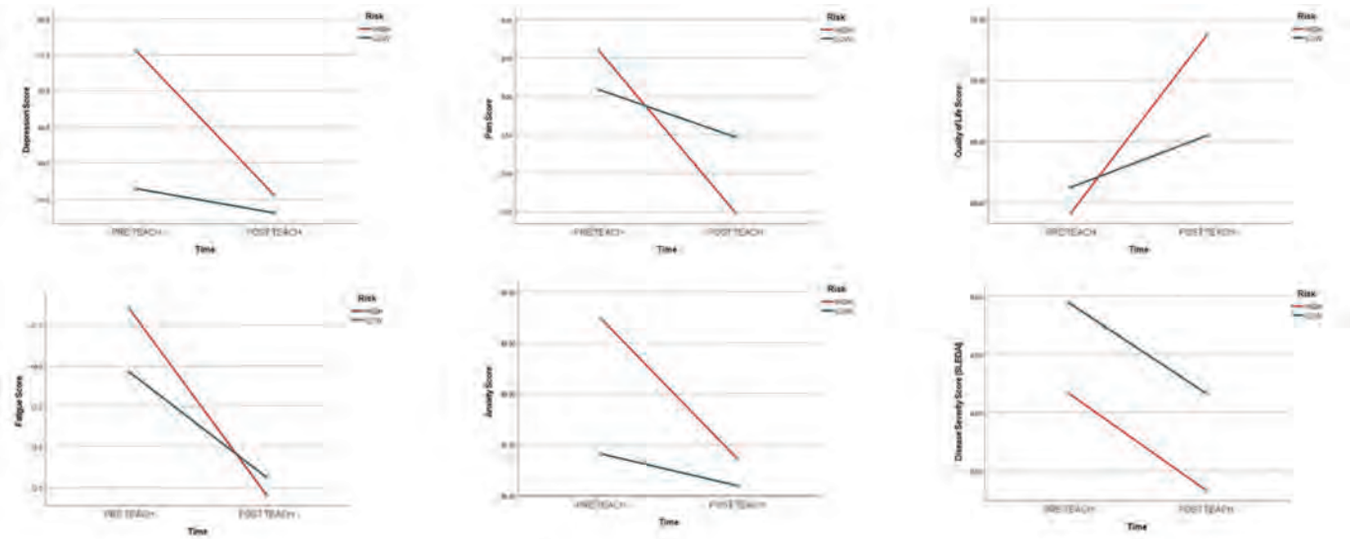


Figure 1. cSLE patient outcomes in low- and high- risk groups pre- and post-TEACH protocol. Depression was measured by Children's Depression Inventory (CDI-II) scores. Fatigue was measured by the Patient Reported Outcome Measurement System (PROMIS) scores. Pain was measured by the Visual Analog Scale (VAS). Anxiety was measured by Screen for Childhood Anxiety Related Emotional Disorders (SCARED). Health-related quality of life was measured by the Pediatric Quality of Life Score (Peds-QL). Disease severity was assessed with the Systemic Lupus Erythematosus Disease Activity Index, or SLEDAI score.

Table 1
Repeated Measures Analysis of Variance Comparing Low- and High- Risk Groups Outcomes Pre and Post TEACH

	Low Risk				High Risk							
	Pre TEACH		Post TEACH		Pre TEACH		Post TEACH					
	Mean	***SD***	***Mean***	***SD***	***Mean***	***SD***	***Mean***	***SD***	***df***	***F***	***p***	***ηp2***
Depression	56.43	8.57	53.08	10.90	75.52	3.65	55.58	7.65	1	28.15	<0.001	0.67
Fatigue	59.62	6.26	53.11	6.04	63.53	6.80	52.02	7.83	1	1.45	0.25	0.09
Pain	3.09	2.35	2.47	1.90	3.60	1.96	1.48	0.84	1	1.84	0.20	0.12
Anxiety	34.10	15.18	31.00	17.52	47.33	13.56	33.67	20.91	1	2.71	0.12	0.16
HRQoL	61.27	8.49	65.48	10.72	59.19	8.20	73.67	16.12	1	2.10	0.20	0.20
Disease Severity	7.78	6.02	4.67	5.57	4.67	5.03	1.33	2.31	1	0.01	0.94	0.001

Note: N= 10 for Low-Risk and N= 6 for High-Risk; *SD*= Standard Deviation; *df*= Degrees of Freedom (hypothesis); *F*= F-distribution; *p*= Significance for confidence interval of 95%; *ηp2* = Partial Eta Squared

Conclusion: There is very little literature on nonpharmacologic treatments for cSLE like CBT (4). These data reveal interesting trends that CBT may be more effective in cSLE patients with high levels of fatigue and depression. Some results may be driven by the fact that the high-risk group begins with higher baseline symptoms. Lack of significant difference between groups may be attributed to a small sample size and these analyses should be replicated with a larger data set (e.g., upon completion of the RCT) in the future. Although the high-risk group shows more improvement overall, this should not shadow that both high- and low- risk groups had improved outcomes with the TEACH protocol.

1. Jones JT, Cunningham N, Kashikar-Zuck S and Brunner HI. Pain, fatigue, and psychological impact on health-related quality of life in childhood-onset lupus. Arthritis Care Res 2016; 68(1): 73–80.
2. Donnelly C, Cunningham N, Jones JT, Ji L, Brunner HI, Kashikar-Zuck S. Fatigue and depression predict reduced health-related quality of life in childhood-onset lupus. Lupus. 2018 Jan;27(1):124-133. doi: 10.1177/0961203317716317.
3. Cunningham NR, Fussner LM, Moorman E, Avar Aydin PO, Brunner HI and Kashikar-Zuck S. Development and pilot testing of the treatment and education approach for childhood-onset lupus (TEACH): a cognitive behavioral treatment. Pediatr Rheumatol 2019; 17(1): 9, DOI: 10.1186/ s12969-019-0307-8.
4. Ross E, Abulaban K, Kessler E, Cunningham N. Non-pharmacologic therapies in treatment of childhood-onset systemic lupus erythematosus: A systematic review. Lupus. 2022 Jun;31(7):864-879. doi: 10.1177/09612033221094704. Epub 2022 Apr 20. PMID: 35442103; PMCID: PMC9191876.

Disclosure: E. Ross: None; **k. abulaban**: None; **E. Kessler**: None; **A. Knight**: None; **N. Cunningham**: None.

Abstract Number: 058

High Levels of Psychological Distress, Depression, and Anxiety Symptoms in Children with Pediatric Rheumatologic Diseases

Natalie Rosenwasser[1], Tamar Rubinstein[2], Andrea Knight[3], Natoshia Cunningham[4], Aimee Hersh[5], Vincent Del Gaizo[6] and **Erin Treemarcki**[5], [1]Seattle Children's Hospital, seattle, WA, [2]Children's Hospital at Montefiore, New York, NY, [3]The Hospital for Sick Children, Toronto, ON, Canada, [4]Michigan State University, Grand Rapids, MI, [5]University of Utah, Salt Lake City, UT, [6]Childhood Arthritis & Rheumatology Research Alliance (CARRA), Whitehouse Station, NJ

SESSION INFORMATION

Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Mental health problems are common in children with pediatric rheumatologic diseases (PRDs) and are associated with worsened quality of life and poorer disease-related outcomes. Psychological distress results from exposure to stress, may be exacerbated in response to traumatic events (e.g., COVID-19 pandemic), and can lead to significant mental health problems. We aimed to determine the rates of psychological stress in PRDs and assess relationships to other measures of psychosocial functioning including anxiety, depressive symptoms, and COVID-related distress.

Methods: Eligible patients in this ongoing cross-sectional study are between 8-17 years and enrolled in the CARRA Registry with a diagnosis of JIA, jSLE, or JDM at one of 3 centers. Consented participants completed a one-time survey during a scheduled rheumatology visit, including Patient-Reported Outcomes Measurement Information System® (PROMIS) measures for psychological stress, physical stress, and depressive symptoms, in addition to the NIH-Toolbox Perceived Stress survey, clinically validated measures of depression (PHQ-9) and anxiety (SCARED), a visual analog scale for COVID-related distress, and a questionnaire to assess acceptability of mental health screening. Descriptive statistics were used for patient characteristics. The proportion of patients with a positive screen based on clinical cutoffs was determined for each measure.

For the PROMIS and NIH-Toolbox measures, 1 standard deviation above the mean of the reference population (T-score 50) indicated high levels of that measure. The relationship between psychological distress and other measures was determined by Pearson Correlation Coefficient.

Results: The 71 patients who completed the survey had a mean age of 13.2 years (SD=2.6) and a diagnosis of JIA in 67 (94%) (Table 1). Psychological stress experiences were elevated in 39% and physical stress experiences in 43% (Table 2). High levels of perceived stress were reported in 26% of patients aged 13-17 years and 15% of those aged 8-12 years. While increased depressive symptoms were seen in only 26% on the PROMIS measure, 54% of patients had a positive PHQ-9 depression screen. Half of the cohort had SCARED scores concerning for anxiety disorder. A majority of patients endorsed mild distress from the COVID-19 pandemic (median 2, IQR 0,5); only 4 (6.0%) endorsed severe distress. Psychological stress was highly correlated with physical stress, perceived stress, depressive symptoms (PROMIS and PHQ-9), and anxiety (Table 3). Patients felt ready to discuss mental health with their rheumatology provider (72%) and moderately confident in discussing mental health concerns (73%).

Table 1. Demographic Information.

Patient Characteristics	
Age, Years (Mean, SD)	13.2 (2.6)
Sex (N, %)	
Female	49 (69.0)
Male	22 (31.0)
Diagnosis (N, %)	
JDM	3 (4.2)
JIA	67 (94.4)
SLE	1 (1.4)

Table 2. Patient-Reported Outcomes

	Complete (N)	Mean T-Score (SD)	Score >60 (N, %)	Mean Score (IQR)	Positive Screen (N, %)
Psychological Stress Experiences	71	57.05 (9.90)	27 (38.6)		
Physical Stress	71	57.70 (10.82)	30 (42.9)		
Perceived Stress					
Age 13-17 (Self)	43	52.73 (11.72)	11 (25.6)		
Age 8-12 (Parent)	26	49.88 (12.12)	4 (15.4)		
Depressive Symptoms	69	53.25 (11.32)	18 (26.1)		
PHQ-9 (Age 11-17)	53			6.42 (0.75, 10.25)	Mild: 14 (26.9) Moderate: 6 (11.5) Moderately Severe: 6 (11.5) Severe: 2 (3.8)
SCARED	67			25.93 (14.25, 36.75)	Score 25+: 33 (50.0) Score 30+: 27 (40.9)

For the PROMIS/NIH measures, T-score for the reference population is 50 and the standard deviation is 10. One standard deviation above the reference population indicates more of the measure. For the measures above, this would indicate increased mental health symptoms.

Table 3. Relationship between PROMIS Psychological Stress Experiences and other measures of psychosocial functioning

Measure	Pearson Correlation Coefficient (P)
Physical Stress Experiences	0.799 (<0.05)
Perceived Stress	
Age 13-17 (Self)	0.900 (<0.05)
Age 8-12 (Parent)	0.753 (<0.05)
Depressive Symptoms	0.870 (<0.05)
PHQ-9 (Age 11-17)	0.825 (<0.05)
SCARED	0.706 (<0.05)
COVID VAS	0.475 (<0.05)

Conclusion: Children with PRDs experience greater psychological distress that is related to physical stress, perceived stress, depressive symptoms, and anxiety. Patients want to and feel confident discussing mental health concerns with their rheumatology providers. Next steps include expanding this cohort to include increased patients with JDM and jSLE and determining the relationship between mental health symptoms and health-related outcomes including disease activity measures.

Disclosure: N. Rosenwasser: None; **T. Rubinstein**: None; **A. Knight**: None; **N. Cunningham**: None; **A. Hersh**: None; **V. Del Gaizo**: None; **E. Treemarcki**: None.

Abstract Number: 059

Update of Clinical and Laboratory Features of the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Systemic Juvenile Idiopathic Arthritis-Associated Lung Disease (SJIA-LD) Cohort

Esraa Eloseily[1], Min-Lee Chang[2], Mary Ellen Riordan[3], allan Russell[4], Marc Natter[2], Yukiko Kimura[5] and Grant Schulert[1], [1]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, [2]Boston Children's Hospital, Boston, MA, [3]Hackensack Meridian Health/ Joseph M. Sanzari Children's Hospital, Hackensack, NJ, [4]Duke Clinical Research Institute, Durham, NC, [5]Hackensack Meridian School of Medicine, Hackensack, NJ

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Systemic juvenile idiopathic arthritis (SJIA) associated lung disease (SJIA-LD) is an emerging and life-threatening clinical problem. Despite recent advances, there remain key unanswered questions regarding prevalence, pathogenesis, influence of biologics, and outcomes, and prospective clinical and laboratory characterization of SJIA-LD is needed.

The objective is to define baseline clinical and laboratory features of patients with SJIA-LD enrolled in the CARRA Registry SJIA-LD cohort.

Methods: Existing or newly enrolled CARRA Registry patients with SJIA and suspected, probable, or definite SJIA-LD were included in the cohort. In addition to standard Registry data, lung disease specific data was obtained at baseline and at 6-month follow-up visits using a standardized case report form through REDCap Cloud. This study was approved by the DCRI Reliant IRB and/or IRB of all Registry sites.

Results: As of January 2023, 36 patients were enrolled in the cohort from 16 CARRA Registry sites. 46% had definite (biopsy-proven), 36% probable, and 18% suspected SJIA-LD. 22 patients had LD data available. Of those, 23% underwent lung biopsy: all had pulmonary alveolar proteinosis (PAP) and interstitial inflammation, and 40% had collagenous fibrosis. 77% had at least one definite episode of macrophage activation syndrome (MAS) (including 64% which met the 2016-SJIA-MAS criteria), 73% had more than one MAS episode, and 32% had subclinical MAS. MAS occurred prior to SJIA-LD diagnosis in 68% and coincided with it in 18%. The demographic and clinical features at the baseline CARRA Registry visit are shown in Table 1. Median (IQR) values of selected labs at the baseline visit were as follows: AST: 42 U/L (34-62), CRP: 0.5 mg/dl (0.9-3.8), ESR: 16.5 mm/hr (8-56), Wbcs: 10*10^^{9}/L) (7.6-14.7) Hb: 11.6 g/dl (10.9-12.7), Plt: 310 *10^^{9}/L) (268-401), IL-18: 24,336 pg/mL (4,147- 49,275).

Table 1: Demographic and clinical features of patients in the SJIA-LD cohort

Sex	62% F, 38%M
Age at enrollment*	4.4 years
Age at LD onset*	3.3 years
Age at SJIA onset*	1 years
LD duration**	0.3 (2.1) years
SJIA disease duration**	1 (2.75) years
SJIA duration at LD diagnosis**	1.6 (1.4) years
Clinical features at LD diagnosis	50% cough 50% clubbing 46% tachypnea 36% dyspnea on exertion 23% digital erythema 18% hypoxemia requiring supplemental oxygen
Baseline chest CT findings	55% ground glass opacities 41% peribronchovascular thickening 41% septal thickening 23% peripheral consolidation 23% hilar adenopathy
Pulmonary function tests	50% abnormal DLCO 50% abnormal spirometry 0% abnormal spot pulse oximetry
Broncho alveolar lavage	19% PAP 8% signs of infection
Parent/patient overall well-being score at most recent visit**	1(2)
Parent/subject assessment of disease activity at most recent visit**	1 (4.5)
Overall SJIA physician global assessment at most recent visit**	0.5 (2.5)
Physician global assessment of lung disease (PGALD) at most recent visit**	3.5 (3.75)
Health Related quality of life at most recent visit	26% excellent 22% very good 35% good 17% fair
Biologics ever used	92% Anakinra 65% Canakinumab 65% Tofacitinib 58%Tocilizumab
DMARDs ever used	46% Methotrexate 19% Mycophenolate mofetil
Currently on oral steroids	81 %

*Median, ** Median (IQR)

Conclusion: The CARRA SJIA-LD cohort represents a broad spectrum of clinical features which commonly includes clubbing, cough, tachypnea, dyspnea, and PAP in all patients who underwent a biopsy. Recurrent MAS was a common clinical feature in patients who developed LD. We plan to continue enrolling patients to fully characterize the clinical features of SJIA-LD, longitudinal disease progression and trajectories, and associated immune biomarkers and cellular populations associated. This cohort will serve as an ongoing prospective cohort study for future clinical and translational research in this emerging disease.

Disclosure: E. Eloseily: None; **M. Chang**: None; **M. Riordan**: None; **a. Russell**: None; **M. Natter**: None; **Y. Kimura**: None; **G. Schulert**: Novartis, 2, SOBI, 2.

Abstract Number: 060

Are the Levels of Cytokines Good Biomarkers for Smoldering Disease Activity in Childhood-Takayasu Arteritis?

Gleice Clemente[1], Maria Teresa Terreri[2], Bruno Gualano[3], Clovis Silva[4] and Alexandre Wagner De Souza[1], [1]Universidade Federal de São Paulo, São Paulo, Brazil, [2]UNIFESP, São Paulo, Brazil, [3]Universidade de São Paulo, São Paulo, [4]Universidade de São Paulo, São Paulo, Brazil

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Biomarkers for disease activity in adult Takayasu arteritis (TA) have been studied exhaustively, but there are inconsistencies among the studies (1). Childhood-TA (c-TA) differs from adult TA in many aspects, including the high frequency of non-specific symptoms at disease onset, and a more pronounced inflammatory disease (2). Therefore, we aimed to assess the levels of serum cytokines as potential biomarkers for smoldering disease activity in c-TA patients undergoing treatment and considered in remission by clinical scores.

Methods: Cross-sectional study with c-TA patients recruited from three Brazilian reference centers in Pediatric Rheumatology. All patients fulfilled EULAR/PRINTO/PRES criteria and were in clinical remission according to Indian Takayasu clinical activity score (ITAS) 2010 and Paediatric Vasculitis Activity Score (PVAS) (3,4,5). Patients were two half lifetime periods off immunosuppressants, before blood sample collection. The following serum cytokines were measured: interferon gamma (IFN-γ), interleukin-10 (IL-10), interleukin-12p70 (IL-12p70), interleukin 1 receptor antagonist (IL-1ra), interleukin-1 beta (IL-1β), interleukin-6 (IL-6), tumor necrosis factor alpha (TNF-α), vascular endothelial growth factor (VEGF) and platelet-derived growth factor (PDGF). The control group consisted of 14 age- and sex-matched healthy individuals (HC).

Results: Twelve c-TA patients were recruited (66.7% females) with a mean age of 18.7 ±2.84 years, and a median follow-up time of 10 (7.0-11.8) years. Ten from 12 patients had high arterial FDG-uptake (visual score =3) revealed in PET/MRI performed at the same time of the blood collection in a previous study (6). All patients were under therapy: 7 (58%) were on biological therapy (4-infliximab, 2-adalimumab and 1-tocilizumab) associated with synthetic immunosuppressants. No significant differences in serum cytokine levels were observed between c-TA patients and HC (p >0.05) (Table). According to Hata classification, no differences in cytokine levels were found between patients presenting diffuse arterial involvement (i.e. angiographic type V) and localized disease (i.e. angiographic types I, IIa, and III) (7).

Laboratory Markers	Patients N=12	Controls N=14	P value
IFN-γ (pg/ml)	2.3 (1.4-8.7)	1.3 (0.9-2.2)	0.089
IL-10 (pg/ml)	9.4 (4.5-13.4)	7.4 (4.5-10.2)	0.410
IL-12p70 (pg/ml)	3.5 (2.6-9.6)	4.9 (3.2-5.2)	0.502
IL-1ra (pg/ml)	41.6 (32.9-62.3)	39.8 (21.5-43.7)	0.135
IL1-β (pg/ml)	2.1 (1.6-3.3)	2.1 (1.9-2.6)	0.757
IL-6 (pg/ml)	8.0 (3.6-27.0)	9.2 (2.6-14.8)	0.607
TNF-α (pg/ml)	13.7 (6.8)	13.8 (6.5)	0.969
VEGF (pg/ml)	1.9 (1.1-2.6)	1.5 (1.4-2.0)	0.440
PDGF (pg/ml)	44.2 (31.2-674.7)	130.8 (53.9-1010.7)	0.446

Cytokine levels of childhood-onset Takayasu arteritis patients and healthy controls

Conclusion: Similarly to adult TA, the investigation of biomarkers to detect smoldering disease activity is also a challenge in c-TA patients during follow-up, especially in those with long follow-up periods and under long-term immunosuppressants, independently of the extension of the disease. Further longitudinal multicenter and multinational studies, with a large number of patients, are necessary to better analyze biomarkers involved in disease progression in c-TA.

Disclosure: G. Clemente: None; **M. Terreri**: None; **B. Gualano**: None; **C. Silva**: None; **A. De Souza**: None.

Abstract Number: 061

Variation in Treatment Approaches to IVIG- Refractory Kawasaki Disease (KD) Among Pediatric Rheumatologists: The Childhood Arthritis and Rheumatology Research Alliance (CARRA) Treatment of Refractory KD Survey

Daniel Ibanez[1], Bianca Lang[2], Ali Yalcindag[3], Linda Wagner-Weiner[4], Julia Shalen[5], Kenneth Schikler[6], Shoghik Akoghlanian[7], Hulya Bukulmez[8], Kristen Hayward[9], Sivia Lapidus[10], Andrea Ramirez[11], Robert Sundel[1], Cagri Yildirim-Toruner[12] and CARRA Registry Investigators[13], [1]Boston Children's Hospital, Boston, MA, [2]Dalhousie University - Halifax, Halifax, NS, Canada, [3]Hasbro Children's Hospital, Milton, MA, [4]The University of Chicago, Chicago, IL, [5]Johns Hopkins University, Baltimore, MD, [6]University of Louisville School of Medicine Norton Children's Hospital, Louisville, KY, [7]Nationwide Children's Hospital, Columbus, OH, [8]MetroHealth Medical Center, Case Western Reserve, Cleveland, OH, [9]Seattle Children's Hospital/University of Washington School of Medicine, Seattle, WA, [10]Department of Pediatrics, Joseph M. Sanzari Children's Hospital, Hackensack University Medical Center, Hackensack, NJ, [11]Baylor College of Medicine, Houston, TX, [12]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [13]CARRA, Washington, DC

SESSION INFORMATION

Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Kawasaki disease (KD) is the leading cause of acquired heart disease in children in North America. Initial treatment with IVIG has significantly reduced the risk of developing coronary artery aneurysms (CAA). However, 10-20% of children with KD fail to respond to the first dose of IVIG (refractory KD) and have an increased risk of developing

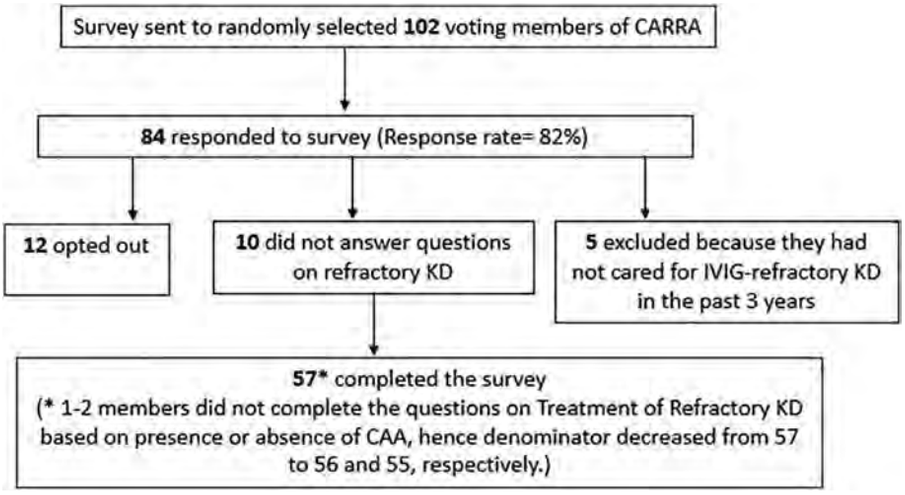


Figure 1. Survey Response and Completion in Numbers

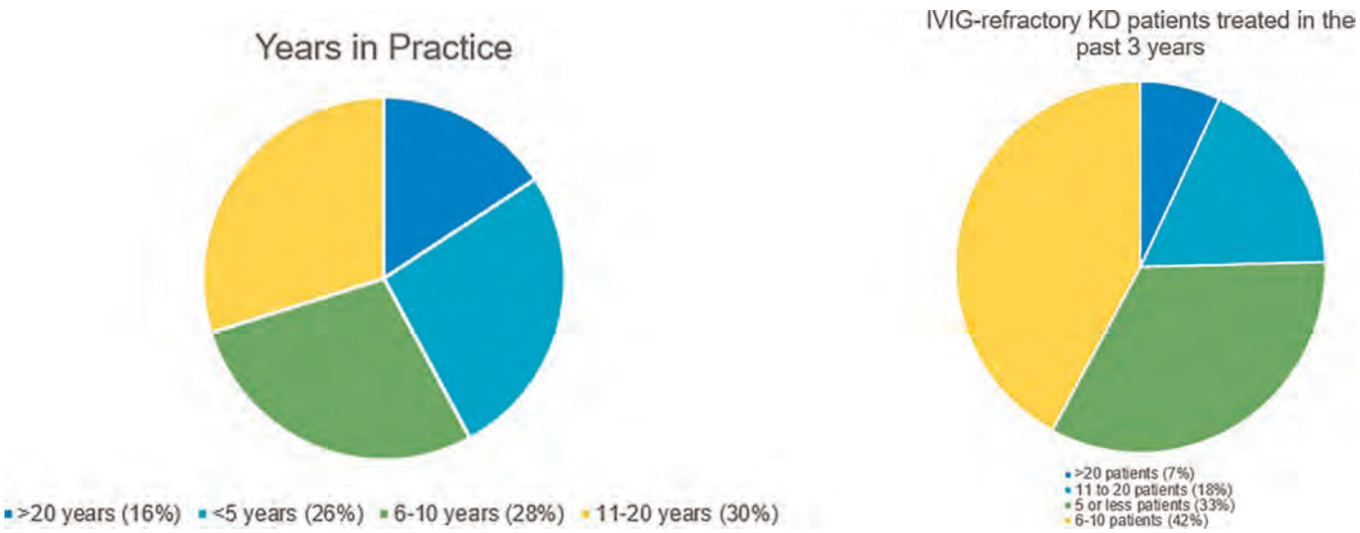


Figure 2. Experience of Survey Respondent Pediatric Rheumatologists

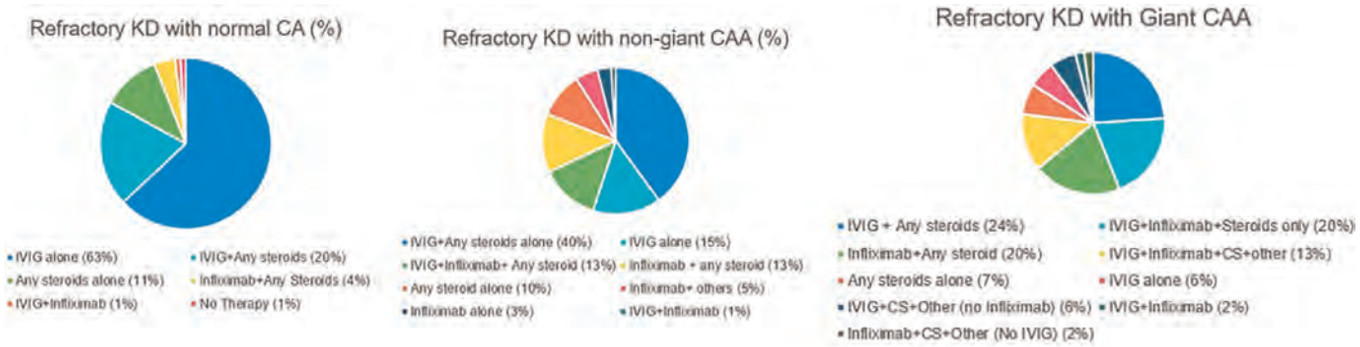


Figure 3. Variability in Treatment of IVIG-refractory KD with and without CAA among Pediatric Rheumatologists

CAA. The best treatment to prevent CAA in IVIG-refractory KD is currently unknown. The purpose of this survey was to determine treatment preferences of pediatric rheumatologists (PR) in North America to inform future development of consensus treatment plans (CTPs) for children with IVIG-refractory KD.

Methods: The CARRA Refractory KD Workgroup developed a 34-item web-based survey with clinical cases and sent it to 102 randomly selected voting members of CARRA in March 2022. The survey asked participants about their practice settings, when patients should be treated for refractory KD and medication choices for treatment of refractory KD. Respondents who had not treated refractory KD in the previous 3 years were excluded.

Results: The response rate was 82%. A total of 57 pediatric rheumatologists completed the refractory KD questionnaire (Figure 1). There was a wide variation of clinical experience including years in practice and number of refractory KD patients treated. Most of the respondents (86%) were consultants and 9% were members of the primary care team for KD at their institution (Figure 2). While 54% of the 57 respondents considered a temperature 100.4 F (38C) at 36 hours after the completion of the first dose of IVIG warranted treatment for IVIG-refractory KD. Another 33% of questionnaire participants considered refractory KD at 24 hours post IVIG, 11% at 48 hours and 2%- other. The therapeutic choices for IVIG-refractory KD varied among respondents, with important differences based on the presence or absence of CAA. Refractory KD with **no CAA:** 83% of n=56 respondents would continue treatment with IVIG, of which 63% would give IVIG alone and 20% would give IVIG with corticosteroids (CS). 11% would give only daily IV/PO

CS. 5% added Infliximab to their treatment regimen in patients with normal CAs. Refractory KD with **non-giant CAA:** 73% of n=55 would continue treatment with IVIG, of which 15% would give IVIG alone, 40% would treat with IVIG and CS, and 13% would treat with IVIG, CS and Infliximab. 13 % would treat with Infliximab and CS without additional IVIG. There were 23 different treatment regimens among the respondents, primarily because of varying CS doses/routes. (Figure 3) Refractory KD with **giant CAA:** 69% of n=55 would continue treating with IVIG, of which 5% would treat with IVIG alone, 24% with IVIG and CS, and 20% would treat with IVIG, CS and Infliximab. 20% would treat with Infliximab and CS, but not IVIG. There were 32 different treatment regimens among the respondents, primarily because of varying CS doses/routes.

Conclusion: Treatment of IVIG-refractory KD varies widely among North American pediatric rheumatologists, particularly in the presence of CAA. The results of this survey support the need to conduct comparative effectiveness research to identify the most effective therapy for the prevention of CAA and to improve outcomes in children with IVIG-refractory KD.

Disclosure: D. Ibanez: None; **B. Lang**: None; **A. Yalcindag**: None; **L. Wagner-Weiner**: None; **J. Shalen**: None; **K. Schikler**: None; **S. Akoghlanian**: None; **H. Bukulmez**: None; **K. Hayward**: None; **S. Lapidus**: None; **A. Ramirez**: None; **R. Sundel**: None; **C. Yildirim-Toruner**: None; **C. Investigators**: None.

Abstract Number: 062

Assessment of Barriers and Facilitators in Implementation of the Childhood Arthritis and Rheumatology Research Alliance Consensus Treatment Plans

Cagri Yildirim-Toruner[1], Daniel Glaser[2], Timothy Beukelman[3], Stacy Ardoin[4], Ahmar Hashmi[5], Rajdeep Pooni[6], Maria Fernandez[5], Vincent Del Gaizo[7], Leslie Hanrahan[7], Mary Ellen Riordan[8], Stacey Tarvin[9] and CARRA Registry Investigators[7], [1]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [2]Yale University School of Medicine, New Haven, CT, [3]University of Alabama at Birmingham, Birmingham, AL, [4]Nationwide Children's Hospital, Columbus, OH, [5]The University of Texas Health Sciences Center at Houston, Institute for Implementation Science, Houston, TX, [6]Stanford University, Palo Alto, CA, [7]CARRA, Washington, DC, [8]Hackensack Meridian Health/ Joseph M. Sanzari Children's Hospital, Hackensack, NJ, [9]Riley Hospital for Children at Indiana University Health, Indianapolis, IN

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Since 2010, the Childhood Arthritis and Rheumatology Research Alliance (CARRA) has developed 12 consensus treatment plans (CTP) with the aim of reducing treatment variability and promoting the conduct of comparative effectiveness research (CER) utilizing the CARRA registry. In November 2021 CARRA created a multi-stakeholder CTP Task Force (TF) charged with assessing the existing CARRA CTP program and making recommendations for improvement and expansion. Here, we report the initial work on systematic assessment of barriers and facilitators to reliable and widespread use of the CARRA CTPs.

Methods: The CTP TF gathered data using surveys, qualitative interviews, brainstorming and feedback sessions. Electronic surveys were developed, beta tested and administered to providers (PR), research coordinators (RC), and caregivers. A clinical implementation science expert with qualitative research experience interviewed a group of PRs and RCs of diverse geography, CARRA site size and experience to further assess barriers and facilitators of CTP use. The interview guides

adhered to implementation frameworks and organizational readiness. The team evaluated CARRA Registry data relevant to CTPs for participants with juvenile idiopathic arthritis (JIA), lupus and juvenile dermatomyositis (JDM). The CARRA Steering Committee provided feedback via structured, small-group discussions.

Results: The initial provider survey (n=154) showed that awareness about most CTPs was high; the vast majority utilized CTPs in clinical practice but strict adherence to CTPs was low. A primary barrier to use was inaccessibility of CTPs at point of care. A second provider survey (n=119) showed that awareness of newer CTPs was lower and affirmed that CTPs were frequently referenced but adhered to with variable fidelity. Survey of research coordinators (n=43) identified high awareness of CTPs but variation in documentation of CTP use in the Registry. Most caregivers were not aware of CTPs but were interested to learn more and support CTP use. Thirteen interviews were conducted (7 PR, 6 RC) and analyzed. Facilitators and

Table 1. Facilitators to Implementing CTPs

Findings from interviewees	Implementation framework constructs (CFIR, organizational readiness)	Suggestions for future CARRA efforts
The majority of providers are amenable to using CTPs	Relative advantage, intervention source, motivation	
Patients respond favorably to evidence-based approach of CTPs	Patient needs and resources, intervention source, relative advantage, motivation	Consider patient materials outlining treatment specifics and duration
Sites with strong research interest/ experience more likely to use CTPs	Implementation climate, readiness	
Site PI can facilitate spaces (e.g., team meetings, journal clubs) to review CTP enrollment criteria and treatment plans	Leadership engagement, network and communication, relative priority, learning climate, readiness	
Coordinators with knowledge of/experience with pediatric rheumatology	Intervention needs and resources	CARRA coordinator trainings for "translating" CTP eligibility requirements, specific treatments, and data reporting
Utilizing electronic medical records to enhance communication	Intervention needs and resources, network and communication, readiness, adaptability	Creating effective messaging modalities through EMRs to facilitate coordinator/provider communication

Table 2. Barriers to Implementing CTPs

Findings from interviewees	Implementation framework constructs (CFIR, organizational readiness)	Suggestions for future CARRA efforts
Limited provider awareness of CTPs	Access to knowledge and information	Identify key avenues to increase awareness among providers about CTPs
Limited access to and usability of CTP treatment guides	Access to knowledge and information	Easy-to-use treatment algorithms for quick reference
Ensuring CTPs are up-to-date	Access to knowledge and information	Identify appropriate time frames for specific CTP updates as treatment modalities can change with new evidence
Unclear reporting of CTP use	Readiness	Clearer guidance on characterizing CTP use for registry data given variability in adherence to prescribed CTPs

Table 3. The CTP TF Recommendations for Improvement

Recommendations for Improvement
Improve awareness, accessibility and dissemination of CTPs in real time (website, CTP App development)
Simplify development of new CTPs and assure timely CTP updates
Develop incentives for CTP use (e.g. Maintenance of Certification (MOC))
Prioritize and optimize collection of critical data elements in the registry which will support outcomes data including CER
Support informatics initiatives including leveraging EHR data collection and extraction
Engage with IS to assess current practices and address barriers to and enhance facilitators of CTP use as outlined in Table 1 and Table 2.

barriers to implementation of CTPs, alignment with implementation framework constructs, and suggestions for future CARRA efforts are outlined in Table 1 and Table 2. CARRA Registry data analysis showed that opportunities exist to improve patient-reported and laboratory data collection. Data quality issues were common regardless of disease or CTP assessed. Site declaration of CTP use was inconsistent and often did not match the treatment received, though overall treatment patterns did not substantially differ between patients with or without a declared CTP. Collection of physician-generated outcomes data is excellent and on par with externally funded CER studies.

Conclusion: CTPs are valued and utilized by the CARRA community as clinical and research tools. Barriers exist to widespread CTP use, particularly accessibility at point of care. Improving data collection in the Registry may provide additional insights. The CTP TF developed a list of recommendations for a multi-year optimization process as outlined in Table 3.

Disclosure: C. Yildirim-Toruner: None; **D. Glaser**: None; **T. Beukelman**: None; **S. Ardoin**: None; **A. Hashmi**: None; **R. Pooni**: None; **M. Fernandez**: None; **V. Del Gaizo**: None; **L. Hanrahan**: None; **M. Riordan**: None; **S. Tarvin**: None; **C. Investigators**: None.

Abstract Number: 063

JAK Inhibition in down Syndrome Associated Arthritis (DA) - Our Experience to Date with Tofacitinib in 5 Patients

Anwar Alkandari and Orla killeen, Children Health Ireland-Crumlin (CHI), Dublin, Ireland

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Down syndrome associated arthritis (DA) is a challenging form of inflammatory arthritis that typically is more erosive and has a greater requirement for biologic agents than JIA (Juvenile Idiopathic Arthritis). There remains a paucity of information on potential therapeutic agents for this aggressive condition. Studies have hypothesized the presence of immune

dysregulation due to interferon (IFN) signalling hyperactivation in patients with Down Syndrome (DS). As Janus kinase (JAK) plays an important role in IFN signalling; using JAK inhibitors such as Tofacitinib which blocks JAK1/JAK 3 pathway therefore block IFN hyperactivity with therapeutic benefit in DA . We present our experience to date of DA patients treated with Tofacitinib.

Methods: All patients with DA treated with Tofacitinib between August 2021 to August 2022 were included. Data was collected at baseline with plan to be collected at 3- and 6-months post treatment but due to COVID-19 backlog in clinic it was collected at 1-3 months for initial visit and then 6-8 months for 2nd clinic visit. The total active joint count, use of non-steroidal anti-inflammatory medication (NSAID), exercise endurance, infection episodes, side effects of medication, overall parental satisfaction with medication and inflammatory markers at each assessment were documented.

Results: A total of 5 patients (2 Male, 3 females), aged between 13-18 years were identified. All patients were of European ethnicity. The duration since diagnosis with DA ranged between 4-11 years. All 5 patients used minimum of 3 biological agents with no or minimal disease control and 2 patients failed trial of Methotrexate in addition to biological therapy. 3 patients (60%) had diagnosis of erosive arthritis and psoriasis. 4 out of 5 patients had improvement in the joint counts (80%). All patients had reduced amount of NSAID use with 3 patients having stopped pain medication altogether. Improvement in exercise endurance noticed by family in 4 patients. There was no reported increase in infection episodes. 2 patients had slightly higher ESR at 2nd follow up visit compared to baseline blood while WBC count, haemoglobin, Platelets, CRP remained unchanged. 2 out of 3 patients with psoriasis had resolution of the rash with Tofacitinib. An overall improvement in clinical status noted by parents was reported between 4-12 weeks after starting the treatment and all 5 families (100%) agreed that Tofacitinib was the most beneficial biologic agent to date.

Conclusion: Down Syndrome associated arthritis is a relatively newly described condition that affects approximately 1in 50 with DS. It is an aggressive form of arthritis that does not respond as well as JIA to conventional medications. To our knowledge this is the first case series outlining the use of JAK inhibitors. Although numbers included were small, results were

Parameter	Patient 1		Patient 2		Patient 3		Patient 4		Patient 5	
Gender	M		F		F		F		M	
Age	15 years		13 years		18 years		14 years		15 years	
Age at diagnosis	9 years		9 years		7 Years		5 Years		5 Years	
Previous treatment	Orencia ,Tocilizumab , Humira ,Enbrel		Enbrel ,Humira , Tocilizumab		MTX , Enbrel ,Humira Ustekinumab ,Tocilizumab		MTX ,Enbrel ,Humira Tocilizumab , Secukinumab		Humira ,Enbrel ,Simponi	
Erosions	No		No		Yes		Yes		Yes	
Psoriasis	No		No		Yes		Yes		Yes	
Co-morbidities	Obstructive sleep Apnoea (OSA) on CPAP ,Operated congenital heart ,Asthma		Epilepsy ,PEG feed , AVSD repair , hearing loss		OSA ,Pacemaker in Situ		Asthma ,Ventricular septal defect repair		OSA on CPAP ,Operated heart disease	
Joint count pre-treatment	2		8		13		4		8	
Joint count post-treatment	2		2		2		1		0	
Use of NSAID	Reduce		Stopped		Nil		Stopped		Stopped	
Infection	COVID-19 2 chest infection		COVID-19		Nil		Nil		Nil	
Exercise endurance	Same		Improved		Improved		Improved		Improved	
Early morning stiffness	Same		Improved		Same		Improved		Not Sure	
Mobility	Same		Improved		Improved		Improved		Improved	
	Pre TX	Post TX	Pre TX	Post TX	Pre TX	Post TX	Pre TX	Post TX	Pre TX	Post TX
WBC	5.2	5.5		3.4	3.1	8.6	4.8	6.8	3.9	3.9
PLT	309	258		357	351	271	362	375	217	196
ESR	8	14		6	-	21	8	16	6	5
CRP	9	6		5	5	5	5	5	5	5

favourable with promising improvements reported and no significant major side effects reported. A larger and prospective study is required to prove the beneficial role of JAK inhibitor in managing patients with DA and changes treatment approach in Down Associated arthritis.

Disclosure: A. Alkandari: None; **O. killeen**: None.

Abstract Number: 064

Towards the Development of Composite Parent-Centered Disease Activity Scores for Juvenile Dermatomyositis

Silvia Rosina[1], Ana Isabel Rebollo-Giménez[2], Letizia Tarantola[3], Roberta Naddei[4], Alessandro Consolaro[2], Angela Pistorio[5] and Angelo Ravelli[6], [1]IRCCS Istituto Giannina Gaslini, Genova, Italy, [2]IRCCS Istituto Giannina Gaslini, UOC Reumatologia e Malattie Infiammatorie, Genova, Italy, [3]Università degli Studi di Genova, Dipartimento di Neuroscienze, Riabilitazione, Oftalmologia, Genetica e Scienze Materno-Infantili (DiNOGMI), Genova, Italy, [4]Università degli Studi di Napoli Federico II, Dipartimento di Scienze Mediche Traslazionali, Napoli, Italy, [5]IRCCS Istituto Giannina Gaslini, Direzione Scientifica, Genova, Italy, [6]IRRCS Istituto Giannina Gaslini and Università degli Studi di Genova, Genova, Italy

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Increasing attention has been recently paid to the development of parent- and child-centered composite DAS for the assessment of health status of children with rheumatic diseases. We aimed to develop and test an entirely parent-centered composite DAS for JDM, named parent Juvenile DermatoMyositis Activity Index (parJDMAI). Two versions of the score were evaluated.

Methods: Both parJDMAI1 and parJDMAI2 include the following items: 1) parent assessment of skin disease activity (Parent Skin Scale) on a 0-5 scale, by giving 1 point to the presence of each of: i) rash on eyelids, ii) nose/cheeks, iii) knuckles, iv) trunk/arms, v) skin ulceration; 2) parent assessment of muscle disease activity (Parent Muscle Scale) on a 0-5 scale, by giving 1 point to the presence of each of: i) fatigue/discomfort, ii) muscle weakness, iii) muscle pain, iv) voice change, v) difficulty swallowing; 3) parent assessment of childs fatigue on a 0-10 visual analog scale (VAS) (0 =no fatigue; 10 = maximum fatigue). As fourth item, the parJDMAI1 includes the parent global assessment of childs wellbeing on a 0-10 VAS (0 = best; 10 = worst), whereas the parJDMAI2 includes the parent global assessment of disease activity on a 0-10 VAS (0 = no activity; 10 = maximum activity). To give the 4 components of the tools the same weight, the scores of the Parent Skin and Muscle Scales were doubled. Thus, the total score of both instruments ranges from 0 to 40. Initial validation was conducted on a multicentric prospective sample of 213 patientsfollowed in standard clinical care (number of visits = 577), and on a monocentric sample including 50 patients, all assessed at baseline and 32 also assessed after a median of 3.9 months (number of visits = 82). Validation analyses included calculation of the correlations between individual parJDMAI items and physician-centered JDM outcome measures, and between the total score of parJDMAI1 and parJDMAI2 with that of the global composite DASs for JDM named JDMAI1 and JDMAI2. Because both JDMAI1 and JDMAI2 include the parent global assessment of childs wellbeing, which is also part of parJDMAI1, we also tested the correlations of parJDMAI1 with reduced versions of JDMAI1 and JDMAI2, that included only the 3 physician-centered items. Spearman correlations were defined as low, moderate or high when rS was 0.4, ≥ 0.4 and ≤ 0.7, or 0.7, respectively.

	Multicentric sample		Monocentric sample	
	Spearman r	N	Spearman r	N
Parent JDMAI1 vs JDMAI1	0.70	402	0.83	68
Parent JDMAI1 vs JDMAI2	0.69	350	0.82	65
Parent JDMAI2 vs JDMAI1	0.69	394	0.80	71
Parent JDMAI2 vs JDMAI2	0.67	347	0.79	68
Parent JDMAI1 vs DAS total	0.51	395	0.72	69
Parent JDMAI2 vs DAS total	0.51	400	0.71	72
Parent JDMAI1 vs Physician Global VAS	0.48	419	0.73	71
Parent JDMAI2 vs Physician Global VAS	0.49	420	0.74	74
Parent JDMAI1 vs JDMAI1-3-Items	0.49	402	0.72	68
Parent JDMAI1 vs JDMAI2-3-Items	0.49	350	0.67	65

Spearman correlation between parJDMAI1 and parJDMAI2 and other JDM outcome measures.

Results: Correlations between individual components of parJDMAI1 and parJDMAI2 and physician-centered JDM outcome measures were low-to-moderate in the multicentric sample, but moderate in the monocentric sample. Likewise, correlations between the scores of parJDMAI1 and parJDMAI2 and that of original and reduced versions of JDMAI1 and JDMAI2 were strong in the monocentric sample, but moderate in the multicentric sample.

Conclusion: The new parent-centered composite DASs revealed satisfactory measurement properties. That correlations with physician-centered outcome measures and original and reduced JDMAI versions were higher in the monocentric sample than in the multicentric cohort indicates that the proposed tools should be further tested in different clinical and cultural environments before their widespread use can be recommended.

Disclosure: S. Rosina: None; **A. Rebollo-Giménez**: None; **L. Tarantola**: None; **R. Naddei**: None; **A. Consolaro**: None; **A. Pistorio**: None; **A. Ravelli**: AbbVie/Abbott, 6, Alexion, 6, Angelini, 6, Bristol-Myers Squibb(BMS), 6, Novartis, 6, Pfizer, 6, Reckitt Benckiser, 6, Roche, 6, Sobi, 6.

Abstract Number: 065

Relationship Between Arthritis and Uveitis Disease Activity in Children with JIA

Meghana Karumuri[1], Megan Quinlan-Waters[2], Alexandra Duell[2], Kelly Rogers[2], Sheila Angeles-Han[3] and Patricia Vega-Fernandez[2], [1]Michigan State University, Novi, MI, [2]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, [3]Cincinnati Children's Hospital, Cincinnati, OH

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Uncontrolled uveitis can lead to visual complications in 50% of children with JIA associated uveitis (JIA-U). While arthritis and uveitis are not considered to run parallel courses, recent studies report that children with active arthritis may have concurrent uveitis[1]. Further, as subclinical synovitis can be detected by musculoskeletal US (MSUS), findings of active arthritis by MSUS may better reflect disease activity and have implications for uveitis activity[2]. This retrospective study aims to evaluate the temporal association between arthritis and uveitis in patients with JIA-U, and whether this association varies when arthritis is detected by MSUS versus clinical examination.

Methods: Medical charts of children with JIA-U who had an ophthalmology examination within 6 weeks, either 3 weeks before or after a rheumatology examination (clinical and/or MSUS) were reviewed. Participants were able to contribute more than one visit. Active uveitis was defined as anterior chamber (AC) inflammation grade of ≥0.5+ cells as per the Standardization of Uveitis Nomenclature Working Group criteria. Active arthritis was defined by 1) joint swelling, or in the absence of joint swelling, limited range of motion in a joint with tenderness; or 2) abnormal MSUS as per OMERACT. Primary outcome was presence of active uveitis within 6 weeks of arthritis examinations.

Results: Eighteen patients contributed 49 total visits (Table 1). Active arthritis by either MSUS or clinical examination was documented in 32 visits and 10 (31%) of these visits also had evidence of active uveitis. Of the 17 visits with inactive arthritis, only 1 (6%) had active uveitis (Fig. 1).

When focusing on visits that had only a clinical examination (n=33) we found that 23 (70%) had active arthritis and that 7 of these 23 visits (30%) also had active uveitis. Of the 10 visits with inactive arthritis on clinical examination, none had active uveitis.

Table 1: Demographics/Characteristics of Patients with JIA-U

Characteristic	N (%)
Ethnicity:	
Hispanic	0
Non-Hispanic	18 (100)
Race:	
White	15 (83)
Black	2 (11)
Multi-Racial	1 (6)
Sex:	
Female	15 (83)
Male	3 (17)
Biomarkers (n = subjects with data available)	
RF positive (n = 13)	0
ANA positive (n = 18)	12 (67)
JIA Subtype:	
Oligoarticular (extended)	3 (17)
Oligoarticular (persistent)	7 (39)
Systemic	0
Polyarticular (RF negative)	4 (22)
Polyarticular (RF positive)	0
Enthesitis Related	0
Psoriatic	2 (11)
Undifferentiated	2 (11)
Patient Medications at Time of Visit (n=49)	
Nonbiologic DMARD	39 (80)
Biological DMARD	25 (51)
Glucocorticoids	32 (65)
Eyedrops	31 (63)
None	3 (6)

Of 16 visits with only an MSUS examination, 9 (56%) had active arthritis and 3 of these 9 also had uveitis (33%). Of the 7 visits with inactive arthritis by MSUS, only 1 (14%) had active uveitis.

Fifteen visits had both an MSUS and clinical examination documented (Fig. 2). Six of these visits (40%) showed arthritis on both examinations with 3 visits (50%) having uveitis also.

Conclusion: Our findings suggest that in children with JIA-U, an arthritis flare may be associated with a concurrent uveitis flare. Therefore, children with JIA-U who present with active arthritis may need an expedited ophthalmology examination. We found no significant difference between arthritis detection by clinical examination vs. MSUS in terms of uveitis activity. Future prospective studies with a larger population are required.

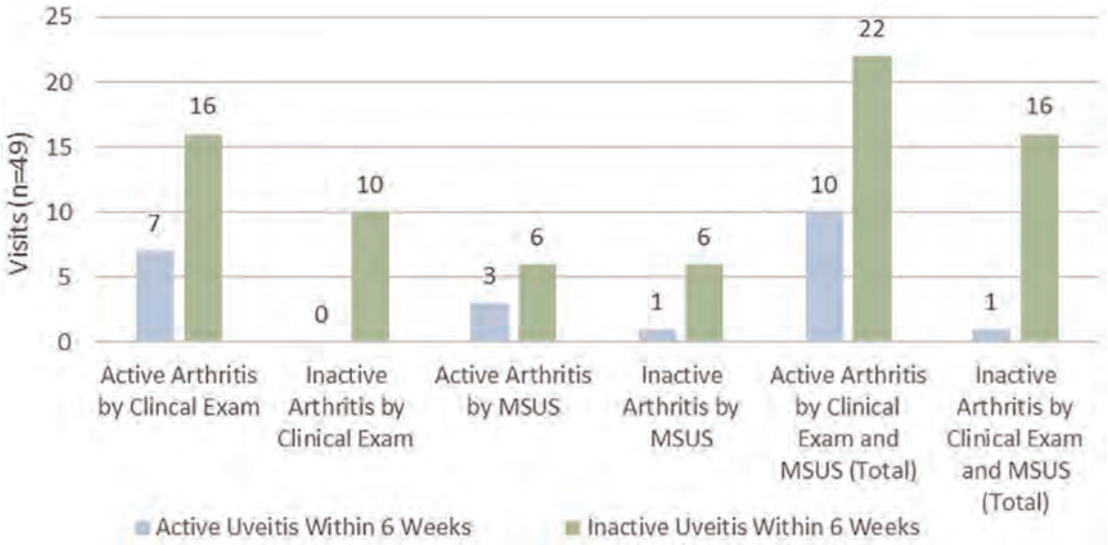


Figure 1: Presence of Uveitis in Patients with Arthritis as Determined by Clinical Examination or MSUS

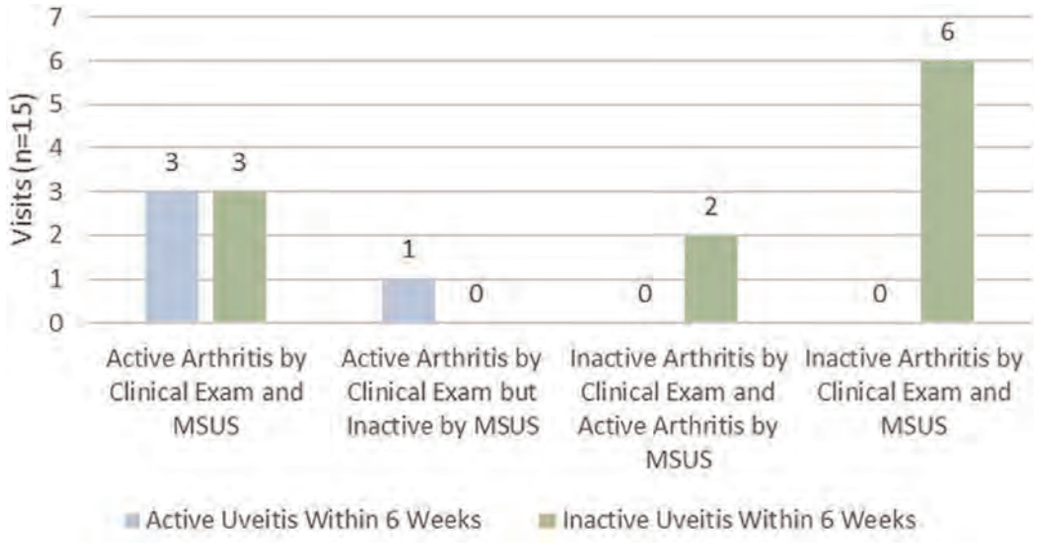


Figure 2: Clinical Exam vs. MSUS Comparison

References

1. Liebling, E., et al. Temporal Relationship Between Juvenile Idiopathic Arthritis Disease Activity and Uveitis Disease Activity. *Arthritis Care Res* 2022;74:349–54.
2. Vega-Fernandez, P., et al. Ultrasonography in Pediatric Rheumatology. *Rheum Dis Clin North Am* 2022;48:217-31.

Disclosure: M. Karumuri: None; **M. Quinlan-Waters**: None; **A. Duell**: None; **K. Rogers**: None; **S. Angeles-Han**: None; **P. Vega-Fernandez**: None.

Abstract Number: 066

Telemedicine Use in the Assessment of Juvenile Myositis: A Mixed-Methods Study of an International Healthcare Provider Experience

Y. Ingrid Goh[1], **Peter Blier**[2], Bianca Lang[3], Marietta De Guzman[4], Julie Fuller[5], Kristin Houghton[6], Kathryn Cook[7], Susan Kim[8], Vanessa Carbone[1], Heather Tory[9], Jo-Anne Marcuz[1], Albert Chow[10], Liza McCann[11], Charalampia Papadopoulou[12], Clarissa Pilkington[13] and Stacey Tarvin[14], [1]The Hospital for Sick Children, Toronto, ON, Canada, [2]Retired, Amherst, MA, [3]Dalhousie University - Halifax, Halifax, NS, Canada, [4]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [5]UT Southwestern, Dallas, TX, [6]University of British Columbia, Vancouver, BC, Canada, [7]Akron Children's Hospital, Akron, OH, [8]UCSF Benioff Children's Hospital, San Francisco, CA, [9]Connecticut Children's Medical Center, S Glastonbury, CT, [10]Loma Linda University, Loma Linda, CA, [11]Alder Hey Children's NHS Foundation Trust, Liverpool, United Kingdom, [12]UCL Institute of Child Health, and Great Ormond Street Hospital NHS Foundation Trust, Section Head Infection, Immunology, and Rheumatology, London, United Kingdom, [13]Great Ormond Street Hospital, London, United Kingdom, [14]Riley Hospital for Children at Indiana University Health, Indianapolis, IN

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Care of patients with juvenile myositis (JM) involves complex assessments performed by specialized healthcare providers (HCPs). Restrictions during the COVID-19 pandemic required the rapid adoption of telemedicine (TM) for evaluation and management of patients. With partial return to in-person care, we sought to understand HCPs current experience with and opinions of TM care.

Methods: A REDCap survey was sent to Childhood Arthritis and Rheumatology Research Alliance (CARRA) and Pediatric Rheumatology European Society (PReS) HCPs from February-September 2022. Quantitative data were analyzed using descriptive statistics and qualitative data were thematically analyzed. Respondents were asked about use of telemedicine, its current place in their provision of JM care, issues, concerns, perceived benefits, and expectations and needs for future use.

Results: There were 361 respondents, although not all answered all questions. Demographic data are presented in Table 1. TM use: Five percent pre-pandemic TM use in JM increased to 87% during the peak of the pandemic but is now 25%. HCPs prefer not to see JM patients by TM; TM visits currently are infrequently offered but often accepted when offered. Most HCPs endorse specific criteria for when a TM visit could be appropriate (e.g., stable disease; family preference; logistics). These align with their opinions on the strengths and weaknesses of telemedicine care. Conducting the visit: TM and in-person visits differ in elements of history taking, organ system evaluation, and use of ancillary data (not shown). There is strong dissatisfaction with the absence of tools for accurate remote assessment of the physical exam, resulting in a shift to use of history information and ancillary studies to determine disease activity (Table 2). Evaluation of the visit: HCPs are generally

Table 1. Characteristics of survey respondents.

Characteristics of survey respondents			
Total respondents:	361	(not all respondents answered all questions)	
Location			
US	180	(85.3%)	34 states represented
Canada	19	(9.0%)	6 provinces represented
Other countries	12	(5.7%)	9 other countries represented
Specialty			
Pediatric rheumatology	199	(94.3%)	
Adult+pediatric rheumatology	12	(5.7%)	
Position/role			
Attending	175	(83.4%)	
Fellow	32	(15.2%)	
Other (NP 2; prefer not to answer 1)	3	(1.4%)	
Practice size			
Large center	145	(68.7%)	
Small center	66	(31.3%)	

Table 2. Usage of telemedicine in the care of JM patients.

Overall assessment of telemedicine care provision and practical use in JM

Question	Strongly agree/ agree	Neither agree/ disagree	Disagree/ strongly disagree
I am comfortable providing care to JDM patients over telemedicine	62.7%	27.5%	9.8%
I am satisfied with the care I provide over TM	44.9%	33.3%	18.8% (3% no ans)
I would consider using TM as part of regular practice	76.0%	12.2%	11.8%
Telemedicine has met my needs as a rheumatologist	41.8%	40.0%	15.2%
Telemedicine has met the needs of my JM patients	46.7%	40%	10.3% (3% no ans)
I feel I can adequately address all issues during a TM visit	28%	20%	52% (3% no ans)

Findings most predictive of JM disease activity (each respondent chose 5 of 18 offered)

During a TM visit	No. choosing	During an In-person visit	No. choosing
Symptomatic muscle weakness	123	Muscle enzymes	112
Muscle enzymes	118	Skin assessment	99
Ability to perform self-care	107	CMAS	98
Ability to perform extracurricular activities	97	Symptomatic muscle weakness	80
Parent/patient global assessment	88	Nailfold capillaroscopy	78

Table 3. Major benefits, challenges, and thematically assessed opinions of telemedicine visits for JM

Major Benefits, Challenges, and Thematically Assessed Opinions of Telemedicine Visits for JM	
Benefits (respondents chose as many as applied)	**No. choosing**
More convenient for patient/family	189
Travel (distance, expense, time)	188
Access to care during pandemic	173
Avoid school/work time lost	165
Monitoring patients who are in remission	157
Continuity of care for families who do not want to travel to in-person visits	145
Ability to see patient's home/living situation, identify challenges and safety issues	92
Challenges (respondents chose as many as applied)	**No. choosing**
Limited ability to perform physical exams (e.g., muscle, skin, other organ)	158
Technology (access, literacy, quality)	99
Licensure/insurance/reimbursement	95
Addressing adolescent health issues (e.g., privacy concerns)	89
Follow-up issues (labs, education, scheduling, multidisciplinary care)	85
Patients unable to assist with examination	66
Patients not knowing how to prepare for visit	43
Identified qualitative themes: deficiencies in TM care for JM	
Inadequacy of obtaining robust physical examination information	
Need for standardization and resources for both families and providers in preparing for and conducting a reliable, quality TM visit	
Insurance, reimbursement, and licensing limitations in the US that limit access to TM care for those who need it	

comfortable providing care over TM, are satisfied with the care provided, and would consider continued use of TM. However they are less satisfied that telemedicine as currently practiced has met their needs or the needs of their patients, and strongly disagree that they can adequately address all issues raised during a TM visit (Table 2). Opinions of TM: HCPs recognize the benefits to patients of TM visits, but outline a consistent set of deficiencies causing dissatisfaction with TM care that fall into 3 themes (Table 3). Respondents offered specific, actionable proposals to address the issues raised (not shown).

Conclusion: TM care of JM patients has greatly diminished since its peak during the pandemic. Providers are comfortable with TM care but are concerned by its limitations, and restrict TM use accordingly. They proposed actionable steps to improve the utility, quality, and acceptability of TM care for patients with JM.

Disclosure: Y. Goh: None; **P. Blier**: None; **B. Lang**: None; **M. De Guzman**: None; **J. Fuller**: None; **K. Houghton**: None; **K. Cook**: None; **S. Kim**: None; **V. Carbone**: None; **H. Tory**: None; **J. Marcuz**: None; **A. Chow**: None; **L. McCann**: None; **C. Papadopoulou**: None; **C. Pilkington**: None; **S. Tarvin**: None.

Abstract Number: 067

Clinical and Demographic Characteristics of Children with Anti-NMDAR Encephalitis

Yike Jiang, Alexander Sandweiss, Timothy Erickson, Eyal Muscal and Kristy Murray, Baylor College of Medicine, Houston, TX

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: The discovery of anti-*N*-methyl-d-aspartate receptor (NMDAR) encephalitis heralded a new class of neuropsychiatric illnesses mediated by autoantibodies, known collectively as autoimmune encephalitis. The clinical characteristics and potential triggers in pediatric cases of anti-NMDAR encephalitis are not well understood. In North America, diagnosis and immunomodulation of children with anti-NMDAR encephalitis are often undertaken by multi-disciplinary teams that include rheumatologists. Our objective is to characterize presentations and potential etiologies of childhood-onset anti-NMDAR encephalitis at a large tertiary urban center that covers a multi-state catchment area.

Methods: After IRB approval, we retrospectively reviewed the initial presentation of patients with anti-NMDAR encephalitis at Texas Children's Hospital between 2009 and 2021.

Results: Our cohort included 76 children who were predominantly female (49/86, 64%) and Hispanic (50/76, 66%). The mean age of onset was 9.5 5.6 years old with a bimodal distribution. This was a larger number than presented annually with neuropsychiatric SLE during the study period (6 vs 2 per year). All children with anti-NMDAR encephalitis had behavioral/cognitive symptoms, 60 (79%) had seizures, 55 (73%) had speech abnormalities, 51 (67%) had movement disorders, and 46 (61%) had memory deficits. Regarding potential etiologies, 4 (5%) had ovarian teratoma (mean age 12.5 1.5 years), and 9 (12%) had a recent history of herpes simplex virus (HSV) encephalitis (mean age 3.7 1.4 years). The remaining 63 (83%) patients had no known causative trigger (mean age 10.1 0.7 years).

Conclusion: In a large single-center pediatric cohort of anti-NMDAR encephalitis, most cases were unrelated to HSV encephalitis or ovarian teratomas. Age of onset had a bimodal distribution representing pre-pubertal and post-pubertal. Female and Hispanic children were disproportionally affected compared to the general inpatient population.

Disclosure: Y. Jiang: None; **A. Sandweiss**: None; **T. Erickson**: None; **E. Muscal**: sobi, 1; **K. Murray**: None.

Abstract Number: 068

Outcomes of Children with Uveitis Associated with Autosomal Dominant Neovascular Inflammatory Vitreoretinopathy (ADNIV)

Ilaria Maccora[1], Arjun Sood[2], Grant Schulert[3], Megan Quilan-Water[3], Alexandra Duell[3], Jennifer Huggins[3], Tiffany Nguyen[2], Cameron Sapp[2], Sumit Sharma[4], Srivastaval Sunil[4] and Sheila Angeles-Han[5], [1]IRCCS Meyer Children's Hospital, Rheumatology Unit, Florence, Italy, [2]Cincinnati Eye Institute, Cincinnati, OH, [3]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, [4]Cole Eye Institute, Cleveland Clinic, Cleveland, OH, [5]Cincinnati Children's Hospital, Cincinnati, OH

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Table 1

Table 1. Characteristics of children with uveitis associated with Autosomal Dominant Neovascular Inflammatory Vitreoretinopathy (ADNIV)	
N (%) unless otherwise stated	n = 8 patients, 16 eyes
Caucasian	8 (100)
Hispanic or Latino	0 (0)
Female	5 (62.5)
Age at ADNIV diagnosis, years, median (range)	14 (9-16)
Duration of ADNIV, mos, median, (range)	12 (6-13)
Asymptomatic at diagnosis	4 (50)
Initial ocular examination and imaging per patient (P) and eyes (E)	
Bilateral	8 (100)
Visual acuity range, worse eye	20/15 – 20/100
Anterior chamber cells present	0 (0)
Vitreous cells present	7 P (87.5) 13 E (81.2)
Macular edema on OCT	3 P (37.5) 6 E (37.5)
Retinal leakage on UWFA	8 P (100) 15 E (93.7)
Neovascularization on clinical exam/UWFA	2 P (25) 4 E (25)
Cataract	2 P (25) 4E (25)
Most recent follow up exam per patient (P) and eyes (E)	
Visual acuity range, worse eye	20/20 – 20/200
AC cells present	0 (0)
Vitreous cells present	6 P (75) 11E (68.7)
Macular edema on OCT	0 (0)
Retinal leakage on UWFA	8 P (100) 15 E (93.7)
Neovascularization on clinical exam/UWFA	2 (25) 4 E(25)
History of Vitreous Hemorrhage	2 P (25) 3 E(18.7)
Cataract	3 P (37.5) 6 E(37.5)
Labs	
Earliest ESR, elevated n=8	0 (100)
Earliest Vitamin D, decreased, n = 7	3 (37.5)
ANA positive, n = 8	0
ACE positive, n = 6	0
Lysozyme positive, n = 3	0
Treatment	
Anti-VEGF	2 (25)
Steroids	6 (75)
Oral	6 (75)
Injection	6 (75)
Time from ADNIV diagnosis to steroid, mos, median (IQR)	0 (0-0.9)
Intraocular Methotrexate	3 (37.5)
Methotrexate	7 (87.5)
Time from ADNIV diagnosis to MTX, mos, median (R)	1.5 (0.5-2)
Duration on methotrexate median (range)	11 (6-13)
Infliximab/inflectra	7 (87.5)
Time from ADNIV to diagnosis to start IFX, mos, median (IQR)	3.2 (2.5-3.1)
Duration on infliximab/inflectra, mos, median (R)	7 (3.5-10)
Patients who stopped Infliximab for inefficacy	5 (71.4)
Tocilizumab	5 (62.5)
Time from ADNIV to diagnosis to start TCZ, mos, median (R)	9 (8-12)
Duration on Tocilizumab, mos, median (R)	3 (0-4)
Abbreviations: ADNIV- Autosomal Dominant Neovascular Inflammatory Vitreoretinopathy; OCT- Optical coherence tomography; UWFA- Ultra-widefield angiography; VEGF- Vascular endothelial growth factor, E= eyes, P=patients	

Background/Purpose: Pediatric uveitis is commonly associated with rheumatic disease and can lead to sight-threatening complications if not properly treated. Systemic immunomodulatory therapy has dramatically changed prognosis of corticosteroid refractory uveitis. Autosomal dominant neovascular inflammatory vitreoretinopathy (ADNIV) is a rare autoimmune condition caused by mutations in *CAPN5*, typically diagnosed in adults, and characterized by intermediate uveitis, retinal degeneration and neovascularization. In the early stages it is asymptomatic but inevitably leads to permanent blindness despite treatment; however routine testing for CAPN5 and ophthalmic screening for uveitis is not regularly done in at-risk children. Proteomic studies have shown that IL6 and VEGF are elevated in the vitreous, suggesting a possible role for targeted therapy to alter disease trajectory. Our aim is to present the short-term outcomes of the first cohort of children diagnosed with ADNIV.

Methods: Cohort study of patients ≤18 years old at diagnosis with (+) *CAPN5* gene mutation (p.Leu244Pro), ocular findings consistent with ADNIV and a minimum follow-up of 6months (m). Treatment response was defined as a decrease in 1) vitreous cells on clinical examination, 2) retinal vascular leakage on ultra-widefield fluorescein angiography (UWFA), and/or 3) macular edema on optical coherence tomography (OCT).

Results: Of 19 children with a family history of ADNIV, 9 were (+) for *CAPN5* mutations and 8 (16 eyes) met the inclusion criteria (Table 1). Sixty-two percent were females, and median age of ADNIV diagnosis was 14 (range 9-16) years. The median follow-up is 12 months (m) (range [R] 6-13). Four children (50%) were asymptomatic and diagnosed by clinical examination/imaging. At diagnosis, visual acuity in the worse eye was 20/100 or better, none had anterior uveitis, while 7 had vitreous cells and vascular leakage (UWFA), 2 neovascularization (UWFA), 3 macular oedema (OCT) and 1 cataract. Laboratory tests (Table 1) were all negative with the exception of Vitamin D (decreased in 3/7). Five of the 8 children were initially treated with oral (n=5) or local/injected corticosteroids (n=4), and anti-VEGF therapy (n=2). Due to persistent inflammation, systemic treatment was started in 7/8 patients. First line treatment was MTX (1 mg/kg [max 20 mg] weekly SQ) that is still ongoing in all (median duration 11 m, R6-12). Because of absent response, IFX (10 mg/kg/dose every 4 weeks) was added in all patients after a median time from diagnosis of 3.2 m (R2.5-3.1) and continued for a median time of 7 m (R3.5-10). However, IFX was ineffective in all patients, and 5/7switched to tocilizumab (10 mg/kg/every 2 weeks IV) after a median time from diagnosis of 9 m (R1-12) with a median duration of 3 m (R0-4). Outcomes at the last available follow-up are reported in Table 1.

Conclusion: We report on the largest series of children with ADNIV treated with systemic immunosuppression. Early testing for CAPN5 gene in at risk children, and regular scheduled screening for uveitis and vasculitis will lead to prompt intervention. MTX and IFX seem ineffective and tocilizumab might be a promising treatment based on underling mechanism.

Disclosure: I. Maccora: None; **A. Sood**: Alimera Sciences, Inc, 12., Carl Zeiss Meditec, Inc, 12., EyePoint Pharmaceuticals, 12.; **G. Schulert**: Novartis, 2, SOBI, 2; **M. Quilan-Water**: None; **A. Duell**: None; **J. Huggins**: None; **T. Nguyen**: None; **C. Sapp**: None; **S. Sharma**: AbbVie/Abbott, 2, bausch/lomb, 2, clearside, 2, eyepoint, 2, Genentech, 2, 5, Gilead, 5, ionis, 5, Regeneration, 2, Roche, 2, 5, Santen, 5; **S. Sunil**: AbbVie/Abbott, 2, Allergan, 5, Bausch and Lomb, 2, Eyepoint, 2, 5, Eyevensys, 2, 5, Novartis, 2, Regeneron, 2, 5, Santen, 5, Zeiss, 2; **S. Angeles-Han**: None.

Abstract Number: 069

Neutrophil-Lymphocyte and Platelet-Lymphocyte Ratios of Patients with Juvenile Systemic Lupus Erythematosus

Rogerio do Prado[1], Fabiola I Suano-Souza[1], Wellington D. R. Rodrigues[2], Lucila Pereira[1], Maria Teresa Terreri[3] and Roseli O. S. Sarni[4], [1]FMABC, São Paulo, Brazil, [2]Unicid, São Paulo, Brazil, [3]UNIFESP, São Paulo, Brazil, [4]Centro Universitário Faculdade de Medicina do ABC (FMABC), Brazil

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: The Neutrophil-Lymphocyte ratio (NLR) and Platelet-Lymphocyte ratio (PLR) are markers of systemic inflammation easily available from the blood count. Previous studies have revealed that elevated NLR and PLR are independently associated with cardiovascular mortality. Patients with Systemic Lupus Erythematosus (SLE) have elevated NLR and PLR values in association with disease activity and renal impairment. Studies in juvenile SLE (jSLE) are scarce and show that both NLR and PLR are correlated with serological indicators and can predict tissue involvement, mainly cutaneous, articular, serositis and hematological involvement. Our aim was to compare the NLR and PLR ratios of patients with jSLE with healthy controls. In the jSLE group we also aimed to verify association of NLR and PLR with SLEDAI-2K, ultrasensitive C-reactive protein (usCRP), ESR, plasma selenium and erythrocyte glutathione peroxidase activity (GPx).

Methods: This was an observational, cross-sectional study of 31 female adolescents with jSLE and 31 healthy females as a control group. Demographic and anthropometric data were obtained from both groups, and clinical data (disease activity measured by SLEDAI-2K and corticosteroid - CTC use) from jSLE group; laboratory data including ESR, plasma selenium, GPx, usCRP and complete blood count (NLR, PLR).

Results: Mean age at diagnosis and mean of disease duration of jSLE group was 15.9±1.7 years and 3.6±2.6 years, respectively. The median NLR was significantly higher in the jSLE group [2.37 (0.43;15.55)] compared to the control [1.62 (0.70;5.80)] (p=0.025). The median PLR was also higher in the jSLE group [221.36 (18.36;1,007.7)] compared to the control [127.8 (51.0;202.1)] (p< 0.001). The NLR did not show a significant correlation with any other variables. There was a significant Correlation between PLR and ESR (rho=0.360; p=0.046), dosis of CTC (mg/kg/day) (rho=0.617; p=0.008) and with GPx (rho=0.360; p=0.047). No association was observed between both NLR and PLR and SLEDAI-2K.

Conclusion: Patients with jSLE have higher values of NLR and PLR. In the jSLE group, only PLR was associated with inflammation and CTC use; however no association was found with SLEDAI-2K.

Disclosure: R. do Prado: None; **F. I Suano-Souza**: None; **W. D. R. Rodrigues**: None; **L. Pereira**: None; **M. Terreri**: None; **R. O. S. Sarni**: None.

Abstract Number: 070

The Impact of the COVID-19 Pandemic on Patients with Juvenile Idiopathic Inflammatory Myopathies

Dawn Wahezi[1], Dominique Jerome[1], Evin Rothschild[1], Jeffrey Dvergsten[2], Stacey Tarvin[3], Susan Kim[4] and Tamar Rubinstein[1], [1]Children's Hospital at Montefiore, New York, NY, [2]Duke University Hospital, Durham, NC, [3]Riley Hospital for Children at Indiana University Health, Indianapolis, IN, [4]UCSF Benioff Children's Hospital, San Francisco, CA

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Since the onset of the COVID-19 pandemic, there have been concerns regarding the risks of SARS-CoV-2 infection in patients with juvenile idiopathic inflammatory myopathies (JIIM), with few reports of disease flare following COVID-19. In this study, we investigated the impact of the COVID-19 pandemic on children and adolescents with JIIM.

Methods: Data were collected using a patient/caregiver survey in English and Spanish via Research Electronic Data Capture (REDCap) database. Participants were eligible if they were diagnosed with JIIM and currently under the age of 21 years old. Caregivers were required to complete surveys for participants under 18 years old. Surveys were distributed via the CureJM organization, social media, the Childhood Arthritis and Rheumatology Research Alliance (CARRA) network and the Dr. Peter Dent Pediatric Rheumatology Bulletin Board.

Results: A total of 84 respondents accessed the survey, with 70 respondents consenting to participate (83%) and 54 completing the full survey (64%). Surveys were primarily completed by parents/caregivers (93%) with the majority of patients reported as having juvenile dermatomyositis (JDM) (95%), female sex (70%), White (74%) and non-Hispanic (64%) (Table 1). Median disease duration of JIIM participants was 4.06 years [IQR: 2.04, 8.70]. Among respondents, 27 out of 57 (47%) reported testing positive for COVID-19, with 7 (12%) testing positive on more than one occasion. The majority of

Table 1: Demographics and JIIM baseline features (n=70)*	N (%)
Sex	
Female	49 (70)
Male	20 (29)
Prefer not to answer	1 (1)
Current age of JIIM patient	
Under 5	5 (7)
6-10 years	22 (31)
11-14 years	20 (29)
15-17 years	11 (16)
18-21 years	12 (17)
Race (n=68)	
White	50 (74)
Black/African American	7 (10)
American Indian	1 (1)
Asian	2 (3)
Other Race	4 (6)
Prefer not to answer	4 (6)
Ethnicity	
Hispanic	22 (31)
Non-Hispanic	45 (64)
Prefer not to answer	3 (4)
JIIM Subtype (n=62)	
JDM	59 (95)
JPM	3 (5)
JIIM clinical manifestations at diagnosis (n=62)	
Skin Disease	52 (84)
Muscle Disease	54 (87)
Gastrointestinal Involvement	7 (11)
Lung Involvement	9 (15)
Heart Involvement	6 (10)
Joint Disease	16 (26)
JIIM autoantibodies (n=63)	
P155/140 (TIF-1)	8 (13)
MJ (NXP-2)	5 (8)
Jo-1 (anti-synthetase)	2 (3)
Mi-2	1 (2)
MDA-5 (CADM-140)	4 (6)
Other	3 (5)
Negative Antibodies	3 (5)
Unknown or not done	37 (60)
Medications during 6 months prior to COVID-19 exposure/infection (n=62)	
Steroids	25 (40)
Methotrexate	34 (55)
Hydroxychloroquine	26 (42)
Intravenous immunoglobulin (IVIG)	25 (40)
Rituximab	11 (18)
Mycophenolate mofetil	16 (26)
Anti-TNF agent	1 (2)
Calcineurin Inhibitors	2 (3)
Other	6 (10)
None of the above	8 (13)

*N= 70 participants, except where otherwise indicated.

patients (89%) reported typical symptoms of fever, cough, headache and fatigue (Figure 1). No patients were hospitalized or received medications to treat COVID-19. Four patients reported a flare of JIIM symptoms after COVID-19 including rash, weakness, myalgias/arthralgias and abdominal pain; three of these patients reported holding immunomodulatory medications in the context of active COVID-19. Among patients who tested positive for COVID-19, there was minimal change in patient global assessment scale (GAS) prior to and post-COVID-19 (median GAS [IQR]: 2 [0,3] and 3 [1,5] respectively; p=0.57) (Figure 2). A total of 69% of participants reported being vaccinated against COVID-19, with 9 (24%) reporting minor vaccine side effects including arm pain, headaches, fever and chills. One patient reported JIIM flare post vaccination. Medication modifications made in response to SARS-CoV-2 exposure, infection and vaccination included medications being held or delayed for 7-14 days in 16%, 26%, 22% respectively. Overall, 16% of respondents reported concerns related to

delayed appointments, difficulty obtaining medication and avoidance of hospital care due to risk of exposure. In addition, 39% reported psychosocial impact including anxiety, depression, stress, social withdrawal, irritability, anger and two participants endorsed suicidal ideation.

Conclusion: Based on our survey, patients with JIIM had mild symptoms related to COVID-19 with none requiring hospitalization. The majority of patients tolerated COVID-19 vaccination well with minor side effects. Few patients report disease flare or worsening GAS post-COVID-19 or vaccination. Mental health concerns were demonstrated in JIIM patients during the COVID-19 pandemic.

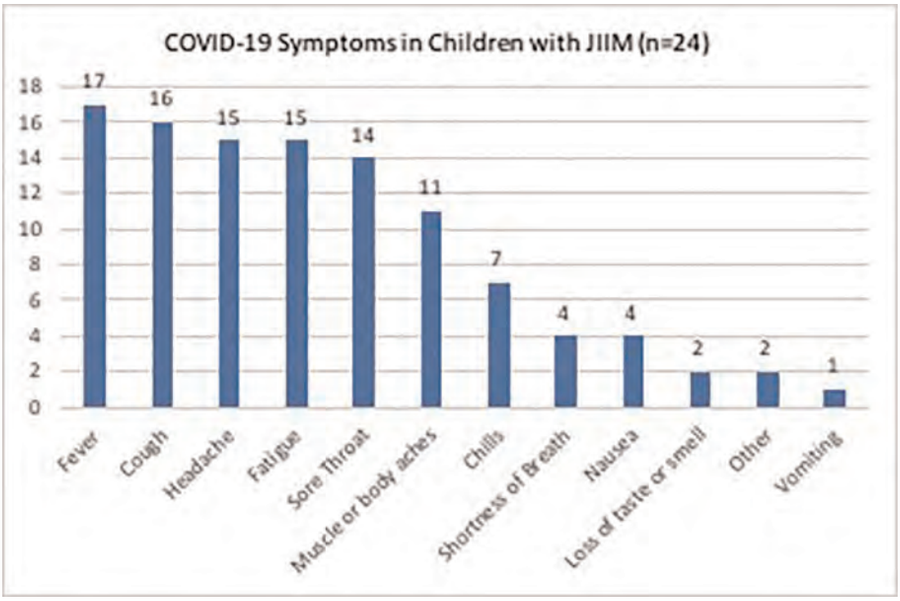


Figure 1: Twenty-four patients out of 27 who tested positive for COVID-19 reported acute COVID-19 symptoms. Total 108 symptoms were reported.

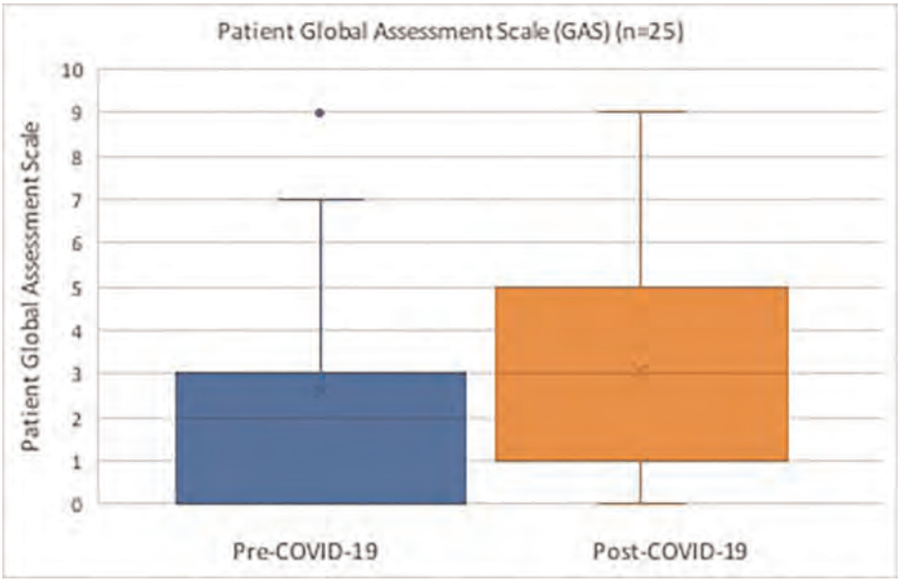


Figure 2: Global assessment scale (GAS) for JIIM disease activity reported by participants pre- and post-COVID-19 (n=25)

Disclosure: D. Wahezi: None; **D. Jerome**: None; **E. Rothschild**: None; **J. Dvergsten**: None; **S. Tarvin**: None; **S. Kim**: None; **T. Rubinstein**: None.

Abstract Number: 071

Clinical Manifestations and Management of Takayasu Arteritis: A Single Center Pediatric Cohort

Ana Luiza Altaffer[1], Alvaro Orjuela[2] and Marietta De Guzman[2], [1]Texas Children's Hospital, Houston, TX, [2]Baylor College of Medicine, Texas Children's Hospital, Houston, TX

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Takayasu arteritis (TA) is a rare granulomatous vasculitis that affects large vessels, including the aorta, its major branches, and the pulmonary artery. Data on the presentation and clinical course of pediatric TA are limited.

Methods: With an institutional review board approval, a retrospective cohort study of children who were diagnosed with TA at Texas Children's Hospital between 2005 and 2022 was performed. Data were abstracted from the electronic medical record. Patient demographics, presenting symptoms and signs, imaging findings, and management data were evaluated using standard descriptive statistics.

Table 1. Clinical Presentation

Symptoms	n (%)
Constitutional symptoms	11 (100)
Fever	4 (36.4)
Weight loss	9 (81.8)
Fatigue	10 (90.9)
Musculoskeletal symptoms	8 (72.7)
Arthralgia	4 (36.4)
Myalgia	5 (45.5)
Arthritis	1 (9.1)
Other[1]	2 (18.2)
Neurologic symptoms	6 (54.5)
Headache	5 (45.5)
Numbness/paresthesias	1 (9.1)
Claudication	1 (9.1)
Exam findings	n (%)
Hypertension	6 (54.5)
Malignant hypertension[2]	2 (18.2)
BP differential[3]	5 (45.5)
Decreased pulse	4 (36.4)
Absent pulse	2 (18.2)
Bruit	3 (27.3)
Carotid artery tenderness	0 (0)

Legend. [1]Other: morning stiffness, muscle spasm
[2]Malignant hypertension: stage 2 hypertension with severe examination or laboratory evidence of ≥ 1 organ damaged (per APP 2017 criteria)
[3]BP differential: difference in systolic BP of ≥ 10 mmHg between any two limbs

Results: Eleven patients were included. All but one were female (90.9%), with a mean age at diagnosis of 13.5 years (range 9-17). Most were Hispanic or Latino (63.6%). Diagnosis was made a median of 4 months following symptom onset (IQR 1-15), and the mean follow up period was 45.7 months (range 7-112). The most common symptoms on presentation were constitutional, with all patients having at least one of fatigue (90.9%), weight loss (81.8%), or fever (36.4%). Only one patient had claudication. Hypertension (54.5%) was the most common exam finding. All patients had an elevated ESR (mean

Table 2. Presenting Lab Findings

	Presentation
Hematuria[1], n (%) 1+ 2+ 3+	6 (54.5) 3 (27.3) 2 (18.2) 1 (9.1)
Proteinuria[2], n (%) 1+ 2+ 3+	3 (27.3) 2 (18.2) 0 (0) 1 (9.1)
Elevated UPC[3], n (%) UPC in mg/mg, mean (SD)	3 (50.0) 0.283 (0.183)
Elevated Cr[4], n (%) Cr in mg/dL, mean (SD)	2 (18.2) 0.6 (0.1)
Anemia[5], n (%) Hemoglobin in g/dL, mean (SD)	6 (54.5) 10.6 (1.6)
Leukocytosis[6], n (%) WBC count x10^3/uL, mean (SD)	3 (27.3) 9.40 (1.59)
Elevated CRP[7], n (%) CRP in mg/dL, mean (SD)	9 (81.8) 5.2 (4.2)
Elevated ESR[8], n (%) ESR in mm/hr, mean (SD)	11 (100) 82 (35)

Legend. [1]Hematuria: 1+ blood or greater

[2]Proteinuria: 1+ protein or greater

[3]UPC: 6 measured; upper limit of normal = 0.2

[4]Elevated Cr: > or = 0.60 mg/dL for females > or = 5 years to < 12 years; > or = 0.80 mg/dL for females > or = 12 years to < 19 years; > or = 1.04 mg/dL for females > or = 19 years; > or = 0.60 mg/dL for males > or = 5 years to < 12 years

[5]Anemia: Hgb < 10.6 for females > or = 6 years to < 12 years; Hgb < 10.8 for females > or = 12 years to < 18 years; Hgb < 10.6 for females > or = 18 years; Hgb < 10.6 for males > or = 6 years to < 12 years.

[6]Leukocytosis: WBC > or = 11.40 for females > or = 6 years to < 12 years; WBC > or = 9.43 for females > or = 12 years to < 18 years; WBC > or = 9.68 for females > or = 18 years; WBC > or = 11.40 for males > or = 6 years to < 12 years.

[7]CRP: upper limit of normal = 1.0 mg/dL

[8]ESR: upper limit of normal = 20 mm/hr

Table 3. Treatments

Medications	N (%)	Time to start in months, median (IQR)
Steroids	11 (100)	0 (0)
Methotrexate	9 (90)	0 (0-0)
Cyclophosphamide	2 (20)	4.5 (n/a)
Mycophenolate Mofetil	1 (10)	10 (n/a)
bDMARDs Infliximab Tocilizumab Adalimumab Etanercept Ustekinumab	11 (100) 7 (63.6) 7 (63.6) 3 (18.2) 2 (18.2) 1 (9.1)	0 (0-2) 0 (0-0) 2 (0-24) 42 (27-47) 14 (n/a) 16 (n/a)

82 mm/hr, range 25-146), and all but two had an elevated CRP (mean 5.2, range 0.2-13.6). All patients had imaging evidence of aortic involvement, including the abdominal (72.7%), ascending thoracic (63.6%), and descending thoracic (54.5%) segments. All patients were started on systemic corticosteroids at the time of diagnosis. Initial steroid sparing immunomodulatory agents included a combination of Methotrexate and Infliximab in most patients (54.5%). Two were started on Tocilizumab alone, two were started on Methotrexate alone (both diagnosed on or prior to 2010), and one was not started on therapy beyond steroids at the initial encounter due to diagnostic uncertainty. All patients were on a biologic DMARD (bDMARD) at some point in their disease course, with a median time to start of 0 months (IQR 0-2). Reasons for addition or change of therapy included poor disease control or flare, inability to wean steroids, new disease manifestations, infusion reactions, side effects, and ease of administration. Four patients (36.4%) required surgical management. At the most recent follow-up visit, only four patients remained on steroids. One patient was off all medications completely and remained in clinical remission. All but three patients were asymptomatic, and there was a statistically significant decrease in ESR (p=0.01) and CRP (p=0.02) from the initial to the final visit.

Conclusion: Like prior studies, patients in our cohort presented predominantly with constitutional symptoms, hypertension, and elevated inflammatory markers. bDMARDs were introduced earlier in disease course than in previously reported cohorts. Outcomes were generally favorable, and most patients were symptom-free off steroids at the most recent follow-up. However, patients were not often able to wean off all medications.

Disclosure: A. Altaffer: None; **A. Orjuela**: None; **M. De Guzman**: None.

Abstract Number: 072

Musculoskeletal Ultrasound Findings in Children with Psoriasis

Laura Nedorezov, Tracy Ting and Patricia Vega-Fernandez, Cincinnati Children's Hospital Medical Center, Cincinnati, OH

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: The presentation of juvenile psoriatic arthritis (JPsA) in children with psoriasis can be insidious and poses a diagnostic challenge. Musculoskeletal ultrasound (MSUS) has emerged in recent years as a sensitive and non-invasive imaging modality to detect joint inflammation. A growing number of evidence has shown that adult psoriasis patients without signs of arthritis have findings of subclinical joint pathology on MSUS. These patients with identified imaging abnormalities are more likely to develop active psoriatic arthritis as demonstrated in longitudinal studies. No such research has been performed among children with psoriasis however, highlighting a critical gap in the understanding of pediatric psoriatic disease. The objective of this investigation is to describe MSUS and nail ultrasound (US) findings in children with psoriasis.

Methods: Grey-scale (B) mode and power Doppler (PD) mode US were utilized in this cross-sectional study to evaluate joint, enthesis, and nail findings in pediatric psoriasis patients (< 18 years old) without clinical symptoms of arthritis. Exclusion criteria included acute joint injury, trauma, or joint surgery within the last 3 months, on systemic treatment or oral corticosteroids in the last 2 months, or pregnancy. Each subject underwent a full musculoskeletal exam and MSUS evaluation of seven sites bilaterally: a) joints/nails – fingernails, MCP, and IP joints of the second and third fingers, as well as any finger with psoriatic nail involvement, patella, tibiotalar and subtalar joints, b) entheses – quadriceps, proximal and distal patellar tendon,

Achilles, and plantar fascia calcaneal insertion. US images were assessed using an established pediatric-specific scoring system for the joints and semi-quantitative scoring approach for the nails. Enthesitis was evaluated according to the OMERACT definition. Demographic, clinical, and laboratory information was collected when available.

Results: Eleven patients were included until now. Demographic and clinical characteristics are shown in Table 1. The mean duration of the MSUS exam was 66 minutes. US characteristics of the nails studied are displayed in Table 2, and findings of the joints and entheses are shown in Table 3. 51 nails (83.6%) had abnormal nail plate structure per the Wortsman classification, and roughly 20% of nails had increased PD activity in either the nail bed or nail matrix classified as grade 2 or 3. Eight out of 11 patients (72.7%) had at least one grade 2 finding of synovitis on B-mode, with the PIP/DIP, suprapatellar, and anterior subtalar joint recesses most frequently affected. Two patients (18.2%) had enthesopathy involving the lower extremities identified on PD examination.

Conclusion: Subclinical inflammatory abnormalities of the nails, joints, and entheses were identified utilizing MSUS in children with psoriasis who did not have clinical manifestations of arthritis on exam. Ongoing studies involving additional pediatric psoriasis patients and healthy controls is underway to more clearly elucidate the significance of these findings.

Table 1: Demographics and clinical characteristics of the patients studied.

Characteristic	Total, n = 11
Male/female	7/4
Age (years)[a]	11.8 ± 2.4
Ethnicity, n (%)	
Caucasian	7 (63.6)
African American	3 (27.3)
Asian	1 (9.1)
Body mass index (kg/m^2)[b]	18.5 (5.2)
Disease duration (years)[b]	5.4 (7.9)
Psoriasis subtype[c], n (%)	
Plaque	5 (45.5)
Guttate	3 (27.2)
Scalp	5 (45.5)
Palmoplantar	2 (18.2)
Clinical nail involvement, n (%)	6 (54.5)
Morning stiffness (minutes)[b]	0 (22.5)
Active joint count, n (%)	0 (0)
Beighton score[d] > 5, n (%)	4 (36.4)
Current treatment[c], n (%)	
Topical corticosteroid	7 (63.6)
Topical vitamin D analog	1 (9.1)
Topical calcineurin inhibitor	1 (9.1)
No treatment	3 (27.3)

[a]Mean ± standard deviation
[b]Median (IQR)
[c]Each patient could have more than one disease subtype or treatment identified.
[d]Measure to assess generalized joint hypermobility.

Table 2: Ultrasound characteristics of the studied nails, n = 61.

Wortsman classification[a]		n (%)
Normal		10 (16.4)
I		11 (18.0)
II		34 (55.8)
III		1 (1.6)
IV		5 (8.2)
Nail parameters		**Mean ± SD**
NP thickness (mm)		0.41 ± 0.11
NB thickness (mm)		1.67 ± 0.22
Matrix thickness (mm)		1.33 ± 0.22
Distal extensor tendon thickness (mm)		0.66 ± 0.16
PD grade[b]	**PD signal nail bed n (%)**	**PD signal nail matrix n (%)**
0	30 (49.2)	32 (52.5)
1	21 (34.4)	16 (26.2)
2	7 (11.5)	6 (9.8)
3	3 (4.9)	7 (11.5)

NP: nail plate, NB: nail bed, SD: standard deviation, PD: power Doppler
[a]I – focal hyperechoic involvement of the ventral plate, II – loosening of the borders of the ventral plate, III – wavy plates, IV – loss of definition of both plates (Wortsman X, Jemec GB. Ultrasound imaging of nails. *Dermatol Clin*. 2006;24(3):323-328. doi:10.1016/j.det.2006.03.014)
[b]PD Grade 0 – normal, 1 – confluent signal in < 25% of the area, 2 – confluent signal in >25% and < 50%, 3 – confluent signal in > 50% (Arbault A, Devilliers H, Laroche D, et al. Reliability, validity and feasibility of nail ultrasonography in psoriatic arthritis. *Joint Bone Spine*. 2016;83(5):539-544. doi:10.1016/j.jbspin.2015.11.004)

Table 3: Ultrasound findings of all joint and enthesis sites.

Joints	**B-mode, n (%)**				**PD mode, n (%)**
Grade[a]	0	1	2	3	0[b]
Fingers					
MCP joint recess[c], n = 123	108 (87.8)	13 (10.6)	2 (1.6)	0 (0)	123 (100)
PIP/DIP joint recess[c], n = 176	140 (79.5)	17 (9.7)	19 (10.8)	0 (0)	176 (100)
Knee, n = 22					
Suprapatellar recess	14 (63.6)	6 (27.3)	2 (9.1)	0 (0)	22 (100)
Medial parapatellar recess	15 (68.2)	6 (27.3)	1 (4.5)	0 (0)	22 (100)
Lateral parapatellar recess	20 (90.9)	2 (9.1)	0 (0)	0 (0)	22 (100)
Ankle, n = 22					
Anterior tibiotalar recess	19 (86.4)	3 (13.6)	0 (0)	0 (0)	22 (100)
Anterior subtalar recess (from medial)	20 (90.9)	0 (0)	2 (9.1)	0 (0)	22 (100)
Posterior subtalar recess (from lateral)	18 (81.8)	3 (13.7)	1 (4.5)	0 (0)	22 (100)
Enthesis, n = 12	**Abnormal B-mode[d], n**				**Abnormal PD mode[d], n**
Quadriceps tendon	0				1
Proximal patellar tendon	0				1
Distal patellar tendon	0				0
Achilles tendon	0				0
Calcaneal plantar fascia	0				1

PD: power Doppler, MCP: metacarpophalangeal, PIP: proximal interphalangeal, DIP: distal interphalangeal
[a]Vega-Fernandez P, Ting TV, Oberle EJ, et al. The MUSICAL pediatric ultrasound examination – a comprehensive, reliable, time efficient assessment of synovitis. *Arthritis Care Res (Hoboken)*. 2021;10.1002/acr.24759. doi:10.1002/acr.24759
[b]None of the joints had PD findings of grade 1, 2, or 3.
[c]Includes dorsal and volar aspects of the MCP and DIP joint recesses, and volar aspect of the PIP joint recess for each examined finger.
[d]Wakefield RJ, Balint PV, Szkudlarek M, et al. Musculoskeletal ultrasound including definitions for ultrasonographic pathology. *J Rheumatol*. 2005;32(12):2485-2487

Disclosure: L. Nedorezov: None; **T. Ting**: None; **P. Vega-Fernandez**: None.

Abstract Number: 073

Golimumab Therapy in Children with Chronic Recurrent Multifocal Osteomyelitis: A Case Series Reviewing Safety and Efficacy

Claire Yang[1], Natalie Rosenwasser[2], Xing Wang[2], Zheng Xu[2], Joshua Scheck[2], Ramesh Iyer[3] and Yongdong (Dan) Zhao[3], [1]University of Washington School of Medicine, Seattle, WA, [2]Seattle Children's Hospital, seattle, WA, [3]University of Washington, Seattle, WA

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Chronic Recurrent Multifocal Osteomyelitis (CRMO) is an autoinflammatory bone disease requiring immunosuppressive therapy in half of patients. Monoclonal Tumor Necrosis Factor inhibitors (TNFi) are second-line off-label therapies, with undesired paradoxical psoriasisoccurring in a subset of patients on TNFi. This can prompt conversion to alternate therapy which can prove challenging, given few TNFi are approved for use in children.

Objective: To determine the efficacy and safety of golimumab, a fully humanized TNFi, in children with CRMO, including those with paradoxical psoriasis following exposure to other monoclonal TNFi.

Methods: A single center retrospective chart review of patients with CRMO who received golimumab between June 1, 2018, and December 21, 2020, was conducted. Patients diagnosed < 21 years old with ^{3} 1 follow-up and ^{3} 3 months of treatment with golimumab were included.

Data including whole-body MRI (WB-MRI) lesion counts, prior treatment, concomitant medications, demographics, clinically relevant data, laboratory results, patient-reported outcomes (PRO), pain scores, and psoriasis burden were extracted. Linear mixed models with log-transformed outcomes were used to assess changes over time with a random effect included to account for within-subject correlation of repeated measures. Confidence intervals of 95% and p-values were reported and considered significant if ≤ 0.05.

Results: Eighteen patients were included, fourteen of whom were previously treated with disease-modifying antirheumatic drugs (DMARDs) and seventeen receiving other TNFi. Medianbone lesion count, physician global assessments, pain scores, and erythrocyte sedimentation rate decreasedat 3- and 6-months follow-up. Five of the nine patients with baseline paradoxical psoriasis had improvement or resolution of paradoxical psoriasis over time. Two patients had worsening of baseline paradoxical psoriasis and one patient who did not previously have TNFi-induced psoriasis developed psoriasis at 3-months. One patient discontinued therapy at 4 months given lack of radiographic improvement and persistence or worsening of pre-existing paradoxical psoriasis. Two children received dual therapy with both golimumab and ustekinumab with clinical improvement. No serious infections or adverse events (AE) were observed during this study.

Conclusion: Golimumab is safe and effective to treat children with CRMO with a low risk of paradoxical psoriasis and improvement in baseline paradoxical psoriasis induced by other TNFi. Additional long-term surveillance with larger sample size is needed to further characterize the safety and efficacy of golimumab and overall risk of paradoxical psoriasis.

Disclosure: C. Yang: None; **N. Rosenwasser**: None; **X. Wang**: None; **Z. Xu**: None; **J. Scheck**: None; **R. Iyer**: None; **Y. Zhao**: Bristol-Myers Squibb(BMS), 5.

Abstract Number: 074

Clinical Characteristics of Juvenile Systemic Sclerosis in Korea: A 30-year Single Center Study

Jieun Jeong[1], Minji Kim[1], Jiwon Jung[2], Seon Hee Lim[3] and Seong Heon Kim[1], [1]Department of Pediatrics, Seoul National University Children's Hospital, Seoul, Republic of Korea, [2]Department of Pediatrics, Asan Medical Center Children's Hospital, University of Ulsan College of Medicine, Seoul, Republic of Korea, [3]Department of Pediatrics, Pusan National University Children's Hospital, Yangsan, Republic of Korea

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Juvenile systemic sclerosis (jSSc) is a rare autoimmune, chronic, multisystem, connective tissue disease characterized by progressive tissue fibrosis of the skin and internal organs. To date, there have been few published studies on the clinical features of jSSc in East Asian population. The aim of this study was to describe clinical and laboratory characteristics at diagnosis, therapeutic drugs, and prognosis of jSSc at a single, tertiary-care hospital in South Korea.

Methods: The study was a retrospective analysis of jSSc patients having age of onset less than 16 years and underwent evaluations at Seoul national university hospital, between January 1992 and July 2022. All patients met the PReS/ACR/EULAR provisional classification criteria for jSSc, and patients with localized scleroderma (morphea) was excluded.

Results: Among the 13 jSSc patients, 8 patients (62%) were female, and the mean age of symptom onset was 10 years (range 2–13 years). The median time from onset of symptoms to diagnosis was 2 years (range 0.4–5 years). Proximal skin sclerosis (13 patients, 100%), Raynaud's phenomenon (RP) (11, 85%), and sclerodactyly (9, 69%) were presented at the

Table 1. Demographic data of patients

Patient characteristics [a]	Total	Clinical subtypes of systemic sclerosis		
		Overlap	Diffuse SSc	Limited SSc
	(N=13)	(N=6)	(N=4)	(N=3)
Female - No. (%)	8 (61.5)	2 (33.3)	3 (75)	3 (100)
Age of symptom onset - Median, years	10 [2-13]	4.5 [2-12]	12.5 [4-13]	12 [5-13]
Age at diagnosis - Median, years	11 [3-17]	7.5 [3-12]	13 [6-15]	15 [7-17]
Duration from symptom onset to diagnosis - Median, years	2 [0.4-5]	1.1 [0.4-5]	1.5 [0.5-2]	2 [2-5]

a. Continuous data are presented as median [range]. Categorical data are presented as number (No.) and percentage (%).

Table 2. Frequency of clinical features at symptom onset, diagnosis, and during follow-up

	Patients (N=13)	At symptom Onset - N (%)	At diagnosis - N (%)	Newly developed during the course - N (%)
Cutaneous	Proximal skin sclerosis	5 (38)	13 (100)	0 (0)
	Sclerodactyly	0 (0)	9 (69)	0 (0)
Peripheral	Raynaud's phenomenon	10 (77)	11 (85)	0 (0)
	Telangiectasia	0 (0)	2 (15)	0 (0)
	Digital tip ulcer	0 (0)	5 (38)	2 (15)
Cardiac	Arrhythmias	0 (0)	0 (0)	1 (8)
Renal	Proteinuria	0 (0)	0 (0)	1 (8)
Respiratory	Interstitial lung disease	0 (0)	5 (38)	2 (15)
	Pulmonary arterial hypertension	0 (0)	1 (8)	1 (8)
Musculoskeletal	Arthritis	1 (8)	5 (38)	1 (8)

time of diagnosis. However, the first symptom patients reported the most was RP which was present in 10 patients (77%). In comparison, proximal skin sclerosis was present only in 5 (38%).

A total of 13 patients (100%) had positive result of ANA test. Anti Scl-70 antibody (ab) was positive in 7 (54%) out of 13 patients tested, and anti RNP ab was positive in 2 out of 11 patients tested. Autoantibody tests including, RF, anti CCP ab, anti-dsDNA ab, anti-Smith ab, ACA, ANCA, and ASCA were performed, but all were negative.

At the time of diagnosis, 5 patients showed mild restrictive pattern on pulmonary function tests (PFT) or chest CT findings of interstitial lung disease, of which 3 felt dyspnea on exertion, and 1 out of 3 complained chronic cough. In the other 2, there was no subjective dyspnea despite abnormal findings.

Glucocorticoids were the most prescribed drug as an initial therapy, which was used in 8 patients, followed by Methotrexate in 6 patients. As a second-line therapy, 5 patients used MMF and 2 patients used CPM to control severe symptoms. For severe RP, 12 patients took calcium channel blockers.

During follow-up, 3 patients developed lung disease, 1 patient had impaired renal function, and 1 patient developed heart disease.

Conclusion: In the early phase of jSSc, the diagnosis is very difficult due to insidious and subtle onset of skin changes. In our population, proximal skin sclerosis, which is crucial in diagnosis of jSSc, was not present in the early phase of disease. Considering the importance of timely diagnosis and treatment in the early phase of disease, clinical suspicion is essential for diagnosis of jSSc with RP especially when ANA is positive. Since all autoantibodies other than ANA were negative in 38% of patients, jSSc cannot be excluded even if all autoantibodies other than ANA are normal in children with RP, and close physical examination is required. PFT and chest CT should be performed at diagnosis as 38% of patients showed lung involvement at the initial evaluation with or without respiratory symptoms.

Disclosure: J. Jeong: None; **M. Kim**: None; **J. Jung**: None; **S. Lim**: None; **S. Kim**: None.

Abstract Number: 075

Emapalumab Treatment Followed by Hematopoietic Stem Cell Transplantation in Systemic Juvenile Idiopathic Arthritis Complicated by Recurrent Macrophage Activation Syndrome

Claudia Bracaglia[1], Manuela Pardeo[1], Giulia Marucci[2], Simona Riccio[2], Francesco Quagliarella[3], Ivan Caiello[2], Giusi Prencipe[2], Pietro Merli[3], Franco Locatelli[3] and Fabrizio De Benedetti[1], [1]Division of Rheumatology, IRCCS Ospedale Pediatrico Bambino Gesù, Roma, Italy, [2]Division of Rheumatology, IRCCS Ospedale Pediatrico Bambino Gesu', Roma, Italy, [3]Ospedale Pediatrico Bambino Gesù, Department of Hematology/Oncology, Cell and Gene Therapy, Roma, Italy

SESSION INFORMATION

Session Date: Thursday, March 30, 2023

Session Title: Posters: Clinical and Therapeutic I

Session Type: Poster Session A

Session Time: 6:00PM–7:00PM

Background/Purpose: Macrophage activation syndrome (MAS) is a life-threatening complication of different rheumatic diseases, particularly of systemic juvenile idiopathic arthritis (sJIA).

Methods: We report the case of 17-year-old girl with sJIA complicated by recurrent severe MAS episodes who received emapalumab (anti-IFNg antibody) in two subsequent MAS episodes and then underwent an uncomplicated hematopoietic stem cell transplantation (HSCT) while receiving emapalumab and anakinra granting complete control of inflammatory activity of the underlying disease.

Results: A 13-year-old White Caucasian girl presented with fever, rash and hepato-splenomegaly. Laboratory parameters were consistent with full-blown MAS (table 1). In the absence of clear evidence of an underlining condition, a diagnosis of secondary HLH was made, treatment with high dose of intravenous (IV) methylprednisolone (mPDN) and oral cyclosporine (CYC) was started with progressive improvement. After one year, still on CYC, she presented with fever, rash and arthritis with laboratory parameters consistent with MAS (table 1). Diagnosis of sJIA complicated by MAS was made. In 24 hours, she rapidly worsened and was admitted in ICU. High dose of IV mPDN (7 pulses of 30 mg/kg/day) as well as IV CYC (5 mg/kg/day) did not yield a

Table 1. Laboratory parameters and cytokine levels during disease course.

Laboratory parameters	Range	First MAS episode	Second MAS episode	Third MAS episode	HSCT	1 year after HSCT
GB (10^3 /uL)	5.5-15	10.87	8.87	3.41	1.87	8.54
PLT (10^3 /uL)	150-450	80	195	147	184	327
Ferritin (ng/ml)	13-150	13.088	27.396	5.921	230	26
ALT (UI/L)	<33	50	81	505	12	13
AST (UI/L)	<32	89	125	865	24	22
LDH (UI/L)	135-225	1717	1623	941	175	234
Fibrinogen (mg/dl)	190-430	400	500	193	341	428
Triglyceride (mg/dl)	<170	197	208	220	148	106
IL-18 (pg/ml)	<300	189764	27761	36606	995	341
CXCL9 (pg/ml)	<612	112895	22488	29403	50	50

response. Emapalumab was started, in the NI-0501-06 trial, (6 mg/kg initial dose followed by 3 mg/kg every 3 days) for 11 infusions. Conditions progressively improved. In order to prevent flares of the underlining sJIA, anakinra (2 mg/kg/day) was started. After 2 years in clinical remission, while she receiving anakinra every other day, she presented with fever, vomiting and diarrhea. Anakinra was immediately increased to daily dosing. Stool analysis showed Salmonella infection and antibiotic therapy was started. Nevertheless, she rapidly worsened, laboratory parameters were again consistent with full-blown MAS (table 1). She required ICU admission for multiorgan failure. Anakinra was administered IV and the dose increased up to 12 mg/kg/day. IV MPDs (8 pulses of 30 mg/kg/day) as well as IV CYC (5 mg/kg/day) were started with partial response. Based on her previous response to emapalumab, emapalumab was started again (compassionate use) with marked and rapid improvement. Because of recurrent MAS episodes and particularly for their rapidly evolution, the patient underwent an ex-vivo T cell-depleted haploidentical HSCT from her mother. The conditioning regimen was based on a Thiotepa-Treosulfan-Fludarabine scheme. Emapalumab was continued 1 month after HSCT together with anakinra. The patient achieved full donor engraftment with complete donor-derived immune reconstitution after 3 months. One year after HSCT, she is in excellent clinical condition on anakinra every other day, with complete remission of sJIA/MAS, also confirmed by persistently normal levels of IL-18 and CXCL9 (table 1).

Conclusion: This case provides further evidence of the efficacy of emapalumab in MAS, of the potential benefit of HSCT in difficult to treat sJIA patients. Notably, full control of inflammatory activity with emapalumab and anakinra may help to obtain a successful HSCT and reduce the risk of rejection.

Disclosure: C. Bracaglia: Sobi, 2, 6; **M. Pardeo**: SOBI, 2, 6; **G. Marucci**: None; **S. Riccio**: None; **F. Quagliarella**: None; **I. Caiello**: None; **G. Prencipe**: None; **P. Merli**: Sobi, 2; **F. Locatelli**: Sobi, 2; **F. De Benedetti**: AbbVie/Abbott, 2, Novartis, 2, Novimmune, 2, Pfizer, 2, Roche, 2, Sobi, 2.

Abstract Number: 076

COVID-Distress in Children with Systemic Lupus Erythematosus During the COVID-19 Pandemic

Brooke Rezmer[1], Michelle Adler[2], Tamar Rubinstein[3], Andrea Knight[4] and Natoshia Cunningham[5], [1]Michigan State University College of Human Medicine, Grand Rapids, MI, [2]Michigan State University, East Lansing, [3]Children's Hospital at Montefiore, New York, NY, [4]The Hospital for Sick Children, Toronto, ON, Canada, [5]Michigan State University, Grand Rapids, MI

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Psychological symptoms are common in childhood-onset systemic lupus erythematosus (cSLE) and may impact other psychological and health-related outcomes. Mental health problems such as anxiety have increased throughout the COVID-19 pandemic and may be correlated to pandemic-related distress. The current study examines COVID-related distress in youth with cSLE, the relationship between anxiety and COVID-distress, and whether COVID-distress is associated with other aspects of mental health and health-related functioning when controlling for patient-reported anxiety symptoms.

Methods: Fifty-nine participants between the ages of 12 and 22, diagnosed with cSLE per American College of Rheumatology Classification Criteria, were recruited from August 2020-November 2022 across seven pediatric rheumatology clinics in the United States and Canada as part of a larger psychological treatment study. Youth completed a baseline assessment and reported COVID-related distress on a visual analog scale with scores ranging from 0 indicating "no distress" to

100 indicating "extreme distress." Several mental health and health-related factors were also measured via validated questionnaires: anxiety (*Screen for Child Anxiety Related Disorders, [SCARED]*), depressive symptoms *(Childrens Depression Inventory 2, [CDI-2]* or *Beck Depression Inventory II, [BDI-II]*), disease activity (*SLE Disease Activity Index, [SLEDAI]*), pain (0-10 numeric rating scale), and fatigue (*Pediatric Fatigue Short Form* or *Adult Short Form, [PROMIS]*). Descriptive data and bivariate correlations were conducted. COVID-distress was compared based on socio-demographic factors including race, ethnicity, and income. Then, separate multiple regressions were conducted to examine the impact of COVID-related distress in predicting depression, fatigue, disease activity, and pain when accounting for patient-reported anxiety.

Results: The average age of participants was 16.34 years (SD = 1.99), 94.9% of which were female. Participants reported an average CDI of 63.54 (SD = 9.68), BDI of 62.19 (SD = 12.46), and SCARED of 33.64 (SD = 14.94) with a mean SLEDAI of 4.43 (SD = 5.15; median = 2.00). Participants reported moderate levels of COVID-related distress (Mean = 51.58, SD = 23.30), which was correlated with anxiety ($r = 0.35$, $p < 0.05$) and depressive symptoms ($r = 0.28$, $p < 0.05$), but not other study variables. COVID-distress rates were comparable between race, ethnicity, and income groups. When examining the simultaneous impact of both COVID-related distress and anxiety on outcomes via multiple regression analyses, the overall models were significant for depressive symptoms, $F(2, 36) = 4.84$, $p < 0.05$, and fatigue, $F(2, 36) = 5.26$, $p < 0.05$, but not for pain nor disease activity. Further, only anxiety (and not COVID-distress) significantly predicted depressive symptoms (beta = 0.45, $p < 0.01$) and fatigue (beta = 0.49, $p < 0.01$).

Conclusion: Findings suggests moderate levels of COVID-related distress in cSLE patients, which was correlated with other aspects of mental health functioning. Interestingly, anxiety and not COVID-distress was a more robust predictor of depression and fatigue in cSLE.

Disclosure: B. Rezmer: None; **M. Adler**: None; **T. Rubinstein**: None; **A. Knight**: None; **N. Cunningham**: None.

Abstract Number: 077

Clinical Characteristics of Chronic Recurrent Multifocal Osteomyelitis or Chronic Nonbacterial Osteomyelitis in Pediatrics; Single Center Study in Korea

Minji Kim[1], Jieun Jeong[1], Jiwon Jung[2], Seong Heon Kim[1] and **Seon Hee Lim**[3], [1]Department of Pediatrics, Seoul National University Children's Hospital, Seoul, Republic of Korea, [2]Asan Medical Center Children's Hospital, Seoul, Republic of Korea, [3]Department of Pediatrics, Pusan National University Children's Hospital, Yangsan, Republic of Korea

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Clinical and Therapeutic I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: CRMO is an autoinflammatory bone disease which has a recurrent bone pain as a symptom. It causes problems in various organs, including joints, skin, and intestines, and causes complications due to persistent pain, bone weakness, and deformation. However, there are difficulties in diagnosis and treatment because of the lack of information due to its low incidence. The purpose of this study is to identify the clinical features and prognosis of CRMO in children and adolescents.

Methods: We retrospectively analyzed the medical records of pediatric CRMO patients who were diagnosed in Seoul National University Children's Hospital between January 2004 to December 2022.

Results: Among 58 patients, male were 62.1% (n=36), the age at diagnosis was 11.13.4 years, and the follow-up duration was 34.540.1 months. The duration from the onset of symptoms to the visit to the hospital was 12.1 16.3 months, and the first symptom site was the knee (n = 34), followed by the ankle (n = 17) and hip (n = 15), with the involvement of average 1.8 1.1 bony sites. Accompanying symptoms were in order of trauma history (n=18), fever (n=16), and morning symptoms (n=7). At the time of the first diagnosis, blood tests showed an average ESR of 30.531.8 mm/h and CRP of 1.94.7 mg/dL. Patients were treated with NSAIDs, DMARDs, bisphosphonates, biologics, etc. The average duration of usage was 18.728.5 months for NSAIDs, 10.824.8 months for DMARDs, and 7.822.0 months for steroids. Bisphosphonate was used by 1 patient and biologics was used by 7 patients. At the final follow-up, 48.3% (n=28) had a clinical improvement, 31.0% (n=18) had improved MRI results, and 32.8% (n=19) had improved blood tests. Complications such as genu varum and leg length discrepancy were found in 12 patients, and 5 of them underwent surgery. Nine out of 28 people, who measured initial and final height, showed a decreased final height percentile compared to the initial height percentile.

Conclusion: CRMO is a rare inflammatory disease that occurs in children and adolescents. Therefore, it is difficult to understand its clinical features, diagnosis, treatment, and prognosis. The purpose of this study is to improve the understanding of the disease by identifying the clinical characteristics of Korean pediatric patients.

Disclosure: M. Kim: None; **J. Jeong**: None; **J. Jung**: None; **S. Kim**: None; **S. Lim**: None.

Abstract Number: 078

Periodic Fever Syndrome and Myelodysplastic Syndrome: Possible Connections Between Two Disorders

Ivanna Romankevych[1], Nicole Torres[2], Imani Sanders[1], Laurent King[1] and Pedro Garcia Rodriguez[1], [1]Jackson Health System, University of Miami Miller School of Medicine, Miami, FL, [2]University of Miami Miller School of Medicine, Miami, FL

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Genetics and Pathogenesis I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Background: associations between autoimmune, immunodeficiency and hematological disorders are well known as well as with autoinflammatory conditions. Modern genetic testing allows to link these conditions not only clinically but on genetic level also.

Methods: literature review for possible connections between autoinflammatory and autoimmune disorder with myelodysplastic syndrome associated with SRP72 gene polymorphism.

Results: L., 3-year-old boy with past medical history of periodic fever, myelodysplastic syndrome (MDS) associated with pathologic polymorphism of gene SRP72 (C1384C T), multiple café au lait spots and autism spectrum disorder was admitted to our hospital due to fever. Fever episodes occurred since 6-month-old (up to 40C), every 3 months and lasted 5-15 days. The episodes associated with rash, conjunctivitis, arthralgias, reactive lymphadenopathy, oral and duodenal ulcers, Raynaud syndrome and elevation of C-reactive protein level during fever episodes. The initially managed the fever

with Prednisone due to concerns for an autoimmune condition or PFAPA. The fever has improved to steroid in the early course of disorder but following episodes hasnt had any response. Extensive work up has been done. Autoimmune, immunology, oncology evaluation was negative, but genetic testing (microarray and Invitae primary immunodeficiency panel) was positive for pathologic polymorphism of gene SRP72. At 18-months of age, he was diagnosed with neutropenia first time. Family history was significant for periodic fever and lupus-like symptoms in his mother and rheumatoid arthritis in maternal uncle. Tests results were inconclusive for possible reason of his symptoms. Next comprehensive evaluation has been performed at National Institute of Health in June 2022. Full genome sequencing results are pending.

While definitive diagnosis is still not clear, we decided to look for possible links between MDS and his periodic fevers. In adults MDS associates with various autoimmune and autoinflammatory processes like vasculitis, inflammatory arthritis, autoimmune cytopenia. MDS associated with SRP72 polymorphism/mutation has familial involvement and usually early onset. The SRP72 is gene encoding one of the subunits of SRP (signal recognition particle) responsible for targeting of secretory protein to the endoplasmic reticulum (ER).

SRP72 cleaved during apoptosis possibly links to impaired tolerance to own peptides and self-autoantibodies formation that triggering the autoimmune diseases. Inappropriate protein production and degradation might be another non-immunologic mechanism triggering inflammation. Adequate function od ER and protein homeostasis are necessary for muscle tissue stability. There is association of SRP72 with idiopathic inflammatory myopathies (IIM). However, process of SRP72 synthesis and IIM in not fully known investigated. There are not know connections between MDS and periodic fever syndromes.

Conclusion: myelodysplastic syndrome associated with SRP72 gene polymorphism has strong association with IIM but not with autoinflammatory syndromes. Patient should have close monitoring for developing features of IIM

Disclosure: I. Romankevych: None; **N. Torres**: None; **I. Sanders**: None; **L. King**: None; **P. Garcia Rodriguez**: None.

Abstract Number: 079

Dynamics of Neutrophil Activation in Repeated TLR-9-Induced Mouse Model of Macrophage Activation Syndrome

Natsumi Inoue, Richard Chhaing, Sanjeev Dhakal, Thuy Do and Grant Schulert, Cincinnati Children's Hospital Medical Center, Cincinnati, OH

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Genetics and Pathogenesis I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Macrophage activation syndrome (MAS) is a life-threatening complication of rheumatic diseases including systemic juvenile idiopathic arthritis (SJIA). SJIA shows prominent neutrophil activation with expansion of proinflammatory neutrophils in the active phase and persistent activation even in clinically inactive disease, but how neutrophils contribute to the pathogenesis of SJIA and MAS remains unclear. Repeated toll like receptor (TLR)-9 induced MAS mouse model is the most characterized model of MAS; however, the dynamics of neutrophils in this system are largely unexplored. The objective of this study is to define neutrophil activation in the TLR9-induced mouse model in both acute MAS and MAS resolution.

Methods: Wild type C57BL/6 mice were injected with CpG intraperitoneally five times in 10 days and sacrificed one day after last injection (acute MAS) or after 21 days (resolution). Neutrophils were identified as Ly6G+CD11b+, and maturity was assessed using CD101 and CXCR2. For gene expression analysis, neutrophils from peripheral blood were isolated via magnetic negative selection. Total RNA extracted from neutrophils were processed using the Ampliseq Transcriptome system and Ion Torrent S5 next-generation sequencing system. Sequence data was analyzed and visualized using AltAnalyze.

Results: During acute MAS, pancytopenia, splenomegaly and elevated plasma IL-18 were observed, which largely normalized at resolution. Neutrophil counts were significantly decreased during acute MAS and still low at resolution. Immature neutrophils markedly increased during acute MAS only.

We performed gene expression profiling on purified neutrophils during acute MAS and resolution. Compared to control neutrophils, we found that during acute MAS, 167 genes were upregulated and 196 genes were down regulated, while in resolution phase 3 genes were upregulated and 412 genes were down regulated (rawp < 0.05, fold change >2.0). The most enriched upregulated gene ontology (GO) pathways at MAS included Defense response (Z score=14.83, adjp=2.18×10^{-18}) and Cellular Response to IFNγ(Z score=14.28, adjp=1.75×10^{-6}), including *CXCL9* and *CXCL10*, while there were no significantly enriched downregulated pathways.

During MAS resolution, there were no significantly upregulated GO pathways, while the most enriched downregulated GO included Protein binding (Z score=8.75, adjp=2.27×10^{-13}) and Immune System Process (Z score=7.26, adjp=8.39×10^{-7}), including *C1QB*, *CXCL2,IRF4* and *TLR9*. When we compared upregulated genes during experimental MAS to our previous study on human neutrophils from patients with active SJIA, which showed numerous upregulated inflammasome related genes, we found very little overlap.

Conclusion: TLR9-induced experimental MAS induced overall neutropenia but increased immature neutrophil production and IFNγ-driven neutrophil transcriptional activation, while resolution shows persistent neutropenia and transcriptional profiles of downregulated chemokine production. Together, this model demonstrates long-standing alternations in the neutrophil compartment that are markedly different than seen in children with SJIA.

Disclosure: N. Inoue: None; **R. Chhaing**: None; **S. Dhakal**: None; **T. Do**: None; **G. Schulert**: Novartis, 2, SOBI, 2.

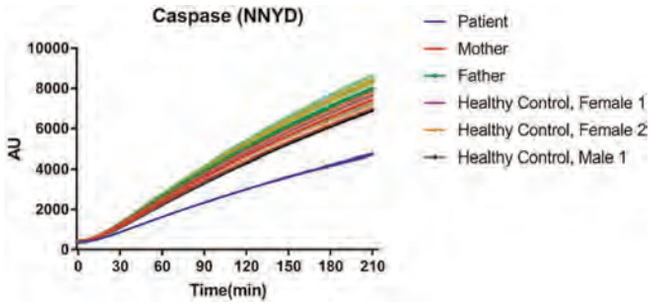
Abstract Number: 080

Analysis of Proteasomal Activity – a Potential Diagnostic Tool for Proteasome-associated Autoinflammatory Syndromes (PRAAS)

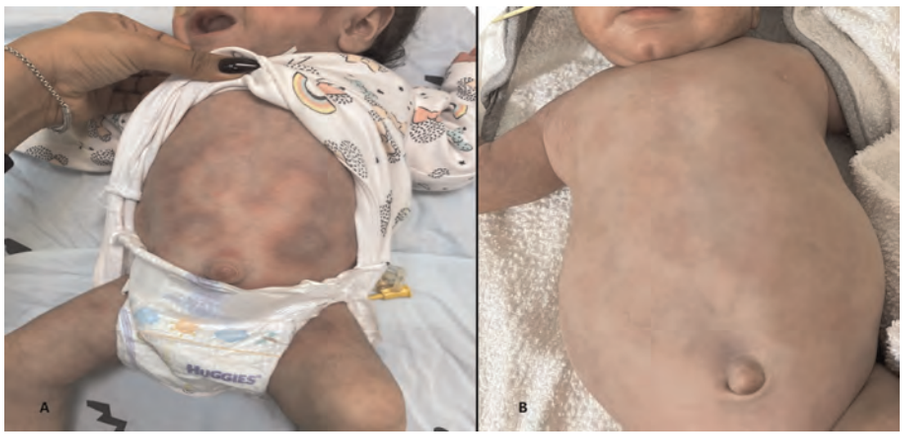
Yoel Levinsky[1], Oded Scheuerman[2], Rotem Tal[3], Gil Amarilyo[3] and Liora Harel[3], [1]Schneider Children's Medical Center of Israel, Tel Aviv University, Petach Tikva, Israel, [2]Pediatric B department, Schneider children's medical center of Israel, Petach Tikva, Israel, [3]Pediatric rheumatology clinic, Schneider children's medical center of Israel, Petach Tikva, Israel

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Genetics and Pathogenesis I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Interferonopathies are a recently recognized group of genetic syndromes associated with uncontrolled activation of interferon. PRAAS (proteasome-associated autoinflammatory syndromes) is an interferonopathy caused by proteasomal dysfunction. Clinical characteristics include early onset inflammation, nodular rashes, hepatosplenomegaly,



Reduced β1 proteasomal subunit activity as measured by caspase reaction, compared to healthy conrtols



The patient, before (A) and 2 weeks after (B) starting the treatment with JAK inhibitor (baricitinib)

myositis, panniculitis, lipodystrophy and basal ganglion calcifications. Our patient presented at age of 3 months with features strongly suggestive of PRAAS. Genetic testing revealed a novel proteasome variant classified as a variant of unknown significance (VUS). Functional testing of the proteasome was undertaken inorder to support the suspicion of PRAAS.

Methods: Samples of whole blood were taken from the patient, parents and 3 healthy controls. PBMC were lysed using activity preserving methods (active extraction). Samples were normalized using BCA enzymatic assay. Activity of the proteasomal subunits was measured using peptides specific to different subunits (caspase, chymotrypsin and trypsin acticity). In addition, interferon signature was obtained and whole exome sequencing was analyzed for the patient and her parents.

Results: Interferon signature was abnormally high, suggestive of an interferonopathy. A pilot analysis of proteasome activity showed a 50% reduction in the catalytic activity of proteasome subunit β1, indicating severe impairment of proteasomal activity, with compensatory hyper-activation of the immunoproteasome subunit β1i (figure 1). Whole exome sequencing revealed a novel heterozygous variant in a PSMB4, the gene that encodes for β7, a structural non –catalytic subunit of the proteasome classified as a variant of unknown significance (VUS). The apparent dysfunction of β1 and hyper activation of β1i suggested a proteasomal assembly dysfunction. Using the above findings, the patient commenced baricitinib treatment with remarkable clinical improvement (figure 2).

Conclusion: Proteasomal activity testing may serve as a useful tool for the diagnosis of PRASS in suspicious cases. Since JAK inhibitors may provide an adequate therapeutic response, it is of great importance to timely diagnose this disease in cases where the genetic results are equivocal.

Disclosure: Y. Levinsky: None; **O. Scheuerman**: None; **R. Tal**: None; **G. Amarilyo**: None; **L. Harel**: None.

Abstract Number: 081

Predicting Extension in Juvenile Idiopathic Arthritis

Megan Simonds[1], Kathleen Sullivan[2] and **AnneMarie Brescia**[1], [1]Nemours Children's Health, Wilmington, DE, [2]Children's Hospital of Philadelphia, Philadelphia, PA

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Genetics and Pathogenesis I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease of childhood and carries a risk of permanent joint damage and disability [1]. In adult rheumatoid arthritis (RA), early aggressive treatment can lead to remission. This may also be true for JIA in children; however, treatment paradigms in JIA remain reactive rather than proactive due to lack of reliable methods to predict extension to a polyarticular course. Our objective was to validate cell subpopulations using flow cytometry to predict which patients have a high likelihood of extending.

Methods: JIA FLS cell lines from oligoarticular (oligo), extended-to-be (ETB), and polyarticular (poly) types were cultured. Flow cytometry was performed by Raybiotech, Inc. scRNA-seq was performed by Genewiz according to 10x Genomics Chromium protocols. SeuratR package was used for QC, analysis, and exploration of data.

Results: Fibroblast-like synoviocytes are heterogeneous. Cells from scRNA-seq were annotated using SingleR data package. Subpopulation percentages were calculated from cell counts (Figure 1). Smooth muscle cell-like cells (SMC) decreased in poly (p 0.05 in oligo v poly). Fibroblast-like cell (FLS) percentage was higher in ETB than oligo (p 0.05). Chondrocyte-like cells (CH) percentage increased in poly compared to oligo and ETB (p 0.05). We performed flow cytometry on normal cell lines to determine if traditional markers for CH, SMC, and FLS can distinguish these cell types from one another. These markers did not confidently isolate these cell types (Figure 2). As a result, we analyzed our scRNA-seq data for markers that could distinguish JIA subtypes via flow cytometry (CH – SMOC2, SOX9; FLS – IGFBP4, VCAN; SMC – CD309). These markers could not distinguish FLS from CH using flow. We revisited gene data from scRNA-seq. Seurat single analysis identified the top genes of each projected cell type for each subtype. Genes that contribute the most variation among cells within

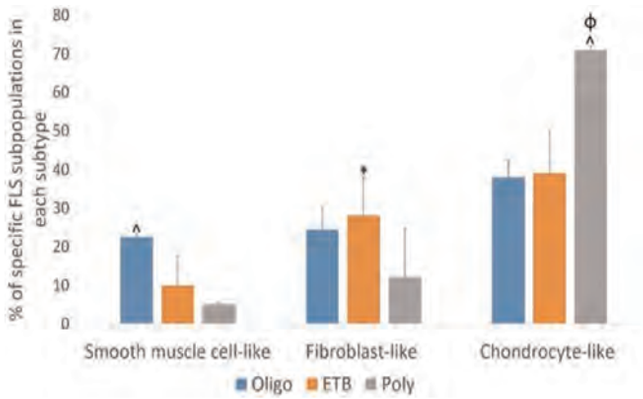


Figure 1. Percentage of FLS subpopulations in cultured FLS from patients with oligoarticular (Oligo), extended-to-be (ETB), and polyarticular (Poly) JIA. The percentage of smooth muscle-cell-like cells decreases in more severe subtypes of JIA; fibroblast-like cells significantly increased in ETB compared to Oligo; and chondrocyte-like cells increases in more severe subtypes of JIA. ^p<0.0006 Oligo vs. Poly; *p=0.03 Oligo vs. ETB; p=0.0044 ETB vs. Poly.

Table 1. Traditional markers from literature to distinguish cell types. Expected outcomes based on literature (A). Actual outcomes based on flow cytometry (B).

A.

	FLS	CH	SMC
CD55	+++	++	++
CD271	Negative	Negative	Negative
CD309	Negative	Negative	+++
CD146	Negative	+	++
CD45	Negative	Negative	Negative
CD166	+++	+++	+++
CD44	+++	+	+++
CD105	+++	+	++
CD31	Negative	Negative	+++
CD90	+++	+++	+

B.

	FLS	CH	SMC
CD55	+	+	++
CD271	Negative	Negative	Negative
CD309	+	Negative	+
CD146	+	+	+
CD45	Negative	Negative	Negative
CD166	+++	+++	+++
CD44	+++	+++	+++
CD105	++	++	+++
CD31	Negative	Negative	Negative
CD90	+++	+++	++

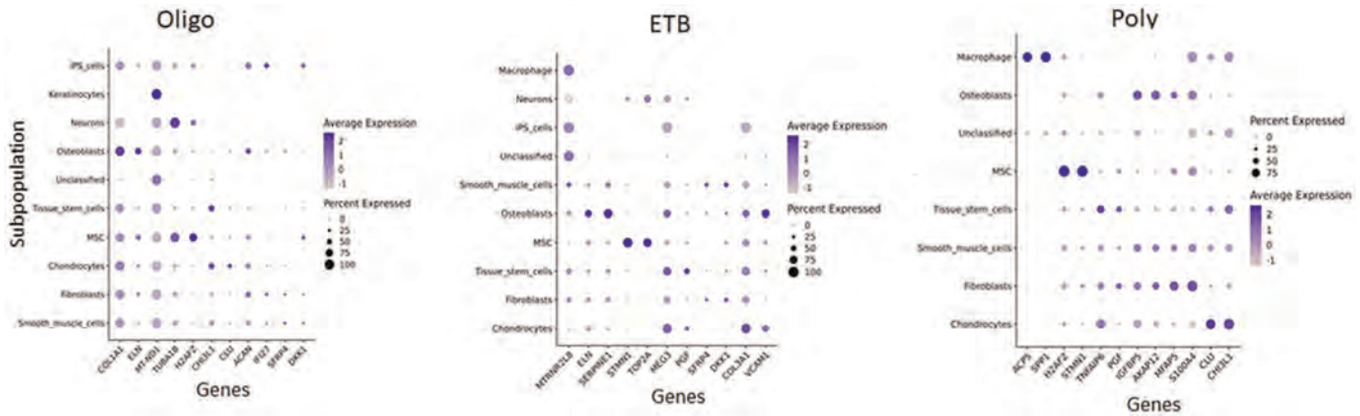


Figure 3. Genes that are the highest contributors to variation among subpopulations. Genes that contribute the most variation among cell subpopulations are as follows: oligo - COL1A1, MT-ND1, TUBA1B, ACAN, and IFI27, ETB - MTRNR2LB, SERPINE1, TOP2A, MEG3, COL3A1, and VCAM, poly - ACP5, SPP1, TNFAIP6, IGFBP5, AKAP12, MFAP5, and S100A4

a JIA subtype are as follows: oligo - COL1A1, MT-ND1, TUBA1B, ACAN, and IFI27, ETB - MTRNR2LB, SERPINE1, TOP2A, MEG3, COL3A1, and VCAM, poly - ACP5, SPP1, TNFAIP6, IGFBP5, AKAP12, MFAP5, and S100A4 (Figure 3).

Conclusion: While flow cytometry has proven unable to distinguish between cell subpopulations within FLS cultures, these cell subpopulations have unique genetic fingerprints with transcript expression levels that can be used to distinguish JIA subtypes.

1. Manners, P.J. and C. Bower, *Worldwide prevalence of juvenile arthritis why does it vary so much?* The Journal of rheumatology, 2002. **29**(7): p. 1520-1530.

The authors wish to acknowledge CARRA and the ongoing Arthritis Foundation financial support of CARRA.

Disclosure: M. Simonds: None; **K. Sullivan**: None; **A. Brescia**: None.

Abstract Number: 082

Globalization and Real-World Implementation of an International Pediatric Rheumatology Learning Resource

Mercedes Chan[1], Tamara Tanner[2], Mutibah Al-Essi[3], Deepthi Abraham[4] and Daire O'Leary[5], [1]BC Children's Hospital, Vancouver, BC, Canada, [2]Children's Hospital at Montefiore, New York, NY, [3]King Fahad Hospital of the University, [4]Stellenbosch University, Cape Town, South Africa, [5]UCD Centre for Arthritis Research, Dublin, Ireland

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Quality, Health Services, and Education I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: The Pediatric Rheumatology Learning Modules (PRML) were developed in 2015 as an academic curriculum for pediatric rheumatology fellowship training at a single center. With new discoveries and demand for use in different global contexts, we formed a virtual working group (WG) to update and modify content. We describe our process in adapting a learning resource, and our early experiences and feedback from piloting our work.

Methods: The WG (n=5) includes patients, trainees, and faculty from 5 countries and 4 continents. Literature reviews; the Textbook of Pediatric Rheumatology, 8th Edition; and ongoing user feedback on clarity and relevance, guide included content. Global adaptation draws on frameworks for internationalizing medical school curricula: plain language use, removal of cultural identifiers, generic drug names, practice variability due to resource inequity, e.g., biologics. We created faculty guides by request to address challenging questions and to include prompts for discussion. Strategies were discussed and refined after a pilot review of 3/37 modules including initial review, user feedback, learning objectives, references, literature review, and quizzes. Proposed changes are independently reviewed by group members; if universally agreed upon, they are accepted prior to a bi-weekly meeting where discrepancies are discussed. Final accepted changes require 75% consensus. 10-item multiple choice quizzes assess baseline knowledge and application of clinical knowledge. All work is facilitated online (virtual meetings, cloud-based file sharing, real-time group file editing). The updated PRLM are piloted as available at 2 pediatric rheumatology (PR) centers. Quiz results assess knowledge change and feedback from trainees and faculty are collated (email, comments on electronic documents) are used to refine the PRLM.

Results: The WG met 26 times over 18 months with 31 of the original 37 modules and quizzes revised. Two new modules were developed in response to gaps identified (transition, therapeutics). At the pilot centers, modules are reviewed weekly. Trainees (PGY4-6) complete quizzes then attempt module questions. Module answers are prepared for discussion with faculty, with the same module quiz repeated afterwards. Quiz answers are then discussed. Mean baseline quiz scores (n=240) from 13 trainees show pre- and post-modules scores of 71.6% and 91.6%, and 63.2% and 80.7% at centers 1 and 2. PGY-4 trainees gain more knowledge from training than PGY-6 trainees (23% vs 9%). Feedback from trainees and faculty includes ensuring quiz and module content align with learning objectives and ways to improve question clarity. Trainees prefer group discussion with PRLM to didactic teaching and feel modules help prepare them for board examinations.

Conclusion: The PRLM is the first evidence-based educational resource developed for PR trainees for teaching and learning in a group setting designed for any global context. Developing the PRLM requires time, learning, knowledge-sharing, and stable internet connections. Real-world user feedback was limited to 2 centers. More widespread use and assessment of PRLM as a tool are needed to make meaningful conclusions for future application.

Disclosure: M. Chan: None; **T. Tanner**: None; **M. Al-Essi**: None; **D. Abraham**: None; **D. O'Leary**: None.

Abstract Number: 083

Running out of Rheum: Where Are the Pediatric Rheumatology Faculty?

McKenzie Vater[1], Miriah Gillispie-Taylor[2], Emma Austenfeld[3] and Julia Shalen[4], [1]Vanderbilt University Medical Center, Nashville, TN, [2]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [3]MCWAH, Wauwatosa, WI, [4]Johns Hopkins University, Baltimore, MD

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Quality, Health Services, and Education I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: The pediatric rheumatology workforce shortage has significant downstream effects on pediatric trainees and patients. Currently, 9 out of 50 states do not have a board-certified pediatric rheumatologist. Earlier identification and treatment of rheumatic disease leads to improved outcomes. Without recognition of these signs by general providers, patients are not referred in a timely manner, but without proper education, we cannot expect them to understand the protean presentation of rheumatic disease. A 2004 study showed that 40% of US pediatric residency programs lacked on-site pediatric rheumatologist, while one-third of medical schools had the same deficiency. The purpose of this project was to identify whether increased attention to workforce shortage issues impacted the number of US and Puerto Rico pediatric residency training programs with associated pediatric rheumatology faculty from 2004 to present.

Methods: One research team member (MV) identified all pediatric residency programs accredited for the 2022-2023 academic year through the Accreditation Council for Graduate Medical Education (ACGME) website. MV then reviewed all websites to identify those that had affiliated pediatric rheumatology faculty. In cases that were unclear, MV emailed programs with the address provided on the ACGME report. Another team member (JS) discussed programs that were unable to be categorized based on their website or via email at a meeting of the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Medical Education Workgroup. Members of the working group, all practicing pediatric rheumatologists or rheumatology fellows, were queried and able to categorize most remaining programs.

Results: 212 pediatric residency programs are accredited for the 2022-2023 academic year by the ACGME in the US and Puerto Rico. 181 programs indicated presence/absence of pediatric rheumatology faculty on their website. Of the remaining 31 programs, 18 did not respond to email inquiries. Of the 212 total accredited pediatric residency programs, 132 (62.3%) have pediatric rheumatology faculty, 77 (36.3%) do not have pediatric rheumatology faculty and 1.4% remain unknown.

Conclusion: Over one-third of all pediatric residency programs do not have a pediatric rheumatologist on faculty, unchanged since 2004. This creates a significant barrier to providing appropriate education in rheumatology for trainees and limits exposure to patients with rheumatologic conditions. Lack of robust rheumatology knowledge impacts patient care and limited exposure to the field likely contributes to declining fellow match rates—most recently 62.8% positions filled for 2022, down from 69.2% in 2021. Next steps include establishing areas of essential knowledge in pediatric rheumatology for pediatric residents with the goal of creating precise learning objectives so programs can focus their educational efforts and better prepare pediatric residents for identifying and managing pediatric rheumatologic issues after graduation. Additionally, efforts should continue to understand resident motivation to pursue fellowship in pediatric rheumatology with the goal of improving recruitment given the known workforce shortage.

Disclosure: M. Vater: None; **M. Gillispie-Taylor**: Pfizer, 5; **E. Austenfeld**: None; **J. Shalen**: None.

Abstract Number: 084

Rheum to Improve: Patient-reported Transition Readiness in a Large Pediatric Rheumatology Clinic

Kristiana Nasto[1], David McDonald[1], Kyla Fergason[1], Mary Robichaux[1], Bernard Danna[1], Monique Maher[1], Alexander Alexander[1], Danielle Guffey[1], Miriah Gillispie-Taylor[2] and Tiphanie Vogel[1], [1]Baylor College of Medicine, Houston, TX, [2]Baylor College of Medicine, Texas Children's Hospital, Houston, TX

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Quality, Health Services, and Education I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Transition of adolescents with chronic healthcare needs to adult care may result in poor outcomes. We have developed a program to improve the transition of pediatric rheumatology patients to adult rheumatology providers, which includes periodic assessment of patient self-reported transition readiness using the validated Adolescent Assessment of Preparation for Transition (ADAPT) survey. Our long-term goal is to identify factors that can predict successful transition; here, we report initial findings following electronic medical record (EMR) automation of ADAPT delivery.

Methods: Return patients 14 years and older were surveyed, irrespective of diagnosis, from July 2021-November 2022. ADAPT survey distribution was automated for all clinic visits using the EMR. Patients/caregivers could respond during electronic clinic check-in, which is available for both in-person and telehealth visits. ADAPT responses were automatically collated using EMR data extraction on the first of each month. Three composite scores, out of 100, for (1) self-management, (2) prescription management, and (3) transfer planning were manually calculated from the ADAPT responses and compared across demographics. Mann-Whitney, Wilcoxon and Kruskal-Wallis tests were used to compare composite scores.

Results: 462 unique patients returned 670 surveys. 87% of returned surveys (586/670) were scorable for at least 1 composite score, and 401 patients (87%) returned a survey with at least 1 calculatable composite score. Most survey respondents were female (75%), aged 14-17 years (83%), Caucasian (69%), non-Hispanic (64%), and spoke English (90%). Overall mean scores for self-management, prescription management, and transfer planning on initial survey were 35 (n=401), 59 (n=288), and 16.6 (n=367), respectively. Scores for self-management (mean 20.4 at age 14 years, increasing to 63.6 at 18+ years) and transfer planning (mean 1 at age 14 years, increasing to 49 at 18+ years) improved across age (both p 0.0001), but scores for prescription management were high for all patients (mean 59 at age 14, 66 at 18+ years, p=0.0442). Scores did not differ by sex or race; Hispanic patients scored higher in self-management (44.5 vs. 30.7, p 0.0001). 97 patients (21%) completed 2 surveys, 39 patients (8.4%) completed 3 or more. The average time between surveys 1 and 2 was 4 months (range 1 day-16 months). There was no difference in this timeframe in self-management (33 vs. 35.3, p=0.444, n=87) or prescription management (60 vs. 58.6, p=0.908, n=54), but there was an increase in scores for transfer planning (13.6 vs. 21, p=0.008, n=90).

Conclusion: In our rheumatology transition program, patient self-reported transition readiness is assessed using the ADAPT survey. Analysis thus far indicates participation in the transition pathway can rapidly improve transfer planning scores; however, opportunities remain to improve readiness in all domains. We look forward to distributing surveys in Spanish, a current EMR limitation. In the future, we will assess which aspects of readiness correlate with successful transfer to adult care.

Disclosure: K. Nasto: None; **D. McDonald**: None; **K. Fergason**: None; **M. Robichaux**: None; **B. Danna**: None; **M. Maher**: None; **A. Alexander**: None; **D. Guffey**: None; **M. Gillispie-Taylor**: Pfizer, 5; **T. Vogel**: Moderna, 2, Novartis, 2, Pfizer, 2, sobi, 2.

Abstract Number: 085

Identification of Barriers to Care Experienced by Children with Rheumatic Disease: A Qualitative Study

Olivia Kwan[1], Gloria Garcia[2], Kiana Johnson[3], Melissa Oliver[1], Stacey Tarvin[4], Alvaro Tori[1], Brandi Stevens[1] and Martha Rodriguez[4], [1]Indiana University, Indianapolis, IN, [2]Indiana University School of Medicine, Indianapolis, IN, [3]East Tennessee State University, Johnson City, TN, [4]Riley Hospital for Children at Indiana University Health, Indianapolis, IN

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Quality, Health Services, and Education I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Pediatric rheumatic diseases are known to have disparities in disease outcomes, but many drivers of these inequities are unknown. Social determinants of health are a source of health inequities and can present as barriers to care. To better understand impacts of social determinants of health we aim to identify barriers to care children with rheumatic disease face from a patient and family perspective.

Methods: Children and young adults (YA) aged 11-22y diagnosed with juvenile systemic lupus erythematosus and juvenile idiopathic arthritis and their caregivers were recruited from Riley Childrens Hospital outpatient pediatric rheumatology clinic during clinic visits or via phone call and email over a period of approximately four months. Semi-guided interviews were conducted via Zoom and consisted mostly of small focus groups of similarly aged children/YA and groups of caregivers. Interviews were conducted in English and Spanish. Interviews were recorded, transcribed, and thematic analysis conducted utilizing NVIVO software.

Results: 19 children/YA and 16 parents participated in group interviews (Table 1). Common barriers to care are described in Table 2. The most common barriers to care identified by caregivers were medication issues, financial, access to pediatric rheumatology care, access for other healthcare needs, and family needs including sibling and parent health. A surprising common theme was caregivers negative perception of their own self efficacy in healthcare and in both their own and the publics health literacy surrounding childhood rheumatic disease. The most common barriers identified by children/YA were medication issues, financial, and access to pediatric rheumatology care. Children and YA focused more on quality of life (Table 3) including pain and fatigue, school attendance and accommodations, and concerns about future independence. The COVID 19 pandemic did not present a large barrier to healthcare for most patients. Many families identified protective factors such as early introduction of the patient being an active participant in their own care. There was a strong desire for more family and peer support groups from both parents and children/YA.

Table 1: Patient Demographics

Race and Ethnicity	
	7 Hispanic
	6 White
	3 African American
	1 Asian
	1 Native American
	1 Unknown
Diagnosis	
	12 SLE
	7 Polyarticular JIA
Sex	
	16 Female
	3 Male

Table 2: Common barriers and examples

Barrier	Examples
Medication Issues	Efficacy, adherence, side effects, injection discomfort, and supply issues
Financial	Inadequate insurance, cost of medications, cost of visits
Rheumatology access	Distance, transportation costs, wait times
Access to other healthcare needs	Primary care, multiple specialists, emergency care
Family needs	Childcare, other children's healthcare, parent healthcare
Self-efficacy	Empowerment, perceived lack of health literacy
Time to reach a diagnosis	
Dietary Adherence	Following a low salt diet, self-prescribed diet

Table 3: Common Quality of Life Issues

Concerns	Examples
Physical Manifestations	Pain, fatigue, morning stiffness
School issues	Attendance, accommodating needs
Future Independence	Higher education, job/career attainment
Social life	Peer relations, involvement in extra-curriculars, hobbies
Mental health	Depression, anxiety

Conclusion: Barriers to care are common for children with rheumatic disease and common themes are identified by both parents and children/YA. An overarching theme is that families prioritize their childrens health to overcome barriers. Peer and parent support groups are of great interest to many patients and families and are a potential solution to perceived poor self-efficacy and health literacy. More studies are needed to understand the relationship between the experience of barriers to care to disease activity, health related quality of life and healthcare satisfaction. Future steps include distribution of surveys utilizing the information discovered from the interviews to a larger cohort of patients/families. Survey data will be examined to explore differences in experiences of barriers to care by race/ethnicity to better assess relationships to health inequities. Patient/family perspectives will also be compared to the perspectives of providers.

Disclosure: O. Kwan: None; **G. Garcia**: None; **K. Johnson**: None; **M. Oliver**: None; **S. Tarvin**: None; **A. Tori**: None; **B. Stevens**: Sermo, 2; **M. Rodriguez**: None.

Abstract Number: 086

Systemic Sclerosis Overlap Syndrome: A Case Series from a Single Large Pediatric Center

Jessica Nguyen[1], Miriah Gillispie-Taylor[1], Eyal Muscal[1] and Marietta Deguzman[2], [1]Baylor College of Medicine, Houston, TX, [2]Baylor College of Medicine, Texas Children's Hospital, Houston, TX

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Quality, Health Services, and Education I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: Childhood-onset systemic sclerosis (SSc) is a rare but potentially life-threatening autoimmune condition with features including immune, fibrotic, and vascular manifestations affecting the skin and other internal organs, such as the cardiac, pulmonary, and gastrointestinal systems. Compared to adults with SSc, a greater proportion of pediatric SSc patients have

overlap syndrome. Information about pediatric SSc overlap syndrome is lacking given the overall rarity of this condition and there is a notable gap in knowledge. We report on 4 patients diagnosed with SSc overlap syndrome from a single large pediatric center.

Methods: This project was conducted under IRB approval from Baylor College of Medicine. A retrospective chart review of patients who fulfilled the 2007 PRES/ACR/EULAR juvenile SSc provisional classification criteria between 2000 to 2022 was conducted. Patients with SSc overlap syndrome who also fulfilled the 2017 EULAR/ACR classification criteria for juvenile dermatomyositis (JDM) or 2012 SLICC criteria for lupus were identified. Information, including demographics, clinical features, diagnostic findings, and outcomes were collected.

Results: During the inclusion period, a total of 21 patients were diagnosed with SSc and 4 (19%) were classified as overlap syndrome. All had features of JDM with one patient having additional lupus features. All patients were female with a mean age at diagnosis of 12.5 years (range 10 – 14 years). The average time between symptom onset and time of diagnosis was 9.5 months (5 – 16 months). JDM features included cutaneous Gottrons rash and myositis based on clinical exam, labs, and/or imaging, and lupus features included hematologic manifestations and serositis. All patients had positive antinuclear antibodies (ANA), while 3 of 4 had a positive U3-RNP antibody. Two patients had hypocomplementemia. Renal disease was not seen in the patient with lupus features, and no patient developed scleroderma renal crisis. All patients received systemic corticosteroids and additional immunomodulatory therapies including rituximab, IVIG, and mycophenolate mofetil. Improvement of the SSc cutaneous features and resolution of the JDM skin changes were noted in all patients. All patients had either recurrences or progression of gastrointestinal and cardiopulmonary manifestations throughout their course despite treatment. Adverse events included prolonged hospitalizations, infections, and death (Table 1).

Conclusion: SSc overlap syndrome is a rare, difficult disease associated with significant morbidity and even mortality in pediatric patients. Cutaneous manifestations improved with multimodal pharmacotherapy while cardiopulmonary and gastrointestinal involvement continued to be a source of morbidity despite treatment. Renal manifestations were not seen throughout the clinical course. Treatment with systemic corticosteroids was well tolerated and choice of immunomodulatory agents was tailored to both the scleroderma and overlapping inflammatory disorder. Further studies need to be done to determine if there are differences in clinical course and prognosis between juvenile SSc and SSc overlap syndrome.

Demographics	Duration of Symptoms Prior to Diagnosis	Overlap Syndrome	Clinical Features						Immunologic Features	Treatment		Course
			Cutaneous	Vascular	Cardiopulmonary	Gastrointestinal	Renal	Hematologic		Corticosteroids	Immunomodulatory	
14 yo Caucasian F	5 months	JDM	Skin tightness, Gottron's over elbows and knees	Telangiectasia, Raynaud's	Restrictive pattern on PFTs	Progressive weight loss due to malabsorption	None	None	ANA, PM/Scl-100	Yes	Rituximab, IVIG, methotrexate, hydroxychloroquine	Continued to have persistent weight loss requiring G-tube - improved, resolved cutaneous features; Adverse Events: PJP pneumonia
13 yo Asian F	9 months	JDM	Skin tightness, edema, heliotrope rash, Gottron's over MCPs, PIPs, DIPs, darkening of neck and shoulders	Telangiectasia, Raynaud's with digital ulceration, nailfold capillary changes	Severe pulmonary hypertension, mildly obstructive disease with severely decreased DLCO on PFTs	Weight loss, SMA syndrome (asymptomatic), GE reflux	None	Thrombocytopenia	ANA, U3 RNP, normal C3 and C4	Yes	Mycophenolate mofetil, tocilizumab, IVIG, hydroxychloroquine	Continues to have persistent weight loss, pulmonary hypertension well controlled, resolved cutaneous features
10 yo African American F	16 months	JDM	Skin tightness, edema, Gottron's over MCPs and PIPs	Telangiectasia, Raynaud's with digital ulceration, nailfold capillary changes	Severe pulmonary hypertension, atrial tachycardia and PVCs	Small intestinal bacterial overgrowth, esophageal dysmotility/hypomotility	None	Pancytopenia, occlusive DVT	ANA, U3 RNP, low C3 and C4, normal CH50	Yes	Rituximab, mycophenolate mofetil, tocilizumab, IVIG, hydroxychloroquine	Pulmonary hypertension well controlled, resolved cutaneous features, appropriate weight trend; Adverse Events: prolonged hospitalization, cardiac arrest x 2, recurrent UTIs
13 yo Hispanic F	8 months	JDM, SLE	Skin tightness, edema, Gottron's over PIPs and DIPs, skin ulcerations of elbows and knees	None	Interstitial lung disease with bronchiectasis, pericardial effusion	Weight loss, dilated and thickened esophagus	AKI	TMA	ANA, U3 RNP, Scl-70, SM, RNP, SSA Ro52, SSA Ro60, SSB, low C3, C4, and CH50, APL, MDA5	Yes	Rituximab, mycophenolate mofetil, tocilizumab, IVIG, hydroxychloroquine, eculizumab	Multiorgan failure specifically pulmonary hemorrhage and pneumothoraces after intubation leading to prolonged hypoxemia; Adverse Events: CLABSI, Deceased

Disclosure: J. Nguyen: None; **M. Gillispie-Taylor**: Pfizer, 5; **E. Muscal**: sobi, 1; **M. Deguzman**: None.

Abstract Number: 087

Delays in Care, Declines in Health, and Food Insecurity in Pediatric Systemic Lupus Erythematosus Patients During the COVID-19 Pandemic

Rebecca Hetrick[1], Maria Pereira[2] and Marietta De Guzman[3], [1]Indiana University School of Medicine, Indianapolis, IN, [2]Baylor College of Medicine, Houston, TX, [3]Baylor College of Medicine, Texas Children's Hospital, Houston, TX

SESSION INFORMATION
Session Date: Thursday, March 30, 2023
Session Title: Posters: Quality, Health Services, and Education I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Background/Purpose: The COVID-19 pandemic created dramatic societal disruptions. Social distancing and measures to reduce disease spread rapidly reshaped healthcare delivery. Recognizing the burden of frequent monitoring and multiple medications in children with Systemic Lupus Erythematosus (SLE), our study sought to describe the effect of pandemic-related disruptions on the health and wellbeing of these families.

Respondent Demographics (Total N=40)		
Language	English	31 (78%)
	Spanish	9 (22%)
Race/Ethnicity	Mixed race/ethnicity	2 (5%)
	Asian	9 (23%)
	Black, not Hispanic	6 (15%)
	Hispanic	19 (48%)
	White, not Hispanic	3 (7%)
	Prefer Not to Answer	1 (2%)
Income	<30K	12 (30%)
	30K - 49K	10 (25%)
	50K - 74K	8 (20%)
	>75K	4 (10%)
	I don't know	6 (15%)
Education	Some high school or less	6 (15%)
	High school graduate or GED	9 (23%)
	Technical school or some colle	12 (30%)
	College graduate	7 (17%)
	Master's level or higher	6 (15%)
Marital Status	Married or Cohabitating	25 (63%)
	Single or Separated/Divorced	15 (37%)
Number of families reporting delays in care		
Missed or delayed medical appointments		15 (37%)
Problems seeing a doctor		10 (25%)
Missed or delayed procedure/surgery		4 (10%)
Missed or delayed lab draws		3 (7%)
Loss of health insurance coverage		2 (5%)
Missed or delayed immunizations		1(2%)
Telehealth Utilization		
Used telehealth pre-pandemic		8 (20%)
Used telehealth post-pandemic		32 (80%)

Respondent Demographics and Healthcare Utilization

Methods: The study consisted of a cross-sectional survey from a convenience sample of caregivers of pediatric systemic lupus erythematosus (SLE) patients at Texas Childrens Hospital from April 2021 to March 2022. Surveys were administered through REDCap in English and Spanish. Participants were recruited during clinic visits and via email from a roster of158 active pediatric SLE patients maintained by the Rheumatology Division.

Results: 40 respondents completed the survey. Sample demographics are shown inTable 1.

Barriers to Care (Table 1)

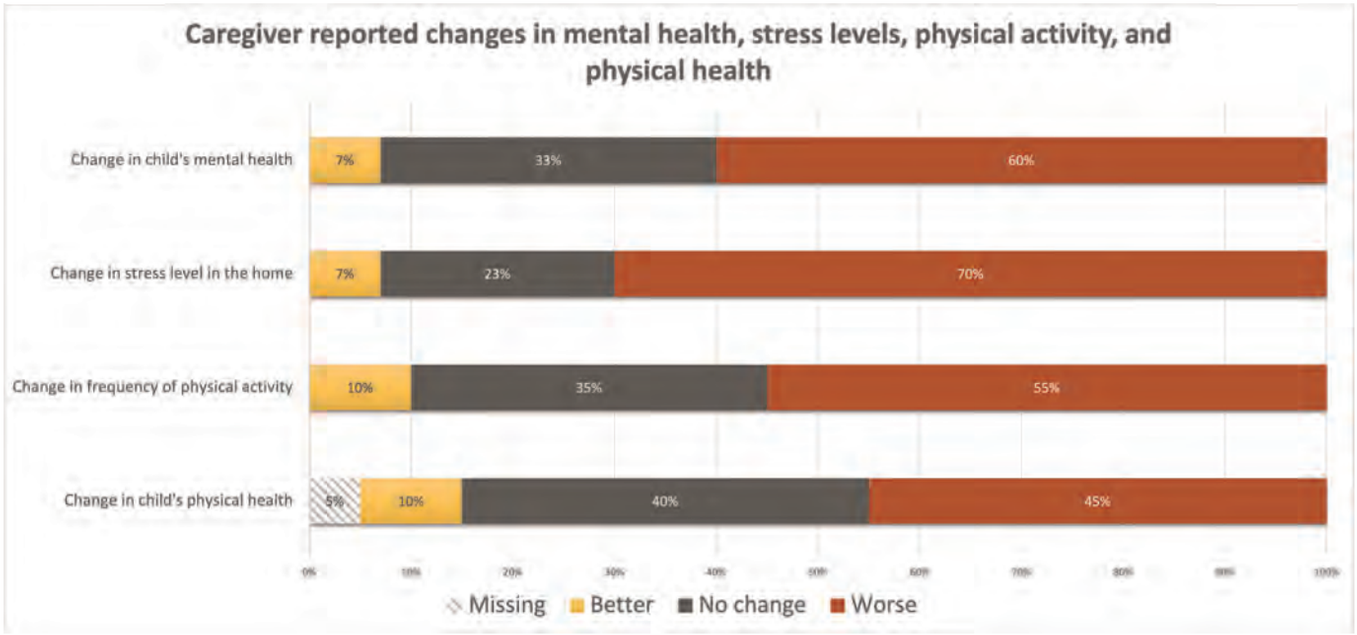
Despite increased telehealth use, many families reported missed or delayed care for their child: 15 (37%) reported missed or delayed medical appointments, 10 (25%) reported problems seeing a doctor, 4 (10%) reported missed or delayed procedures or surgeries, 3 (7%) reported missed or delayed lab draws, 2 (5%) reported loss of health insurance coverage, and 1 (2%) missed or delayed immunizations.

Changes in Mental and Physical Health (Fig. 1)

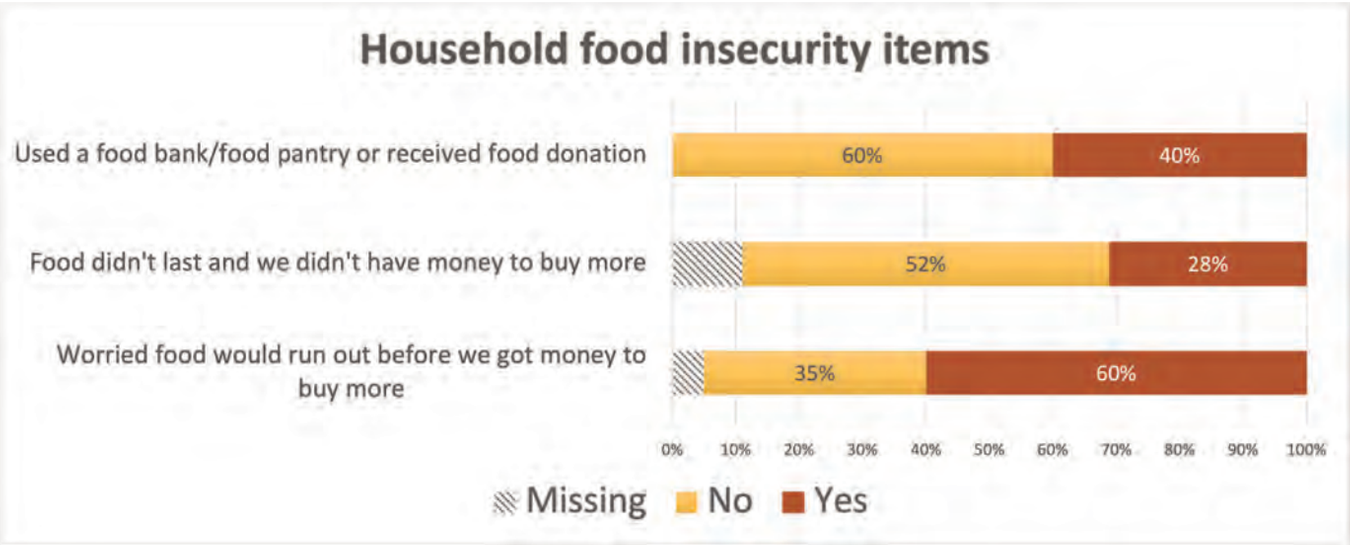
Respondents reported declines in child mental and physical health. 57% reported a decline in their childs mental health, and 79% reported higher levels of stress in the home during the pandemic. Similarly, 46% reported a decline in their childs physical health, and 57% reported that their child exercised less often during the pandemic.

Food Insecurity (Fig. 2)

Our sample demonstrated a high prevalence of food insecurity: 64% endorsed concerns that food would run out before getting money to buy more, 32% endorsed that food had run out without money to purchase more, and 39% reported using a food pantry or receiving a food donation.



Changes in Physical and Mental Health



Household Food Insecurity

Conclusion: Our survey captured important experiences of families with pediatric SLE patients during the COVID-19 pandemic. Compared to national data, our sample may have demonstrated greater declines in child physical and mental health and higher rates of food insecurity.[1,2] Our sample demonstrated a dramatic increase in the use of telehealth. Many delays in care were identified, most notably missed or delayed medical appointments in over one-third.

Given the cross-sectional design and convenience sampling, this data may not be representative of pediatric SLE patients nationally. Limited conclusions can be drawn from comparison to national data given differing data collection methods. However, in this vulnerable population, pandemic-related limitations in care are concerning and warrant further study.

Works Cited: Lebrun-Harris LA, Ghandour RM, Kogan MD, Warren MD. Five-Year Trends in US Childrens Health and Well-being, 2016-2020. *JAMA Pediatr*. 2022;176(7):e220056.Coleman-Jensen A, Rabbitt MP, Gregory CA, Singh A. *Statistical Supplement to Household Food Security in the United States in 2021*. Washington, DC: U.S. Department of Agriculture, Economic Research Service; 2022. AP-105.

Disclosure: R. Hetrick: None; **M. Pereira**: None; **M. De Guzman**: None.

Abstract Number: 088

Quality Improvement Cycles to Actualize Distribution of the Adolescent Assessment of Preparation for Transition (ADAPT) Survey

David McDonald[1], Kristiana Nasto[1], Kyla Fergason[1], Mary Robichaux[1], Bernard Danna[1], Monique Maher[1], Alexander Alexander[1], Ariel Coleman[2], Anne Dykes[2], JaLeen Rogers[2], Miriah Gillispie-Taylor[1] and Tiphanie Vogel[1], [1]Baylor College of Medicine, Houston, TX, [2]Texas Children's Hospital, Houston, TX

SESSION INFORMATION

Session Date: Thursday, March 30, 2023
Session Title: Posters: Quality, Health Services, and Education I
Session Type: Poster Session A
Session Time: 6:00PM–7:00PM

Table 1: ADAPT survey distribution and return per PDSA cycle

Cycle Number	Cycle Length (months)	Method of Distribution	Personnel Requirement to Distribute/Collect	# Surveys Distributed	# Surveys Completed	Return Rate
1	3.5	Email	Moderate	36	452	8%
2	2.5	Paper/Email	Heavy	99	329	30%
3	9	Email	Moderate	169	1365	12.4%
4	9	EMR	Low	na	61	na
5	8	EMR	None (Automatic)	597	608	98%

EMR: electronic medical record, na: not available

Background/Purpose: The challenges of transitioning patients with chronic rheumatic diseases from pediatric to adult care can lead to increased morbidity and mortality following transfer. Therefore, approaches to assess transition readiness are urgently needed, and should ideally be practical and sustainable. The Adolescent Assessment of Preparation for Transition (ADAPT) is a validated survey of patient self-reported transition readiness. Our overall goal was to implement a low-labor strategy by which 90% of transition-eligible patients complete an ADAPT survey at each visit.

Methods: Transition-eligible patients are age 14 years or older and attending a return clinic appointment. We conducted Plan-Do-Study-Act (PDSA) cycles using different methods of ADAPT survey distribution to eligible patients. Numbers of surveys distributed and rates of surveys returned during each PDSA cycle were calculated and compared.

Results: Our initial PDSA cycle distributed ADAPT surveys through a Research Electronic Data Capture (REDCap) database using email, requiring moderate weekly personnel effort yet achieving only 8% survey return rate (Table 1). In cycle 2, in-person coordinators increased the return rate to 30%. In March 2020, pandemic-related clinic closures and transition to telehealth necessitated a return to survey distribution by email (cycle 3). However, we observed an increase in survey return rate from cycle 1 by more than half (12.4%) despite using the same delivery method and have attributed this finding to increased patient/family familiarity with electronic clinic communications. The launch of telehealth inspired efforts to coordinate survey distribution using the electronic medical record (EMR). In cycle 4, providers could manually assign surveys to eligible patients while in the EMR. Results for returned surveys were obtained by monthly data pull, but did not capture the denominator of eligible patients. In cycle 5, the EMR was adjusted so that surveys are automatically launched to eligible patients prior to both in-person and telehealth appointments for completion via electronic clinic check-in. This method requires no personnel effort. Results are provided by monthly data pull and yielded 597/608 (98%) returned surveys in the first 8 months.

Conclusion: Using quality improvement cycles, we have created a fully automated method for assessment of patient self-reported transition readiness. Distribution through the EMR prior to appointments is efficient and effective. In the future, the ADAPT data we generate will allow us to assess for correlations between patient self-reported transition readiness and successful healthcare transition.

Disclosure: D. McDonald: None; **K. Nasto**: None; **K. Fergason**: None; **M. Robichaux**: None; **B. Danna**: None; **M. Maher**: None; **A. Alexander**: None; **A. Coleman**: None; **A. Dykes**: None; **J. Rogers**: None; **M. Gillispie-Taylor**: Pfizer, 5; **T. Vogel**: Moderna, 2, Novartis, 2, Pfizer, 2, sobi, 2.

Abstract Number: 089

The Mosaic of Mental Health: Perceived Impact of a Workshop Empowering Pediatric Rheumatology Clinicians in Routine Screening and Effective Management of Mental Health Problems

Tala El Tal[1], Kaveh Ardalan[2], Natoshia Cunningham[3], Megan Curran[4], Mariel Dela Paz[5], Suzanne Edison[6], Michelle Itczak[7], Susan Kim[8], Alana Goldstein-Leever[9], Sharon Lorber[1] and **Andrea Knight**[1], [1]The Hospital for Sick Children, Toronto, ON, Canada, [2]Duke University School of Medicine, Durham, NC, [3]Michigan State University, Grand Rapids, MI, [4]University of Colorado, Denver, CO, [5]UCSF, San Francisco, CA, [6]Cure JM Foundation, [7]University of Indianapolis, Indianapolis, IN, [8]UCSF Benioff Children's Hospital, San Francisco, CA, [9]Nationwide Children's Hospital, Columbus, OH

SESSION INFORMATION

Session Date: Thursday, March 30, 2023

Session Title: Posters: Quality, Health Services, and Education I

Session Type: Poster Session A

Session Time: 6:00PM–7:00PM

Background/Purpose: Mental health (MH) problems, particularly anxiety and depression, are common in children and adolescents with pediatric rheumatologic diseases, and impact disease-related outcomes. Pediatric rheumatology providers often lack training to effectively address these problems. As part of a quality improvement project, we developed a workshop series to equip pediatric rheumatology providers with MH assessment skills and management strategies. We measured participant-reported perceived impact of the workshops on future MH practices.

Methods: Two 2-hour, Continuing Medical Education (CME)-certified virtual workshops were delivered in October 2022. Speakers included 3 pediatric rheumatologists, 2 social workers, 1 parent, 2 psychologists, and 1 art therapist/licensed mental health counselor. The learning objectives were to: 1) recognize the impact of MH for pediatric rheumatology patients, 2) identify early signs/symptoms of anxiety and depression, 3) perform routine screening for anxiety and depression, suicide

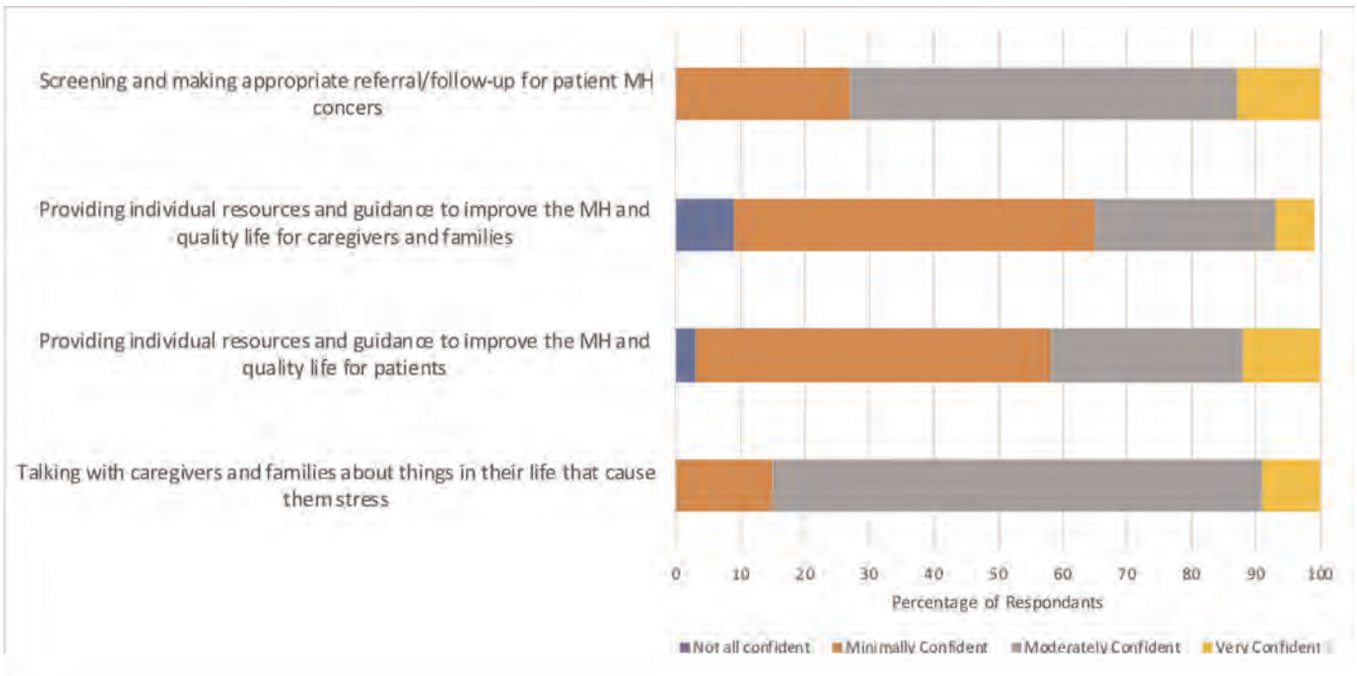


Figure 1: Pre-activity survey for participant-reported level of confidence in MH care, n=38

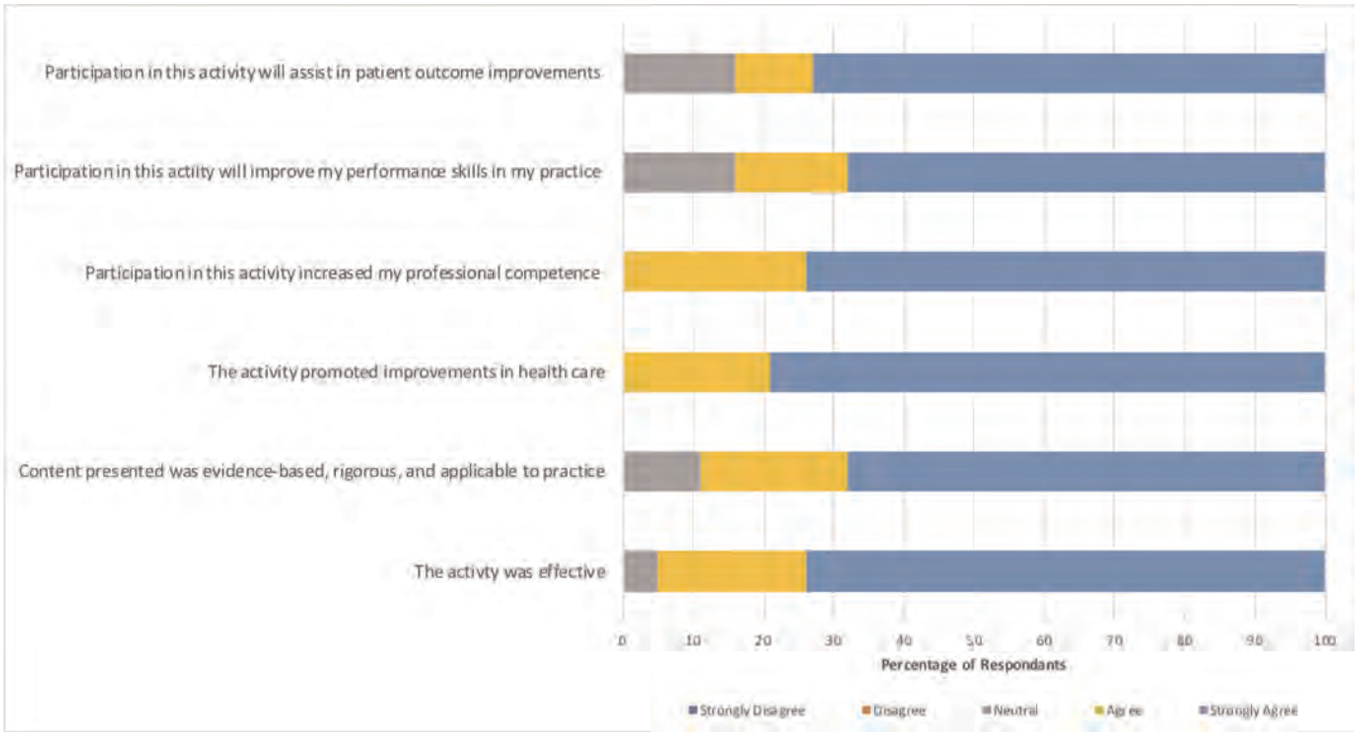


Figure 2: Post-activity survey for participant-reported feedback on workshop series, n=19

risk assessment, and safety planning, and 4) understand the role of different MH interventions (e.g., coping strategies, cognitive behavioral therapy, art/play therapy). Each workshop had a 30-minute breakout discussion of participants MH care experience, barriers and facilitators. Participants completed pre- and post-activity surveys using Likert scales to indicate their level of confidence in MH care, and perceived impact of the workshops. Participant demographics were summarized using descriptive statistics, and Likert responses were tabulated.

Results: Eighty participants registered for both workshops from across the world (Canada, Colombia, Italy, Mexico, Spain, United Kingdom, and United States) including 46 physicians (57.5%), 7 nurse practitioners (8.8%), 6 nurses (7.5%), 5 social workers (6.3%), 3 psychologists (3.8%), and 13 others (with roles as physical therapists, physician assistants and administration). Forty-eight percent (n=38) of registered participants attended both workshops. Pre-activity surveys indicated that 97.3% (n=37) of attendees agreed it is important to routinely screen patients for depression and anxiety in rheumatology clinic. The most common cited barriers were limited time, MH resources and funds. While 76% felt at least moderately confident about talking with patients/families about MH, less than half felt at least minimally confident about providing MH resources/guidance (Figure 1). Post-activity surveys showed that 90% and 85% felt that the workshops were effective and increased their competence level, respectively (Figure 2). Anticipated changes in practice included better implementation of MH screening, suicide risk assessment and safety planning.

Conclusion: This data supports the feasibility of a multidisciplinary MH educational workshop series for equipping pediatric rheumatology providers with knowledge and skills to screen and manage MH problems. Further refinement and dissemination of provider educational workshops may improve MH outcomes for youth with pediatric rheumatology diseases.

Disclosure: T. El Tal: None; **K. Ardalan**: None; **N. Cunningham**: None; **M. Curran**: None; **M. Dela Paz**: None; **S. Edison**: None; **M. Itczak**: None; **S. Kim**: None; **A. Goldstein-Leever**: None; **S. Lorber**: None; **A. Knight**: None.

Abstract Number: 090

Safety Outcomes of Combined Biologics Use in Pediatric Rheumatology: A Single Center Experience

Angela Chun[1], MaiLan Nguyen[1], Marietta De Guzman[2] and Andrea Ramirez[3], [1]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [2]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [3]Baylor College of Medicine, Houston, TX

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Posters: Clinical and Therapeutic II

Session Type: Poster Session B

Session Time: 5:00PM–6:00PM

Background/Purpose: The management of pediatric rheumatic disease has been forever changed by the advent of biologic drugs and the pursuit of targeted therapy. There is growing literature on the safety and efficacy of combining synthetic and biologic disease modifying anti-rheumatic drugs (DMARDs). However, in rare instances, patients continue to have breakthrough disease activity despite escalation of conventional therapies, and rheumatologists must consider the

Table 1: Patient outcomes while on combined biologics regimen Abbreviations used: BID- twice daily, CBR- combined biologics regimen, CVC- central venous catheter, DC- discontinuation, IAC- intra-articular corticosteroids, JIA- Juvenile Idiopathic Arthritis, MAS- Macrophage Activation Syndrome, Mos- months, PO- oral, q- every, QD- daily, SJIA-LD- Systemic Juvenile Idiopathic Arthritis Related Lung Disease, subQ- subcutaneous, Yrs- years

Patient	Rheum Diagnosis	Age at Diagnosis (yrs)	Time from symptom onset to initial therapy (mos)	Previously failed therapies	Combined biologics regimen (CBR) [+Concomitant synthetic DMARDs]	Age at CBR (yrs)	Outcomes while on CBR							Follow up time since CBR initiation (mos)
							Off chronic PO steroids	Infection triggering a flare	Infection requiring antimicrobials	Infection requiring hospitalization	Other adverse outcomes	Flare requiring hospitalization	Clinical & serologic remission for >6 mos	
1	Systemic JIA with history of MAS and SJIA-LD	1	6 (Steroids) 11 (Tocilizumab)	-Etoposide -Tocilizumab -Tofacitinib	-Canakinumab 150 mg (~7 mg/kg) q28 days -Tofacitinib 3.2 mg BID	3	N	N	N	N	Mild transaminitis	Y	N	26
2	Systemic JIA with history of MAS	4	6 (Anakinra)	-Anakinra -Canakinumab -Tacrolimus -Tocilizumab	-Canakinumab 150 mg (~8mg/kg) q21 days -Tofacitinib 5 mg BID	6	N	N	N	N	N	Y	N	10 prior to DC
					-Emapalumab -Tofacitinib 5 mg BID	7	N	N	Y (Axillary lymphadenitis)	Y (Viral gastroenteritis x2)	Pneumothorax x2 associated with CVC	Y	N	24
3	Systemic JIA with history of MAS	9	7 (Anakinra)	-Anakinra -Canakinumab -IAC injections -Methotrexate -Tofacitinib -Tocilizumab	-IV Tocilizumab 10mg/kg q14 days -Tofacitinib 10 mg BID [+PO Methotrexate 20 mg weekly]	15	Y	Y (Herpes Zoster)	Y (Herpes Zoster)	Y (Herpes Zoster)	N	Y	N/A (CBR DC)	2 prior to DC
4	Systemic JIA	3	3 (Steroids) 10 (Tocilizumab)	-Infliximab -Methotrexate -Tocilizumab -Tofacitinib	-Anakinra 100 mg (~3mg/kg) QD -Tofacitinib 5 mg BID [+SubQ Methotrexate 15 mg weekly]	8	Y	N	N	N	N	N	N	5
5	Seronegative polyarticular JIA	3	N/A	-Abatacept -Adalimumab -Etanercept -Infliximab -IAC injections -Tocilizumab -Tofacitinib	-IV Tocilizumab 10mg/kg q28 days -Tofacitinib 5 mg BID [+SubQ Methotrexate 12.5mg weekly]	6	Y	N	N	N	Mild transaminitis	N	N	30
6	Seronegative polyarticular JIA	1.5	5 (Steroids) 6 (Methotrexate)	-Anakinra -Etanercept -Golimumab -Hydroxy-chloroquine -IAC injections -Infliximab -Methotrexate -Tocilizumab	-Anakinra 100 mg BID (~4mg/kg) -Tofacitinib 5 mg BID [+Hydroxychloroquine 100 mg QD]	13	Y	N	N	N	N	N	N	15
7	Systemic JIA with history of MAS and SJIA-LD	2	20 (Steroids) 21 (Anakinra)	-Anakinra -Canakinumab -Tocilizumab -Tofacitinib	-Anakinra 100 mg QD (~6mg/kg) -Emapalumab [+Cyclosporine 4mg/kg QD]	3	Y	N	N	N	N	N	N	10 prior to DC
					-Anakinra 100 mg QD (~5mg/kg) -Ruxolitinib 5 mg BID [+Cyclosporine 4mg/kg QD]	4	Y	N	N	N	N	N	N	2
8	Systemic JIA	3	2 (Steroids) 13 (Anakinra)	-Adalimumab -Anakinra -Canakinumab -Etanercept -IAC injections -Methotrexate -Tocilizumab	-Tofacitinib 5 mg BID -Canakinumab 150 mg (~2mg/kg) q28 days [+Sulfasalazine 500 mg BID]	10	Y	N	N	N	N	Y	N	18

Table 1: Patient outcomes while on combined biologics regimen
Abbreviations used: BID- twice daily, CBR- combined biologics regimen, CVC- central venous catheter, DC- discontinuation, IAC- intra-articular corticosteroids, JIA- Juvenile Idiopathic Arthritis, MAS- Macrophage Activation Syndrome, Mos- months, PO- oral, q- every, QD- daily, SJIA-LD- Systemic Juvenile Idiopathic Arthritis Related Lung Disease, subQ- subcutaneous, Yrs- years

synergistic use of combined biologics. Currently, there is a paucity of literature describing the use of combined biologics in pediatrics. We aim to describe the short-term safety outcomes of a cohort of pediatric rheumatology patients treated with a combined biologics regimen (CBR).

Methods: In this single-center retrospective chart review, we identified pediatric patients treated with combined biologics for a systemic autoimmune disease between 2020-2022 at Texas Childrens Hospital. With IRB approval, we extracted clinical features, immunomodulation use, and outcomes for these patients.

Results: Eight patients were identified: 6 with systemic Juvenile Idiopathic Arthritis (JIA) and 2 with seronegative polyarticular JIA. Median age at time of CBR initiation was 6.5 years (range 3-18). All patients had severe, refractory disease, and all had failed multiple synthetic and biologic DMARDs prior to CBR initiation. Two patients were trialed on more than one CBR.

Table 1 summarizes CBR-related outcomes. While on CBR, two patients were hospitalized for infection (viral gastroenteritis and cutaneous herpes zoster respectively), and CBR was subsequently discontinued for one. One patient was hospitalized for pneumothoraces associated with implanted port access. Two patients are monitored for mild transaminitis attributed to pharmacotherapy.

Though 6 patients were able to come off of chronic oral steroids after CBR initiation, none of the patients were able to maintain clinical and serologic remission for longer than 6 months.

Conclusion: Majority of patients did not develop serious infections or adverse events requiring hospitalization while on CBR. Additional prospective cohort studies are needed to determine the efficacy and long-term outcomes for pediatric patients on combined biologics.

Disclosure: A. Chun: None; **M. Nguyen**: None; **M. De Guzman**: None; **A. Ramirez**: None.

Abstract Number: 091

Achieving Remission in Childhood-onset Systemic Lupus Erythematosus: Rapid Implementation of an EMR-integrated Dashboard to Measure Disease Activity and Remission Rates

Kaleo Ede[1], Nikita Goswami[2], Elisa Wershba[2], Michael Shishov[2], Samantha Casselman[2], Pierina Ortiz[2] and Vinay Vaidya[2], [1]Phoenix Children's Hosptial; University of Arizona College of Medicine- Phoenix, Phoenix, AZ, [2]Phoenix Children's Hospital, Phoenix, AZ

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Children with childhood-onset systemic lupus erythematosus (cSLE) experience more severe disease than their adult counterparts, in addition to high rates of clinical depressive symptoms (30%) and fatigue (65%) (1). A "high- risk" profile categorized by clinically elevated depressive symptoms and fatigue has been shown to be related to reduced health-related quality of life and poorer patient outcomes in cSLE patients (2). The **T**reatment and **E**ducation **A**pproach for **CH**ildhood-onset lupus (TEACH) is a tailored cognitive behavioral therapy (CBT) for youth with cSLE that

may address these symptoms. (3). TEACH is currently being tested in an RCT with promising results after 6 weeks. However, it is unknown how the "high- risk" group may respond to this program. This study aims to assess the impact of risk status on the outcomes of cSLE patients undergoing CBT.

Methods: This study uses data from an ongoing multisite clinical trial of youth (average age of 17 years, 100% female, 70% identifying as from a minority group, n=46/60 currently completed evaluation for the study). All of these patients met the ACR criteria for cSLE. 16 of these youth have thus far been randomized to and completed TEACH. They were grouped into low- or high-risk categories based on baseline depression and fatigue t-scores (>70; >2SD from mean) (3). Repeated measures ANOVAs were conducted to compare the low- and high-risk groups pre and post 6 week TEACH protocol for main study outcomes (e.g. depressive symptoms (CDI-II), fatigue (PROMIS), and pain (VAS)) and additional exploratory outcomes

DORIS

Table definitions

Date	cSLEDAI 2K SCORE	PGA	Pred dose mg/day	SLE MEDS	Clinical Status	COMMENTS/RX

Figure 1: Example of DORIS remission criteria table in patient EMR clinic note

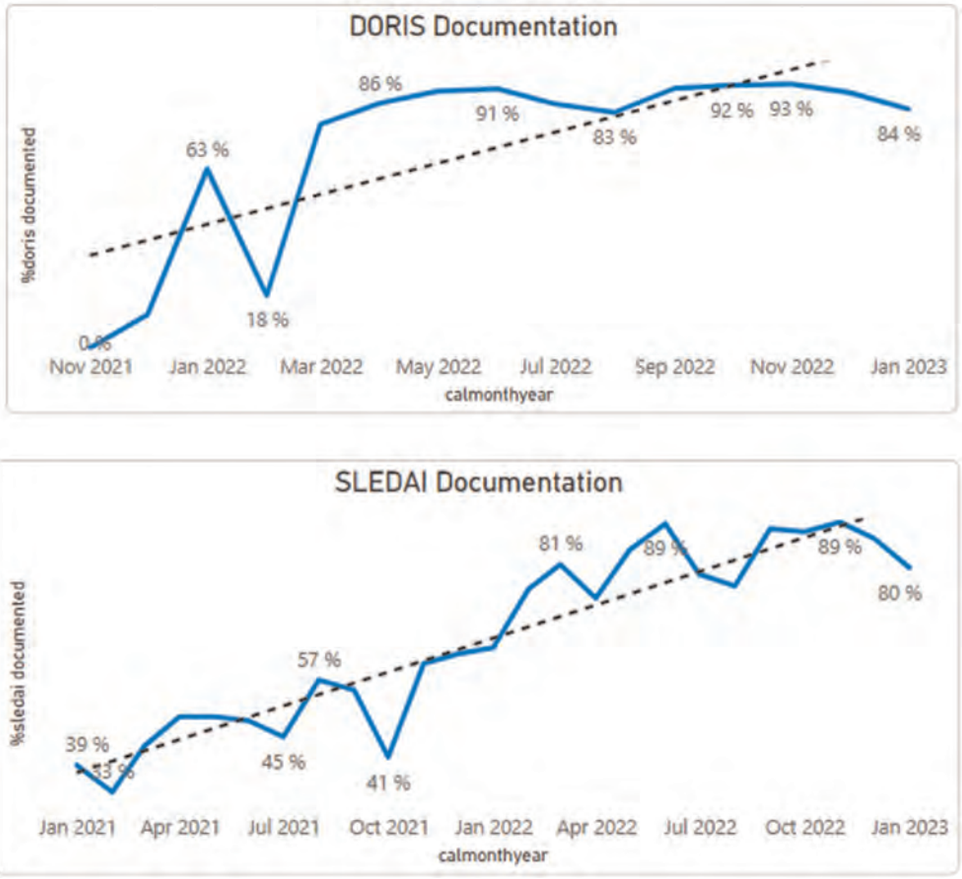


Figure 2a: % of charts with cSLEDAI documented at last visit by month Figure 2b: % of charts with DORIS remission criteria documented at last visit by month

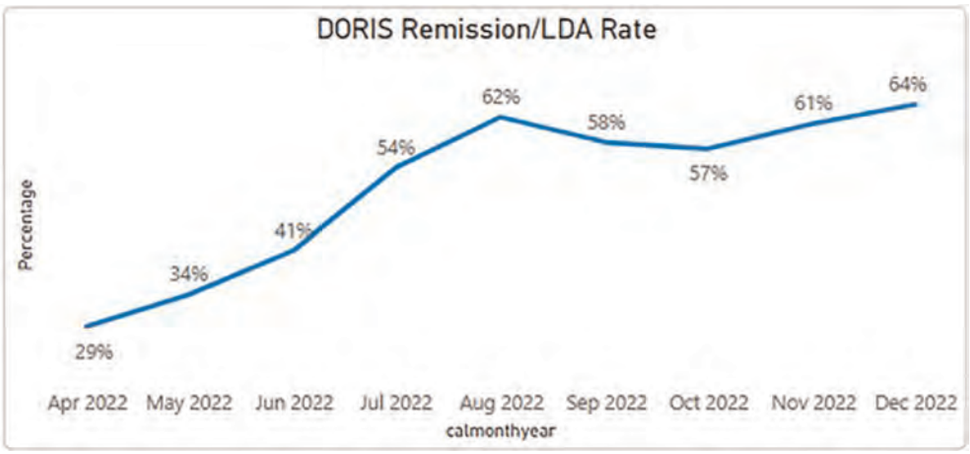


Figure 3: % of patients with DORIS/LLDA at last visit by month

(anxiety (SCARED), disease severity (SLEDAI), and health-related quality of life (Peds-QL)). Independent samples t-test was used to determine if baseline outcome measures were significantly different between low- and high-risk groups.

Results: Out of the 16 patients who have completed the TEACH protocol, 6 (37.5%) met criteria for inclusion in the high-risk group. There was no significant difference between risk groups based on age, disease duration, or self-reported race. There was a significant difference between groups in baseline depression scores only ($p = 0.005$). At week 6 after completion of TEACH, depression scores decreased by 26% in the high- risk group, while the low- risk group only decreased 6% ($p < 0.001$; Table 1). Fatigue, anxiety, health-related quality of life, and disease severity followed the same trends (see Figure 1). Despite these trends, there were no significant differences found between the low- and high-risk groups in any outcomes other than depressive symptoms.

Conclusion: There is very little literature on nonpharmacologic treatments for cSLE like CBT (4). These data reveal interesting trends that CBT may be more effective in cSLE patients with high levels of fatigue and depression. Some results may be driven by the fact that the high-risk group begins with higher baseline symptoms. Lack of significant difference between groups may be attributed to a small sample size and these analyses should be replicated with a larger data set (e.g., upon completion of the RCT) in the future. Although the high-risk group shows more improvement overall, this should not shadow that both high- and low- risk groups had improved outcomes with the TEACH protocol.

Disclosure: K. Ede: None; **N. Goswami**: None; **E. Wershba**: None; **M. Shishov**: AbbVie/Abbott, 6, Novartis, 6; **S. Casselman**: Sanofi, 6; **P. Ortiz**: None; **V. Vaidya**: None.

Abstract Number: 092

Procalcitonin Levels in Patients with Juvenile Dermatomyositis Compared to Healthy Controls

Christopher Costin[1], Gabrielle Morgan[2], Lutfiyya Muhammad[3], Amer Khojah[4], Marisa Klein-Gitelman[1], Yvonne Lee[5] and Lauren Pachman[6], [1]Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, [2]Ann & Robert H. Lurie Children's Hospital of Chicago and Northwestern University Feinberg School of Medicine, Chicago, IL, [3]Northwestern University Feinberg School of Medicine, Chicago, IL, [4]Umm Al-Qura University, Makkah, Saudi Arabia, [5]Northwestern University, Chicago, IL, [6]Northwestern's Feinberg School of Medicine. Ann and Robert H. Lurie Children's Hospital of Chicago; Stanley Manne Children's Research Institute of Chicago, Lake Forest, IL

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Patients with JDM are at increased risk of infection due to increased aspiration risk and immunosuppression. Procalcitonin (PCT) is a biomarker of infection, that has a growing role in both diagnosis of bacterial infection and antibiotic management. PCT rises in response to bacterial infections such as pneumonia or sepsis; however, studies have shown that PCT is also elevated in sJIA, MAS or SLE even in the absence of infection. The objectives of this study were to investigate whether PCT is elevated in patients with active JDM and if PCT correlates with JDM disease activity. If PCT is not associated with JDM activity, it may remain a reasonable choice to screen for bacterial infection. If PCT is associated with JDM activity, it may serve as a tool to monitor JDM activity; however, its utility for bacterial infection detection would be reduced.

Methods: Data and serum samples were obtained from the Cure JM biorepository and registry. Control samples were obtained from previously stored samples (housed in the biorepository) of allergy clinic patients under 18 years of age with non-allergic rhinitis. Inclusion criteria for the JDM samples included a diagnosis of JDM by the Bohan and Peter criteria and availability of clinical data and a serum sample within 4 months of enrollment. Exclusion criteria were age greater than 18 years. PCT concentration of the serum samples was determined by the central clinical laboratory through electrochemiluminescence. Laboratory (muscle enzymes, MSAs, NK cells, inflammatory markers) and clinical characteristics (demographics, Childhood Myositis Assessment Scale (CMAS), Disease Activity Scores) for the JDM patients were obtained from the registry from the closest collection point to serum acquisition. A Fishers exact test was used to compare the proportions of JDM and control groups with PCT levels above and below the detection threshold of 0.1. Spearman correlation was used to assess the correlation between PCT values and markers of JDM disease activity among patients with detectable PCT levels.

Results: We obtained the serum of 44 patients with recently diagnosed JDM along with 21 sex-matched healthy controls. Of the 44 JDM patients, 6 patients had a detectable PCT levels with the highest PCT value obtained being 0.39 ng/ml. All of the controls had undetectable PCT levels. The proportion of patients with detectable PCT levels was not significantly elevated in JDM compared to controls (p= 0.17). Among the 6 patients with abnormal PCT, there was no correlation between PCT level and other markers of JDM disease activity.

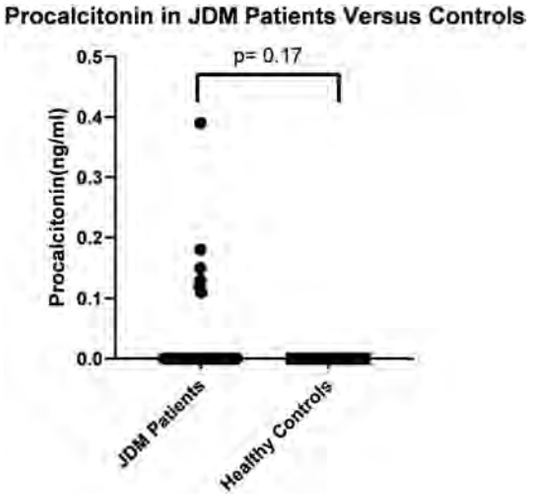


Figure 1: Box Plot Comparing PCT levels by Fisher Exact Test Between JDM Patients and Healthy Controls

Clinical and Laboratory Characteristics of the Elevated and Normal Procalcitonin Groups.

Laboratory Findings	Reference Range	Elevated PCT Group (N = 6)	Normal PCT Group (N = 38)
C Reactive Protein (mg/dl)	0-0.8	0.20 ± 0.10	0.24 ± 0.27
Erythrocyte Sedimentation Rate(mm/hr)	0-20	13 ± 6.3	15.1 ± 21.35
Neopterin (nmol/l)	0-10	17.7 ± 12.2	14.1 ± 10.8
NK cells (CD16+/CD56+)	121-1581	166.2 ± 94	138 ± 92
von Willebrand factor antigen (%)	36-241	148 ± 104	132 ± 46
Creatine phosphokinase (IU/L)	26-279	716 ± 1347	2765 ± 10387
Aspartate aminotransferase (IU/L)	18-65	308 ± 510	136 ± 327
Lactate dehydrogenase (IU/L)	147-438	433 ± 230	492 ± 410
Aldolase (U/L)	3.4-11.8	9.93 ± 5.05	12.7 ± 9.7
Clinical Findings	**Reference Range**	**Elevated PCT Group (N = 6)**	**Normal PCT Group (N = 38)**
Age(years)	0-18	6.12 ± 4.5	8.18 ± 4.68
Duration of Untreated Disease(months)	n/a	7.11 ± 10.5	5.24 ± 4.92
Disease activity score—total	0-20	7.75 ± 4.73	30.65 ± 14.28
Disease activity score—skin	0-9	3.33 ± 2.58	5.00 ± 2.29
Disease activity score—muscle	0-11	4.42 ± 3.17	3.91 ± 2.83
Childhood Myositis Assessment Scale	0-52	38 ± 12.5	34.85 ± 14.31

Conclusion: The proportion of children with detectable PCT levels was not significantly higher among those with JDM compared to controls, suggesting that elevated PCT levels are not a marker of JDM. This study suggests that moderately or highly elevated PCT levels in JDM patients are unlikely to be due to their autoimmune disease. PCT did not correlate with typical markers of JDM disease activity although this analysis was limited due to the small number of JDM patients with abnormal PCT. Further studies including a higher proportion of JDM patients with elevated PCT are needed to explore potential associations that may not have been captured in this study.

Disclosure: C. Costin: None; **G. Morgan**: None; **L. Muhammad**: None; **A. Khojah**: None; **M. Klein-Gitelman**: AbbVie/Abbott, 5, Bristol-Myers Squibb(BMS), 5, Pfizer, 5, Up to Date, 12, Author; **Y. Lee**: Cigna-Express Scripts, 11, Eli Lilly, 12, Medical Writer, Pfizer, 5, Sanofi, 12, Medical Writer; **L. Pachman**: None.

Abstract Number: 093

Comparison Between Induction with Rituximab and Cyclophosphamide in Treatment of Childhood-Onset ANCA-Associated Vasculitis

Samuel Gagne[1], Kimberly Morishita[2], Else Bosman[3], Vidya Sivaraman[4], David Cabral[5], For the PedVas Investigators[6] and Brett Klamer[7], [1]Nationwide Children's Hospital, Columbus, OH, [2]University of British Columbia, Vancouver, BC, Canada, [3]UBC, Vancouver, BC, Canada, [4]Nationwide Children's Hospital/ The Ohio State University, Columbus, OH, [5]BC Children's Hospital and University of British Columbia, Vancouver, BC, Canada, [6]PedVas, [7]Biostatistics Resource at Nationwide Children's Hospital (BRANCH), Nationwide Children's Hospital, Columbus, OH

SESSION INFORMATION

Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA) are a group vasculitides with significant morbidity and mortality requiring toxic therapy. Clinical trials in adults have identified rituximab (RTX) as being an effective alternative to cyclophosphamide (CYC) for remission induction. As disease rarity in children limits the feasibility of pediatric trials, treatment data has been extrapolated from adult studies although efficacy and toxicity may differ in children. Here, we evaluate the efficacy and toxicity of CYC versus RTX through pragmatic, registry-based comparative evaluation.

Methods: From ARChiVe, a registry of childhood vasculitis, we identified patients with MPA or GPA who received induction with either RTX or CYC. We compared outcomes including rate of infection or drug-related hospitalization at 6 months and rate of remission and Pediatric Vasculitis Damage Index (pVDI) at 6 & 12 months. Remission was defined as Pediatric Vasculitis Activity Score (PVAS) ≤ 2 (inactive or low disease activity). Regression analysis was used for comparative evaluation of outcomes.

Table 1: Patient Characteristics by Induction Medication

Characteristic	Rituximab, N = 45[1]	Cyclophosphamide, N = 48[1]	Combination, N = 11[1]	p-value[2]	Overall, N = 104[1]
Sex				0.7	
Female	31 (69%)	33 (69%)	6 (55%)		70 (67%)
Age at diagnosis (years)	14.0 (11.8, 16.0)	14.0 (12.0, 15.0)	16.0 (15.0, 17.0)	**0.033**	14.0 (12.0, 16.0)
Parents' ethnicity				0.6	
White	31 (69%)	25 (52%)	8 (73%)		64 (62%)
Hispanic/Latino	3 (6.7%)	4 (8.3%)	1 (9.1%)		8 (7.7%)
Asian	4 (8.9%)	9 (18.8%)	1 (9.1%)		14 (13.5%)
Black	1 (2.2%)	0 (0%)	1 (9.1%)		2 (1.9%)
Other	6 (13.3%)	10 (20.1%)	0 (0%)		16 (15.4%)
Country of Residence					
United States	22 (49%)	10 (21%)	7 (64%)		39 (38%)
Canada	12 (27%)	23 (48%)	0 (0%)		35 (34%)
Europe	11 (24.4%)	9 (18.8%)	4 (36.3%)		24 (23.1%)
Asia	0 (0%)	4 (8.3%)	0 (0%)		4 (3.8%)
Argentina	0 (0%)	2 (4.2%)	0 (0%)		2 (1.9%)
Diagnosis				0.077	
Granulomatosis with Polyangiitis	37 (82%)	41 (85%)	6 (55%)		84 (81%)
Microscopic Polyangiitis	8 (18%)	7 (15%)	5 (45%)		20 (19%)
Days between symptom onset and diagnosis	41 (16, 119)	35 (23, 78)	55 (14, 100)	0.7	36 (22, 94)
Renal involvement	35 (78%)	45 (94%)	10 (91%)	0.066	90 (87%)
Chest involvement	21 (47%)	24 (50%)	8 (73%)	0.3	53 (51%)
Postinduction visit				>0.9	
Refractory disease	16 (36%)	18 (38%)	4 (36%)		38 (37%)
Low disease activity	8 (18%)	10 (21%)	2 (18%)		20 (19%)
Remission	21 (47%)	20 (42%)	5 (45%)		46 (44%)
Remission or low disease activity	29 (64%)	30 (62%)	7 (64%)		66 (63%)
PVAS score at diagnosis	19 (14, 24)	19 (17, 24)	21 (18, 26)	0.4	19 (16, 24)

[1]n (%); Median (IQR)
[2]Fisher's exact test; Kruskal-Wallis rank sum test; Pearson's Chi-squared test

Results: 104 patients (median age 14 years) were included (81% with GPA). 43% received RTX, 46% CYC, and 11% received both RTX and CYC. Ninety of 104 had 12-month follow-up data. Initial PVAS score, diagnosis, and age at onset were similar across treatment groups. Overall, 63% and 72% of patients achieved remission at 6 and 12 months, respectively. There was no significant difference between treatment groups (p >0.9). Patients treated with RTX had a slightly higher odds of remission at 6 months, but this was not statistically significant (OR 1.07, 95% CI: 0.45,2.52). Hospitalization occurred in 18%; 22% of patients on RTX versus 10% on CYC although this difference was not statistically significant (OR 2.29, 95% CI: 0.73,7.16). The median pVDI at 12 months was one in both groups; patients receiving RTX had a slightly higher odds of a greater VDI, but this was not statistically significant (OR 1.06, 95% CI 0.47,2.36).

Conclusion: This is the first study comparing CYC and RTX for induction in pediatric GPA and MPA. No significant differences were shown between induction regimens with respect to rates of remission, severe adverse events, or degree of organ damage. Patients receiving RTX had more hospitalizations than those on CYC, but this was not statistically significant though there were a low number of hospitalizations overall. This study was limited by its sample size, retrospective nature, and lack of longitudinal adverse drug-related event data. Larger studies with longer follow-up are needed for increased precision in estimated differences between treatment regimens and reduction of uncontrolled confounding. These results represent an important early evaluation.

Disclosure: S. Gagne: None; **K. Morishita**: None; **E. Bosman**: None; **V. Sivaraman**: None; **D. Cabral**: None; **F. PedVas Investigators**: None; **B. Klamer**: None.

Abstract Number: 094

Provider Assessment of the Temporomandibular Joint in Juvenile Idiopathic Arthritis

Anna Costello[1], Marinka Twilt[2] and Melissa Lerman[1], [1]Children's Hospital of Philadelphia, Philadelphia, PA, [2]Alberta Children's Hospital, Calgary, AB, Canada

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: TM joint arthritis is an underrecognized complication of JIA that can cause long-term problems including decreased mandibular growth, altered facial morphology, and orofacial pain. It is estimated that the TM joint may be affected in 40-80% of children with JIA. Standardized physical exam and imaging are important ways to accurately assess for this phenomenon. Little is known about the rate at which providers evaluate for TM joint involvement in their clinical practice.

Methods: Data was obtained from the New Childhood Arthritis and Rheumatology Research Alliance (CARRA)Registry. Data fields related to assessment for TM joint arthritis were added in 2019. Patients were included in the study if they had a diagnosis of JIA and had data recorded between January 2020 and August2021. Standard descriptive statistics were used to describe demographic and clinical features.

Results: 17761 visits were reviewed for a total of 7473 patients with JIA. 52.7% of patients ever had maximal mouth opening (MMO) recorded as finger breadths or total incisal distance (TID). Only 8% had TID measured (Figure 2). 5.0%had MRI with contrast performed. 939 patients had a diagnosis of TM joint arthritis. Of these, 28.5% have an MRI documented,

	Total in Sample (n=7473)	MMO Ever Recorded	p-value	MRI with Contrast Ever Recorded	p-value
Sex					
Female	5290 (70.8%)	3533 (66.8%)	0.64	297 (5.6%)	<0.001
JIA Subtype			0.013		<0.001
ERA*	771 (10.3%)	528 (68.5%)		43 (5.6%)	
Oligoarthritis	2712 (34.3%)	1831 (67.5%)		134 (4.5%)	
Poly (RF neg) *	2227 (29.8%)	1491 (67.0%)		132 (5.9%)	
Poly (RF pos)*	469 (6.2%)	287 (61.2%)		15 (3.2%)	
Psoriatic*	613 (8.2%)	430 (70.1%)		35 (5.7%)	
Systemic*	518 (6.9%)	325 (62.7%)		6 (1.2%)	
Undifferentiated*	156 (2.1%)	109 (69.9%)		7 (4.5%)	
Race/Ethnicity					
Native American	131	87 (66.4%)		4 (3.1%)	
Asian	304	190 (62.5%)		11 (3.6%)	
Black	387	265 (68.5%)		23 (5.9%)	
White	6036	4082 (67.6%)		310 (5.1%)	
Hispanic	769	477 (62.0%)		24 (3.1%)	
Middle Eastern	54	28 (51.9%)		2 (3.7%)	
Native Hawaiian	43	31 (72.1%)		1 (2.3%)	
Other	101	70 (69.3%)		5 (4.9%)	
No answer	166	118 (71.1%)		6 (3.6%)	
Age			0.765		<0.001
0 to 5	3317 (44.4%)	2205 (66.5%)		110 (3.3%)	
6-12	2745 (36.7%)	1882 (68.6%)		150 (5.5%)	
12+	1407 (18.8%)	916 (65.1%)		112 (8.0%)	
Diagnosis of TMJ arthritis					
Yes	833 (11.4%)	702 (84.2%)	<0.001	234 (28.1%)	<0.001
ANA Status			0.53		0.25
Positive	3596 (54.2%)	2435 (67.7%)		199 (5.5%)	
Negative	4345 (48.2%)	2706 (62.3%)		261 (6.0%)	
Anti-CCP			0.13		0.71
Positive	462 (13.8%)	305 (66.0%)		22 (4.8%)	
Negative	4040 (86.4%)	2647 (65.5%)		315 (7.8%)	
HLA-B27			0.37		0.50
Positive	638 (17.2%)	458 (71.7%)		33 (5.2%)	
Negative	4226 (82.8%)	2786 (65.9%)		359 (8.5%)	
Rheumatoid Factor			0.26		0.07
Positive	531 (9.6%)	350 (65.9%)		19 (3.8%)	
Negative	6748 (89.8%)	4399 (65.2%)		536 (7.9%)	

Legend: ERA: Enthesitis related arthritis; Poly: Polyarticular; Psoriatic: Psoriatic arthritis; Systemic: Systemic Arthritis; Undifferentiated: Undifferentiated arthritis.

Figure 1

83% have an MMO documented, and 40% have TID measured. During the study, 51 patients were newly diagnosed with TM joint arthritis. 84% have MMO recorded which was evaluated as TID in 51%. 41%had an MRI with contrast. Few patient-level characteristics were statistically related to having MMO assessed. It was assessed in ~2/3 of patients in all subtypes (Table 1). MRI was more likely to be obtained in older patients and in female patients. It was significantly more likely that MMO was recorded at the patients baseline visit than at a follow up visit. There was variance in assessments between the 59 sites. MMO was recorded at the baseline visit on 80% of patients at 3 sites and it was never recorded at 11 sites. MRIs were infrequently performed at all sites with 38 sites having no MRIs ordered (Figure 3).

Conclusion: Maximal mouth opening is not consistently measured in patients with JIA, and it is rarely measured quantitatively. Similarly, TMJ MRIs are rarely obtained in these patients. Site of care is more associated with TMJ assessments than patient level characteristics. It is possible that abstractions of TMJ data into the database is incomplete and does not reflect clinical assessment. The data fields were newly added, and it is possible that completion rates might increase in later time

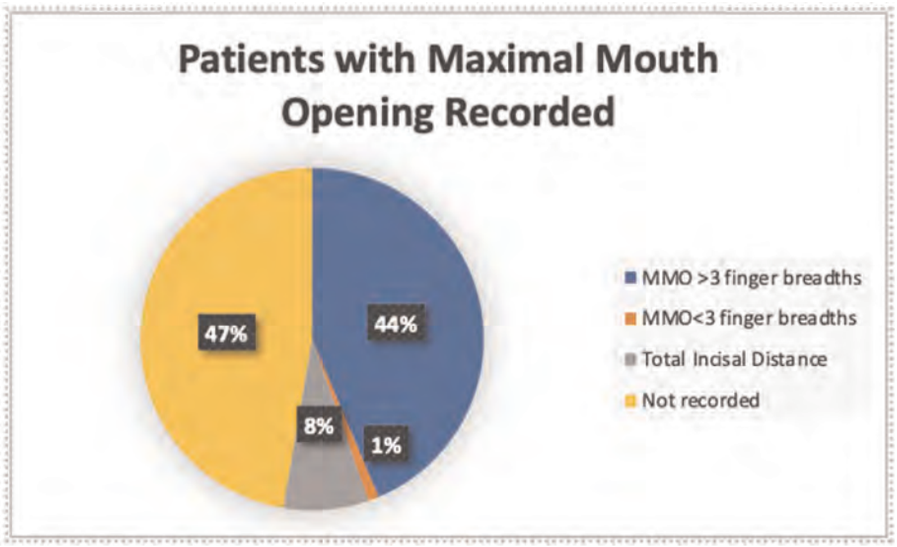


Figure 2

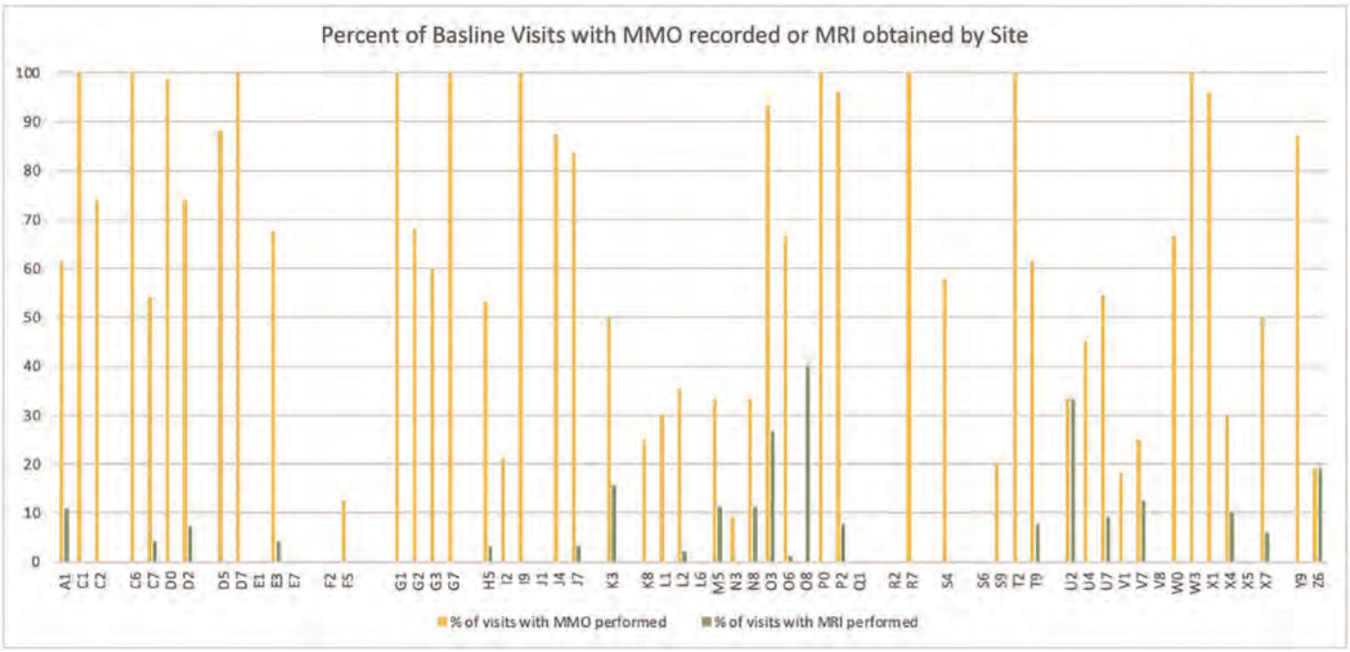


Figure 3

periods. However, this data suggests that education of providers is needed to improve the assessment of the TM joint in patients with JIA to help prevent long-term complications.

Disclosure: A. Costello: None; **M. Twilt**: None; **M. Lerman**: None.

Abstract Number: 095

Systemic Juvenile Idiopathic Arthritis Associated Lung Disease in Europe

Claudia Bracaglia[1], Francesca Minoia[2], Christoph Kessel[3], Sebastiaan Vastert[4], Manuela Pardeo[1], Alessia Arduini[1], Sarka Fingerhutova[5], Irina Nikishina[6], Ozge Basaran[7], Nural Kiper[8], Mikhail Kostik[9], Mia Glerup[10], Roberta Caorsi[11], AnnaCarin Horne[12], Giovanni Filocamo[2], Helmut Wittkowski[3], Marija Jelusic[13], Jordi Anton[14], Samira Khaldi-Plassart[15], Alexandre Belot[15], Gerd Horneff[16], Seraina Palmer Sarrott[17], Elvira Cannizzaro Schneider[18], Lampros Fotis[19], Pavla Dolezalova[5], Angelo Ravelli[20], Seza Ozen[7] and Fabrizio De Benedetti[1], [1]Division of Rheumatology, IRCCS Ospedale Pediatrico Bambino Gesù, Roma, Italy, [2]Fondazione IRCCS Ca' Grande Ospedale Maggiore Policlinico, Milano, Italy, [3]Department of Pediatric Rheumatology & Immunology, WWU Medical Center (UKM), Muenster, Germany, [4]Wilhelmina Children's Hospital, Department of Pediatric Immunology and Rheumatology, Utrecht, The Netherlands, Utrecht, Netherlands, [5]Centre for Paediatric Rheumatology and Autoinflammatory Diseases, Department of Paediatrics and Inherited Metabolic Disorders, 1st Faculty of Medicine, Charles University and General University Hospital in Prague, Prague, Czech Republic, [6]V.A. Nasonova Research Institute of Rheumatology, Moscow, Russia, [7]Department of Pediatrics, Division of Pediatric Rheumatology, Hacettepe University, Ankara, Turkey, [8]Department of Pediatrics, Division of Pediatric Pulmonology, Hacettepe University, Ankara, Turkey, [9]Saint-Petersburg State Pediatric Medical University, Saint-Petersburg, Russia, [10]Department of Pediatrics, Aarhus University Hospital, Aarhus, Denmark, [11]Department of Pediatrics and Rheumatology, IRRCS Istituto G. Gaslini, Genova, Italy, [12]Department of pediatric rheumathology Karolinska University Hospital and Department of pediatrics, Karolinska Institute, Stockholm, Sweden, [13]Department of Paediatrics, University of Zagreb School of Medicine, University Hospital Centre, Zagreb, Croatia, [14]Pediatric Rheumatology, Hospital Sant Joan de Déu, Universitat de Barcelona, Barcelona, Spain, [15]Pediatric Nephrology, Rheumatology, Dermatology Unit, Hôpital Femme Mère Enfant, Hospices Civils de Lyon, Lyon, France, [16]Pediatrics, Asklepios Clinic Sankt Augustin, Sankt Augustin, Germany, [17]Paediatric Rheumatology University Children's Hospital Zurich, Zurich, Zurich, Switzerland, [18]Paediatric Rheumatology University Children's Hospital Zurich, Zurich, Switzerland, [19]Pediatric Rheumatology Division, 3rd Department of Pediatrics, National and Kapodistrian University of Athens, "ATTIKON" General University Hospital, Athens, Greece, [20]IRRCS Istituto Giannina Gaslini and Università degli Studi di Genova, Genova, Italy

SESSION INFORMATION

Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Chronic parenchymal lung disease (LD) is a new emerging severe life-threatening complication of sJIA. The number of sJIA patients with LD is apparently increasing and interestingly they are reported more frequently in North America. Data regarding frequency and features of sJIA-LD in Europe are not available. The aim of this study was to evaluate the burden of sJIA associated LD in Europe.

Methods: Patients with diagnosis of sJIA with LD, including pulmonary alveolar proteinosis (PAP), interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH), followed in European paediatric rheumatology centres were identified through a survey sent to the members of the MAS/SJIA Working Party.

Results: Data from 49 JIA-LD patients, diagnosed in 17 European paediatric rheumatology centres between 2007 and 2022, were collected. 48 patients were Caucasian and 1 was African-American; 31 were female. The median age at sJIA onset was 7 years and LD occurred after a median time of 3 years. 27 patients had a chronic persistent sJIA course, 21 had a polycyclic course and only 1 patient had a monocyclic course; 38 patients (77%) had active sJIA at time of LD diagnosis. During the disease course, 41 (84%) patients developed MAS, 18 (44%) of whom had MAS at sJIA onset and 24 (58%) had full-blown MAS at time of LD diagnosis; 31 (76%) patients had 1 MAS episode. 42 (86 %) patients were treated with at least one IL-1 or IL-6 inhibitor before LD diagnosis: 33 with anakinra, 26 with canakinumab, and 23 with tocilizumab; 22 (45%) patients experienced drug adverse reaction to a cytokine inhibitor: 14 to tocilizumab and 6 to anakinra, and 1 patient to cyclosporine. 39 (80%) patients developed ILD, 6 (12%) PAP and 4 (8%) PAH. 22 (45%) patients presented

acute digital clubbing; 18 (37%) patients developed hypoxia and 9 (18%) developed pulmonary hypertension. A chest CT scan was performed in all patients with evidence of septal thickening, peri-bronchovascular thickening and ground glass opacities in the majority of patients (39, 25 and 28 patients respectively). In 22 patients a bronchoalveolar lavage was performed and 15 underwent a lung biopsy. The histopathological pattern was alveolar proteinosis in 5 patients, endogenous lipoid pneumonia in 5, vasculitis in 1 and fibrosis in 1. 45 out of 49 patients were treated with glucocorticoids (GCs) at time of LD diagnosis, and 39 received IL-1 and/or IL-6 inhibitor after the diagnosis (25 anakinra, 20 canakinumab, 16 tocilizumab). Around half of the patients (23, 47%) required ICU admission and 9 (18%) died.

Conclusion: Lung involvement is an emerging life-threatening complication of sJIA, patients are also reported in Europe. Prompt recognition is crucial and new therapeutic strategies are needed to reduce the risk and improve the outcome of this complication.

This abstract has been submitted on behalf of MAS/sJIA Working Party of PReS.

Disclosure: C. Bracaglia: Sobi, 2, 6; **F. Minoia**: Sobi, 2; **C. Kessel**: Novartis, 2, 5, Sobi, 6; **S. Vastert**: Novartis, 6, SOBI, 5, 6; **M. Pardeo**: SOBI, 2, 6; **A. Arduini**: None; **S. Fingerhutova**: None; **I. Nikishina**: Ipsen, 6, Merck/MSD, 6, Novartis, 6, Pfizer, 6, Roche, 6, R-Pharm, 6, Sobi, 6; **O. Basaran**: None; **N. Kiper**: None; **M. Kostik**: None; **M. Glerup**: None; **R. Caorsi**: Novartis, 2, Sobi, 2; **A. Horne**: None; **G. Filocamo**: Sobi, 2; **H. Wittkowski**: None; **M. Jelusic**: None; **J. Anton**: AbbVie/Abbott, 5, Amgen, 5, Bristol-Myers Squibb(BMS), 5, Eli Lilly, 5, GlaxoSmithKlein(GSK), 2, 5, 6, Novartis, 2, 5, 6, Novimmune, 2, 5, 6, Pfizer, 2, 5, 6, Roche, 5, Sanofi, 5, Sobi, 2, 5, 6; **S. Khaldi-Plassart**: None; **A. Belot**: Novartis, 2, Pfizer, 2, Roche, 2, Sobi, 2; **G. Horneff**: AbbVie/Abbott, 6, Chugai, 6, Eli Lilly, 6, Merck/MSD, 5, Novartis, 5, 6, Pfizer, 6, Roche, 5, Sanofi, 6; **S. Palmer Sarrott**: None; **E. Cannizzaro Schneider**: None; **L. Fotis**: None; **P. Dolezalova**: None; **A. Ravelli**: AbbVie/Abbott, 6, Alexion, 6, Angelini, 6, Bristol-Myers Squibb(BMS), 6, Novartis, 6, Pfizer, 6, Reckitt Benckiser, 6, Roche, 6, Sobi, 6; **S. Ozen**: Novartis, 2, 6, Sobi, 2, 6; **F. De Benedetti**: AbbVie/Abbott, 2, Novartis, 2, Novimmune, 2, Pfizer, 2, Roche, 2, Sobi, 2.

Abstract Number: 096

DADA2 – a Case Series from North India

Manjari Agarwal, Jyotsna verma, ratna puri and Sujata Sawhney, Sir Ganga Ram Hospital, New Delhi, India

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: We present a series of children previously diagnosed and managed as Polyarteritis nodosa at our unit. Due to ease of availability of mutation analysis and ADA2 enzyme assay, we were able to give a confirmatory etiological diagnosis to some of these children. Aims: To study in detail children recently diagnosed as DADA2 at our unit

Methods: A retrospective review was done of all children presenting as polyarteritis nodosa who were later confirmed as DADA2 on genetic analysis.

Results: So far 10 children have been confirmed as DADA2 at our unit. 5 boys and 5 girls formed our cohort.All these children belong to Agarwal community originating from North India.

Table 1: Clinic profile

Clinical features	
Fever Pain abdomen Hypertension Stroke Nerve palsy Livedo Headache2	6/10 9/10 8/10 1/10 2/10 10/10 2/10
CT angiography abdomen demonstrating aneurysms	7/10
Renal scarring	5/10
Initial treatment Steroids Cyclophosphamide IVIG Azathioprine	10/10 7/10 2/10 8/10
Genetic analysis ADA2 gene exon 2 homozygous for variant c.139G A (p.Gly47Arg)	9 children homozygous 1 child is compound heterozygous

Median age at first presentation was 6 years(IQR 1.25-10). Most common presentation was fever, pain in abdomen and livedoid rash. ADA2 enzyme assay was available on 8 children, 2 had 0% activity of ADA2 in comparison to total ADA. Remaining 6 also had low activity of ADA 2 to less than 12% as compared to 40% in age matched control samples. 2 children succumbed after diagnosis while being treated at different centres. TNF inhibitors have been initiated for 8 children.

Conclusion: DADA2 may present as varied clinical scenarios The pain in abdomen and fever subsided after corticosteroid therapy in all children but the livedo and nodular rash persisted or reappeared on tapering steroids. A high index of suspicion is needed. Livedoid rash in a young child in the absence of antiphospholipid antibody is an important clinical clue that was present in all these children. Early diagnosis and treatment might prevent institution of therapy with conventional disease modifying agents. This is an initial data set and these children shall be followed up longitudinally.

Disclosure: M. Agarwal: None; **J. verma**: None; **r. puri**: None; **S. Sawhney**: None.

Abstract Number: 097

Survey of Covid-19 Immunization and Infection in Patients with Systemic Juvenile Idiopathic Arthritis and Adult Onset Still's Disease

Mariana Correia Marques[1], Paul Subrata[2], Carol Lake[3], Ly-Lan Bergeron[4], Rashmi Sinha[5], Luciana Peixoto[6], Marinka Twilt[7] and Michael Ombrello[8], [1]Translational Genetics and Genomics Section, National Institute of Arthritis and Musculoskeletal and Skin Diseases/Children's National Hospital, Bethesda, MD, [2]NIAID Collaborative Bioinformatics Resource (NCBR), National Institute of Allergy and Infectious Diseases, National Institutes of Health, Bethesda, MD, [3]NIH, GAITHERSBURG, MD, [4]NIH/NIAMS, Vienna, VA, [5]Systemic JIA Foundation, Cincinnati, OH, [6]Systemic JIA Foundation, Cincinnati, [7]Alberta Children's Hospital, Calgary, AB, Canada, [8]Translational Genetics and Genomics Section, National Institute of Arthritis and Musculoskeletal and Skin Diseases, National Institutes of Health, North Bethesda, MD

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Immunization is one of the most important tools for the control of the Covid-19 pandemic. The safety and effectiveness of the Covid-19 immunizations have been established for the general population and in patients with autoimmune and autoinflammatory conditions. However, given it's rarity, patients with systemic juvenile idiopathic arthritis (sJIA) and adult onset Still's disease (AOSD) have been under-represented in these studies. Therefore, we performed a survey with patients with sJIA and AOSD to inquire about the safety and tolerability of the Covid-19 immunization in this population.

Methods: We performed an anonymous online survey on closed Facebook groups for patients and parents with sJIA and AOSD from June 27th until August 30th, 2022 on the SurveyMonkey platform. It consisted of 27 close-ended questions in English, with a space for further comments. Inclusion criteria included a diagnosis of sJIA or AOSD self-reported by the respondent. Consent was obtained at the beginning of the survey.

Results: Of a total of 167 responses, 16 were excluded (2 respondents were 18 years old,14 failed to complete the survey). Table 1 and Table 2 show the demographics of the cohort, divided by vaccination status. Most patients received the Pfizer/Biontech vaccine (88%), followed by Moderna (18%) and Oxford/Astra Zeneca (6%). Fifty three percent of patients had inactive and 38% of patients had active disease at the time of the immunization, and 4% had the vaccine prior to the diagnosis. Twelve patients paused medications for the immunization. Thirty-nine patients reported vaccine side effects (Table 2). The most commonly listed reasons for patients to not receive the Covid-19 vaccine were concern about potential side effects from the vaccine related to sJIA/AOSD (58%) and not related to sJIA/AOSD diagnosis (46%), vaccine unavailable for age group/location (26%), medical provider recommendation to not receive the vaccine (12%), concern about potential interaction of the vaccine with patient's medications including loss of effectiveness (11%) and allergy to a vaccine component (6%). Forty-seven (48%) immunized patients tested positive for Covid-19 at some point, compared with 33 (66%) non-immunized patients. Most patients had asymptomatic (18%), or mild/moderate (58%) symptoms. However, one immunized patient required hospitalization in a general ward, one non-immunized patient required ICU admission for the Covid-19 infection.

Table 1. Demographics

	Unvaccinated (n=51)	Vaccinated (n=100)
Age (years)	8 (4 - 9.5)***	12 (8-18)***
Age of onset (years)	2 (1-4.5)***	6.5 (2-12.25)***
Female Sex	29 (57%)***	80 (80%)***
Country		
USA	39 (76%)	74 (74%)
United Kingdom	1 (2%)	8 (8%)
Canada	1 (2%)	7 (7%)
Australia	3 (6%)	6 (6%)
Other	7 (14%)	4 (4%)
Diagnosis		
sJIA	50 (98%)	82 (82%)
AOSD	1 (1%)	18 (18%)
History of complications		
MAS	25 (49%)	50 (50%)
Lung Disease	15 (29%)	18 (18%)
Arthritis	26 (50%)	51 (51%)
Pericarditis/myocarditis	4 (7%)	10 (10%)
History of Disease flare or severe side effects with other immunizations	12 (24%)***	4 (4%)***
Total number of medications ever used beyond NSAIDs	5.940*	4.75*

*p ≤ 0.05, ** p ≤ 0.01, ***p ≤ 0.001. MAS: macrophage activation syndrome, sJIA: systemic juvenile idiopathic arthritis, AOSD: adult-onset Still's disease, NSAIDs: non-steroidal anti-inflammatory drugs

Table 2. Medications ever used to treat sJIA/AOSD

Medications ever used	Unvaccinated (n=51)	Vaccinated (n=100)
NSAIDS	39 (76%)	84 (84%)
Steroids		
Oral	46 (90%)	89 (89%)
IV	35 (68%)	59 (59%)
IL-1 inhibitors		
Anakinra	41 (80%)	72 (72%)
Canakinumab	29 (56%)	51 (51%)
Rilonacept	2 (3%)	0 (0%)
Tocilizumab	33 (64%)	46 (46%)
Methotrexate	23 (45%)	46 (46%)
JAK inhibitors total	32 (62%)	27 (27%)
Baracitinib	0	2 (%)
Tofacitinib	20 (39%)	17 (17%)
Other JAKi	12 (23%)	8 (8%)
Cyclosporin	14 (27%)	26 (26%)
Tacrolimus	13 (25%)	11 (11%)
Abatacept	6 (11%)	9 (9%)
TNF inhibitors	13 (25%)	16 (31%)
Adalimumab	5 (10%)	9 (9%)
Etanercept	5 (10%)	3 (3%)
Infliximab	3 (6%)	4 (4%)
Emapalumab	10 (20%)	2 (2%)
Cyclophosphamide	2 (4%)	3 (3%)
Azathioprine	0 (0%)	2 (2%)
Bone marrow transplant	1 (2%)	1 (1%)
IL-18 inhibitor	0 (0%)	1 (1%)
Hydroxychloroquine	1 (2%)	4 (4%)
Thalidomide/lenalinomide	0 (0%)	2 (2%)
Leflunomide	0 (0%)	3 (3%)

Table 3. Characteristics of vaccine side effects

	n=39
Type of reaction	
Local symptoms at injection site)	25
Fever	20
Chills	12
Fatigue/ tiredness	29
Muscle pains	17
Headache	19
Nausea	6
Lymph node swelling	8
Rashes	5
Myocarditis (inflammation of the heart)	0
Disease flare	3
MIS-C/MIS-A	1
Other (please specify)	7
Atrial fibrillation	1
Pericarditis	1
Delayed flare	1
Flare of Crohn's disease	1
Fever and URI symptoms	1
Food sensitivities	1
Coughing	1
Duration of reaction	
≤2 days	16
3-7 days	15
≥ 8 days	7
Highest level of care required	
No medical care required	23
Called physician	9
Visited physician in clinic	3
Emergency department/urgent care	2
Hospital admission	1
Intensive care unit admission	0

Another non-immunized patient died of severe disease flare after the Covid-19 infection. There was a trend toward higher risk of disease flare after Covid-19 infection among non-immunized patients (43%), compared to immunized patients (25%), but it was not statistically significant.

Conclusion: The Covid-19 vaccine was well tolerated by sJIA and AOSD patients even in this group of patients with high incidence of macrophage activation syndrome and lung disease. Most vaccine side effects were mild and lasted less than 7 days. There was a low incidence of disease flare with immunization (3%). There was a trend towards lower risk of flare with the Covid-19 infection in the immunized group. The only ICU admission and only death occurred in unvaccinated subjects.

Disclosure: M. Correia Marques: None; **P. Subrata**: None; **C. Lake**: None; **L. Bergeron**: None; **R. Sinha**: None; **L. Peixoto**: None; **M. Twilt**: None; **M. Ombrello**: None.

Abstract Number: 098

Characterization of Pulmonary Nodules in Juvenile-onset Systemic Sclerosis: A Single Center Case-Series

Jonathan Li[1], Franziska Rosser[1], Sameh Tadros[1] and Kathryn Torok[2], [1]UPMC Children's Hospital of Pittsburgh, Pittsburgh, PA, [2]University of Pittsburgh, Pittsburgh, PA

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Given the high prevalence of intestinal lung disease (ILD) in systemic sclerosis (SSc) and the low sensitivity of pulmonary function testing for diagnosing ILD, current guidelines recommend ILD-screening with high resolution computed topography (HRCT) at diagnosis. Although HRCT is critical for appropriate ILD diagnosis and management, these screenings may lead to the discovery of pulmonary nodules, an infrequently described feature of SSc. Unlike adults, incidental pulmonary nodules in children produce a clinical dilemma as there are no evidence-based management guidelines and they rarely represent malignancy. Children with Juvenile-onset SSc (jSSc) have risk factors for pulmonary inflammation and therefore nodule development, such as increased prevalence of esophageal dysfunction (e.g. reflux aspiration), usage of immunosuppression (e.g. infection), and systemic inflammation (e.g. early ILD), although the prevalence of nodules in jSSc is unknown. We sought to evaluate the prevalence and characteristics of pulmonary nodules in jSSc.

Methods: Data was curated in January 2022 from the National Registry for Childhood Onset Scleroderma (IRB PRO11060222), a prospective research registry of patients with jSSc evaluated at a single tertiary care institution with an associated multidisciplinary jSSc clinic. All patients fulfilled the 2013 ACR/EULAR criteria for SSc. Demographic, serologic,

Table 1: Select characteristics of juvenile systemic sclerosis registry participants with at least one CT scan (N=32)

Clinical characteristics	No Nodules	Nodule(s)	P value
N	20	12	
Age of JSSc-onset, years	9.8 (±4.0)	9.0 (±3.5)	0.49
Age at initial CT or nodule diagnosis, years*	13.6 ± 3.9	10.8 (±3.3)	**0.047**
Time from jSSc onset to nodule diagnosis, years	3.8 (± 3.1)	1.8 (±1.7)	**0.04**
Diagnosed with jSSc-overlap disease	9 (40)	7 (58)	0.31
Diagnosed with interstitial lung disease	2 (10)	5 (42)	0.07
Any abnormal finding on CT chest, yes	2 (10)	12 (100)	**<0.001**
Infectious disease evaluation at time of initial nodule(s)	1 (5)	1 (5)	0.40
Serologies			
ANA positive	19 (95)	9 (75)	0.10
Anti-Scl-70 positive	7 (35)	3 (25)	0.70
Anti-PM-Scl positive	4 (20)	4 (33)	0.43
Anti-centromere positive	1 (5)	1 (8)	1.00
Anti-RNA Pol III positive	0 (0)	1 (8)	0.38
Anti-U1/U3RNP positive	2 (10)	1 (8)	1.00
Anti-histone positive	2 (10)	0 (0)	0.52
Antibody negative	3 (25)	7 (35)	0.70
Gastrointestinal disease features			
Structural esophageal abnormalities	2 (10)	3 (25)	0.34
GERD	14 (70)	7 (58)	0.70
Upper GI dysmotility	8 (40)	4 (33)	1.00
On PPI or H2B therapy	11 (55)	4 (33)	0.29
Soft tissue and vascular disease features			
Raynaud's	14 (70)	11 (92)	0.15
Nailfold capillary abnormalities	13 (65)	10 (83)	0.26
Digital ulceration/pitting, yes	9 (45)	6 (50)	0.78
Sclerodactyly	7 (35)	9 (75)	**0.03**
Clubbing	0 (0)	1 (8)	0.38
Myositis	0 (0)	2 (17)	0.13
Modified rodman skin score (mRSS)	6.4 (±9.8)	7.0 (±7.8)	0.28
DMARD at time of CT			
On any DMARD at time of nodule diagnosis	16 (80)	6 (50)	0.08
Mycophenolate mofetil	10 (50)	3 (25)	0.27
Methotrexate	6 (30)	3 (25)	1.000
Prednisone	9 (45)	2 (17)	0.14
Cyclosporine	3 (15)	0 (0)	0.27
Hydroxychloroquine	10 (50)	3 (25)	0.27
Intravenous Immune Globulin	2 (10)	1 (8)	1.000
Rituximab	2 (10)	1 (8)	1.000
Tocilizumab	1 (5)	0 (0)	1.000

Results displayed as N (%) or mean (SD). P value obtained from X^2 or Fischer's Exact Test (where appropriate), or Wilcoxon rank sum. **Bold** = p value < 0.05.
* Age of initial chest computed tomography (CT) for those without nodules and age of initial CT chest demonstrating nodules for those found to have nodules.

radiological, clinical, and laboratory data were extracted. Chest HRCTs and/or reports were reviewed by a pediatric pulmonologist and radiologist. Bivariate analysis was used to compare select measures between children with and without nodules on imaging (significance defined a $p < 0.05$).

Results: Of 40 children enrolled in the registry, 32 children received a HRCT chest and 15 had reports of nodule(s). Independent review confirmed nodules in 12 (38%) of 32 children. Most had > 1 nodule and at least one follow-up HRCT. Bivariate analysis results are reported in **Table 1**. Those with nodules were younger at initial imaging, had less time between diagnosis and onset of symptoms, and higher prevalence of sclerodactyly. More children with nodules had ILD and decreased usage of disease-modifying anti-rheumatic drugs compared to children without nodules, although neither was statistically significant. No association with gastrointestinal features were observed. Most nodules were < 5mm in size and stable over time. No nodules were biopsied.

Conclusion: Pulmonary nodules may be a more common pulmonary feature as over one third of jSSc patients in a tertiary referral center registry revealed nodule(s) on HRCT chest. Most nodules were small and demonstrated stability over time. The etiology of the pulmonary nodules is unknown however may be related to systemic inflammation given the association with younger age, earlier symptoms, and sclerodactyly. Larger cohort studies are needed to better characterize the prevalence and natural course of pulmonary nodules in jSSc, including the association with ILD.

Disclosure: J. Li: None; **F. Rosser**: None; **S. Tadros**: None; **K. Torok**: None.

Abstract Number: 099

The Impact of the COVID-19 Pandemic on Juvenile Dermatomyositis

Tresa Ambooken[1], Sangati Kadakia[1], Tara Lozy[1], Brianna Bulbin[2], Suhas Ganguli[3], Dawn Wahezi[4] and Sivia Lapidus[1], [1]Department of Pediatrics, Joseph M. Sanzari Children's Hospital, Hackensack University Medical Center, Hackensack, NJ, [2]Hackensack Meridian School of Medicine, Hackensack, NJ, [3]Department of Pediatrics, K. Hovnanian Children's Hospital, Jersey Shore University Medical Center, Neptune City, NJ, [4]Children's Hospital at Montefiore, New York, NY

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

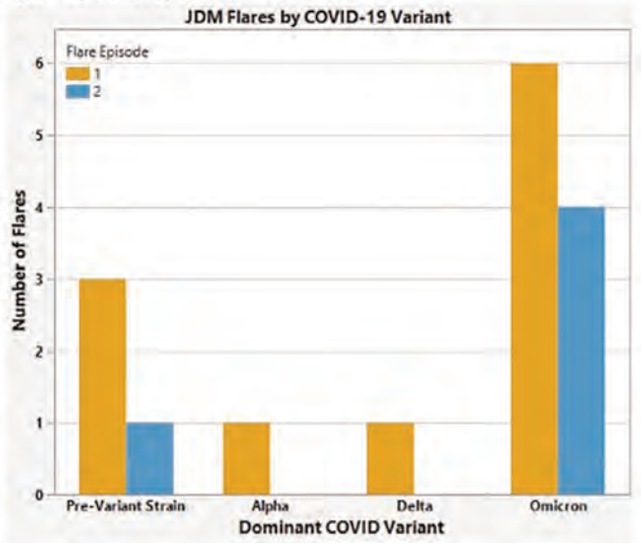
Background/Purpose: Juvenile Dermatomyositis (JDM) is theorized to occur in a genetically susceptible individual as a response to an environmental trigger, leading to autoimmunity and inflammation. Several viruses have been linked to the underlying pathophysiology of JDM, with few reports of SARS-CoV-2 infection associated with both initial episodes and flares of JDM around the world. In this study, we sought to investigate the impact of the COVID-19 pandemic on our population of JDM patients and evaluate the number of new JDM diagnoses and JDM flares during the pandemic, as compared to 5 years pre-pandemic.

Methods: Biomedical Informatics identified patients 21 years and younger using the ICD-10 code M33, who were diagnosed between June 2015 and December 2022. Data were collected retrospectively comparing manifestations of JDM patients initial presentations and flares pre-pandemic (6/1/15-2/28/20) as well as during the pandemic (3/1/20-12/30/22). Information about known COVID-19 exposures and infections preceding a flare or initial diagnosis was assessed. Flare episodes were characterized by clinical symptoms, physical exam findings, pertinent labs, and medications at the time of flare. Exploratory data analysis was used to explore potential relationships between flares occurring before and during the pandemic using summary statistics, univariate and bivariate analysis.

Table 1: JDM Patient Flares

	JDM Patient Flares	
	JDM Diagnosis Pre Pandemic (n=9)	JDM Diagnosis Pandemic (n=6)
Timing of Flare		
Pre Pandemic	2 (22.2)	N/A
Pandemic	7 (77.8)	6 (100.0)

Figure 1: JDM Flares By COVID-19 Variant



Results: Seventeen patients diagnosed with JDM were identified; 8 pre-pandemic and 9 during the pandemic. Fifteen flares were captured from 12 patients, of which 87% (13/15) occurred during the pandemic (Table 1). Of those JDM patients diagnosed pre-pandemic, 78% of their flares occurred during the pandemic. Of the 12 patients who experienced flares, 80% of these patients experienced at least one flare, while 25% experienced more than one flare during the pandemic. Fifteen percent of patients who experienced flare had a medication (Methotrexate) held for COVID-19 vaccination; 55% of our patients were vaccinated. Two patients had documented COVID-19 infections preceding flare. The majority of flares occurred during the time period when Omicron variants of COVID-19 were predominant (12/1/21 to 12/30/2022) (Figure 1).

Conclusion: The majority of flares in this study period occurred during the pandemic. Although a minority of patients had documented COVID-19 infection preceding flare, most flares occurred during the time when Omicron variants were predominant; therefore, these patients may have had undocumented or asymptomatic COVID-19 infections that potentially triggered flares. Holding immunomodulating medications to optimize immune response to COVID-19 vaccination may have also potentially contributed to flares. An increase in telehealth visits during the pandemic may have also led to suboptimal monitoring and disease control in JDM potentially modulating the number of JDM flares during the pandemic. Future investigation of the COVID-19s impact in larger cohorts would elucidate correlations between the pandemic, COVID-19 infections, and vaccinations with JDM flares.

Disclosure: T. Ambooken: None; **S. Kadakia**: None; **T. Lozy**: None; **B. Bulbin**: None; **S. Ganguli**: None; **D. Wahezi**: None; **S. Lapidus**: None.

Abstract Number: 100

The Brazilian Registry of Juvenile Dermatomyositis (JDM): II – A Longitudinal Assessment of Muscle Strength by Manual Muscle Test (MMT) and Childhood Myositis Assessment Scale (CMAS) Tools

Darcisio Antonio[1], **Taciana Fernandes**[1], Adriana Elias[2], Teresa Robazzi[3], Ana Julia Moraes[4], Sheila Oliveira[5], Flavio Sztajnbok[6], Luciana Carvalho[7], Luciana Marques[8], Silvana Sacchetti[9], Maria Teresa Terreri[10], Simone Appenzelle[11], Roberto Marini[12], Carlos Rabello Jr[13], Cristina Magalhaes[14], Melissa Fraga[15], Marcia Bandeira[16], Iloite Scheibel[17], Isabela Daud[2], Beatriz Carneiro[2], Claudio Len[18], Clovis Silva[19] and Claudia Magalhaes[20], [1]Universidade Estadual Paulista (UNESP) Botucatu, Brazil, [2]Instituto da Criança - Universidade de São Paulo (USP), São Paulo, Brazil, [3]Universidade Federal da Bahia, Brazil, [4]Universidade Federal do Para, Brazil, [5]Universidade Federal do Rio de janeiro, Rio de Janeiro, [6]Universidade Estadual do Rio de Janeiro, Rio de Janeiro, Brazil, [7]Universidade de Sao Paulo- Ribeirao Preto, São Paulo, Brazil, [8]Hospital Albert Sabin, Brazil, [9]Santa Casa de Sao Paulo, Brazil, [10]UNIFESP, São Paulo, Brazil, [11]University of Campinas, Campinas, Sao Paulo, Brazil, [12]UNICAMP, São Paulo, Brazil, [13]Hospital Geral de Fortaleza, Brazil, [14]Hospital Jose de Alencar - Brasilia, Brazil, [15]Hospital Darcy Vargas, Brazil, [16]Hospital Pequeno Principe- Curitiba, Brazil, [17]Hospital Conceição de Porto Alegre, Brazil, [18]Universidade Federal de São Paulo - Unifesp, São Paulo, Brazil, [19]Universidade de São Paulo, São Paulo, Brazil, [20]São Paulo State University, Pediatric Rheumatology Division, Botucatu, Brazil

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Posters: Clinical and Therapeutic II

Session Type: Poster Session B

Session Time: 5:00PM–6:00PM

Background/Purpose: Muscle weakness is often progressive and persistent in Juvenile Dermatomyositis (JDM). Muscle strength testing is useful for evaluating severity of muscle weakness. There is age limitation for testing muscle strength; the Manual Muscle Test (MMT) and the Childhood Myositis Assessment Scale (CMAS) were selected as myositis core set of outcome measures in adults and children (Miller FW Rheumatology 2001, 40:1262) (Ruperto N Rheumatology 2003, 42: 1452). Both tests were carried out according to the Brazilian Registry of JDM protocol. Muscle strength scored by MMT and CMAS in newly diagnosed JDMwas compared, at onset and follow up for 2 years .

Methods: All 96 patients selected in the JDM registry with diagnoses certified by the attending physician were elicited to perform the tests according to age and willing to participate, under supervision of a physician or physiotherapist; at baseline, 6, 12, 18 and 24 month follow up. The Manual Muscle Test (MMT) (Rider LG Arthritis Care Res 2010; 62:465) version was developed based on Kendall method using a summary of 8 muscle groups and a potential range of 0-80, being 1 axial, 5 proximal (2 upper extremity and 3 lower extremity) and 2 distal muscles (upper and lower extremity). Childhood Myositis Assessment Scale (CMAS) (Lovell DJ Arthritis Rheum 1999, 42: 2213-9) is composed by 14 manoeuvres testing proximal muscle strength and the version of 0 to 53 - final score was selected (Huber A Arthritis Care Res 2014, 66:648). Attending physicians were trained by a video performance (recorded by Dr R Rennebohn). The scores were compared by Poisson model and Wald test for repeated measures with significance set at 5% or p 0.05.

Results: The number (n) of performed tests was variable for each of the longitudinal assessments. MMT (mean± SD) scores were: Baseline (n=92) (37.5±34.6)*, 6m (n= 60) (58.5±29)*, 12m (n=44) (66.6±23.7), 18m (n= 26) (72.8±16.7) and 24m (n=17)(74.8±7.8), the difference among visits was significant with p=0.015*; and the only significant difference (Wald test) was from the baseline to six months. CMAS test scores had no significant variation between visits. The mean± SD scores of CMAS scores (0-53), were Baseline (n=60) (29.5±11.4), 6m (n=51) (32.3±11.4), 12m (n=41) (34.2±5.8), 18m (n=23) (34 ± 6) and 24 m (n=15) (33.3±5.4) p = 0.06 (NS).

Conclusion: We tested the feasibility and validity of MMT and CMAS performed by the attending physician or physiotherapist in a national JDM registry . Discontinued follow up was the main limitation. There was marked improvement in MMT scores during the first 6 months, compared to only mild improvement of CMAS, that persisted stable with moderate weakness indicating persistent functional impairment up to 2 years.

Disclosure: D. Antonio: None; **T. Fernandes**: None; **A. Elias**: None; **T. Robazzi**: None; **A. Moraes**: None; **S. Oliveira**: None; **F. Sztajnbok**: None; **L. Carvalho**: None; **L. Marques**: None; **S. Sacchetti**: None; **M. Terreri**: None; **S. Appenzelle**: None; **R. Marini**: None; **C. Rabello Jr**: None; **C. Magalhaes**: None; **M. Fraga**: None; **M. Bandeira**: None; **I. Scheibel**: None; **I. Daud**: None; **B. Carneiro**: None; **C. Len**: None; **C. Silva**: None; **C. Magalhaes**: None.

Abstract Number: 101

Differences in Clinical and Patient-reported Outcomes in Juvenile Dermatomyositis by Race and Ethnicity

Susan Kim[1], **Rebecca Olveda**[2] and Jessica Neely[2], [1]UCSF Benioff Children's Hospital, San Francisco, CA, [2]UCSF, San Francisco, CA

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Table 1: Baseline demographic characteristics by race and ethnicity

Variable	n	All (n = 219)	Black, African American, African, or Afro Caribbean (n = 25)	Hispanic, Latino, or Spanish origin (n = 44)	White (n = 150)	p-value
Age at diagnosis, y, median (IQR)	216	8.1 (4.5 – 12.0)	10.2 (7.8 – 13.8) n = 25	8.6 (5.4 – 13.8) n = 44	7.6 (4.1 – 11.3) n = 147	0.01
Female sex, % (n)	219	67.1 (147)	76 (19)	75 (33)	63.3 (95)	0.21
Median income by US zip code (US $, IQR)	217	84,506 (64,505 – 111,599)	63,770 (50,274 – 82,192)	80,099 (60,037 – 103,189)	88,250 (68,589 – 114,881)	0.0009
Highest Parental/Guardian Education, % (n)	185		19	38	128	0.000
Graduated high school or less		27.6 (51)	31.6 (6)	52.6 (20)	19.5 (25)	
Some college or more		72.4 (134)	68.4 (13)	47.4 (18)	80.5 (103)	
Insurance type, % (n)	214		25	43	146	0.000
Public insurance		29.9 (64)	44.0 (11)	60.5 (26)	18.5 (27)	
Private insurance		67.3 (144)	52.0 (13)	37.2 (16)	78.8 (115)	
No insurance		2.8 (6)	4.0 (1)	2.3 (1)	2.7 (4)	

Background/Purpose: Previous studies in juvenile dermatomyositis (JDM) have shown that patients from minoritized ethnicities and those with lower family income are more likely to have worse clinical outcomes. Patient-reported outcome measures (PROs) are valuable indicators of disease activity but are less well-studied in JDM. This study aimed to investigate differences in clinical outcomes and PROs based on race and ethnicity at baseline enrollment, in a cohort of patients enrolled in the Childhood Arthritis and Rheumatology Research Alliance (CARRA) registry.

Methods: This was a retrospective cohort study using data from children diagnosed with JDM and enrolled into the CARRA Registry between February 2018 and November 2021, for whom data regarding race and ethnicity were available. Participants without valid US zip codes were excluded. The primary predictor was self-identified race and/or ethnicity. The primary

Table 2: Baseline disease characteristics by race and ethnicity

	n	All (n = 219)	Black, African American, African, or Afro Caribbean (n = 25)	Hispanic, Latino, or Spanish origin (n = 44)	White (n = 150)	p-value
Clinical Characteristics						
Time to diagnosis, months, median (IQR)	194	3 (1 – 6)	2 (1 – 6)	2.5 (1 – 5)	3 (1 – 6)	0.70
Physician global assessment, median (IQR)	181	4 (2 – 5)	5 (3 – 6)	4 (2.75 – 6)	3 (2 – 5)	0.03
Physician global assessment* > 2, % (n)	181	71.3 (129)	81.0 (17)	80.6 (29)	66.9 (83)	0.18
Physician global assessment of skin disease activity > 2, % (n)	123	56.9 (70)	61.5 (8)	50.0 (15)	58.8 (47)	0.67
Proximal muscle weakness, % (n)	214	75.2 (161)	91.7 (22)	88.1 (37)	68.9 (102)	0.005
Presence of elevated muscle enzymes, % (n)	208	76.9 (160)	91.3 (21)	86.1 (37)	71.8 (102)	0.03
Calcinosis ever, % (n)	211	6.2 (13)	4.4 (1)	4.6 (2)	6.9 (10)	0.90
Baseline steroid use, % (n)	182	80.8 (147)	82.6 (19)	86.1 (31)	78.9 (97)	0.66
Patient-reported outcomes						
Patient/parent global assessment, median (IQR)	198	3 (1 – 5)	4 (2 – 7)	3 (1 – 7)	2 (0 – 4)	0.05
Patient/parent global assessment > 2*, % (n)	198	51.5 (102)	73.9 (17)	57.9 (22)	46.0 (63)	0.03
CHAQ[i], median (IQR)	190	0.67 (0 – 1.63)	1.51 (0.5 – 2.25)	1 (0.125 – 1.9)	0.38 (0 – 1.37)	0.006
Pain interference[ia], mean ± SD	112	14.1 ± 8.7	18.5 ± 9.2	12.3 ± 9.0	13.9 ± 8.3	0.11
Physical function[ib] – mobility, median (IQR)	163	25 (15 – 32)	18 (8 – 26)	25 (13 – 29)	27 (17 – 32)	0.03
Physical function[ic] – upper extremity, median (IQR)	146	27 (12 – 32)	22 (6 – 31)	27.5 (19 – 31)	27 (12 – 32)	0.60
Global health assessment (IQR)	167	23 (19 – 25)	22 (16 – 24)	21 (18 – 24)	23 (20 – 25)	0.04

Table 3: Unadjusted logistic regression of patient/parent global assessment > 2 at baseline

Race and ethnicity	Odds ratio	95% confidence interval	p-value
White	1.00 (Reference)	--	--
Black	3.33	1.24 – 9.00	0.02
Hispanic	1.62	0.78 – 3.34	0.20

outcome was the Patient/Parent Global Assessment Score (PGA) as a dichotomous variable of score < /= 2 (lower disease activity) or > 2 (higher disease activity). Secondary outcomes included proximal muscle weakness, elevated muscle enzymes, and physician global assessment of skin disease activity. Descriptive statistics were used to summarize each variable, and demographic and baseline disease activity measures were evaluated across racial and ethnic categories using Chi-squared, Kruskal-Wallis, and one-way ANOVA tests. Demographic variables with significant association with race and ethnicity were included in a multivariate model analyzing PGA score greater than 2 at baseline.

Results: 219 participants identified as Black (11.4%), Hispanic (20.1%), or White (68.5%). Demographic characteristics are listed in table 1. Median income by US zip code was lowest among Black participants, while White participants had the highest median income (p = 0.04) (Table 1). Black and Hispanic participants were more likely to have proximal muscle weakness (p = 0.005), elevated muscle enzymes (p = 0.03), and higher physician global assessment scores indicating worse disease activity (p = 0.03) compared to White participants (Table 2). Black participants had 3.3 times the odds of having a PGA > 2 at baseline compared to White participants (95% CI 1.2 to 9.0, p = 0.02). After adjusting for age at diagnosis, sex, income, education, and insurance status, this association was no longer statistically significant, however, those with private insurance had 74.6% lower odds of having a PGA > 2 compared to those with public insurance.

Conclusion: In JDM participants enrolled in the CARRA Registry, Black and Hispanic participants tended to have lower income and education levels. While Black participants had a higher odds of having worse PGA at baseline, this association diminished after adjusting for age at diagnosis, sex, income, education, and insurance status. Our study was limited by a smaller proportion of participants from minoritized racial and ethnic groups. More research is needed to determine how patient outcomes are influenced by social determinants of health, including income, education, and the effects of systemic racism, in order to inform future interventions to improve health disparities

Disclosure: S. Kim: None; **R. Olveda**: None; **J. Neely**: None.

Abstract Number: 102

Preliminary Results from a Survey of Psychological Resilience Among JIA Patients

Daniella Schocken and Tracy Ting, Cincinnati Children's Hospital Medical Center, Cincinnati, OH

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Psychological resilience – an individuals capacity to adapt in the face of stressors and recover after adverse events – has been linked to a number of outcome measures impacting patient functioning and well-being among populations living with chronic illness. To date, no research has been conducted into the resilience of children and

adolescents with JIA. This study aims to define the level of resilience observed among JIA patients to better characterize this psychological feature in the population using a validated psychometric tool.

Methods: Participants between the ages of 10 and 17 with an established diagnosis of JIA were recruited from an outpatient pediatric rheumatology clinic to complete a series of questionnaires including the Connor-Davidson Resilience Scale (CD-RISC-25) and the Pain and Symptom Assessment Tool (PSAT). Participants parents were additionally requested to complete their own CD-RISC-25 and provide information regarding recent participant educational achievement and psychiatric health history. Study staff conducted chart review to confirm participants JIA subtype, duration of disease since diagnosis, treatment status, and physical exam findings on the date of study participation. The Pearson correlation coefficient was calculated to detect relationships between CD-RISC-25 scores and other parametric measures collected. Welchs t-test was applied for comparison of CD-RISC-25 scores between subgroups of participants.

Results: Among 52 participants enrolled in the study, CD-RISC-25 scores ranged from 39 to 100 (out of 100 possible points) with a mean of 75.42. Scores on the PSAT consistent with a diagnosis of juvenile FM were present in 13% of the population surveyed, but there was no statistically significant difference between CD-RISC-25 scores among participants with and without evidence of FM ($t = -1.85$, p-value = 0.11). CD-RISC-25 scores also did not differ to a statistically significant degree between participants with and without a history of depression or anxiety ($t = -0.26$, p-value = 0.80). There was no statistically significant correlation detected between CD-RISC-25 score and age at time of JIA diagnosis ($r = 0.75$, p-value = 0.46), time since JIA diagnosis ($r = -0.48$, p-value = 0.63), or parental CD-RISC-25 score ($r = 0.46$, p-value = 0.65).

Conclusion: A convenience sample from a single ambulatory care clinic demonstrated a broad range of psychological resilience levels among JIA patients. No relationships were identified between resilience and duration of disease, age at diagnosis, parental resilience, or a reported history of anxiety or depression. The trend identified in the comparison of resilience among JIA patients with and without clinical features of juvenile FM is compelling and suggests that this preliminary study was not sufficiently powered to capture such a relationship given the relatively low rate of participants reporting features of FM within the data collected. Further study is warranted to determine whether resilience among JIA patients relates to the presence of comorbid juvenile FM or to functional outcomes observed in that patient population.

Disclosure: D. Schocken: None; **T. Ting**: None.

Abstract Number: 103

Clinical, Serologic, and Imaging Findings of Rhupus Syndrome in the Pediatric Population: A Systematic Literature Review

Muriel Velez, Bryan Nicolalde, Kevin Moreno-Montenegro, Gabriela Carolina Carrera-Barriga, Camila Gallegos and Beatriz Leon, Universidad San Francisco de Quito, Quito, Ecuador

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Pediatric Rhupus syndrome is a complex autoimmune disease characterized by overlapping clinical and immunological features of Juvenile Idiopathic Arthritis (JIA) and juvenile Systemic Lupus Erythematosus (jSLE). Pediatric presentation of Rhupus is rare. The need for established clinical and serological diagnostic criteria and the low incidence rate

make Rhupus identification difficult. Through a systematic literature review, we aim to exhibit common clinical and serological characteristics in children presenting this disease.

Methods: We performed a systematic literature review of clinical features of pediatric Rhupus syndrome through Pubmed, Scopus, EBSCO, and Google Scholar following PRISMA guidelines (Figure 1). Our search terms included:((Rhupus) AND (pediatric OR pediatrics OR children OR adolescence OR paediatric OR paediatrics OR child)). Inclusion criteria were: population with an initial manifestation of JIA with simultaneous or subsequent onset of Rhupus syndrome with an age less than 18 years old. Only cross-sectional, retrospective and prospective cohort studies, case reports, and case series studies published between 2000 and 2023 were considered.

Results: Among 564 articles collected using Pubmed, Scopus, Google Scholar, and the EBSCO database, only 13 articles met the inclusion criteria. Just 8 case reports and 5 case series with 26 patients of whom the clinical manifestations, serologic, and imaging findings in pediatric Rhupus patients were recorded. The mean age of patients with Rhupus syndrome was 11 years old, 80% of females and 19% of males (Figure 2). Overall, the first manifestation was articular involvement with the predisposition of erosive patterns, including big articulations. Erosive arthritis was presented in 73% of patients, followed by juxta-articular osteopenia in 21%, chronic synovitis in 15%, joint space narrowing in 10%, and 5% of patients presenting synovial thickening, effusions, and no joint erosions. Ankle arthritis was reported in 65% of patients, followed by arthritis of knees in 50%, wrists in 25%, PIP in 10%, DIP in 10%, and hip involvement in 5%. Of the 26 patients reported, 69% had Rheumatoid factor +, 26% were anti-CCP+, 73% had ANA+, 46% had ds-DNA positive, 19% were anti-Smith+, and 11% were antiphospholipid positive. After being diagnosed with Rhupus Syndrome, a systemic involvement was presented in most patients: 64% had renal involvement, 11% exhibited neurological complications, 11% presented hematologic manifestations, 17% had other sequels, and 23% reported no disease complications. These findings are presented in Table 1.

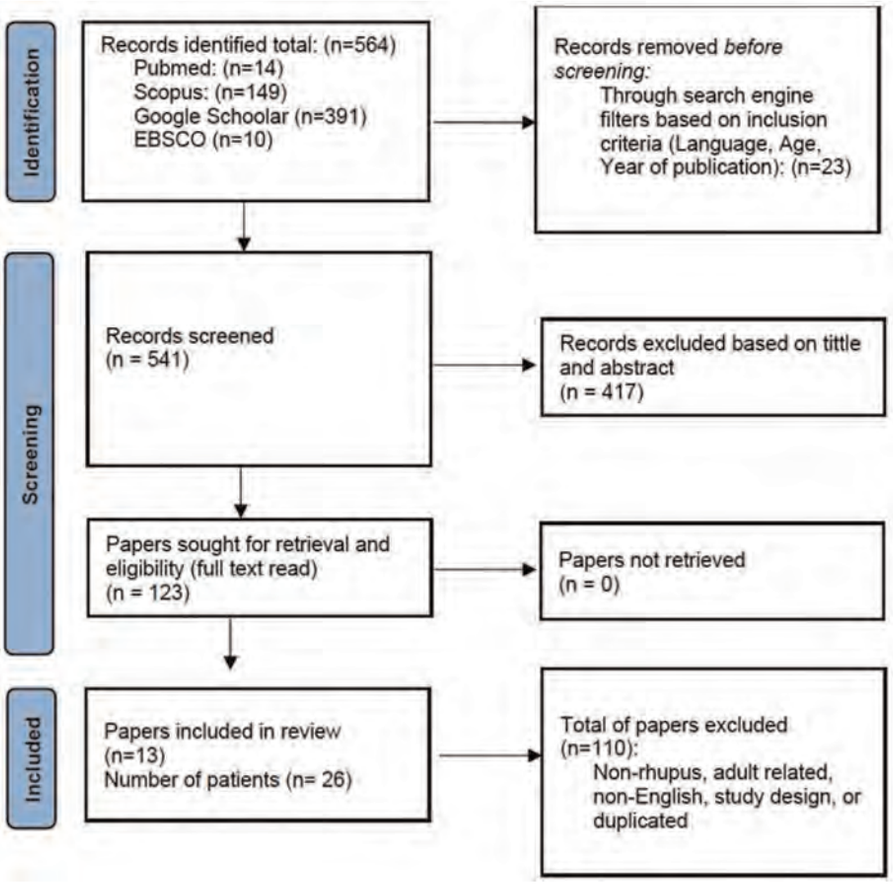


Figure 1. Prisma Flowchart regarding Rhupus diagnostic findings in pediatric patients a Systematic Literature Review

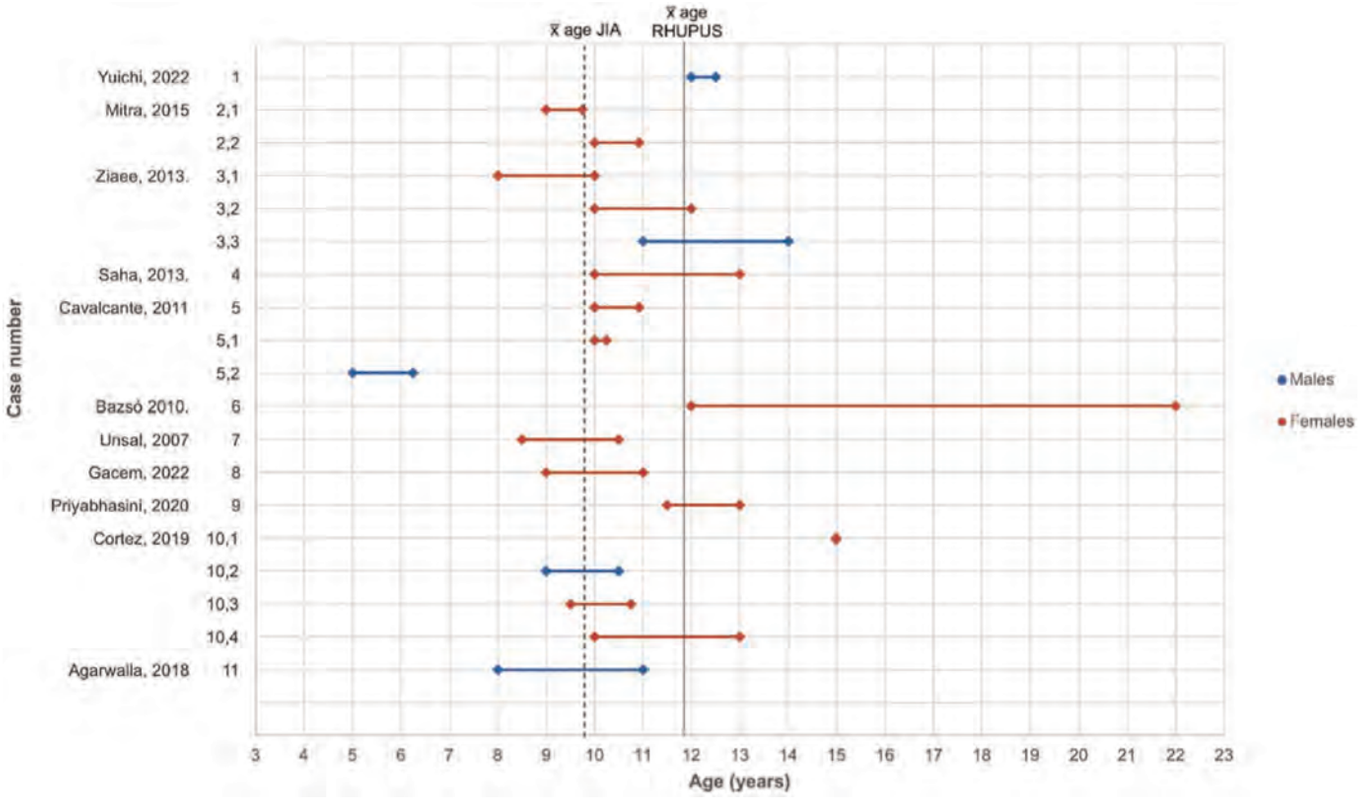


Figure 2. The first diamond shape per case corresponds to the onset of JIA, and the second diamond represents the onset of Rhupus. The dashed line shows the mean age of JIA diagnosis (9 years, 10 months). The full line shows the mean age for the development of Rhupus (10 years, 10 months).

Table 1. Detailed clinical, serology, and imaging findings of pediatric patients with Rhupus

Study	*Patient*	**Serology**	**Imaging findings**	**First join affected**	**Complications**
Yuichi, 2022	1	RF+, anti-CCP+, ANA+ dsDNA+, Anti-Smith +	Chronic inflammation with synovial thickening	Knees, hips, elbows, proximal interphalanged joints	Lupus nephritis class II
Mitra, 2015	2	1. RF+, ANA+, dsDNA+, Coombs+ 2. RF+, ANA+, dsDNA+, Coombs+	1. Chronic synovitis 2.	1 and 2: bilateral deforming polyarthritis in wrist and knees	1. Lupus nephritis class II, lupus myelitis, hemolytic anemia 2. Lupus nephritis grade III
Sakamoto, 2016	6	1. RF+ 2. RF+ 3. RF + 4. RF - 5. RF- 6. RF-	1-6. Erosive arthritis	1-6. Hands and ankles	
Gormezano 2015.*	1	RF+	Erosive arthritis		
Ziaee, 2013.	3	1. RF+, ANA+, dsDNA+ 2. ANA+, ds-DNA+, Direct Coombs+ 3. RF+, ANA+, dsDNA+	1. Erosive arthritis both knees 2. MRI: Erosive arthritis of knees 3. No joint erosions	1. Both knees, left ankle 2. Stiffness in both knees and ankles 3. Swelling of right knee and swelling in the back of the ankle	1. Lupus nephritis (Not described) 2.Heart failure. Recurrent infections, patient dies 3. Non-complication
Saha, 2013.	1	ANA+, dsDNA, anti-CCP+	MRI: knee effusion, mild erosion	Distal interphalangeal joints, knee, ankle joints	Lupus nephritis class V
Cavalcante, 2011	3	1. RF +, ANA+, anti RO+, anti-LA+ 2. RF+, ANA+ 3.RF+, ANA+, anti-SM+, anti-RO	Juxtaarticular osteopenia, joint space narrowing 2. Juxtaarticular osteopenia, erosion 3.Juxtaarticular osteopenia, joint space narrowing, erosions	1. Cervical Spinal, shoulder, elbow, MTC, DIP, bilateral ankle, bilateral tarsal joint, bilateral wrists 2. wrist, MCP, ankle 3. Temporomandibular, PIP, tarsal	1.Lupus nephritis (Not described) 2.Neo-complication 3.Lupus nephritis (Not described)
Bazsó 2010.	1	RF+, ANA+, HLAB27+, antiCCP+	Erosive arthritis	NA	Lupus nephritis class II
Unsal, 2007	1	ANA+,dsDNA+,	Erosive arthritis	Knees	Hashimoto
Gacem, 2022	1	RF+, ANA +, anti-Smith +	Juxtaarticular osteopenia, chronic synovitis of the wrists	Bilateral polyarthritis deformans in wrists, hands, and feet	NA
Priyabhasini, 2020	1	RF+, ANA +, coombs+		Pain, swelling and contracture Bilateral knees, hands, feet, restricted movements	Lupus nephritis class IV
Cortez, 2019	4	1. ANA +, dsDNA +, antiCCP +, anti-Smith +, RF -, anti-Ro -, antiphospholipid + 2. ANA +, dsDNA+, antiCCP +, anti-Smith -, RF+, antiRO -, antiphospholipidic - 3 ANA +, dsDNA+, antiCCP +, anti-Smith +, RF +, antiRO + antiphospholipid + 4. ANA +, dsDNA+, antiCCP +, anti-Smith -, RF +, antiRO -, antiphospholipidic +		1. Oligoarticular 2-3-4. Polyarticular erosive arthritis, morning stiffness, decrease joint range of motion	1.Lupus nephritis (Not described) 2. Non-complication 3. Non-complication 4.Neurological condition non-specified
Agarwalla, 2018	1	ANA+		1. swelling bilateral ankle. Left knee	Lupus nephritis (Not described)

Conclusion: This comprehensive review invites physicians to consider Rhupus Syndrome in patients with erosive arthritis that is unusually seen in jSLE. Despite our extensive systematic review, only a few cases were found which suggests the underreport or underdiagnosis of these cases. We invite more specialists to report the nature of this illness to reach a consensus on diagnosis and management for preventing delayed sequels of this pathology. Close follow-up with extensive clinical, serological, and imaging studies is essential to improve these patients' medical care and quality of life, especially in the setting of erosive arthritis in patients with lupus.

Disclosure: M. Velez: None; **B. Nicolalde**: None; **K. Moreno-Montenegro**: None; **G. Carrera-Barriga**: None; **C. Gallegos**: None; **B. Leon**: None.

Abstract Number: 104

Prevalence of Celiac Disease Among Children and Adolescents with Systemic Lupus Erythematosus (SLE)

Oscar Mwizerwa[1], Andrea Knight[2], Daniela Dominguez[2], Deborah Levy[2], Holly Convery[2], Kendal Thompson[2], Nicholas Gold[2], Catharine Walsh[2] and Linda Hiraki[2], [1]University of Toronto, The Hospital for Sick Children, Toronto, ON, Canada, [2]The Hospital for Sick Children, Toronto, ON, Canada

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Systemic lupus erythematosus (SLE) and celiac disease (CD) are autoimmune diseases characterized by the presence of specific autoantibodies. We aimed to investigate the prevalence of CD in a multiethnic cohort of children and adolescents with SLE.

Methods: We completed a retrospective cohort study of patients under 18 years of age who were diagnosed and followed for SLE at The Hospital for Sick Children between January 2010 and June 2022. We restricted to patients screened for CD with immunoglobulin A antibodies against tissue transglutaminase (anti-tTG-IgA) within one year of SLE diagnosis. We recorded anti-tTG-IgA titers and reviewed medical records for additional investigations, including duodenal biopsy results. CD was confirmed by biopsy according to the Marsh-Oberhuber classification (Marsh ≥3). Demographic and SLE disease features were extracted from the dedicated Lupus database. We calculated the prevalence of positive anti-tTG serology and biopsy-confirmed CD.

Results: CD screening was completed in 93% (374/404) of children diagnosed with SLE. Sixteen (4.2%) had positive anti-tTG-IgA serology (ranging from 31 to 100 U/ml [positive 8U/ml] for Enzyme-linked immunoassay testing and 34 to 4965 CU [positive 30 CU] using Chemiluminescent Immunoassay). 81% (13/16) also had positive anti-endomysial antibodies. Eleven of 16 (68.8%) patients with positive anti-tTG autoantibodies had an endoscopy and duodenal biopsy. Of those biopsied, nine had histopathologic evidence of CD which represents 56% of patients with positive serology, and 2.4% of the screened SLE population. Less than half (45%) of patients with CD had gastrointestinal complaints at screening.

Conclusion: Biopsy-confirmed CD was diagnosed in 2.4% of children and adolescents with SLE, which is higher than the prevalence in the general population of 1.4% by seroprevalence and 0.7% by biopsy. Less than half of patients with biopsy confirmed CD had GI symptoms, highlighting the utility of screening in the childhood SLE population. Future analyses will compare SLE features between those with and without CD.

Disclosure: O. Mwizerwa: None; **A. Knight**: None; **D. Dominguez**: None; **D. Levy**: Janssen, 1, Roche, 5, Sobi, 1, 5; **H. Convery**: None; **K. Thompson**: None; **N. Gold**: None; **C. Walsh**: None; **L. Hiraki**: None.

Abstract Number: 105

Awareness of Multisystem Inflammatory Syndrome in Children Among U.S. Parents: A Cross-Sectional Survey

Lyndsey Cole[1], E. Adrianne Hammershaimb[2], Yuanyuan Liang[2], Megan Hendrich[3], Dhiman Das[3], Robert Petrin[3], James Campbell[2], Sean O'Leary[1] and Jessica Cataldi[1], [1]University of Colorado School of Medicine, Denver, CO, [2]University of Maryland School of Medicine, Baltimore, MD, [3]Ipsos US Public Affairs, Washington, DC

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Little is known about parental awareness of multisystem inflammatory syndrome in children (MIS-C), a rare but severe sequela of SARS-CoV-2 infection. We sought to describe parental knowledge of and attitudes toward MIS-C, identify demographic variables associated with knowledge of MIS-C, and assess associations with parents perceived COVID-19 disease severity and susceptibility in children.

Methods: Via a nationally representative, cross-sectional survey of U.S. parents conducted via Ipsos KnowledgePanel® from October-November 2021, we used bivariate and multivariable analyses to identify correlates of MIS-C awareness and examine associations with perceived COVID-19 severity and susceptibility.

Results: Survey response rate was 64.2% (3,230/5,034). Thirty-two percent of respondents had heard of MIS-C. After adjustment, higher educational level (compared to high school degree, "some college" Odds Ratio [OR]=2.00 [95% confidence interval 1.44,2.77]; "bachelor's degree or higher" OR=3.14 [2.26,4.35]), being a healthcare worker (OR=1.82 [1.37,2.42]), having a child with a chronic medical condition (OR=1.62 [1.22,2.14]), and experience with more severe COVID-19 (OR=1.46 [1.14,1.86]) were associated with MIS-C awareness. Respondents with a child aged 12-17 years were less likely to be aware of MIS-C compared to those without (OR=0.78 [0.63,0.96]), as were male respondents (OR=0.56 [0.46,0.69],) and respondents aged 18-34 years (OR=0.72 [0.54,0.94]) compared to 35-44. Awareness of MIS-C was associated with higher perceived COVID-19 severity and susceptibility (regression coefficients 0.18 [0.10,0.25], $p< 0.001$ and 0.19 [0.11,0.28], $p< 0.001$, respectively).

Conclusion: This survey highlights the need to increase parental awareness of MIS-C. Future studies should explore how education regarding MIS-C as a complication of SARS-CoV-2 infection could improve understanding of pediatric disease severity and susceptibility.

Table 1: Bivariate Analysis of Characteristics Associated with Awareness of MIS-C Among All Respondents

Characteristic		Aware of MIS-C (Weighted %)	Odds Ratio (95% CI)	*P* value
Age of parent (years)	35-44	34	1	
	18-34	26	0.69 (0.54, 0.87)	0.002
	45-54	34	0.97 (0.80, 1.19)	0.80
	55+	30	0.82 (0.58, 1.17)	0.28
Gender	Female	35	1	
	Male	27	0.69 (0.57, 0.82)	<0.001
Age of children				
0-4 years	No	31	1	
	Yes	33	1.13 (0.94, 1.37)	0.12
5-11 years	No	31	1	
	Yes	32	1.03 (0.86, 1.22)	0.77
12-17 years	No	33	1	
	Yes	30	0.84 (0.71, 1.00)	0.05
Race/Ethnicity	White, Non-Hispanic	35	1	
	Black, Non-Hispanic	25	0.60 (0.42, 0.87)	0.007
	Other, Non-Hispanic	35	1.00 (0.70, 1.43)	1
	Hispanic	22	0.53 (0.41, 0.68)	<0.001
	2+ Races, Non-Hispanic	46	1.59 (1.00, 2.53)	0.05
Survey language	English	33	1	
	Spanish	14	0.34 (0.22, 0.52)	<0.001
Education	High school	18	1	
	Less than high school	15	0.83 (0.52, 1.33)	0.44
	Some college	32	2.19 (1.61, 2.96)	<0.001
	Bachelor's degree or higher	44	3.51 (2.67, 4.62)	<0.001
Healthcare worker	No	30	1	
	Yes	50	2.36 (1.79, 3.11)	<0.001
Household income	$25,000-$74,999	24	1	
	< $25,000	18	0.70 (0.49, 1.02)	0.06
	≥ $75,000	37	1.85 (1.50, 2.27)	<0.001
Census region	Northeast	32	1	
	Midwest	30	0.91 (0.69, 1.20)	0.52
	South	31	0.95 (0.73, 1.23)	0.68
	West	33	1.01 (0.77, 1.33)	0.93
Urbanicity	Suburban	31	1	
	Urban	33	1.09 (0.90, 1.33)	0.37
	Rural	32	1.05 (0.82, 1.34)	0.69
Child with chronic medical condition	No	30	1	
	Yes	42	1.70 (1.30, 2.21)	<0.001
Personal COVID-19 experience[a]	None/no or mild symptoms only	23	1	
	Moderate symptoms, hospitalization, death	34	1.73 (1.37, 2.17)	<0.001

[a] Most severe level of illness due to COVID-19 in the respondent, adults they knew, and/or children they knew; **Abbreviations:** CI, confidence interval

Table 2: Unadjusted Effect of Awareness of MIS-C on Perceived COVID-19 Susceptibility and Severity Among All Respondents and By Age Group of Children

Perceived COVID-19 Severity								
Aware of MIS-C	All Respondents		Children Aged 0-4		Children Aged 5-11		Children Aged 12-17	
	Coeff[a] (95% CI)	*P* value	Coeff[a] (95% CI)	*P* value	Coeff[a] (95% CI)	*P* value	Coeff[a] (95% CI)	*P* value
Yes vs. No	0.20 (0.12, 0.27)	<0.001	0.16 (0.03, 0.30)	0.02	0.20 (0.10, 0.30)	<0.001	0.15 (0.05, 0.25)	0.003
Perceived COVID-19 Susceptibility								
Aware of MIS-C	All Respondents		Children Aged 0-4		Children Aged 5-11		Children Aged 12-17	
	Coeff[a] (95% CI)	*P* value	Coeff[a] (95% CI)	*P* value	Coeff[a] (95% CI)	*P* value	Coeff[a] (95% CI)	*P* value
Yes vs. No	0.17 (0.09, 0.25)	<0.001	0.21 (0.07, 0.35)	0.004	0.19 (0.08, 0.29)	0.001	0.06 (-0.05, 0.17)	0.26

[a] Weighted regression coefficient represents the difference in the mean composite COVID-19 severity (or COVID-19 susceptibility) score (range 1-4). For example, a coefficient of 0.2 indicates that the mean perceived COVID-19 severity score was 0.2 units higher among respondents aware of MIS-C compared to those not aware of MIS-C.

Abbreviations: CI, confidence interval; coeff, weighted regression coefficient

Disclosure: L. Cole: None; **E. Hammershaimb**: Moderna, 5, Novavax, 5; **Y. Liang**: None; **M. Hendrich**: None; **D. Das**: None; **R. Petrin**: None; **J. Campbell**: Moderna, 5, Novavax, 5; **S. O'Leary**: None; **J. Cataldi**: None.

Abstract Number: 106

Caregivers' Perspectives on Barriers to Care in Juvenile Localized and Systemic Scleroderma

Leigh Stubbs[1], Andrew Ferry[2], Danielle Guffey[1], Christina Loccke[3], Erin Moriarty Wade[3], Pamela Pour[3], Kaveh Ardalan[4], Peter Chiraseveenuprapund[5], Ingrid Ganske[6], Daniel Glaser[7], Gloria Higgins[8], Nadia Luca[9], Katharine Moore[10], Vidya Sivaraman[11], Katie Stewart[1], Natalia Vasquez Canizares[12], Raegan Hunt[1], Renata Maricevich[1], Kathryn Torok[13] and Suzanne Li[14], [1]Baylor College of Medicine, Houston, TX, [2]Oregon Health and Science University, Portland, OR, [3]n/a, [4]Duke University School of Medicine, Durham, NC, [5]University of California - San Diego, San Diego, CA, [6]Boston Children's Hospital and Harvard Medical School, Boston, MA, [7]Yale University School of Medicine, New Haven, CT, [8]Nationwide Childrens Hospital/ The Ohio State University, Columbus, OH, [9]University of Calgary, Calgary, AB, Canada, [10]University of Colorado / Children's Hospital Colorado, Denver, CO, [11]Nationwide Children's Hospital/ The Ohio State University, Columbus, OH, [12]Children's Hospital at Montefiore, New York, NY, [13]University of Pittsburgh, Pittsburgh, PA, [14]Hackensack Meridian School of Medicine, Joseph M. Sanzari Children's Hospital, Hackensack, NJ

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Posters: Clinical and Therapeutic II

Session Type: Poster Session B

Session Time: 5:00PM–6:00PM

Background/Purpose: Juvenile localized scleroderma (LS) and systemic sclerosis (SSc) are rare rheumatic diseases often associated with severe morbidities. Delays in diagnosis are common, putting children at risk for permanent damage and worse outcomes. This study investigated caregiver perspectives on barriers they encountered while seeking a diagnosis and care for their child's scleroderma.

Methods: In this cross-sectional study, researchers recruited parents of juvenile LS or SSc patients from a virtual family scleroderma educational conference and a juvenile scleroderma online support group over three months to survey them about their child's condition and factors affecting diagnosis and treatment.

Results: The response rate was 61% (73/120). The respondents comprised 38 parents of LS patients and 31 parents of SSc patients. Most patients were female (80%), and more than half were non-Hispanic white (55%). The majority of families had at least one person with a college education or higher (87%), traveled < 2 hours to see their doctor (83%), and had private insurance (75%). Nearly half had an annual household income > $100,000 (46%). Caregivers surveyed identified these main barriers to care: lack of knowledge about scleroderma in the medical community, finding reliable information about scleroderma in children, long wait times for a rheumatologist/specialist appointment, balance of school/work and child's healthcare needs, medication side effects, and identifying effective medications. Respondents said the lack of knowledge about juvenile scleroderma was their most challenging barrier. Diagnosis and systemic treatment initiation were more than one year from initial presentation for approximately 28% and 36% of patients, respectively.

Conclusion: Caregivers of children with LS or SSc surveyed reported many common barriers to the diagnosis, treatment, and ongoing care of juvenile scleroderma. The biggest problem highlighted was the medical community's lack of knowledge of scleroderma. However, most respondents had relatively high socioeconomic status, so additional studies are needed to reach a broader audience, including caregivers with limited English proficiency, geographical limitations, and financial constraints, to evaluate whether these problems are universal. Identifying care barriers will help direct efforts to address needs better, reduce disparities in care, and ultimately improve patient outcomes.

Disclosure: L. Stubbs: None; **A. Ferry**: None; **D. Guffey**: None; **C. Loccke**: None; **E. Moriarty Wade**: None; **P. Pour**: None; **K. Ardalan**: None; **P. Chiraseveenuprapund**: None; **I. Ganske**: None; **D. Glaser**: None; **G. Higgins**: None; **N. Luca**: None; **K. Moore**: None; **V. Sivaraman**: None; **K. Stewart**: None; **N. Vasquez Canizares**: None; **R. Hunt**: None; **R. Maricevich**: None; **K. Torok**: None; **S. Li**: Merck/MSD, 11.

Abstract Number: 107

Validation of Serious Adverse Event Reporting in a Multicenter Registry

Matthew Basiaga[1], Rajdeep Pooni[2], Caitlan Pinotti[3], Lisa Buckley[4], Alysha Taxter[5] and CARRA Registry Investigators[6], [1]Mayo Clinic, Rochester, MN, [2]Stanford University, Palo Alto, CA, [3]Duke, Durham, NC, [4]Vanderbilt University Medical Center, Nashville, TN, [5]Nationwide Children's Hospital, Columbus, OH, [6]CARRA, Washington, DC

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Children with rheumatic disease frequently require management with immune suppressing medications. The benefits of these interventions often outweigh the risks, however serious adverse events (SAE) will occur in a subset of patients. Accurately capturing SAEs is a cornerstone of pharmacovigilance. The Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry is a multicenter collaboration to maintain such data. It is unknown if underreporting exists in registry site self-reported event data in the CARRA registry. This study aims to identify novel ways to identify SAEs using a study sites electronic health record (EHR) tools and compare to the Registry.

Table 1: Comparison of Abstracted Hospitalizations and Reported Hospitalizations in the Registry

	Patients Reviewed at Each Site	Total Patient Observation Period (years)	Number of Hospitalizations Identified	Hospitalizations for Infection Identified	Hospitalizations Reported in Registry	Hospitalizations for Infection Reported in Registry	Unreported Hospitalizations for Infection
Site 1	48	92.4	11	1	0	0	1
Site 2	18	29.4	6	3	4	1	2
Site 3	30	111.4	57	8	3	2	6
Site 4	30	60	3	2	0	0	3
Site 5	30	60	0	0	0	0	0
Aggregate	156	353.2	77	14	7	3	12

Comparison of Abstracted Hospitalizations and Reported Hospitalizations in the Registry

Methods: Five CARRA sites each performed a chart review of approximately thirty randomly selected children enrolled in the Registry. The most recent 24 months, or the duration of registry enrollment if 24 months, were examined. SAEs identified using a variety of methods in the electronic health records including creating filters, keyword search, manually reviewing all patient encounters, and reviewing outside medical records when available. The variables were collected in a standardized template and aggregate data were examined. Admissions prior to CARRA enrollment were excluded. Routine hospitalization for medication infusion, sedation for diagnostic testing, sedation for joint injections, or admissions unrelated to their primary diagnosis were excluded. The CARRA database was then reviewed for the same patients to collect reported SAEs adverse events over the same time frame.

Results: A total of 156 patients were included for analysis with 353 patient years of observation. Although Admission filters were available, 30% of identified admission included non-event admissions, such as sedation or a scheduled procedure. Although some clinic note templates were standardized to capture event data, none were available in a discrete, reportable field. Only one site leveraged pre-visit patient-questionnaires to directly ask patients these data . We identified 77 hospitalizations with 14 directly related to infection in our patient sample (Table 1). A rate of 3.96 infection related hospitalizations per 100 patient years was calculated compared to 1.98 infection related hospitalizations reported.

Conclusion: We identified significant underreporting of SAE in a random sample of patients enrolled in the CARRA from five participating sites utilizing tools in the electronic health record. This may lead to mischaracterization of the risks of treatment in this patient population. Future efforts leveraging existing tools in the electronic health record may streamline the reporting process and minimize the prevalence of SAE under reporting.

Disclosure: M. Basiaga: None; **R. Pooni**: None; **C. Pinotti**: None; **L. Buckley**: None; **A. Taxter**: None; **C. Investigators**: None.

Abstract Number: 108

Development of Mental Health Guidance Statements for Pediatric Rheumatology

Ashley Danguecan[1], Natoshia Cunningham[2], Samantha Ely[3], Yaa Amponsah[4], Alaina Davis[5], Suzanne Edison[6], Alicia Halbert[4], Julia Harris[7], Alicia Hoffman[8], Jordan Jones[7], Alana Leever[9], Catherine Levalee[4], Alison Manning[10], Anne McHugh[11], Sam Mendoza[4], Crystal Mui[4], Ekemini Ogbu[12], Nikki Reitz[4], Martha Rodriguez[13], Natalie Rosenwasser[14], Alyse Tankanow[4], Erin Treemarcki[15], Tracy Van Ness[16], Katie Winner[11], Tamar Rubinstein[8] and **Andrea Knight**[1], [1]The Hospital for Sick Children, Toronto, ON, Canada, [2]Michigan State University, Grand Rapids, MI, [3]Michigan State University, East Lansing, [4]CARRA Mental Health Taskforce, Washington, DC, [5]Vanderbilt Children's Hospital, Nashville, TN, [6]Cure JM Foundation, [7]Children's Mercy, Kansas City, KS, [8]Children's Hospital at Montefiore, New York, NY, [9]Nationwide Children's Hospital, [10]Duke University School of Medicine, Durham, NC, [11]Dayton Children's Hospital, Dayton, OH, [12]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, [13]Riley Hospital for Children at Indiana University Health, Indianapolis, IN, [14]Seattle Children's Hospital, Seattle, WA, [15]University of Utah, Salt Lake City, UT, [16]Pace University, New York, NY

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Posters: Clinical and Therapeutic II

Session Type: Poster Session B

Session Time: 5:00PM–6:00PM

Background/Purpose: There are high rates of anxiety and depression in youth with rheumatologic diseases, with known impact on health-related outcomes. Thus, it is important to effectively screen and manage mental health concerns in the context of pediatric medical care. We developed guidance statements for pediatric rheumatologists to support assessment and management of mental health concerns. These statements were developed by a multi-disciplinary team of experts, including patient/caregiver stakeholders, drawing from mental health research in rheumatology and the broader pediatric literature.

Table 1. Timeline of the Development of the Mental Health Guidance Statements

Date	Milestone
April 12th, 2019	Initial discussion of key concepts and drafted statements.
April – October 2019	Open call and invitation sent to participate in the Mental Health Taskforce.
October 22nd, 2019	Finalized taskforce roster and divided into screening and management subgroups.
Late October 2019 – Early April 2020 (~24 weeks)	Subgroups reviewed relevant literature and modified statements based on findings. Statements were assigned a grade based on the OCEBM levels of evidence.
April 9th, 2020	Refined statements were distributed to the taskforce for further feedback.
Early April 2020 – Late August 2020 (~20 weeks)	Statements were revised with feedback from the taskforce.
October 14th, 2020 – October 21st, 2020	Taskforce members voted on statements and provided additional feedback.
Late October 2020 – Late April 2021 (~27 weeks)	Statements were refined with taskforce feedback.
May 13th, 2021 – June 30th, 2021	Revised statements disseminated to broader CARRA organization for feedback in an open comment period.
Early July 2021 – Late October 2021 (~17 weeks)	Feedback from the CARRA open comment period was incorporated into the statements.
January 10th, 2022 – March 18th, 2022	Statements were voted on by a random sample of the rheumatologists and an enriched sample of behavioral health providers, parents at patients part CARRA organization using RAND methodology.
March 22nd, 2022 – August 31, 2022	Review and discussion of data among taskforce from first voting period.
September 30, 2022	Presentation of initial results and discussion with taskforce leadership to identify additional items of importance for a second vote
December 5 2022- January 15 2023	Panel queried for second vote on four selected items pertaining to management of mild
January 17, 2023	mental health symptoms and identification of a mental health champion
January 2023	Analysis of results from second voting period

Table 2. Consensus Data for Statements Addressing Pediatric Mental health in Pediatric Rheumatological Populations

Statement	Responders (n)	Appropriateness Category (n, %)		
		Inappropriate 1-3	Uncertain 4-6	Appropriate 7-9
Screening for anxiety, depression, and suicidal ideation (SI) should occur as soon as is feasible following the initial rheumatology clinic visit or onset of a new rheumatologic diagnosis. (Grade 2)	76	3, 3.95%	12, 15.79%	61, 80.26%
Screening should occur annually, at a minimum, for individuals aged 12 years and up, and more frequently as deemed necessary. (Grade 1)	76	2, 2.63%	10, 13.16%	64, 84.21%
Additional screenings should occur more frequently during periods of increased disease activity, psychosocial changes, and periods of transition (e.g., a move, starting a new school). (Grade 2)	76	0, 0%	11, 14.47%	65, 85.53%
Screening measures should be developmentally appropriate, patient-reported psychometrically validated measures, when possible. (Grade 2)	76	1, 1.32%	5, 6.58%	70, 92.11%
Screening for SI should occur at least annually using a clinically validated tool with an item to assess for SI, in a setting where there are resources in place to address positive screens. (Grade 2)	76	4, 5.26%	11, 14.47%	61, 80.26%
Discussion of screening results should take place at the patient visit to gain additional understanding of symptoms reported. (Grade 5)	76	1, 1.32%	10, 13.16%	65, 85.53%
Discussion of mental health concerns should be completed privately with the patient when appropriate as this may yield more reports of anxiety, depression, and SI. (Grade 5)	76	0, 0%	11, 14.47%	65, 85.53%
Provider concerns about mental health symptoms or issues should prompt further evaluation despite screening results. (Grade 5)	76	1, 1.32%	7, 9.21%	68, 89.47%
Caregiver concerns about mental health symptoms or issues should prompt further evaluation despite screening results. (Grade 5)	76	1, 1.32%	4, 5.26%	71, 93.42%
Providers should assess the possibility that neuropsychiatric manifestations of rheumatic disease, developmental and behavioral issues, and substance abuse may cause or contribute to anxiety, depression, or SI. (Grade 5)	76	1, 1.32%	6, 7.89%	69, 90.79%
Anxiety and depression may present as issues with medication adherence, medical visit attendance, needle phobia, somatic symptoms, etc. In the presence of such behaviors, medical providers should query for anxiety and depression in the context of the developmental status of the patient. (Grade 5)	76	2, 2.63%	6, 7.89%	68, 89.47%
Minoritized groups are often less likely to be diagnosed and referred for mental health treatment. Providers should understand and assess for inequities in evaluation and access to mental health resources, especially in minoritized groups, when implementing mental health screening. (Grade 3)	76	0, 0%	8, 10.53%	68, 89.47%
Patients who demonstrate evidence of clinically significant symptoms of anxiety, depression, or SI at a prior screening should be reassessed at subsequent visits. (Grade 2)	76	0, 0%	4, 5.26%	72, 94.74%
Mental health specialists (e.g., social worker, therapist, psychologist, psychiatrist, psychiatric nurse, adolescent medicine physician), should be involved with screening processes and additional evaluation in instances of positive screens, if those resources are available (Grade 5)	76	1, 1.32%	2, 2.63%	73, 96.05%
Mental health specialists should assess family and socioeconomic factors contributing to the burden of disease on the family system. (Grade 5)	76	1, 1.32%	3, 3.95%	72, 94.74%
Mental health specialists should assess stress and coping strategies adopted by the family member. (Grade 5)	76	1, 1.32%	4, 5.26%	71, 93.42%
Given that patients often feel stigmatized regarding mental health, rheumatologists are encouraged to facilitate discussions about options for mental health intervention (e.g. seeing a therapist, psychopharmacologic treatments) to normalize such treatments and increase patient/caregiver buy-in regarding potential referrals. (Grade 5)	76	3, 3.95%	6, 7.89%	67, 88.16%
For patients who demonstrate mild symptoms (and no SI): Recommend that the patient follow-up with a primary care provider to help monitor symptoms. (Grade 3)	76 *(54)*	2, 2.63% (2, 3.70%)	15, 19.74% *(8, 14.81%)*	59, 77.63% *(44, 81.48%)*

For patients who demonstrate mild symptoms (and no SI): Refer to a mental health specialist, re-assess at the next visit, and advise patient and caregiver(s) to monitor symptoms. (Grade 3)	76 *(54)*	6, 7.89% *(2, 3.43%)*	15, 19.74% *(11, 20.37%)*	55, 72.37% *(41, 75.92%)*
For patients who demonstrate mild symptoms (and no SI): Provide preventative education/resources regarding co-morbidity of mental health concerns and rheumatic disease. (Grade 5)	76 *(54)*	3, 3.95% *(1, 1.85%)*	14, 18.42% *(8, 14.81%)*	59, 77.63% *(45, 83.33%)*
For patients who demonstrate moderate to severe symptoms (with or without SI): An urgent assessment by a mental health specialist is preferred to advise on evidence-based treatments, including possible options for medication management to be used in conjunction with psychological intervention. (Grade 1)	76	0, 0%	0, 0%	76, 100%
For patients who demonstrate moderate to severe symptoms (and no SI): A face-to-face (e.g., in-person, video) follow-up assessment of mental health symptoms should occur with an identified member of the clinical team to guide treatment planning and ongoing monitoring. (Grade 5)	76	3, 3.95%	8, 10.53%	65, 85.53%
If SI is endorsed or detected by screen (regardless of depression severity), a comprehensive assessment of the risk of suicidality on the same day should be used. (Grade 5)	76	1, 1.32%	3, 3.95%	72, 94.74%
If a patient is determined to be actively suicidal and unable to commit to a safety plan, an immediate referral to the closest Emergency Department or mental health crisis center should be initiated. If a non-psychiatric medical cause is suspected, consider referring to the closest medical emergency department. (Grade 5)	76	0, 0%	0, 0%	76, 100%
Communicate positive mental health screening results to appropriate members of the care team (primary care physician, rheumatologist, mental health professional), to promote shared care. (Grade 5)	76	1, 1.32%	6, 7.89%	69, 90.79%
Plans of care for mental health should be made collaboratively with the patient and appropriate members of the health care team. (Grade 5)	76	1, 1.32%	1, 1.32%	74, 97.37%
Rheumatologists should provide education to all patients and families in clinics about common mental health issues in rheumatologic/chronic disease in a developmentally appropriate context. (Grade 5)	75	1, 1.33%	20, 26.67%	54, 72.00%
Educational information regarding mental health should be provided to patients and families on an ongoing basis (e.g., annually). (Grade 5)	75	2, 2.67%	27, 36.00%	46, 61.33%
In an effort to provide appropriate follow-up to patients with mental health concerns, pediatric rheumatology practices should strive to maintain a current list of local community mental health resources that is updated annually. (Grade 5)	75	4, 5.33%	10, 13.33%	61, 81.33%
Identification of a mental health champion within the pediatric rheumatology clinic may be helpful for maintaining an up-to-date local resource list for mental health which can include online or telehealth resources. (Grade 5)	75 *(53)*	3, 4.00% *(0, 0%)*	14, 18.67% *6, 11.32%*	58, 77.33% *(47, 88.67%)*
To facilitate discussion about mental health, consider inquiring about multiple aspects/domains of functioning including: medication adherence, sleep hygiene, nutrition, family relationships, social engagement, and school functioning. (Grade 5)	75	1, 1.33%	6, 8.00%	68, 90.67%
On-going discussions and education about mental health should occur (e.g., annually), including discussion of warning signs and symptoms of mental health disorders. Information should be given in a manner consistent with the patient's developmental stage and socio-cultural background. (Grade 3)	75	2, 2.67%	10, 13.33%	63, 84.00%
To facilitate discussion about mental health, patients who have been given a new diagnosis should be offered resources (e.g., fact sheets, brochures, websites, peer support) that address common mental health concerns in rheumatologic/chronic disease. (Grade 3)	75	5, 6.67%	10, 13.33%	60, 80.00%
Mental health educational sessions should be provided at national rheumatology meetings. (Grade 5)	75	3, 4.00%	5, 6.67%	67, 89.33%

Methods: Key concepts and content areas for the statements were first developed by the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Mental Health Workgroup at the 2019 annual CARRA meeting. Subsequently, the CARRA Mental Health Taskforce panel was formed, consisting of 10 rheumatologists, 6 pediatric behavioral health providers, 4 patients, 3 parents, and 3 members who were both behavioral/healthcare providers and parents. The Taskforce (divided into

screening and management subgroups) convened multiple times between October 2019 and March 2021 to review relevant literature and modified statements based on findings. The statements were graded based on the Oxford Centre for Evidence-Based Medicine (OCEBM) levels of evidence, then disseminated to the entire Taskforce for additional modification/review before being shared with the broader CARRA membership for open review from May June 2021. Feedback was integrated by Taskforce leadership before a final set of 34 statements was sent for evaluation by a random sample of the broader CARRA membership (n=123) using a well-established method of consensus gathering (modified Delphi method) from January 2022 January 2023. A timeline of activities is provided in Table 1. An enriched sample of all active behavioral health providers (n=11) and parents/patients (n=12) in CARRA was included to ensure their voice was represented. This entire group was invited to submit asynchronous consensus ratings for each statement by email. Panel members rated their agreement on the draft statements on a scale from 1 (inappropriate) to 9 (appropriate). Items that failed to achieve consensus were reviewed and an additional round of voting was conducted with select items. To be approved as final guidance statements, median agreement ratings were required to be at 80% or greater (with an appropriate median rating of 7-9).

Results: Of the 123 queried, 80 responded (65%) and nearly all of the respondents (n=77, 96%) agreed to participate. A second round of voting occurred on 4 selected items, with greater than 70% participation. Of the 34 statements, 31 statements achieved consensus (Table 1). Bolded statements in Table 2 reflect those that achieved final consensus.

Conclusion: Consensus was achieved for all the mental health screening statements, as well as for a majority of statements related to the management of mental health concerns and clinic environment/education. It is the goal that these recommendations be used to help improve both mental health- and health-related outcomes for youth with rheumatological conditions.

Disclosure: A. Danguecan: None; **N. Cunningham**: None; **S. Ely**: None; **Y. Amponsah**: None; **A. Davis**: None; **S. Edison**: None; **A. Halbert**: None; **J. Harris**: None; **A. Hoffman**: None; **J. Jones**: None; **A. Leever**: None; **C. Levalee**: None; **A. Manning**: None; **A. McHugh**: None; **S. Mendoza**: None; **C. Mui**: None; **E. Ogbu**: None; **N. Reitz**: None; **M. Rodriguez**: None; **N. Rosenwasser**: None; **A. Tankanow**: None; **E. Treemarcki**: None; **T. Van Ness**: None; **K. Winner**: None; **T. Rubinstein**: None; **A. Knight**: None.

Abstract Number: 109

Long-term Safety of Biologics versus Conventional Synthetic Treatments in Systemic Juvenile Idiopathic Arthritis Patients

Ana Isabel Rebollo-Giménez[1], Luca Carlini[2], Yulia Vyzhga[3], Silvia Rosina[4], Ekaterina Alexeeva[5], Charlotte Myrup[6], Silvia Magni Manzoni[7], Maria Trachana[8], Valda Stanevicha[9], Constantin Ailioaie[10], Elena Tsitsami[11], Alexis-Virgil Cochino[12], Chiara Pallotti[13], Silvia Scala[13], Angela Pistorio[14], Sebastiaan Vastert[15], Joost F. Swart[16] and Nicolino Ruperto[17], [1]IRCCS Istituto Giannina Gaslini, UOC Reumatologia e Malattie Infiammatorie, Genova, Italy, [2]IRCCS Istituto Giannina Gaslini, UOC Reumatologia e Malattie Autoinfiammatorie, Genova, Italy, [3]IRCCS Istituto Giannina Gaslini, UOC Reumatologia e Malattia Infiammatorie, Genova, Italy, [4]IRCCS Istituto Giannina Gaslini, Genova, Italy, [5]Federal State Autonomous Institution "National Medical Research Center of Children's Health", Ministry of Health of the Russian Federation, Moscow, Russia, [6]Rigshospitalet, Pediatric rheumatology unit 4272, Copenhagem, Denmark, [7]IRCCS Ospedale Pediatrico Bambino Gesù, Division of Rheumatology, Roma, Italy, [8]Hippokration General Hospital, Thessaloniki University School of Medicine, First Department of pediatrics, Pediatric Immunology and Rheumatology Referral Center, Thessaloniki, Greece, [9]Riga Stradins University, Children University Hospital, Riga, Latvia, [10]Alexandru Ioan Cuza University of Iasi, Iasi, Romania, [11]Aghia Sophia Childrens Hospital, First Department of Pediatrics, University of Athens Medical School, Athens, Greece, [12]Institute for Mother and Child Care, Pediatrics, Bucharest, Romania, [13]IRCCS Istituto Giannina Gaslini, U.O.C. Pediatric and Rheumatology Clinic, PRINTO, Genova, Italy, [14]IRCCS Istituto Giannina Gaslini, Direzione Scientifica, Genova, Italy, [15]Wilhelmina Children's Hospital, Department of Pediatric Immunology and Rheumatology, Utrecht, The Netherlands, Utrecht, Netherlands, [16]Wilhelmina Children's Hospital, Department of Pediatric Immunology and Rheumatology, Utrecht, Netherlands, [17]IRCCS Istituto Giannina Gaslini, UOSID Centro Trial, PRINTO, Genova, Italy

SESSION INFORMATION

Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: The better understanding of systemic Juvenile Idiopathic Arthritis (sJIA) pathogenesis and availability of new drugs, such as biologic disease-modifying anti-rheumatic drugs (bDMARDs) specifically dedicated to sJIA, have led to treatment advances that have ameliorated the disease outcome and reduced the use of glucocorticoids. However, published evidence on long term safety data regarding biologic therapies in sJIA is still limited.The objective of the study was to compare the adverse events (AEs) of at least moderate intensity and serious AEs in sJIA patients treated with biologics compared with a cohort treated with csDMARDs (conventional synthetic disease-modifying anti-rheumatic drugs).

Table. Demographics characteristics, number of AEs and frequencies in the complete set. Data presented as n (%). Events with a frequency of > 30 by overall SOC are presented. AEs: adverse events; SOC: system organ class; bDMARDs: biologic disease-modifying anti-rheumatic drugs; csDMARDs conventional synthetic disease-modifying anti-rheumatic drugs.

	bDMARDs N=353	csDMARDs N=627	Overall N=980	p-value
Male	175 (49.6%)	279 (44.5%)	454 (46.3%)	-
Mean (SD) age at onset (years)	6.21 (4.24)	5.77 (4.04)	5.92 (4.12)	-
Age groups (years)				
<2	65 (18.4%)	115 (18.3%)	180 (18.4%)	-
>=2 to 12	242 (68.6%)	444 (70.8%)	686 (70.0%)	-
>=12 to 18	46 (13.0%)	68 (10.9%)	114 (11.6%)	-
>= 18	0 (0%)	0 (0%)	0 (0%)	-
Age at diagnosis	6.56 (4.34)	6.17 (4.13)	6.31 (4.21)	-
Age at final visit	12.05 (5.39)	12.41 (5.13)	12.28 (5.22)	-
Disease duration	5.84 (4.61)	6.65 (4.63)	6.36 (4.64)	-
Origin				-
Europe	320 (90.7%)	466 (74.3%)	786 (80.2%)	-
Other	33 (9.3%)	161 (25.7%)	194 (19.8%)	-
Concomitant steroid therapy (>=6 months)	165 (46.7%)	414 (66.0%)	579 (59.08%)	<0.0001
SOCs				
Infections and infestations	146 (41.4%)	176 (28.1%)	332	<0.0001
Gastrointestinal disorders	33 (9.3%)	51 (8.1%)	84	0.59
Injury, poisoning and procedural complications	28 (7.9%)	50 (8.0%)	78	0.99
Skin and subcutaneous tissue disorders	40 (11.3%)	27 (4.3%)	67	<0.0001
Blood and lymphatic system disorders	25 (7.1%)	42 (6.7%)	67	0.92
Endocrine disorders	13 (3.7%)	46 (7.3%)	59	0.03
Immune system disorders	14 (4.0%)	36 (5.7%)	50	0.28
Musculoskeletal and connective tissue disorders	13 (3.7%)	37 (5.9%)	50	0.17
General disorders and administration site conditions	29 (8.2%)	17 (2.7%)	46	<0.0001
Investigations	16 (4.5%)	29 (4.6%)	45	0.99
Nervous system disorders	15 (4.2%)	16 (2.6%)	31	0.21

Methods: Patients with sJIA, classified according to the International League of Associations for Rheumatology (ILAR), enrolled in the Pharmachild Registry since 2011 and followed up until 31 December 2022 were included. All patients had received at least one bDMARDs or csDMARDs. Those who switched to more than one treatment during the period of observation were assigned only to one group according to the longest time of drug exposure. AEs are reported according to the latest release of the Medical Dictionary for Regulatory Activities (MedDRA, Version 23.1) and were grouped into highest term System Organ Classes (SOCs). Frequency of SOCs were analysed. Events repeated for the same SOC in the same patient were counted once in the analysis.

Results: A total of 980 sJIA patients were enrolled, 353 treated with bDMARDs and 627 treated with csDMARDs. The distribution of demographic data was similar in both cohorts, with the exception that the prescription of csDMARDs compared to bDMARDs was more frequent in other continents compared to Europe. As shown in the table, patients in the bDMARDs group developed more frequently infections and infestations (41.4%), skin and subcutaneous tissue disorders (11.3%) and general disorders and administration site conditions (8.2%) than patients treated with csDMARDs (all $p< 0.0001$). On the other hand, endocrine disorders were more frequent in the csDMARDs group (7.3%, $p=0.03$). The distribution between other SOCs was similar in children treated with bDMARDs compared with csDMARDs.

Conclusion: Patients in the bDMARD group had a higher prevalence of infections and skin manifestations when compared with the csDMARDs cohort.

Disclosure: A. Rebollo-Giménez: None; **L. Carlini**: None; **Y. Vyzhga**: None; **S. Rosina**: None; **E. Alexeeva**: AbbVie/Abbott, 5, Amgen, 5, Bristol-Myers Squibb(BMS), 5, Centocor, 5, Eli Lilly, 5, Merck/MSD, 5, Novartis, 5, 6, Pfizer, 5, 6, Roche, 5, 6, Sanofi, 5; **C. Myrup**: None; **S. Magni Manzoni**: None; **M. Trachana**: None; **V. Stanevicha**: None; **C. Ailioaie**: None; **E. Tsitsami**: None; **A. Cochino**: None; **C. Pallotti**: None; **S. Scala**: None; **A. Pistorio**: None; **S. Vastert**: Novartis, 6, SOBI, 5, 6; **J. F. Swart**: None; **N. Ruperto**: Amgen, 6, AstraZeneca, 6, Aurinia, 6, Bayer, 6, Bridge, 6, Brystol Myers and Squibb, 5, 6, Cambridge Healthcare Research, 6, Celgene, 6, Domain Therapeutic, 6, Eli Lilly, 5, 6, EMD Serono, 6, F Hoffmann-La Roche, 5, Glaxo Smith Kline, 6, Idorsia, 6, inMed, 6, Janssen, 6, Novartis, 5, 6, Pfizer, 5, 6, Sobi, 5, 6, UCB, 6.

Abstract Number: 110

Medications Affect Antibody Responses to COVID-19 Vaccinations in Children with Autoimmune Diseases

Janna Shapiro[1], Florence Choi[2], Amy Xu[3], Trang Duong[4], Tania Watts[1], Anne-Claude Gingras[5], Sasha Bernatsky[6], Susanne Benseler[7] and **Rae Yeung**[8], [1]Department of Immunology, University of Toronto, Toronto, ON, Canada, [2]Division of Rheumatology, Department of Paediatrics, The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada, [3]The Hospital of Sick Children, Department of Paediatrics, University of Toronto, Division of Rheumatology; Cell Biology Program, The Hospital for Sick Children Research Institute, Toronto, ON, Canada, [4]Cell Biology Program, The Hospital for Sick Children Research Institute, Toronto, ON, Canada, [5]Lunenfeld-Tanenbaum Research Institute at Mount Sinai Hospital, Sinai Health; Department of Molecular Genetics, University of Toronto, Toronto, ON, [6]Centre for Outcomes Research and Evaluation (CORE), Research Institute of the McGill University Health Centre, Montreal, Québec, Canada; Division of Rheumatology, McGill University Health Centre, Montreal, QC, Canada, [7]Division of Rheumatology, Department of Paediatrics, The Alberta Children's Hospital, Calgary, AB, [8]The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Posters: Clinical and Therapeutic II

Session Type: Poster Session B

Session Time: 5:00PM–6:00PM

Background/Purpose: To compare antibody responses to COVID-19 vaccines among children with autoimmune diseases taking different classes of immunosuppressants

Methods: A prospective observational study was conducted at Canadian paediatric centers. Children under the age of 18 with autoimmune disease (rheumatic and/or inflammatory bowel disease) were eligible for the study regardless of treatment. Antibodies against SARS-CoV-2 spike and receptor-binding domain were measured in either serum or dried blood spots collected after each COVID-19 vaccine dose. The kinetics and magnitude of antibody response following each vaccine dose were compared between treatment groups, using mixed-effects multivariable regression models. Participants who had been diagnosed with COVID-19 were excluded from the current analysis.

Results: From clinical and research registries, 2199 potential subjects were identified and 1941 were contacted by letter and telephone or during a clinic visit. From these, 239 individuals consented to participate in the study. For the current analyses, 116 participants contributed 57 samples post-COVID-19 vaccination dose 2, 59 samples post-dose 3, and 22 samples post-dose 4. It was found that anti-spike IgG responses were both reduced in magnitude and waned faster for participants treated with biologic DMARDs (disease modifying anti-rheumatic drugs) than for those who were treatment naive or receiving traditional DMARD therapy post-dose 2. For post-dose 3, IgG responses remained lower in the biologics group, but there were no clear differences in waning over time. After dose 4, no differences between the groups were observed. Among the biologics, anti-TNF therapies and rituximab had the greatest effect on IgG titers.

Conclusion: Antibody responses to COVID-19 vaccination in children with autoimmune diseases was lowest in individuals treated with biologics, particularly with anti-TNF agents and rituximab. Third and fourth vaccine doses are necessary to yield robust and durable antibody responses.

Disclosure: J. Shapiro: None; **F. Choi**: None; **A. Xu**: None; **T. Duong**: None; **T. Watts**: None; **A. Gingras**: None; **S. Bernatsky**: None; **S. Benseler**: None; **R. Yeung**: None.

Abstract Number: 111

Monitoring for Hypogammaglobulinemia After B-Cell Therapy in an Academic Pediatric Center

Omar Mostafa, **Sharon Bout-Tabaku**, Buthaina Al-Adba, Ahmad Kaddourah, Abubakr Imam, Ibrahim Shatat and Mohammed Yousuf Karim, Sidra Medicine, Ar-Rayyan, Qatar

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Hypogammaglobulinemia is an under-recognized complication of B-cell targeted therapies (BCTT) in both autoimmune diseases (AID) and malignancy. Hypogammaglobulinemia may be transient or persistent, and may be associated with increased infection risk. While in 2019 and 2021, guidance was published for hypogammaglobulinemia in patients receiving BCTT, the majority of the primary literature quoted in these guidance articles is based on adult studies. Here we describe immunoglobulin (Ig) monitoring in our pediatric cohort receiving BCTT.

Methods: We retrospectively screened for all patients, including both AID and malignancy, who had received BCTT at a pediatric academic tertiary center, between 2016-22. Patients were identified by pharmacy records. The frequency of Ig testing and measurements were extracted from the electronic medical records. Frequency of hypogammaglobulinemia and the need for immunoglobulin replacement (IGRT) were noted. These findings were compared against the monitoring guidance in the 2019 and 2021 publications.

Results: Fifty-seven patients were included in the study: nephrotic syndrome 28, SLE 12, other rheumatological diseases 6, neurological diseases 6, malignancy 5. Pre-BCTT Ig results were available in 49/57 patients (85.9%), of which 13/49 (26.5%) had low IgG levels. During follow-up, 3/13 patients remained low, 6/13 normalized, and 4/13 did not have Igs repeated. Overall 39/57 (68.4%) patients had Ig testing after BCTT. The range of Ig measurements per patient was between 1-10 Ig over a follow-up of 1-36 months. Post BCTT, 16/39 (41%) patients developed low Igs, of which 2 were transient; one SLE patient developed low Igs after only a single BCTT cycle, subsequent investigations suggesting common variable immunodeficiency (CVID). However, no patients required initiation of IGRT.

Conclusion: Baseline Ig measurements were almost always performed per the guidance, and indeed baseline Ig's were abnormal in 26.5% patients. This confirms the importance of the baseline timepoint, whereby low baseline levels could be disease-related or due to other medications. Otherwise low Igs during follow-up might be incorrectly attributed to BCTT. However, monitoring of Igs was less strictly followed compared with the guidance. The importance of monitoring is demonstrated by the unmasking of CVID in an SLE patient after a solitary BCTT cycle. Development of hypogammaglobulinemia did not by itself require IGRT, in the absence of recurrent infections.

Disclosure: O. Mostafa: None; **S. Bout-Tabaku**: None; **B. Al-Adba**: None; **A. Kaddourah**: None; **A. Imam**: None; **I. Shatat**: None; **M. Karim**: Takeda Ltd, 6.

Abstract Number: 112

Adverse Childhood Experiences: Prevalence and Relationship to Disease in Childhood-onset Lupus

Olivia Hendrikx[1], Stephanie Fevrier[1], Ibrahim Mohamed[1], Chelsea DeCoste[2], Paris Moaf[1], Lawrence Ng[1], Deborah Levy[1], Linda Hiraki[1], Alene Toulany[1] and Andrea Knight[1], [1]The Hospital for Sick Children, Toronto, ON, Canada, [2]IWK Health Centre, Halifax, NS, Canada

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Adverse Childhood Experiences (ACEs) measure traumatic experiences in childhood. ACEs are associated with epigenetic changes, are known to increase stress response and inflammation, and can contribute to the onset and progression of chronic disease. Childhood-onset systemic lupus erythematosus (cSLE) is a chronic autoimmune disorder characterized by inflammation of multiple organs in children. ACEs may therefore play a role in cSLE, however, there is a lack of studies on the impact of ACEs in cSLE. This study aimed to describe the prevalence of ACEs and to determine the association between ACEs and i) disease activity, and ii) disease damage in patients with cSLE.

Methods: A retrospective cohort study of 106 adolescents aged 9-18 years with cSLE who were followed in the SickKids lupus clinic and seen by Adolescent Medicine between July 2018-July 2020 was conducted. The presence of ACEs was determined through medical chart review in EPIC and Chartmaxx. Disease activity was measured by the SLEDAI-2K, and disease damage by the SLICC damage index (SDI defined as score >0). Separate multivariable logistic regression models were used to examine the relationship between the presence of ACEs and the disease measures, adjusting for age of diagnosis, gender, comorbid psychiatric diagnosis, and comorbid medical diagnosis.

Results: 37% of patients reported at least 1 ACE, of which 90% experienced the ACE >1 year before diagnosis. The most common ACE was separated/divorced parents (n=21). In multivariable regression analyses, there were no significant associations between ACEs and disease activity or damage. ACEs were significantly associated with comorbid psychiatric diagnoses.

Conclusion: We found that ACEs were prevalent in our cSLE cohort, and associated with psychiatric diagnosis, but not cSLE disease measures. Future study is needed to better understand the impact of ACEs on inflammation and neuropsychiatric function in cSLE.

Disclosure: O. Hendrikx: None; **S. Fevrier**: None; **I. Mohamed**: None; **C. DeCoste**: None; **P. Moaf**: None; **L. Ng**: None; **D. Levy**: Janssen, 1, Roche, 5, Sobi, 1, 5; **L. Hiraki**: None; **A. Toulany**: None; **A. Knight**: None.

Abstract Number: 113

Proportion of Patients with a Polyphasic Disease Course in Systemic-onset Juvenile Idiopathic Arthritis May Be Higher in the Age of Cytokine Inhibitors

Itay Marmor[1], Rotem Semo Oz[2], Amir hendel[3], Guy Hazan[4], Kevin Baszis[5], Anthony French[5], Cuoghi Edens[6], Irit Tirosh[7], Yonatan Butbul Aviel[8], Liora Harel[9] and Gil Amarilyo[10], [1]Dana-Dwek Children's Hospital, Hod Hasharon, Israel, [2]Sheba medical center, Herzelyia, Israel, [3]Tel Aviv University, Tel Aviv, Israel, [4]Soroka University Medical Center, Be'er Sheva, Israel, [5]Washington University School of Medicine, St Louis, MO, [6]University of Chicago, Chicago, IL, [7]Sheba Medical Center, Savyon, Israel, [8]Rambam Medical center, Haifa, Israel, [9]Scheiders Children Medical Center of Israel, Petah-Tiqva, Israel, [10]Schneider Children's Medical Center of Israel, Petach Tikva, Israel

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Systemic-onset juvenile idiopathic arthritis (sJIA) is a pediatric autoinflammatory condition, known for significant variability between patients in its severity and long-term outcomes. The classification of disease course into monophasic, polyphasic (intermittent) and persistent disease has been commonly used, with polyphasic disease usually recognized in a small portion of patients[1,2] . However, this proportion was established according to data mostly collected before the biologic IL-1 and IL-6 inhibitors were available. It has been recently suggested [3,4] that these medications, now the mainstay of treatment in sJIA, can potentially alter the long-term course of the disease, especially with earlier use.

Methods:: A multi-center, retrospective chart review was conducted from 3 hospitals in Israel and 2 in the US, involving patients diagnosed with sJIA between 1998-2019, with a minimum follow-up of 1 year. Disease course classification was done according to previously released definitions.[1] Drug-free remission was defined as inactive disease (no active signs or

symptoms and no elevation of inflammatory markers) while not receiving any medications for a period of at least 3 months. Polyphasic disease was defined as at least one disease relapse after a period of drug-free remission. Persistent disease was defined as at least 2 years of disease (either active or in remission on immunosuppressive medication) with no drug-free remissions. Monophasic disease was defined as a single episode lasting less than 2 years.

Results: 85 patients met the inclusion criteria, with a median follow up time of 2.8 years (IQR 1.1-1.7). 54 (63.5%) of the patients were female; mean age at diagnosis was 6.3 ± 4.3 years. 67 (78.9%) were diagnosed in 2012 or later, when IL-1 and IL-6 inhibitors became widely used. 52 (61.2%) were treated with an IL-1/6 inhibitor during their disease course. The rates of monophasic, polyphasic and persistent disease were 41.2%, 44.7% and 14.1%, respectively, with a higher-than-expected rate of polyphasic disease and a lower rate of persistent disease than previously published. [1,2,5]

Conclusion: In the age of IL-1 and IL-6 inhibitors, polyphasic sJIA disease course may be more common than previously described, suggesting that cytokine blockers may potentially alter the natural history of this disease.

Disclosure: I. Marmor: None; **R. Semo Oz**: None; **A. hendel**: None; **G. Hazan**: None; **K. Baszis**: None; **A. French**: None; **C. Edens**: None; **I. Tirosh**: None; **Y. Butbul Aviel**: None; **L. Harel**: None; **G. Amarilyo**: None.

Abstract Number: 114

Clinical Significance of SSA and SSB Antibodies in Pediatric SLE Patients: A Single Center Cohort

Yiressy Pina[1], Dawn Janysek[1], Danielle Guffey[2], Maria Pereira Palacios[3] and Marietta De Guzman[1], [1]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [2]Baylor College of Medicine, Houston, TX, [3]Baylor College of Medicine, Texas Children's Hospital, Houston, TX

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Anti-nuclear antibodies (ANA) are important in the diagnosis of Systemic Lupus Erythematosus (SLE). Of the disease specific ANAs, anti-dsDNA and anti- Smith antibodies carry the most diagnostic power and clinical application. Other specific ANAs which include antibodies to SSA, SSB, and Scl70 are intrinsic component to the understanding of the immune dysregulation of this disease. In adult lupus, these autoantibodies have been described to closely ally with particular clinical features or findings. In this study we sought to explore the clinical significance of SSA and SSB antibodies in pediatric SLE, as to specific disease manifestation and disease severity.

Methods: With BCM IRB approval, we performed a retrospective chart review of patients diagnosed with SLE as per SLICC and EULAR/ACR classification criteria, between 2018 and 2021. We collected clinical and laboratory features, ANAs (per University of Missouri ANA Laboratory) and SLEDAI at presentation, 6 and 12 months and last follow up. Study group consisted of patients with anti-SSA or SSB, while control group without these ANAs. Characteristics are summarized by group and compared using independent t-test, Wilcoxon rank sum, Chi-squared, or Fisher's exact test.

Results: There were 137 patients diagnosed with SLE during the study period. There were 40 patients in the study group, and 39 in the control group. Mean age was similar between these groups, as well as percentages of Hispanic patients. More African American patients was noted in the study group (37.5% vs 17.9%). Patients in the study group presented with dry

Table 1: Patient Characteristics in Control vs Study Group at Presentation

	Control (N=39)	Study group (N=40)	P Value
Demographics			
Age	Mean (SD) 13.5 (3.2)	Mean (SD) 13.1 (3.2)	0.564
Gender *Male* *Female*	N (%) 4 (10.3) 35 (89.7)	N (%) 7 (17.5) 33 (82.5)	0.518
Ethnicity *Non Hispanic White* *Non Hispanic AA* *Asian/PI/Other* *Hispanic*	N (%) 5 (12.8) 7 (17.9) 5 (12.8) 22 (56.4)	N (%) 3 (7.5) 15 (37.5) 2 (5.0) 20 (50.0)	0.197
Clinical Symptoms			
 Dry Eyes *Dry Mouth* *Parotid Gland Swelling*	N (%) 1 (2.6) 4 (10.3) 1 (2.6)	N (%) 2 (5) 4 (10) 1 (2.5)	 1.000 1.000 1.000
Renal Changes			
% of LN	N (%) 14 (35.9)	N (%) 18 (45.0)	0.494
Renal interstitial changes	N (%) 12 (30.8)	N (%) 17 (42.5)	0.352
Pertinent Laboratory and Markers of Disease Activity			
Total IgG	Mean (SD) 1436.8 (675.2)	Mean (SD) 1735.4 (507.8)	0.036**
SLEDAI	Median (IQR) 11 (7.0, 22.0)	Median (IQR) 15.5 (9.5, 22.5)	0.112

SD: standard deviation, AA: African American, PI: Pacific Islander, LN: lupus nephritis, IQR: interquantile range ** Represents p values <0.05

eyes (5%), dry mouth (10%), parotid gland swelling (2.5%), none of which was statistically different from control. Mean IgG was higher in the study group when compared to controls (1735.4 v 1436.8, p =0.036). SLEDAI was higher among study patients at presentation, although not statistically different (15.5 vs 11, p=0.112); no significant difference was observed at 6 or 12 months. There was a similar percentage of lupus nephritis between study group and control at presentation (45% vs 35.9%, p=0.494). There were increased interstitial changes in the study group, although not statistically significant (42.5% vs 30.8%, p=0.352). Table 1 summarizes pertinent clinical features and characteristics between groups.

Conclusion: This study showed no increased incidence of sicca symptoms or salivary gland swelling among pediatric SLE patients with anti-SSA and SSB, findings well defined among adults. The study showed that patients with these ANAs have significantly higher IgG at presentation than control. Having SSA and SSB antibodies is not associated with a higher percentage of lupus nephritis. Our study is limited by its retrospective nature and small sample size. A large scale, multicenter study is recommended to study the significance of these antibodies in pediatric SLE.

Disclosure: Y. Pina: None; **D. Janysek**: None; **D. Guffey**: None; **M. Pereira Palacios**: None; **M. De Guzman**: None.

Abstract Number: 115

Development of an Electronic Clinical Phenotype to Identify Potential Study Subjects with Juvenile Arthritis

Alysha Taxter[1], Marc Natter[2], Min-Lee Chang[2], Laura Schanberg[3], Valarie Morrow[4], Eveline Wu[5], Tedryl Bumpass[4], Alex Fist[4], Meg Waite[6], Vincent Del Gaizo[7], Melanie Kohlheim[7] and CARRA Registry Investigators[7], [1]Nationwide Children's Hospital, Columbus, OH, [2]Boston Children's Hospital, Boston, MA, [3]Duke University Medical Center, Durham, NC, [4]Duke University, Durham, NC, [5]UNC Chapel Hill, Chapel Hill, NC, [6]Boston Children's Hospital, Boston, MA, [7]CARRA, Washington, DC

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: The LIMIT-JIA trial is the first study of the use of biologic therapy to prevent disease extension in children with newly diagnosed, uncomplicated, oligo-articular course (“early limited”) juvenile idiopathic arthritis (JIA). Study recruitment requires extensive efforts to identify and approach eligible patients. Potential subject identification was initially conducted manually by study staff. This project developed and implemented a computable phenotype to enhance timely identification of potentially eligible subjects for this randomized trial.

Methods: Monthly reports of potentially eligible subjects for the trial were generated by developing, testing, and then applying an “early limited JIA” computable phenotype to The Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry records throughout 2022. The algorithm inclusion criteria included individuals diagnosed with oligoarticular JIA within the preceding six months; exclusion criteria included extension to polyarticular course, uveitis, treatment with DMARDs, or comorbid diagnoses of psoriasis, inflammatory bowel disease, or sacroiliitis. Study sites received monthly reports and annotated these reports with actual eligibility status of potential subjects from the CARRA Registry. Several sites configured on-demand reports using their electronic health record (EHR) software to identify potential subjects. All sites were surveyed about enrollment using CARRA Registry and their EHR reports. Data analyses were conducted and descriptive statistics were compiled.

Results: There were 15 study sites with 50 potentially eligible subjects identified by the computable phenotype during the study period. The median age was 6.7 [3.6, 12.1] years, and 36 (72%) were female. Thirty-two (64%) were not eligible for the study. The most common reasons that subjects were ineligible were related to temporal factors and included: 13 (41%) had recruitment delayed beyond the six-month enrollment window, 8 (25%) had started systemic therapy, and 5 (16%) no longer met oligoarticular JIA criteria (e.g. developed psoriasis or sacroiliitis) or had a change of diagnosis. Other ineligibility reasons included inability to be recruited virtually, language barriers, and no follow-up scheduled within the enrolment time window. Of the 18 eligible subjects, 9 (50%) declined enrollment. Nine study sites responded to monthly surveys; only three were using their EHR reporting tools to complement CARRA Reports, and most sites reviewed EHR reports monthly. Sixteen subjects were identified using EHR tools, 3 were eligible for the study, but none were approached for the study.

Conclusion: Novel tools to identify subjects for study recruitment are necessary to improve study recruitment, particularly in studies with time-sensitive inclusion criteria and pediatric prevention trials with active treatment arms. Development of accurate computable phenotypes to assist with subject identification requires accurate data and timely recruitment strategies.

Disclosure: A. Taxter: None; **M. Natter**: None; **M. Chang**: None; **L. Schanberg**: Bristol-Myers Squibb(BMS), 5, Sanofi, 12, DSMB member, UCB, 12, DSMB chair; **V. Morrow**: None; **E. Wu**: Enzyvant, 2, Pharming Healthcare, Inc, 2; **T. Bumpass**: None; **A. Fist**: None; **M. Waite**: None; **V. Del Gaizo**: None; **M. Kohlheim**: None; **C. Investigators**: None.

Abstract Number: 116

Clinical Significance of Anti-Scl-70 Antibodies in Pediatric Lupus Patients: A Single Center Cohort

Dawn Janysek[1], Yiressy Pina[2], Danielle Guffey[3] and Marietta De Guzman[1], [1]Baylor College of Medicine, Texas Children’s Hospital, Houston, TX, [2]Baylor College of Medicine, Texas Children’s Hospital, Houston, TX, [3]Baylor College of Medicine, Houston, TX

SESSION INFORMATION

Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Production of autoantibodies is a hallmark of SLE, with ANAs as a required diagnostic feature and anti-dsDNA and anti-Smith antibodies being disease-specific. Anti-Scl-70 antibodies are clinically and prognostically associated with SSc, a disease characterized by cutaneous and visceral fibrosis and microvascular damage. The most common symptoms of SSc include Raynaud's phenomenon, skin thickening, telangiectasias, and pulmonary, gastrointestinal, and renal disease. Adult studies have demonstrated an increased risk for pulmonary hypertension and kidney disease in SLE patients with anti-Scl-70 compared to those without. This study aims to determine the significance of anti-Scl-70 in pediatric SLE patients.

Methods: We performed a retrospective review of the electronic medical records of patients diagnosed with SLE, as per SLICC and EULAR/ACR classification criteria, at Texas Children's Hospital between 2018 and 2021. Twenty-seven had positive anti-Scl-70 as reported by the University of Missouri lab. The data collected included clinical and diagnostic features at presentation, 6 months, and 12 months. Patients with anti-Scl-70 were compared to patients without anti-Scl-70. Characteristics were summarized by group and compared using an independent t-test, Wilcoxon rank-sum test, Chi-squared test, or Fisher's exact test, as appropriate.

Results: Twenty-seven patients with anti-Scl-70 were analyzed against 52 patients without. The two groups were of similar demographics (Table 1). Study patients were more likely to have anti-dsDNA and were noted to have higher SLEDAI scores and IgG levels at diagnosis. These patients were noted to have a higher prevalence of lupus nephritis and acute cutaneous features, such as malar rash and photosensitivity.

Table 1: Pediatric SLE Controls vs. Anti-Scl-70 at Presentation

	Control (N=52)	Study group (N=27)	P Value
Demographics			
Age	Mean (SD) 13.1 (3.3)	Mean (SD) 13.7 (2.9)	0.391
Gender *Male* *Female*	N (%) 5 (9).6 47 (90.4)	N (%) 6 (22.2) 21 (77.8)	0.172
Ethnicity *Non Hispanic White* *Non Hispanic AA* *Asian/PI/Other* *Hispanic*	N (%) 6 (11.5) 14 (26.9) 5 (9.6) 27 (51.9)	N (%) 2 (7.4) 8 (29.6) 2 (7.4) 15 (55.6)	0.961
Clinical Symptoms			
Raynaud's Phenomenon	N (%) 8 (15.4)	N (%) 1 (3.7)	0.155
Skin Tightening	0 (0)	0(0)	
Connective Tissue Disease	0 (0)	0(0)	
GI Symptoms	15 (28.8)	12 (44.4)	0.213
Pulmonary Disease	7 (13.5)	3 (11.1)	1.000
Acute cutaneous features	9 (17.3)	12 (44.4)	0.015**
Renal Changes			
Lupus Nephritis	N (%) 16 (30.8)	N (%) 16 (59.3)	0.017**
Renal interstitial changes	N (%) 15 (28.8)	N (%) 14 (51.9)	0.053
Pertinent Laboratory and Markers of Disease Activity			
Total IgG	Mean (SD) 1477.4 (680.7)	Mean (SD) 1776.8 (415.6)	0.042**
SLEDAI	Median (IQR) 10.5 (6.5, 19.0)	Median (IQR) 20 (13.0, 28.0)	<0.001**
dsDNA	N (%) 23 (44.2)	N (%) 27 (100)	<0.001**

** Represents p values <0.05

Table 2: Pediatric SLE Controls vs. Anti-Scl-70 After Controlling for Anti-dsDNA

	Control (N=23)	Study group (N=27)	P Value
Demographics			
Age	Mean (SD) 12.3 (3.9)	Mean (SD) 13.7 (2.9)	0.168
Gender *Male* *Female*	N (%) 3 (13.0) 20 (87.0)	N (%) 6 (22.2) 21 (77.8)	0.479
Ethnicity *Non Hispanic White* *Non Hispanic AA* *Asian/PI/Other* *Hispanic*	N (%) 2 (8.7) 6 (26.1) 3 (13.0) 12 (52.2)	N (%) 2 (7.4) 8 (29.6) 2 (7.4) 15 (55.6)	0.933
Clinical Symptoms			
Raynaud's Phenomenon	N (%) 5 (21.7)	N (%) 1 (3.7)	0.082
Skin Tightening	0 (0)	0(0)	
Connective Tissue Disease	0 (0)	0(0)	
GI Symptoms	6 (26.1)	12 (44.4)	0.241
Pulmonary Disease	4 (17.4)	3 (11.1)	0.689
Acute cutaneous features	2 (8.7)	12 (44.4)	0.010**
Renal Changes			
Lupus Nephritis	N (%) 6 (26.1)	N (%) 16 (59.3)	0.024**
Renal interstitial changes	N (%) 6 (26.1)	N (%) 14 (51.9)	0.086
Pertinent Laboratory and Markers of Disease Activity			
Total IgG	Mean (SD) 1588.7 (698.0)	Mean (SD) 1776.8 (415.6)	0.259
SLEDAI	Median (IQR) 11 (8.0, 18.0)	Median (IQR) 20 (13.0, 28.0)	0.001**
dsDNA	N (%) 23 (100)	N (%) 27 (100)	

** Represents p values <0.05

There was no significant difference in Raynaud's phenomenon, skin tightening, pulmonary disease, or gastrointestinal symptoms. When controlling for anti-dsDNA, as shown in Table 2, SLEDAI scores, IgG levels, lupus nephritis, and acute cutaneous features continued to be significantly higher in anti-Scl-70 patients compared to controls.

There was no significant difference in IgG levels or prevalence of lupus nephritis between the two groups at 6-month and 12-month visits. Acute cutaneous features were more prevalent in anti-Scl-70 patients at the 6-month visit ($p = 0.039$) but were no longer significant at subsequent intervals. SLEDAI scores were higher at the 12-month visit ($p = 0.026$) in study subjects but demonstrated no difference at the 6-month visit.

Conclusion: This study showed associations between anti-Scl-70 and higher SLEDAI scores, higher IgG levels, lupus nephritis, and acute cutaneous features in pediatric SLE patients at diagnosis. These higher SLEDAI scores and IgG levels may suggest more active disease. After controlling for anti-dsDNA, there was no change in the relationship between anti-Scl-70 positivity and the above variables. Thus, the presence of dsDNA in anti-Scl-70 positive SLE patients cannot fully explain the relationships demonstrated in this study. Anti-Scl-70 in pediatric SLE patients may predict specific disease presentations and outcomes at 6-month and 12-month visits.

Disclosure: D. Janysek: None; **Y. Pina**: None; **D. Guffey**: None; **M. De Guzman**: None.

Abstract Number: 117

A Descriptive Study of Patients with Multisystem Inflammatory Syndrome in Children

Alexis Begezda, Penn State, State College, PA

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Children with childhood-onset systemic lupus erythematosus (cSLE) experience more severe disease than their adult counterparts, in addition to high rates of clinical depressive symptoms (30%) and fatigue (65%) (1). A "high- risk" profile categorized by clinically elevated depressive symptoms and fatigue has been shown to be related to reduced health-related quality of life and poorer patient outcomes in cSLE patients (2). The **T**reatment and **E**ducation **A**pproach for **CH**ildhood-onset lupus (TEACH) is a tailored cognitive behavioral therapy (CBT) for youth with cSLE that may address these symptoms. (3). TEACH is currently being tested in an RCT with promising results after 6 weeks. However, it is unknown how the "high- risk" group may respond to this program. This study aims to assess the impact of risk status on the outcomes of cSLE patients undergoing CBT.

Methods: This study uses data from an ongoing multisite clinical trial of youth (average age of 17 years, 100% female, 70% identifying as from a minority group, n=46/60 currently completed evaluation for the study). All of these patients met the ACR criteria for cSLE. 16 of these youth have thus far been randomized to and completed TEACH. They were grouped into low-

	Alpha (n=24)	Delta (n=21)	Omicron (n=10)
Mean age (years)	8.3	9.2	7.5
Admitted to the ICU	11 (46%)	13 (62%)	3 (30%)
Required vasopressors	7 (30%)	12 (57%)	3 (30%)
Mean brain natriuretic peptide (BNP) (pg/ml)	10214	11786	8779
Mean CRP (mg/dl)	17	17	16
Mean ferritin (ng/ml)	592	978	806
Patients with elevated troponin	13 (54%)	13 (62%)	3 (30%)
Mean troponin (ng/ml)	0.048	0.073	0.169
Patients with a dilated coronary artery	6 (25%)	3 (14%)	0
Patients with depressed systolic function on echo	10 (42%)	10 (48%)	5 (50%)
Length of hospitalization (days)	8	6	7

Comparison of MIS-C between the COVID variants. Emergence of dominant variants was defined by the CDC's infection rates.

	Alpha Variant Dominance (03/28/2020-07/02/2020)	Delta Variant Dominance (07/03/2021-12/24/2021)	Omicron Variant Dominance (12/25/2021-10/22/2022)
Total number of COVID positive cases	1644	1477	2774
Total number of MIS-C cases	24	21	10

COVID and MIS-C incidence rates for each COVID variant.

	ICU (n=27)	Non-ICU (n=28)	P-value
CRP (mg/dl) (reference range)	18.7 (<0.03)	15.5 (<0.03)	0.18
ESR (mm/hr) (reference range)	53.8 (0-10)	63.9 (0-10)	0.29
Ferritin (ng/ml) (reference range)	1048 (30-400)	518 (30-400)	0.001
	Vasopressor Use (n=22)	No Vasopressor Use (n=33)	
Troponin (ng/ml) (reference range)	0.213 (<0.010)	0.021 (<0.010)	0.05
BNP (pg/ml) (reference range)	15849 (<125)	7023 (<125)	0.006
	Steroids within 24 Hours of Admission (n=35)	Steroids Started After 24 Hours of Never Started (n=20)	
Length of Hospitalization (days)	6.3	9.0	0.02

Comparison of mean laboratory values and length of hospitalization for all patients with MIS-C. Mean CRP, ESR, and Ferritin values are compared between patients admitted and not admitted to the ICU. Cardiac markers (troponin and BNP) are compared between patients who required vasopressors and patients who did not. Length of hospitalization is compared between patients who received steroids within 24 hours of admission and those who received steroids later or not at all.

or high-risk categories based on baseline depression and fatigue t-scores (>70; >2SD from mean) (3). Repeated measures ANOVAs were conducted to compare the low- and high-risk groups pre and post 6 week TEACH protocol for main study outcomes (e.g. depressive symptoms (CDI-II), fatigue (PROMIS), and pain (VAS)) and additional exploratory outcomes

(anxiety (SCARED), disease severity (SLEDAI), and health-related quality of life (Peds-QL)). Independent samples t-test was used to determine if baseline outcome measures were significantly different between low- and high-risk groups.

Results: Out of the 16 patients who have completed the TEACH protocol, 6 (37.5%) met criteria for inclusion in the high-risk group. There was no significant difference between risk groups based on age, disease duration, or self-reported race. There was a significant difference between groups in baseline depression scores only ($p= 0.005$). At week 6 after completion of TEACH, depression scores decreased by 26% in the high- risk group, while the low- risk group only decreased 6% ($p< 0.001$; Table 1). Fatigue, anxiety, health-related quality of life, and disease severity followed the same trends (see Figure 1). Despite these trends, there were no significant differences found between the low- and high-risk groups in any outcomes other than depressive symptoms.

Conclusion: There is very little literature on nonpharmacologic treatments for cSLE like CBT (4). These data reveal interesting trends that CBT may be more effective in cSLE patients with high levels of fatigue and depression. Some results may be driven by the fact that the high-risk group begins with higher baseline symptoms. Lack of significant difference between groups may be attributed to a small sample size and these analyses should be replicated with a larger data set (e.g., upon completion of the RCT) in the future. Although the high-risk group shows more improvement overall, this should not shadow that both high- and low- risk groups had improved outcomes with the TEACH protocol.

Disclosure: A. Begezda: None.

Abstract Number: 118

Serum Hepatocyte Growth Factor in Children with Juvenile Idiopathic Arthritis

Natalia Shevchenko and **Olga Pavlova**, V. N. Karazin Kharkiv National University, Kharkiv, Ukraine

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Only limited data are available on the risk of liver fibrosis in patients with rheumatoid arthritis (RA) and juvenile idiopathic arthritis (JIA) on long-term methotrexate (MTX) treatment, despite MTX association with a range of liver related adverse events there are also controversial results. As a result, common extra-articular manifestation is the leading cause of mortality in RA. Thus, there is a need of liver fibrosis formation predictors and its reparation capacity learning in children with juvenile idiopathic arthritis (JIA) to reduce future risks.

Purpose of our study was to evaluate serum levels of hepatocyte growth factor (HGF) in children with juvenile idiopathic arthritis treated with methotrexate.

Methods: 104 patients with JIA were included in this 4-years prospective study. Almost half of children had polyarthritis (50,96%) JIA variant. In 104 children with JIA (mean age 13.3 yrs, 59,62% female, mean age of JIA onset 7.2 yrs, mean disease duration 5.06 yrs) were treated with MTX 75.96% (vs. 24.04% not treated with MTX, 16,35% were prescribed MTX, but they didnt receive any dose yet on investigation day). Among patients treated with MTX 4.81% had dose less than 10 mg/m^2/week, 32.69% 10-12.5 mg/m^2/week, 31.73% 12.6 - 15 mg/m^2/week, 6.73% over 15 mg/m^2/week. HGF levels were determined by HGF ELISA kits (Elabscience, USA). Serum levels of HGF were analyzed depending on patients gender,

age, and age of JIA onset, its variant, duration, activity, and presence of MTX in treatment and its dose. Levels of rheumatoid factor (RF), antinuclear antibodies (ANA), erythrocyte sedimentation rate (ESR), C-reactive protein (C-RP), circulating immune complex (CIC) and antistreptolysin-O (ASL-O) were analyzed in this study. This work complies with the guiding principles found in the Declaration of Helsinki of the World Medical Association.

Results: We evaluated HGF level for children with JIA (min: 89.13 pg/ml; Me:168.04 pg/ml; max: 629.13 pg/ml). The average level of HGF significantly increased in children with age of JIA onset from 6 to 10years versus age of JIA onset 11–14 years ($p = 0.009$). Mostly HGF was correlated with CIC (among: patients with no activity according to JADAS-27 $r= -0.99$, $p < 0.05$, high disease activity according to JADAS-27 $r=0.95$, $p < 0.05$), with MTX dose (among: girls $r= -0.38$, $p < 0.05$). According to regression analysis, HGF in children with JIA depended on the age of the patients, JADAS 27, ANA, RF and MTX mg/m^2/week.

Conclusion: CIC and MTX dose were corresponded with high HGF level. Lower serum HGF, which can be considered as inhibition of the reparative capabilities of the liver, were mainly in girls, in patients aged 10–13 years, with the JIA onset at 3–5 years, polyarticular variant and high JIA activity according to JADAS-27.

Disclosure: N. Shevchenko: None; **O. Pavlova**: None.

Abstract Number: 119

Laser Flare Photometery in the Pediatric Rheumatology Clinic as a Screening Tool for Juvenile Idiopathic Arthritis Associated Uveitis

Kaleo Ede[1], Michael Shishov[2], Elisa Wershba[2], Nikita Goswami[2], Sabrina Gorry[2], Malin Jospeh[2], Lucia Mirea[2] and James O'neil[2], [1]Phoenix Children's Hosptial; University of Arizona College of Medicine- Phoenix, Phoenix, AZ, [2]Phoenix Children's Hospital, Phoenix, AZ

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Juvenile Idiopathic Arthritis (JIA) is the most common chronic rheumatic disease in childhood, affecting 1 to 22 per 100,000 children. JIA-associated uveitis is known to occur in up to 12% of JIA patients in the United States. Visual complications are present in up to 2/3 of patients, with blindness in as many as 30-50% of affected eyes. The gold standard to diagnose JIA-associated uveitis is slit lamp examination by trained eye specialists using a subjective grading scale (Standardization of Uveitis Nomenclature or SUN) to quantify anterior segment inflammation. Recently, laser flare photometery (LFP) has been used in multiple studies as an objective, non-invasive, and quantitative method to measure anterior chamber inflammation. LFP has been used by ophthalmologists to monitor JIA-associated uveitis patients as well as other forms of pediatric non-infectious chronic anterior uveitis.

The purpose of this study is to investigate if LFP can be used in a pediatric rheumatology office setting to diagnose patients with JIA-associated uveitis.

Methods: This observational study was approved by the Phoenix Childrens IRB, and enrolled patients aged 4-16 years diagnosed with JIA based on the 2001 ILAR classification criteria during 2019-2022. Exclusion criteria were having a history of JIA-associated uveitis or any other form of uveitis, as well as history of pacemaker placement. All patients underwent at

Table 1. Demographic characteristic data at baseline

Characteristic	Overall (N=58 patients)
Gender	
Female	43 (74.1%)
Race	
Asian	2 (3.4%)
Black/African American	2 (3.4%)
Hispanic/Latino	15 (25.9%)
Native American	1 (1.7%)
White/Caucasian	37 (63.8%)
Other	1 (1.7%)
Age at diagnosis (years)	
Mean (SD)	8.4 (4.2)
Median	8.6
Q1, Q3	4.9, 12.2
Range	(1.2, 16.3)
Age at enrollment (years)	
Mean (SD)	11.9 (3.2)
Median	12.4
Q1, Q3	9.9, 14.6
Range	(4.8, 16.5)

Table 2. JIA subtype and treatment characteristic data at baseline

Characteristic	Overall (N=58 patients)
JIA subtype	
Oligoarticular	19 (32.8%)
RF (-) Polyarticular	12 (20.7%)
RF (+) Polyarticular	8 (13.8%)
Systemic	3 (5.2%)
Enthesitis-related arthritis	10 (17.2%)
Psoriatic	4 (6.9%)
Undifferentiated	2 (3.4%)
Treatment at baseline	
Methotrexate	26 (44.8%)
Adalimumab	22 (37.9%)
Etanercept	5 (8.6%)
Tocilizumab	6 (10.3%)
Other biologic	0
None	15 (25.9%)

Table 3. Uveitis activity based on slit lamp exam versus Laser Flare Photometry (LFP)

Slit lamp result or equivalence	*Slit lamp exam N=138*	*LFP exam N=135* [a]
Normal (0.5 to 9 LFP flare)	138 100.0%	131 97.0%
Low amount of flare (10 to 25 LFP flare)	0 0.0%	1 0.7%
1+ to 2- slit lamp reading (76 to 125 LFP Flare)	0 0.0%	3 2.2%
Total false positive rate N, % (95% CI)	--	4 3.0% (0.8%, 7.4%)

[a] 3 patients only had one eye measurement taken at their rheumatology visit
[b] A false positive is defined as a normal result on the slit lamp exam and anything other than a normal result on the LFP exam

least one evaluation of both eyes using a Kowa FM-600 laser flare photometer, as well as a standard slit lamp examination by optometry or ophthalmology during routine clinical care. Data collected at patient visits included demographics, JIA characteristics, treatment medications, LFP readings, and AC cell grade scored by SUN grading system. Data were summarized using descriptive analyses and the uveitis false positive rate was calculated along with 95% confidence interval.

Results: Subjects included 58 pediatric patients diagnosed with JIA with data obtained for 138 eyes at 69 visits. The mean age was 8.4 years (range: 1.2-16.3 years) at diagnosis and 11.9 (range: 4.8-16.5) years at enrollment. Of the 58 patients, the majority 43 (74.1%) were female (table 1). Most common JIA subtypes included 19 (32.8%) patients with persistent oligoarticular JIA, and 12 (20.7%) with RF negative polyarticular JIA. At enrollment, 15 patients (25.9%) were on no medications, with 26 (44.8%) on methotrexate, 22 (37.9%) on adalimumab, 6 (10.3%) on tocilizumab, and 5 (8.6%) on etanercept (table Treatment at baseline included methotrexate (44.8%), adalimumab (37.9%), etanercept (8.6%), tocilizumab (10.3%) and none (25.9%) (table 2). During the study period, no eye exams detected active uveitis based on slit lamp exam with a SUN grade over 0. However, of the 135 LFP readings, 131 (97.0%) were normal, yielding a false positive rate of 3% (95% CI of 0.8%, 7.4%) (table 3).

Conclusion: Laser Flare Photometry is a non-invasive tool that can be utilized in the pedaitric rheumatology clinic to evaluate for JIA-associated uveitis. There is a low false positive rate of LFP when compared with standard slit lamp exam. Future studies will examine LFP to monitor uveitis activity in patients with JIA-associated uveitis to help guide treatment decisions.

Disclosure: K. Ede: None; **M. Shishov**: AbbVie/Abbott, 6, Novartis, 6; **E. Wershba**: None; **N. Goswami**: None; **S. Gorry**: None; **M. Jospeh**: None; **L. Mirea**: None; **J. O'neil**: None.

Abstract Number: 120

The Brazilian Registry of Juvenile Dermatomyositis (JDM): I- Onset Clinical Features and Disease Activity Scores by DAS-20 over 2-Years-Follow Up

Beatriz Carneiro[1], Adriana Elias[1], Teresa Robazzi[2], Ana Julia Moraes[3], Sheila Oliveira[4], Flavio Sztajnbok[5], Luciana Carvalho[6], Luciana Marques[7], Silvana Sacchetti[8], Maria Teresa Terreri[9], Simone Appenzelle[10], Roberto Marini[11], Andre Cavalcante[12], Marcia Bandeira[13], Cristina Magalhaes[14], Melissa Fraga[15], Iloite Scheibel[16], Isabela Daud[1], Darcisio Antonio[17], Claudio Len[18], Clovis Silva[19], Taciana Fernandes[17] and Claudia Magalhaes[20], [1]Instituto da Criança - Universidade de São Paulo (USP), São Paulo, Brazil, [2]Universidade Federal da Bahia, Brazil, [3]Universidade Federal do Para, Brazil, [4]Universidade Federal do Rio de janeiro, Rio de Janeiro, [5]Universidade Estadual do Rio de Janeiro, Rio de Janeiro, Brazil, [6]Universidade de Sao Paulo- Ribeirao Preto, São Paulo, Brazil, [7]Hospital Albert Sabin, Brazil, [8]Santa Casa de Sao Paulo, Brazil, [9]UNIFESP, São Paulo, Brazil, [10]University of Campinas, Campinas, Sao Paulo, Brazil, [11]UNICAMP, São Paulo, Brazil, [12]Hospital Materno-Infantil de Goiania, Brazil, [13]Hospital Pequeno Principe- Curitiba, Brazil, [14]Hospital Jose de Alencar - Brasilia, Brazil, [15]Hospital Darcy Vargas, Brazil, [16]Hospital Conceição de Porto Alegre, Brazil, [17]Universidade Estadual Paulista (UNESP) Botucatu, Brazil, [18]Universidade Federal de São Paulo - Unifesp, São Paulo, Brazil, [19]Universidade de São Paulo, São Paulo, Brazil, [20]São Paulo State University, Pediatric Rheumatology Division, Botucatu, Brazil

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: A national registry was set up, enrolling new onset JDM cases in 18 hospitals, during 3-years (2015-2018) with 2-years follow up, in a low resource country aiming at first evaluating clinical presentation and disease activity scores by DAS-20 Disease Activity Scoring tool over 2-years-follow up.

Methods: Questionnaires inquiring cases, within 6 months of the first symptoms, were filled out by participating physicians with clinical and functional assessments at baseline, 6, 12, 18 and 24 months. Data capture, storage and analysis were carried in one of centres. Protocol ethics approval and informed consent was obtained for all. Disease Activity Score (DAS20) tool training was provided (Bode RK et al. Arthritis Rheum 2003, 49:7-15). Investigations included: muscle enzymes, full blood count, ESR, renal function, MRI, EMG, and muscle biopsy, calcinosis image, overlap features and auto-antibodies.

Results: Ninety six cases were enrolled from 18 referral hospitals, 60 towns (zip code) in the five geographic regions of a continental country (207.75 million population). Of those cases, 61 (64%) were female with mean (SD) age 10.7(4.2) years. Presenting signs and symptoms frequency was: face and extremities rash (97%), generalized muscle weakness (95%), fatigue (88%), myalgia (87%), arthralgia (61%), fever (51%), irritability (41%), facial oedema (47%), body oedema (27%), weight loss (37%), joint oedema (37%), stiffness (37%), dysphagia (40%), dysphonia (27%), dyspnoea (20%), abdominal pain (24%), chest pain (6%). Onset calcinosis was described in 12 of 91 reports (13%). Only 42 cases performed EMG and 32 had a muscle biopsy and MRI was performed in 22 and US in 7. The muscle enzymes values mean (SD) IU was: CK 1835 (3822), LDH 972 (801), AST 204 (622), ALT 118 (208), Aldolase 20 (25,9). Treatment received, included: 81/90 (90%) high dose oral prednisone, 46/90 (51%) IV methylprednisolone, 72/90 (80%), methotrexate, 40/91 (44%) Hydroxycloroquine, 15/91 (16%) IVIG, 3/91 (3%) cyclosporine, 3/90 (3%) azathioprine, 4/90 (4%) IV cyclophosphamide and 1/90 (1%) received rituximab. Disease activity status scored by DAS20 at baseline and follow up indicated wide variation and significant improvement over 2-years. The mean± SD scores of DAS20 were: baseline n=96 (8±5), n=96 (6m) (2.7±4.4), n=96 (12m) (1.5±3.2), n=73 (18m) (1±3) and n=96 (24m) (0.5±2.3). Values compared by Poisson model (p 0.0001) and Wald post-test indicated significant difference between each of the visits, from baseline to 24 months.

Conclusion: This preliminary analysis of a registry included case ascertainment and feasibility of standardized outcome measures for JDM (DAS20), under mainstay treatment with prednisone and methotrexate. It indicated a good performance of clinical assessment by a quantitative tool, performed by physicians in low resource settings, in spite of limited access to all diagnostic pathway tools, imaging and biomarkers.

Disclosure: B. Carneiro: None; **A. Elias**: None; **T. Robazzi**: None; **A. Moraes**: None; **S. Oliveira**: None; **F. Sztajnbok**: None; **L. Carvalho**: None; **L. Marques**: None; **S. Sacchetti**: None; **M. Terreri**: None; **S. Appenzelle**: None; **R. Marini**: None; **A. Cavalcante**: None; **M. Bandeira**: None; **C. Magalhaes**: None; **M. Fraga**: None; **I. Scheibel**: None; **I. Daud**: None; **D. Antonio**: None; **C. Len**: None; **C. Silva**: None; **T. Fernandes**: None; **C. Magalhaes**: None.

Abstract Number: 121

The Effectiveness of Tonsillectomy in Periodic Fever, Aphthous Ulcer, Pharyngitis, and Adenitis Syndrome in Pediatric Patients

John Storwick[1] and Marinka Twilt[2], [1]Alberta Children's Hospital/University of Calgary, Calgary, AB, Canada, [2]Alberta Children's Hospital, Calgary, AB, Canada

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Periodic Fevers with Aphthous Ulcers, Pharyngitis, and Adenitis Syndrome (PFAPA Syndrome) is the most common pediatric periodic fever syndrome. The most recent diagnostic criteria was described by Thomas et al. in 1999 and defines PFAPA as; regular periodic fevers lasting 3 to 7 days, occurring every 3 to 8 weeks, accompanied by

at least one of the following symptoms: aphthous stomatitis, pharyngitis, and cervical adenitis. Between febrile episodes the patient is well and demonstrates normal growth and development. Treatment of PFAPA has considerable heterogenicity between practitioners, and there is no standard of care given a lack of clinical trials. Fortunately, PFAPA is relatively benign and will usually resolve spontaneously over time. In 1989, Abramson and colleagues described successful fever resolution post-tonsillectomy. Post-tonsillectomy, patients show a complete response with immediate cessation of fevers, a partial response with fever cessation followed by recurrence, or a failure with no fever cessation. The role of tonsillectomy in PFAPA treatment remains controversial. Our study aimed to determine the effectiveness of tonsillectomy as curative treatment of PFAPA compared to conservative non-surgical management.

Methods: We conducted a single center retrospective cohort study of children with PFAPA who were seen at the Alberta Childrens Hospital Rheumatology Clinic from January 2015 to July 2020. Inclusion criteria was based on the modified Thomas criteria for PFAPA. Exclusion criteria included patient with the Familial Mediterranean Fever gene variants, monogenic autoinflammatory diseases, and those that underwent tonsillectomy for another reason. We compared the time to fever resolution between the patients who underwent tonsillectomy and those who did not.

Results: We reviewed 113 charts of potential PFAPA patients and identified 34 patients who met inclusion criteria for typical PFAPA syndrome. 13 (38.2%) underwent tonsillectomy and 21 did not. In the tonsillectomy group, 10 were found to have complete response with immediate cessation of their fevers following tonsillectomy, 2 had a partial response, and 1 had no response. Patient characteristics were comparable; average age of fever onset was 2.9 and 3 years in the tonsillectomy and non-tonsillectomy groups and the average duration of PFAPA symptoms was 2.6 years and 3.2 years respectively.

Conclusion: Our study demonstrates that tonsillectomy may be beneficial in shorting the duration of PFAPA symptoms by approximately 6 months. Given the associated surgical risks, careful consideration should be given prior to recommending tonsillectomy as curative treatment.

Disclosure: J. Storwick: None; **M. Twilt**: None.

Abstract Number: 122

Juvenile Arthritis in Minnesota: Geospatial Variability and Environmental Exposures in Juvenile Idiopathic Arthritis

Colleen Correll[1], Austin Rau[2] and Jesse Berman[2], [1]University of Minnesota, Minneapolis, MN, [2]University of Minnesota, Minneapolis

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: It is widely accepted that juvenile idiopathic arthritis (JIA) is caused by a combination of genetic and environmental factors, but the potential environmental triggers are not well-known. Some studies have shown air pollution as a possible risk factor for JIA and rheumatoid arthritis. We mapped children with JIA living in Minnesota and used publicly available geospatial data to assess whether air pollution exposure and/or community social vulnerability were associated with JIA.

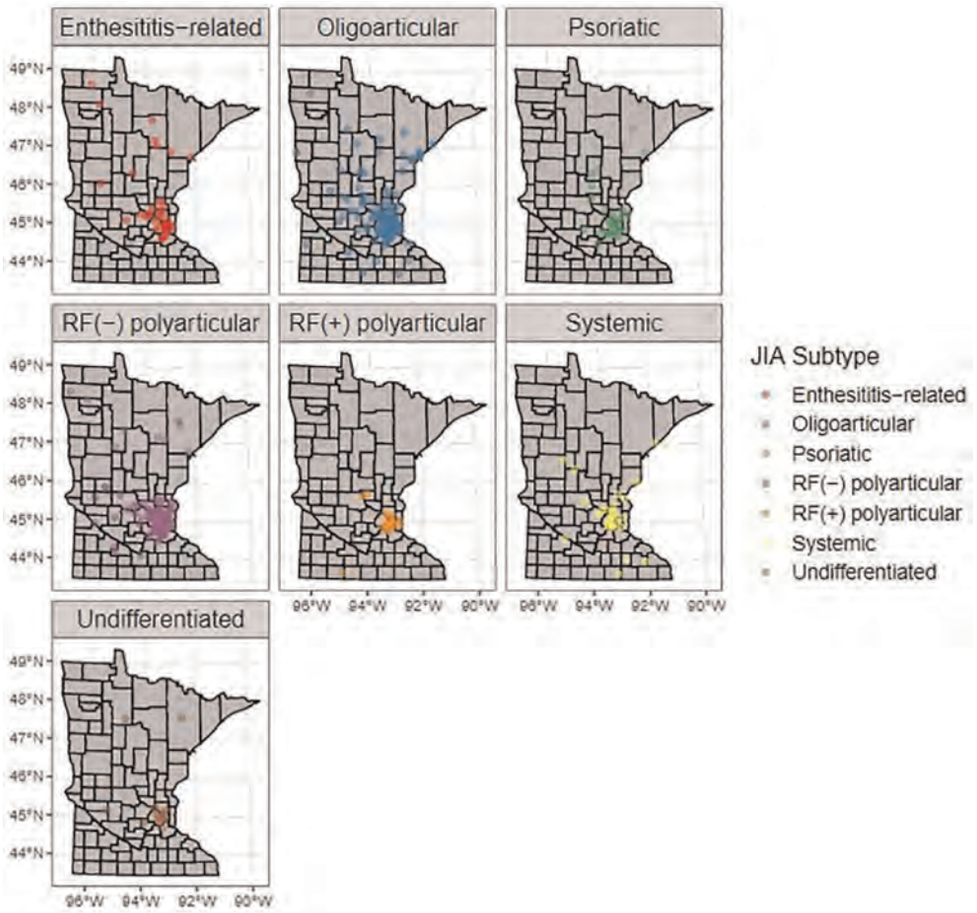


Figure 1. Geospatial distribution of children with JIA living in Minnesota by JIA subtype

Methods: We geocoded 4 years of patients enrolled in the Juvenile Arthritis in Minnesota (JAMinn) study who had a current address and calculated county-level prevalence rates of JIA throughout the state. Enrollment criteria for JAMinn are age < 16, JIA diagnosis by treating rheumatologist, and Minnesota residence at the time of enrollment. The vast majority of patients were enrolled at the University of Minnesota (UMN). We also studied markers of air pollution as factors associated with JIA. To avoid biases due to referral patterns, we limited air pollution exposures to children who lived within the 7-county metro area of Minneapolis and Saint Paul, MN, which is where the UMN clinic is located. We used census tract-level data and compared weighted averages of modeled annual air pollution concentrations including fine particulate matter 2.5 (PM2.5), sulfur dioxide (SO2), and nitrogen dioxide (NO2), and the Centers for Disease Control and Preventions social vulnerability index (SVI) in children with JIA compared to the general population of children in the metro area.

Results: 389 patients with JIA were included in the geospatial mapping throughout the state of Minnesota by JIA subtype (Figure 1). Prevalence was mapped at the county level (Figure 2). The prevalence of JIA in Hennepin County, the most populated county in Minnesota, ranged from 60-74 per 100,000 children. Five counties had relatively higher prevalence rates (75-129) per 100,000 children. Forty-five percent (175/389) of patients with JIA lived in the metro area and were included in the air pollution and SVI comparison (Table 1). There were 230 census tracts with children with JIA out of the 740 census tracts in the metro area. Patients with JIA appeared to have lower air pollution exposures and were less socially vulnerable compared to the general population.

Conclusion: To our knowledge, this is the first study in the US to use geospatial data to assess environmental risk factors for all subtypes of JIA. Prevalence rates varied by county, but this may be secondary to small case counts and variations in population sizes by county; further investigation is needed. The study found that patients with JIA living in the metro area were

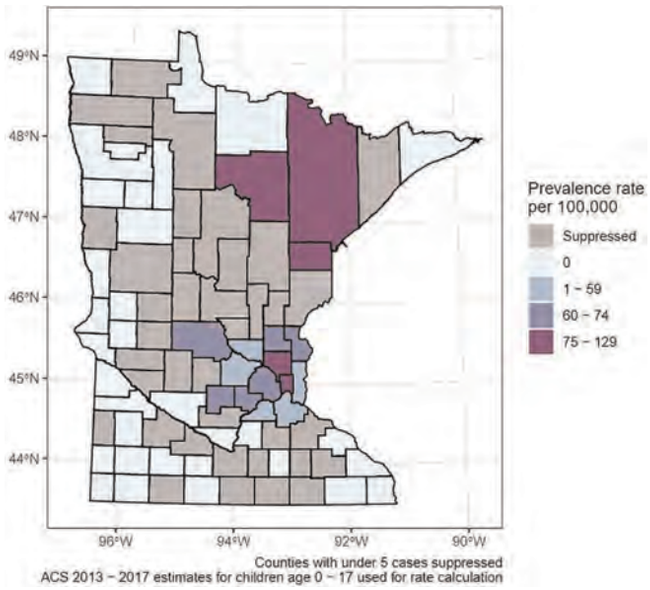


Figure 2. Prevalence rates of JIA by population of children living in Minnesota, by county.

Table 1. Comparison of air pollution exposure and social vulnerability index measures of children with JIA living in the 7-county metro area compared to the general population of children by census tracts.

Variable	Mean in census tracts with JIA patients	Weighted mean in all metro census tracts*
$PM_{2.5}$	6.69	6.79
SO_2	0.87	0.88
NO_2	6.94	7.53
Overall SVI	0.32	0.45

$PM_{2.5}$= Fine particulate matter, SVI = social vulnerability index

*Weights for the weighted means were the proportion of the entire metro area population that each census tract represented

exposed to relatively lower concentrations of $PM_{2.5}$ and NO_2 and lived in census tracts that were relatively less socially vulnerable compared to the metro area as a whole. This was a small preliminary study and our air pollution assessments were not adjusted for demographic data such as race, JIA subtype, or patient age, and we lacked access to healthy controls. Larger national studies that include demographic and clinical data, and that use more refined geospatial techniques are needed to better study these environmental risk factors.

Disclosure: C. Correll: None; **A. Rau**: None; **J. Berman**: None.

Abstract Number: 123

Development of Specific Classification Criteria for Juvenile System Sclerosis Patients: A Scoping Review

Ioana Dobre[1], Suzanne Li[2], Natalia Vasquez Canizares[3], Barbara Reich[4], Xurong Zhao[5], Quinn McCormick[6] and Marinka Twilt[5], [1]Alberta Children's Hospital/University of Calgary, Calgary, AB, Canada, [2]Hackensack Meridian School of Medicine, Joseph M. Sanzari Children's Hospital, Hackensack, NJ, [3]Children's Hospital at Montefiore, New York, NY, [4]Hackensack University Medical Center, Hackensack, NJ, [5]Alberta Children's Hospital, Calgary, AB, Canada, [6]Hackensack Medical Hospital Network, Hackensack, NJ

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Juvenile systemic sclerosis (jSSc) is associated with one of the highest morbidity and mortality rate in pediatric rheumatology, yet care recommendations are based upon low quality data. As no jSSc clinical trials have been done, recommendations are primarily derived from adult trials. However, many differences in disease patterns exist between jSSc and adult SSc, including for morbidity and mortality risks, which makes this approach sub-optimal.

A key tool for conducting clinical trials are sensitive and specific classification criteria. The sensitivity of the 2007 jSSc classification criteria for classifying jSSc is < 70%, making them unfeasible to use in clinical trials. We seek to generate highly sensitive classification criteria to make it possible to conduct jSSc clinical trials. This will enable studies directed towards developing evidence-based care recommendations, and thereby improve long-term outcome for jSSc. An important first step is a comprehensive understanding of clinical features of jSSc and their frequencies. We also seek to better delineate differences between juvenile versus adult SSc.

Methods: A scoping review of jSSc and aSSc literature was conducted in PubMed and EMBASE. Search terms for jSSc and aSSc literature were determined by jSSc experts and an experienced librarian, the search was uploaded to Covidence for reviewer access. Inclusion criteria for jSSc articles were: 1) $\geq$ 3 patients with jSSc as defined by authors and 2) frequency of at least some clinical features reported. Exclusion criteria were 1) Non-English articles and 2) articles of the same study population and research group. For aSSc articles literature, articles that included $\geq$ 500 patients with aSSc as defined by authors and reported frequency of at least some clinical features were included. Non-English articles and articles from the same study population/group were excluded. Data extracted included disease subtype, antibody profile, and frequency of clinical features, including Raynauds phenomenon, interstitial lung disease, gastrointestinal manifestations, arthritis, sclerodactyly, tendon friction rub, heart manifestations and renal crisis.

Results: A total of 2175 articles were screened for jSSc, and 3446 for adult SSc. A higher proportion of jSSc patients had diffuse cutaneous SSc compared to adults (69.9% and 32.1%, respectively; p-value < 0.001) whereas the opposite was true for limited cutaneous SSc (20.8% and 56.9%, respectively). Arthritis was far more prominent in jSSc compared to aSSc (32.5% and 17%, respectively; p< 0.001). 8.1% of jSSc patients had positive Anti-centromere serology compared to 28.7% of aSSc patients (p-value < 0.001). 15.3% of jSSc patients had positive Anti-PM/Scl serology compared to 4.6% in aSSc patients (p-value < 0.001). Rate of renal crisis was far less in jSSc compared to aSSc patients (1.2 % vs. 3.7%, p-value=0.044).

Conclusion: Our study shows that the frequency of disease subtypes, antibody profiles and several clinical characteristics differ significantly between jSSc and aSSc patients. This supports the need for developing jSSc-specific classification criteria, and further research to better understand jSSc.

Disclosure: I. Dobre: None; **S. Li**: Merck/MSD, 11; **N. Vasquez Canizares**: None; **B. Reich**: None; **X. Zhao**: None; **Q. McCormick**: None; **M. Twilt**: None.

Abstract Number: 124

JIA-Associated TMJ Arthritis, Idiopathic Condylar Resorption or Anterior Disc Displacement – a Care Provider Survey

Daria Sosna[1], Nancy Pan[2], Shelly Abramowicz[3], Mara Becker[4], Melissa Lerman[5], Cory Resnick[6], Tova Ronis[7], Matthew Stoll[8], Peter Stoustrup[9], Marinka Twilt[10], CARRA Registry Investigators[11] and For TMJaw[12], [1]Alberta Children's Hospital/University of Calgary, Calgary, AB, Canada, [2]Hospital for Special Surgery, New York, NY, [3]Emory University, Atlanta, GA, [4]Duke University Medical Center/Duke Clinical Research Institute, Durham, NC, [5]Children's Hospital of Philadelphia, Philadelphia, PA, [6]Harvard University, Boston, MA, [7]Children's National Hospital, Chevy Chase, MD, [8]University of Alabama at Birmingham, Birmingham, AL, [9]Aarhus University, Aarhus, Denmark, [10]Alberta Children's Hospital, Calgary, AB, Canada, [11]CARRA, Washington, DC, [12]TMJaw, Fullerton, CA

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: The temporomandibular joint (TMJ) can be affected in juvenile idiopathic arthritis (JIA) patients of any age or subtype. There have been reports of isolated TMJ arthritis. Idiopathic condylar resorption (ICR) and anterior disc displacement (ADD) also affect the TMJ and can be difficult to distinguish from JIA. The MRI appearance of active JIA-associated TMJ arthritis shows significant overlap with patients diagnosed with ADD. A Swiss study showed various degrees of inflammation with joint effusion, synovial thickening, and joint enhancement in both patients with JIA and ADD. Rheumatologists are increasingly asked to see patients with isolated TMJ problems. The confidence level of rheumatologists regarding differentiation between these different conditions is unclear. Interested researchers from the Temporomandibular Jaw working group (TMJaw) and the Childhood Arthritis Rheumatology Research Alliance (CARRA) TMJ interest group surveyed pediatric rheumatologist to assess the burden of isolated TMJ referrals and their confidence level in differentiating these conditions.

Methods: A survey was developed that had both closed and open-ended questions. This survey was distributed to a group of pediatric rheumatologist CARRA members. Participants were asked to identify areas of opportunity in this area including: Increasing awareness among rheumatologists on the different diagnosis, increasing awareness among radiologists, and development of algorithms to help assist to differentiate these conditions. Descriptive statistics were used.

Results: In total, 93 individuals participated in the survey, 94.5% (88) self-identified as pediatric rheumatologists, 5.5% (5) individuals were combined pediatric/adult rheumatologists. On average most of the participants see between 1-5 patients per year for the question of isolated JIA-associated TMJ arthritis. The majority of respondents rated themselves as somewhat confident in evaluating isolated TMJ concerns in children. Survey participants had seen both ICR and ADD patients and felt mostly minimally confident in differentiating between ICR and TMJ arthritis for both mimickers. The majority of participants stated that they have a team to discuss these cases with, most teams included a radiologist or oral and maxillofacial surgeon.

Conclusion: According to the results of this survey, pediatric rheumatologists have an overall level of discomfort regarding differentiation of TMJ mimickers, as many participants stated that they were minimally or somewhat confident in most circumstances at diagnosing these mimickers. However, many participants stated that they have assistance from a multidisciplinary team in aiding them with these diagnoses and mainly this includes a dedicated radiologist. These results highlight the need for additional research to help providers differentiate between these disparate TMJ conditions.

Disclosure: D. Sosna: None; **N. Pan**: None; **S. Abramowicz**: None; **M. Becker**: None; **M. Lerman**: None; **C. Resnick**: None; **T. Ronis**: None; **M. Stoll**: None; **P. Stoustrup**: None; **M. Twilt**: None; **C. Investigators**: None; **F. TMJaw**: None.

Abstract Number: 125

Measurable Outcomes of an Ophthalmology and Rheumatology Coordinated Care Clinic

Catherine Lavallee[1], **Sabrina Gmuca**[2] and Melissa Lerman[2], [1]Virginia Tech Carilion School of Medicine, ROANOKE, VA, [2]Children's Hospital of Philadelphia, Philadelphia, PA

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Non-infectious pediatric uveitis is a vision threatening disease whose treatment involves both ophthalmologists and rheumatologists.In other diseases necessitating multidisciplinary care, coordinated care clinics have shown improved outcomes.While many pediatric uveitis guidelines suggest that coordinated uveitis care is important, there is sparse data on its effectiveness.The objective of this study was to evaluate the impact of an Ophthalmology and Rheumatology Coordinated Care clinic on patients with anterior uveitis by comparing outcomes between those who received traditional care vs. coordinated care.

Methods: This was a retrospective cohort study of children ages ≤ 19 years, with anterior uveitis (AU) included in a registry from pediatric tertiary care center between 2013-2022. Inclusion criteria for the Uveitis Coordinated Care Clinic (coordinated care) cohort: ≥2 visits within the first 6 months of first coordinated care visit with ≥1 additional visit in the subsequent

Table one.

Characteristic	Coordinated Care, N = 61[1]	Traditional Care, N = 61[1]	p-value[2]
Primary Ocular Diagnosis			0.3
Anterior Uveitis	59	55	
Sex			0.7
Female	39 (64%)	41 (67%)	
Male	22 (36%)	20 (33%)	
Age at diagnosis	9.4 (4.5, 13.1)	8.4 (5.9, 12.5)	0.5
Rheumatic diagnosis			0.2
JIA	26 (43%)	27 (44%)	
TINU	3 (4.9%)	4 (6.6%)	
Sarcoid/Blau	2 (3.3%)	0 (0%)	
Idiopathic	22 (36%)	15 (25%)	
Vasculitis/Multiple Sclerosis	0 (0%)	4 (6.6%)	
Missing/Unknown	8 (13%)	11 (18%)	
JIA Subtype			>0.9
Oligo	50 (82%)	50 (82%)	
Poly RF-	4 (6.6%)	3 (4.9%)	
Poly RF+	0 (0%)	0 (0%)	
ERA	3 (4.9%)	3 (4.9%)	
PsA	2 (3.3%)	2 (3.3%)	
Undifferentiated	1 (1.6%)	2 (3.3%)	
Missing/Unknown	1 (1.6%)	1 (1.6%)	
ANA Status			>0.9
Positive	28 (46%)	28 (46%)	
Negative	28 (46%)	27 (44%)	
Not done, unknown	5 (8.2%)	6 (9.8%)	
Baseline AC cell count			0.4
0	21 (34%)	25 (41%)	
0.5+	18 (30%)	13 (21%)	
1+	11 (18%)	11 (18%)	
2+	5 (8.2%)	9 (15%)	
3+	5 (8.2%)	1 (1.6%)	
4+	1 (1.6%)	2 (3.3%)	

[1] n (%); Median (IQR)
[2] Fisher's exact test; Pearson's Chi-squared test; Wilcoxon rank sum test

6 months (minimum of 3 clinic visits within year 1). If criteria were not met, patients were assigned to the non-UCCC (traditional care) cohort. Start time into the coordinated care cohort was the first coordinated care visit in the registry and for the traditional care cohort was the first ophthalmology appointment in the registry. The patient could transition from traditional to coordinated care cohort and time in each could be counted for the disease control analysis. Disease control was defined as ≤0.5+ anterior chamber (AC) cell (as per SUN criteria) and ≤2 topical steroid drops daily. Standard descriptive statistics were used to describe clinical features including sex, age at diagnosis, and primary rheumatic diagnosis. Survival analysis was used to identify differences in cohort disease activity and biologic DMARDs use over time. Chi-square test of independence was used to examine differences in topical steroid use between cohorts at their initial visit and at 6 months.

Results: A total of 122 patients met inclusion criteria. Sixty-one in the coordinated care cohort and 61 in the traditional care cohort. There was no statistical difference between cohorts in the speed at which they achieved disease control (median time 44 (traditional care) vs 59 days (coordinated care), p = 0.3) (Figure 1). No patient level covariates such as baseline AC

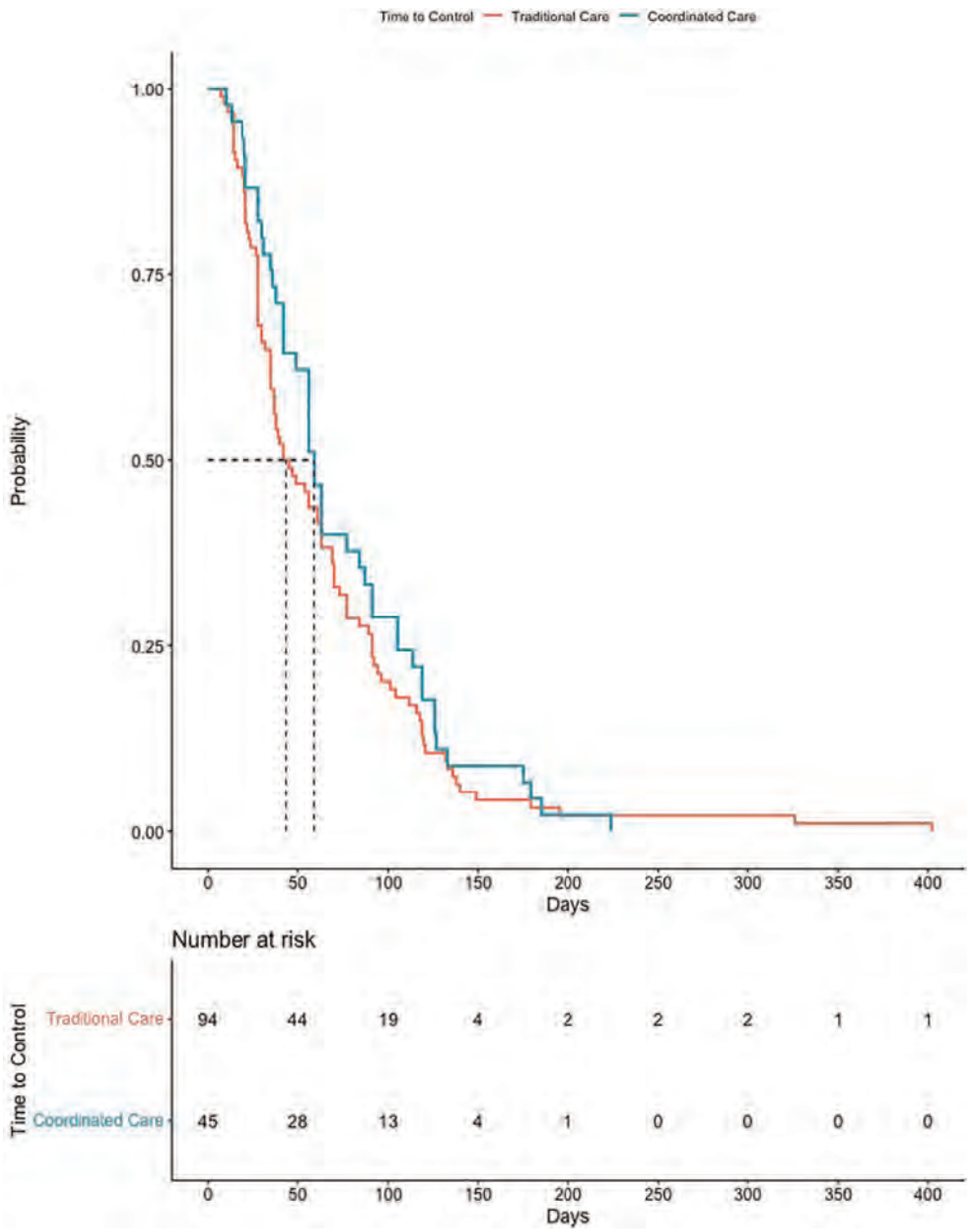


Figure one.

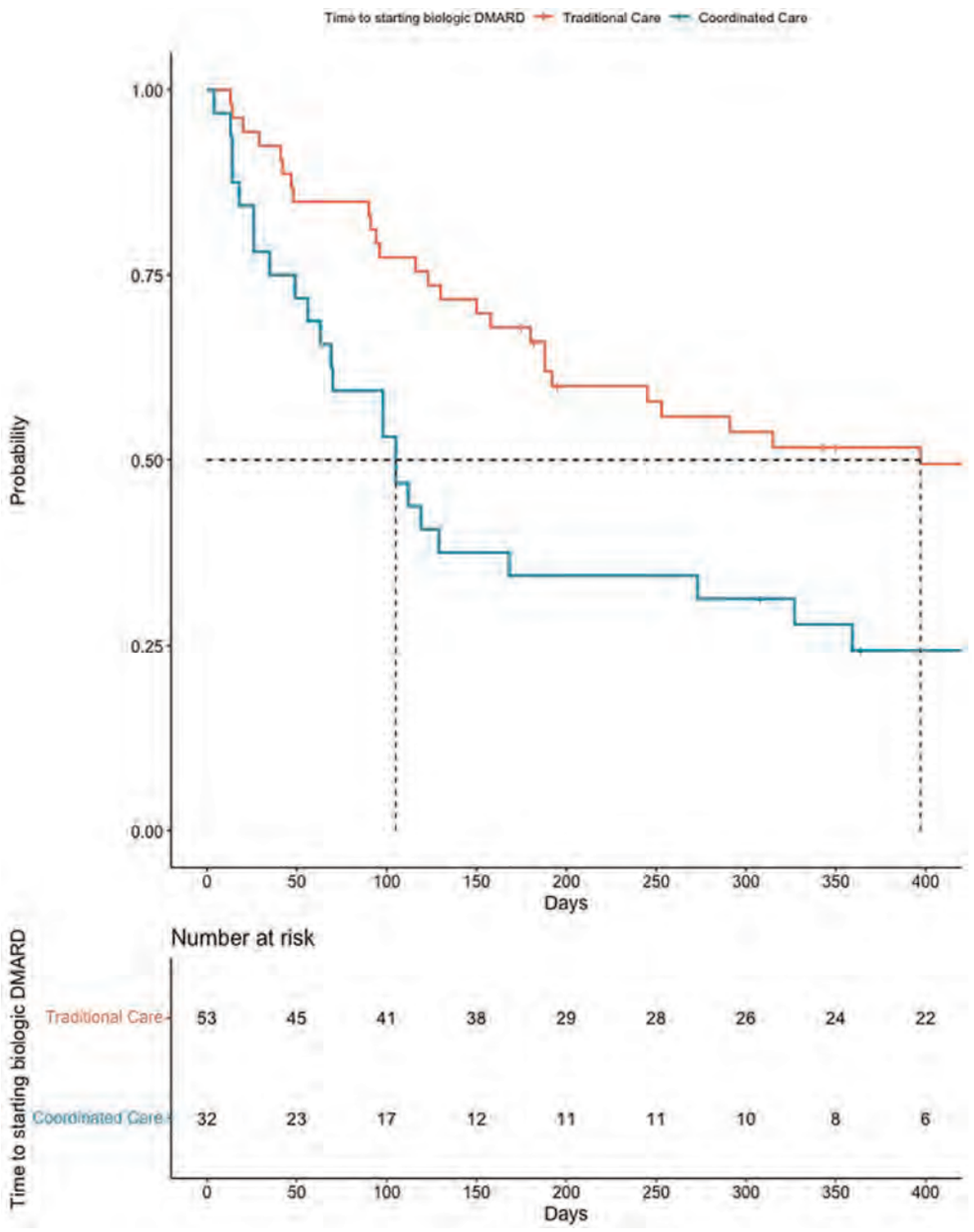


Figure two.

cell or duration of disease before cohort entry statistically impacted the outcome. The coordinated care cohort did start biologic DMARDs faster from entry into cohort than the traditional care cohort (p = 0.016) (Figure 2). The coordinated care cohort was on fewer steroid drops at baseline than the traditional care cohort. There was no difference in topical steroids at six months between cohorts.

Conclusion: In our current cohort, we did not demonstrate improved speed at which they achieved disease control, although they did initiate care sooner with biologics. Further analyses are needed to explore differences between the two cohorts such as number of recurrent disease activity, flares, and how potential confounding factors, such as entry time into each cohort might impact these results.

Disclosure: C. Lavallee: None; **S. Gmuca**: None; **M. Lerman**: None.

Abstract Number: 126

Facilitating Peer-to-Peer Conversations Around Key Clinical Trial Recruitment Barriers in the Limit-JIA Trial Using Low-Fidelity Video Capture

Melanie Kohlheim[1], **Eveline Wu**[2], Laura Schanberg[3], Vincent Del Gaizo[1], Catherine Lavallee[4], Marc Natter[5], Katie Clem[6], Brian Shakley[6] and Kevin Urban[7], [1]CARRA, Washington, DC, [2]UNC Chapel Hill, Chapel Hill, NC, [3]Duke University Medical Center, Durham, NC, [4]Virginia Tech Carilion School of Medicine, ROANOKE, VA, [5]Boston Children's Hospital, Boston, MA, [6]LIFT 1428, Llc, Ooltewah, TN, [7]Business Coaching for Creatives, New York, NY

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Clinical and Therapeutic II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: The LIMIT-JIA trial aims to study if early abatacept treatment can prevent disease extension in children with recent-onset, uncomplicated, and oligoarticular or limited JIA. Barriers to recruitment identified in the first trial phase included 1) families expressed concern over being diagnosed with JIA, 2) confusion about usual treatment of JIA, and 3) fear around administering injections to their children. When LIMT-JIA enrollment reopened in the new COVID-19 environment, these pre-COVID barriers were amplified. Qualifying patients and families were more hesitant to pursue treatment that might be labeled as immune-suppressing. We sought to identify and implement COVID-friendly strategies to minimize these recruitment barriers.

Methods: A group of stakeholders (JIA parents, a pediatric rheumatologist, an experienced interviewer, and an adult JIA patient) met weekly throughout the summer of 2021 to determine target groups to interview, how to best gather video footage, and the look of the final products. As COVID-19 cases spiked again, plans were modified from in-person recorded interviews to recorded marketing-style zoom interviews. Interviewees included 3 parent members from the LIMIT-JIA Stakeholder Advisory Committee, 3 young adult patients 20-25 y.o., and 4 children 8-12 y.o. Interviewees were asked about their experience with JIA and described their own hesitation and experience with their first injection. Interviews were transcribed with key phrases or points of each interview flagged for editing purposes.

Results: Patient and families were familiar with use of Zoom/Teams/Webex style platforms for communicating and hearing information from Tik-Tok, Instagram stories, and reels. This suggested more dynamic resources were needed in a digital format, and several videos were created to address the needs expressed by patient families and clinic staff. To date, 8 video clips have been developed for our patient-facing website, covering topics around injection preparation, diagnosis journey, and JIA. The videos were made available on the

www.limitjia.com website as part of a new recruitment packet. Study is ongoing to determine whether the patient-facing websites and video clips were effective recruitment tools.

Conclusion: Ethnographic interviews of patients, families, and clinical staff informed peer-to-peer recruitment strategies. Casual, low-fidelity video capture is more acceptable and relatable today than highly edited and directed film. Our study team felt this worked to our advantage and allowed us to develop cost-efficient digital tools to address the needs of families and clinic staff. We were able to use creative approaches to address barriers to patient and site engagement amplified by the COVID pandemic.

Disclosure: M. Kohlheim: None; **E. Wu**: Enzyvant, 2, Pharming Healthcare, Inc, 2; **L. Schanberg**: Bristol-Myers Squibb(BMS), 5, Sanofi, 12, DSMB member, UCB, 12, DSMB chair; **V. Del Gaizo**: None; **C. Lavallee**: None; **M. Natter**: None; **K. Clem**: None; **B. Shakley**: None; **K. Urban**: None.

Abstract Number: 127

What's in a Name? A20 Protein Expression in an in Vitro Model of A20 Haploinsufficiency

Patricia Pontes Aires[1], DANIELA GERENT PETRY PIOTTO[1], Andre Cunha[1], Sandro Perazzio[2] and **Maria Teresa Terreri**[3], [1]Universidade Federal de São Paulo, São Paulo, Brazil, [2]Universidade de So Paulo; Fleury Laboratories, São Paulo, Brazil, [3]UNIFESP, São Paulo, Brazil

SESSION INFORMATION

Session Date: Friday, March 31, 2023
Session Title: Posters: Genetics and Pathogenesis II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Tumor necrosis factor alpha (TNF-alpha) induced protein 3 gene, or *TNFAIP3*, encodes the A20 protein, an important regulator of the NF-κB pathway. Since its initial report in 2016, A20 haploinsufficiency has been described as a human autoinflammatory disease caused by heterozygous loss-of-function mutations in *TNFAIP3* leading to insufficient suppression of NF-kB activity. Previous studies suggested that mutated A20 was degraded, and the phenotype was caused by insufficient NF-kB canonical pathway modulation. This study aims to determine the effect of the L227X mutation in A20 protein expression, using different cell lines.

Methods: Green fluorescent protein (GFP)-tagged plasmids were constructed with the *TNFAIP3* gene and a L227X mutation was induced using QuikChange II XL Site-Directed Mutagenesis Kit (Agilent®, Santa Clara, USA). Mutated plasmids were transfected into THP-1 (human monocytic), Jurkat (human lymphoblastic T cells) and HL-60 (human neutrophilic) cells, using a viral vector. THP-1 cells were also transfected with an empty plasmid (EP, only the backbone and GFP) or GFP-tagged wild-type TNFAIP3 plasmid (TNFAIP3-WT). Original genetic content was not altered (no knock-out was performed). Cells were assessed for GFP expression, after multiple passages, to ensure stable plasmid incorporation. A20 was stained by a C-terminal monoclonal antibody against TNFAIP3 (Abcam®, Cambridge, UK) and protein expression assessed by flow cytometry.

Results: A20 expression was systematically reduced in mutated TNFAIP3-L227X plasmid transfected cell lines compared to non-transfected control cells: HL-60 (MFI=572 vs 450, p 0.05, figure 1) and Jurkat (MFI=764 vs 695, p 0.01, figure 2). A20 protein expression was significantly reduced in TNFAIP3-L227X transfected THP-1 cells (MFI=581) compared to non-

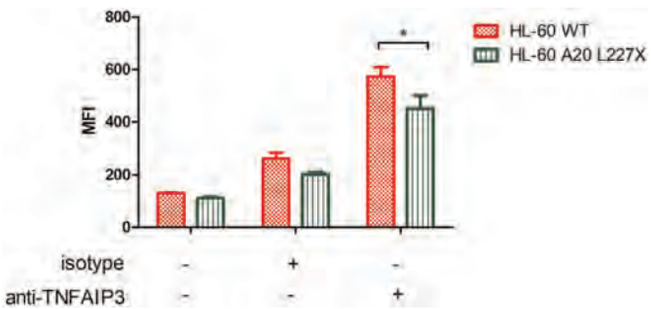


Figure 1: A20 protein expression in HL-60 cells, non-transfected (red) or transfected with TNFAIP3-L227X plasmid (green). * p < 0.05

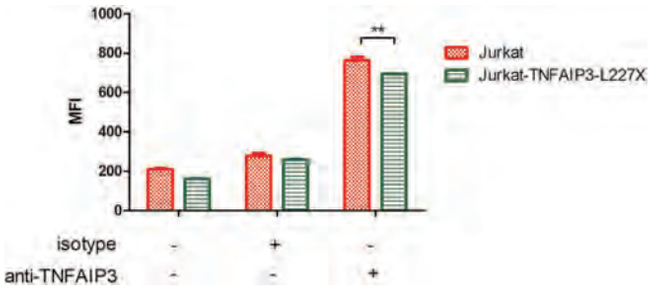


Figure 2: A20 protein expression in Jurkat cells, non-transfected (red) or transfected with TNFAIP3-L227X plasmid (green). ** $p < 0.01$

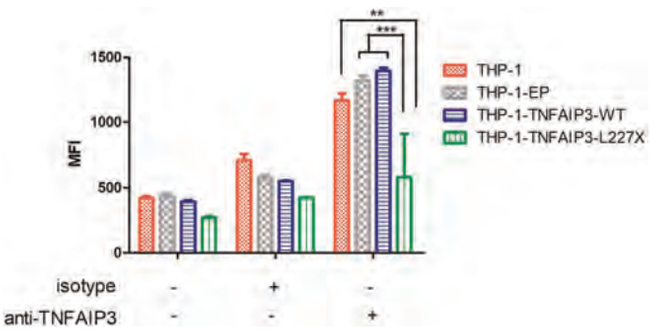


Figure 3: A20 protein expression in THP-1 cells, non-transfected (red), transfected with empty plasmid (grey), transfected with wild-type TNFAIP3 plasmid (blue) or transfected with TNFAIP3-L227X plasmid (green). ** $p<0.01$; ***$p<0.001$.

transfected (MFI=1168), EP-transfected (MFI=1321) or TNFAIP3-WT transfected cells (MFI=1395, p 0.01; figure 3). TNFAIP3-WT showed a modest but significant increase of A20 expression compared to non-transfected THP-1.

Conclusion: Stable TNFAIP3-L227X transfection in human cell lines was able to induce reduction of A20 protein expression, in the absence of gene silencing or knock-out of endogenous TNFAIP3. This suggests TNFAIP3-L227X mutation might act through other mechanisms beyond haploinsufficiency in immune cell lines, which merits further investigation.

Disclosure: P. Pontes Aires: None; **D. PIOTTO**: None; **A. Cunha**: None; **S. Perazzio**: None; **M. Terreri**: None.

Abstract Number: 128

Multisystem Inflammatory Syndrome in Children and Systemic Juvenile Idiopathic Arthritis Share Clinical Phenotypes and Genetic Contributions

Paul Tsoukas[1], Hua Lu[2], Marla Mendes de Aquino[2], Michael Ombrello[3], Lisa Strug[2] and Rae Yeung[4], [1]The Hospital of Sick Children, Department of Paediatrics, University of Toronto, Division of Rheumatology; Cell Biology Program, The Hospital for Sick Children Research Institute, Toronto, ON, Canada, [2]The Hospital for Sick Children, Toronto, ON, Canada, [3]Translational Genetics and Genomics Section, National Institute of Arthritis and Musculoskeletal and Skin Diseases, National Institutes of Health, North Bethesda, MD, [4]The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Genetics and Pathogenesis II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Multisystem inflammatory syndrome in children (MIS-C) is a novel clinical entity presenting following SARS CoV2 infection. This study describes a subgroup of MIS-C patients that develop a systemic juvenile idiopathic arthritis (sJIA)-like illness. Their clinical characteristics, treatment response, patient outcomes, and biologic data are compared to a large cohort of MIS-C patients. No specific diagnostic tests are available to differentiate between MIS-C and sJIA. Polygenic risk score (PRS) analysis will assess whether the genetic burden in sJIA is also seen in MIS-C.

Methods: Clinical data and biospecimens from treatment naïve MIS-C patients were collected prospectively within a single tertiary care center. All MIS-C patients fulfilled the Royal College of Paediatrics and Child Health case definition. 70% of the co-diagnosis patients satisfied CDC MIS-C criteria. Whole genome sequencing was performed on a subset of 249 samples (68 MIS-C, 181 acute COVID controls) using Illumina Nextera Custom Capture Assays and Illumina sequencers. Called variants were identified using Dragen. Publicly available genome-wide association study summary statistics for COVID-19 severity and sJIA were utilized for calculating PRS for each individual via the Pruning+Thresholding method. The top four principal components adjusted the PRS to avoid confounding due to population stratification, followed by logistic regressions to test the association between the constructed scores and MIS-C. Best models were selected based on the highest variability explained using Nagelkerke R-squared as a metric.

Results: Ten MIS-C patients of the 298 studied, were diagnosed and treated with an sJIA-like illness. The evolution of the sJIA-like illness was insidious with a co-diagnosis made a median of 40 days following the initial MIS-C diagnosis. In comparison to the MIS-C patients, the co-diagnosis subgroup had a longer duration of fever, with a greater incidence of arthritis, macrophage activation syndrome and also development of coronary artery lesions. Biochemically, the two groups had notable differences. The sJIA-MISC patients had greater leukocytosis, neutrophilia, more substantial anemia, hyperferritinemia and elevations in ESR. In terms of disease severity, co-diagnosis patients had a longer length of stay and requirement for respiratory support. All 10 co-diagnosis patients required biologic therapy. All received IL-1 inhibition, however, disease was refractory in 30% and subsequently required IL-6 inhibition.

We calculated two PRSs (COVID-19 severity and sJIA), representing distinct biologic pathways to investigate shared genetic contributions with MIS-C. No significant association between MIS-C and acute COVID-19 severity PRS (p=0.451, OR = 1.023, 95% CI [0.9645,1.0845]) was observed. However, SJIA PRS was found to be significantly associated with the MIS-C status (p=0.014, OR = 1.063, 95% CI [1.0136,1.1182]) after Bonferroni correction.

Conclusion: MIS-C and sJIA have overlapping clinical features and treatments, as well as shared genetic contributions suggesting shared immunobiology.

Disclosure: P. Tsoukas: None; **H. Lu**: None; **M. Mendes de Aquino**: None; **M. Ombrello**: None; **L. Strug**: None; **R. Yeung**: None.

Abstract Number: 129

Interrogation of STAT3 Activation in Patients with Polyarticular Juvenile Arthritis (polyJIA)

Stephanie Wood[1], Justin Branch[1], Priscilla vasquez[1], Marietta De Guzman[1], Amanda Brown[2], A. Carmela Sagcal-Gironella[3], Saimun Singla[4], Andrea Ramirez[5] and Tiphanie Vogel[5], [1]Baylor College of Medicine, Texas Children's Hospital, Houston, TX, [2]University of Arkansas for Medical Sciences, Little Rock, AR, [3]Hackensack University Medical Center, Hackensack, NJ, [4]Self, Houston, TX, [5]Baylor College of Medicine, Houston, TX

SESSION INFORMATION

Session Date: Friday, March 31, 2023

Session Title: Posters: Genetics and Pathogenesis II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: A better understanding of the pathogenesis of polyJIA is necessary to guide more effective clinical care, such as the development of data-driven approaches to guide selection of the ideal therapeutic agent for an individual patient. One inflammatory pathway of interest is the JAK-STAT signaling cascade. STAT3 is a transcription factor critical to the differentiation of inflammatory T helper 17 cells (Th17s), as well as influencing T regulatory cell (Tregs) development. Previous studies have demonstrated elevated Th17 cells and activated STAT3 in adult patients with rheumatoid arthritis, but less is known about T cell subsets and STAT3 activation in polyJIA. We hypothesized that Th17 cells and STAT3 activation would be increased in treatment-naïve polyJIA patients compared to pediatric controls.

Methods: Blood from 17 patients with polyJIA was collected at initial diagnosis (treatment-naïve) and after remission was achieved. Pediatric healthy controls were also collected. Peripheral blood mononuclear cells were isolated and CD4+ T cell subsets and STAT activation (phosphorylation) were evaluated using flow cytometry.

Results: Treatment naïve polyJIA patients had increased Th17 cells (CD3+CD4+interleukin(IL)-17+) compared to controls (0.15% v 0.44%, p=0.0371), but, Tregs (CD3+CD4+CD25+FOXP3+) from patients with polyJIA did not differ from controls. We identified dual IL-17+ and interferon (IFN)-gamma+ expressing CD4+ T cells (Th17/1s) in patients, but not controls. Further, both Th17/1s and ex-Th17s (CD3+CD4+CCR6+CD161+IFN-gamma+IL-17neg) were increased in patients post-treatment (0.065% v 0.29%, p=0.0117 and 1.42% v 2.26%, p=0.0195, respectively). The patients with the highest Th17/1 cells post-treatment remained therapy-bound, but other patients were successfully weaned off medications following remission. *Ex vivo* stimulation of CD4+ T cells (using IL-6 or IFN-alpha) from treatment-naïve patients compared to controls demonstrated smaller changes in STAT3 phosphorylation between stimulated and unstimulated cells.

Conclusion: Patients with polyJIA have increased baseline Th17 cells and are less responsive to inflammatory cytokine stimulation *ex vivo*, possibly reflecting higher tonic STAT3 activation *in vivo* . These quantifiable immune markers may diagnostically identify patients that would benefit from upfront, pathway-focused anti-cytokine biologic therapeutics. Our data also suggest that Th17/1s, a subset not detected in controls but increased in samples from treated patients, should be further evaluated as a prognostic tool to stratify patients in clinical remission on medication. Future work will explore both these proposed diagnostic and prognostic biomarkers.

Disclosure: S. Wood: None; **J. Branch**: None; **P. vasquez**: None; **M. De Guzman**: None; **A. Brown**: None; **A. Sagcal-Gironella**: None; **S. Singla**: None; **A. Ramirez**: None; **T. Vogel**: Moderna, 2, Novartis, 2, Pfizer, 2, sobi, 2.

Abstract Number: 130

Levels of Neutrophil Extracellular Traps Correlate with Disease Activity in Pediatric Lupus

Lydia Thomas[1], Jenna Battaglia[2], Bharati Matta[3], Kim Simpfendorfer[4], Joyce Hui-Yuen[5] and Betsy Barnes[3], [1]Cohen Children's Medical Center, Northwell Health, New Hyde Park, NY, [2]Northwell Health, New York, NY, [3]Center for Autoimmune, Musculoskeletal, and Hematopoietic Diseases Research, Feinstein Institutes for Medical Research, Northwell Health, Manhasset, NY, [4]Robert S. Boas Center for Genomics and Human Genetics, Feinstein Institutes for Medical Research, Northwell Health, Manhasset, NY, [5]Cohen Children's Medical Center, Northwell Health, Lake Success, New York; Center for Autoimmune, Musculoskeletal, and Hematopoietic Diseases Research, Feinstein Institutes for Medical Research, Northwell Health, Manhasset, NY

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Genetics and Pathogenesis II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Pediatric lupus (pSLE) is a multisystemic autoimmune disease characterized by autoantibody production leading to organ damage. Neutrophil extracellular traps (NETs) are considered a potential source of antigen, leading to autoantibody production. NETs activate plasmacytoid dendritic cells to produce high levels of interferon-α, a known driver of lupus pathogenesis. Impaired degradation of NETs by DNASEs may play a role in development of lupus, as low DNASE activity and mutations in *DNASEIL3* have been associated with lupus. Here, we investigate levels of circulating NETs in pSLE and healthy children and elucidate mechanisms behind NETs accumulation.

Methods: Plasma from whole blood samples of 13 pSLE patients and 12 healthy children were evaluated for the presence of NETs using our multiplex ELISA and novel immunofluorescence smear assay. DNASE1L3 concentration was measured using ELISA and DNASE1L3 activity by nuclei digest. NET degradation assays were performed using healthy neutrophils stimulated with either pSLE or healthy plasma. Lupus disease activity was measured by SELENA-SLEDAI.

Results: Significantly higher levels of circulating NETs were found in pSLE plasma compared to healthy children, consistent with ELISA and smear assays ($p < 0.05$). The number of NETs were positively correlated with SELENA-SLEDAI scores and anti-double stranded DNA levels. Although DNASE1L3 levels were higher in pSLE patients, enzymatic activity was significantly reduced, compared to healthy children. Moreover, we found that pSLE plasma was not able to degrade NETs as effectively as plasma from healthy children, suggesting that NET degradation was impaired in pSLE, leading to accumulation of NETs.

Conclusion: Decreased DNASE1L3 activity may play a role in impaired clearance of NETs in plasma from pSLE patients leading to NETs accumulation. High levels of DNASE1L3 seen in pSLE patients are likely due to compensatory mechanisms to overcome reduced enzymatic activity in NET clearance. Interestingly, patients with higher SLEDAI scores had higher number of NETs. Thus, level of NETs accumulation in plasma detected by our newly developed assays could potentially be a useful biomarker for pSLE disease severity.

Disclosure: L. Thomas: None; **J. Battaglia**: None; **B. Matta**: None; **K. Simpfendorfer**: None; **J. Hui-Yuen**: None; **B. Barnes**: None.

Abstract Number: 131

Improving Methotrexate Documentation in Electronic Health Records – a Quality Improvement Initiative

Jayne MacMahon[1], Jeanine McColl[2], Alaa Al-Shehab[1], Deborah Levy[3], Ronald laxer[1] and Shirley Tse[1], [1]University of Toronto, The Hospital for Sick Children, Toronto, ON, Canada, [2]University of Alberta, Edmonton, AB, Canada, [3]The Hospital for Sick Children, Toronto, ON, Canada

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Quality, Health Services, and Education II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Prescribing methotrexate,is common practice in rheumatology. Appropriate medication counselling and documentation is important. In our province, as per thephysician regulatory body the College of Physicians and Surgeons of Ontario (CPSO), it is mandated that informed consent is obtained before a treatment is provided. The CPSO provides guidelines on medication documentation in a patients health record. This includes specific risks communicated, any risks unique to the patient, risks of not treating and if consent was obtained.[1]We believed that the consent process for DMARDs is not being documented according to these standards within our hospital.The aim of this project was to develop a Quality Improvement (QI) initiative to improve documentation of methotrexate initiation within our division, to >80%.

Methods: An audit of medication documentation was performed. We identified 10 sequential patients, from general rheumatology clinics, who started methotrexate from June 2018 - November 2021, when electronic health record (EHR) was introduced. We reviewed the health record from the time of initiation of methotrexate. We recorded if documentation was available in the EHR. Where available, we recorded if the documentation fully met CPSO standards (i.e. documented specific risks, risks unique to patient, risks of not treating/alternative treatments and agreement to commence therapy). If all factors were not documented, we recorded this as partial documentation. We organized an education session on CPSO guidelines and shared the results of our audit. We created a smart-phrase,in accordance with CPSO guidelines,for all patients starting methotrexate. A smart phrase is a tool in our EHR that allows users to link a paragraph to a simple phrase. For example, typing .mtxnotedocumentation populates the note with a detailed paragraph on methotrexate. This paragraph was reviewed by all prescribers within the division and revised. We placed visual reminders in clinics where documentation was performed. After implementation, we tracked use of the smart-phrase for a 4 month period to assess uptake.

Results: Of the ten patient charts identified in our audit, one had documentation that was fully adherent to CPSO guidelines (10%). Seven charts had partial documentation and two had no documentation other than stating that methotrexate was prescribed. This was in keeping with our initial hypothesis. In the first month of implementation of the smart-phrase, the number of charts with full documentation increased to 28%. Over the next 3 months, full documentation was measured at 80%, 50% and 75% for April, May and June respectively.

Conclusion: It is important to document precisely what was conveyed to the family when starting treatment in case there are later clinical concerns or medico-legal proceedings. Tools such as smart phrases, can increase documentation in a time efficient manner. Our intervention showed that once the smart phrase was available, it was used broadly. Ongoing education regarding the availability of the smart phrase will be needed to reach our goal of >80% compliance consistently. In the future, we plan to introduce similar smart-phrases for commonly prescribed biologic therapies.

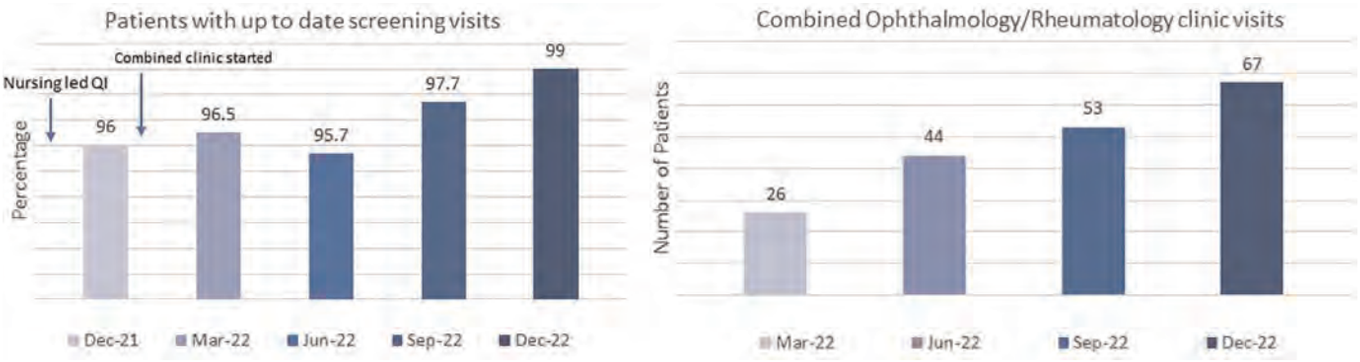
Disclosure: J. MacMahon: None; **J. McColl**: None; **A. Al-Shehab**: None; **D. Levy**: Janssen, 1, Roche, 5, Sobi, 1, 5; **R. Iaxer**: Akros Pharmaceutical, 2, alexion, 2; **S. Tse**: None.

Abstract Number: 132

Quality Improvement Lessons in a New Practice

Farah Shaya, **Sharon Bout-Tabaku** and Buthaina Al-Adba, Sidra Medicine, Ar-Rayyan, Qatar

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Quality, Health Services, and Education II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM



Patients with up to date eye visits improved after nursing engagement in the QI project and continued to improve after the implementation of combined clinics.

Background/Purpose: Children with Juvenile Idiopathic Arthritis (JIA) have better disease outcomes with current medications available, yet there is variability in these outcomes. Quality improvement (QI) processes are dynamic ways to identify gaps, implement goals and positively impact variability in outcomes. Organizations worldwide have identified QI goals for JIA that are important and feasible in their settings. Our practice opened in 2018, as the sole provider of pediatric rheumatology services in a small country. We identified QI goals that were published in the literature, essential for safety, and feasible in our setting with limited resources. We targeted four areas that posed safety risks: laboratory monitoring for DMARDs, tuberculosis screening prior to starting biologics, joint injection referrals, and uveitis screening. We describe the outcomes of our goals from 2018 to 2022.

Methods: From 2018 to 2022, we screened for drug toxicity among patients receiving methotrexate or leflunomide within 3-4 month of receiving methotrexate. We built measures that are captured and retrieved from the electronic medical record (EMR). From September 2021 to 2022, we added new QI goals that could not be captured by the EMR. We engaged dedicated nursing staff to monitor tuberculosis screening prior to starting biologics by ensuring completion of QuantiFERON gold or PPD testing, and/or chest x-ray. Our team developed a joint injection tracker to capture the number of patients having a procedure within 2 weeks of the referral. For uveitis screening and monitoring, we measured the percent of eligible JIA patients with up to date screenings over the previous 6 months. In January 2022, we implemented a monthly combined clinic where patients see Ophthalmology and Rheumatology together. Measures were reviewed quarterly.

Results: Between 2018 and 2022, laboratory monitoring for DMARDs showed that 100% of children receiving methotrexate or leflunomide, were screened for toxicity. However, during a COVID 19 surge, in the first quarter of 2021, 83% were screened. Between 2021 and 2022, 100% of patients had tuberculosis screening prior to starting biologics. Timely performance of joint injections was variable with a median of 53% done within 2 weeks. Prior to the combined clinics, 96% of patients had up to date eye screening visits. After the combined clinics, a median of 96.7% had up to date eye screening visits over 4 quarters.

Conclusion: QI projects can be successful and should start early by choosing and collecting the data needed to monitor improvement. Most of our goals were successful, except for the timeliness of procedures due to scheduling problems, patients canceling due to conflicts or illness. The EMR was able to capture one of our QI goals whereas other goals were not easily extractable from the EMR. Most of our QI measurements required the engagement of nursing staff to maintain the workflow with tracking sheets and reminders. The most successful goals were those that were completed by the rheumatology team at or before the visit. Finally, the implementation of a monthly, combined clinic was effective in having patients seen routinely and promptly. Sustainability is crucial and finding ways to automate cumbersome workflows are needed.

Disclosure: F. Shaya: None; **S. Bout-Tabaku**: None; **B. Al-Adba**: None.

Abstract Number: 133

An Interdisciplinary Team Approach to Implementation of a Social Determinants of Health Screener for Pediatric Rheumatology Patients

Sarah Campbell[1], Rosemary Peterson[2], Sarah Barrientos[3], Elinore Benett[3] and Cori Christenholz[3], [1]University of Texas at Austin Dell Medical School, Austin, TX, [2]Dell Medical School at UT Austin, Austin, TX, [3]Dell Children's Medical Center Department of Rheumatology, Austin, TX

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Quality, Health Services, and Education II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Adolescents with chronic disease often struggle with the transition from pediatric to adult healthcare leading to poor follow-up and negative health outcomes. Social determinants of health (SDoH) such as income, food insecurity, and immigration status likely have a significant impact on transition success. This study aims to successfully implement a standardized approach to SDoH screening and proactive social work involvement for pediatric rheumatology patients.

Social Health Needs Survey

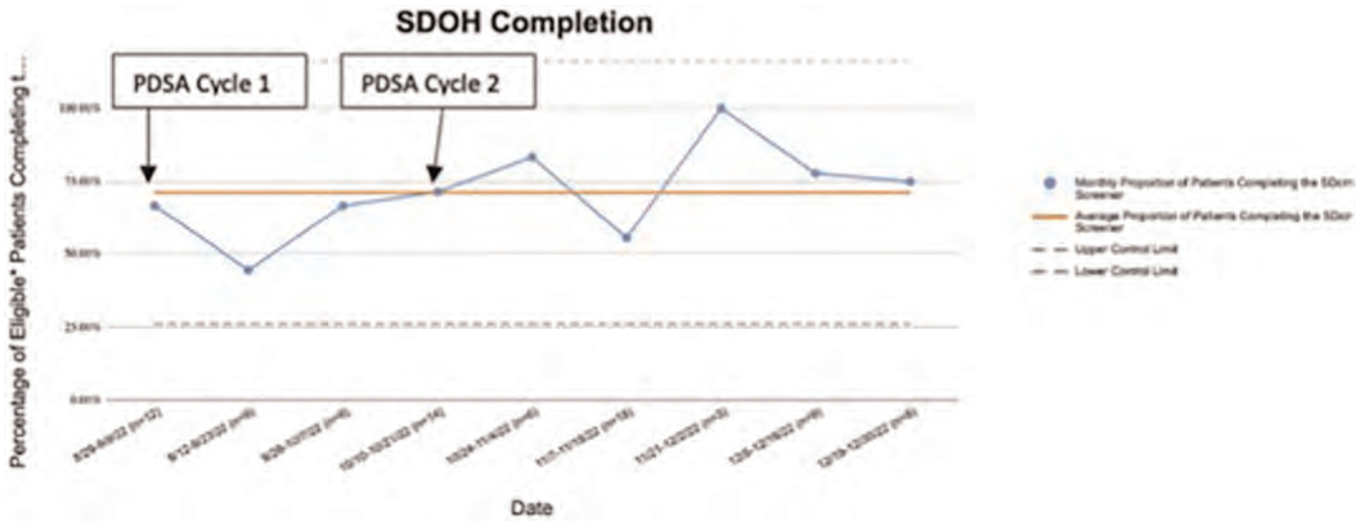
We know that many families in our community have trouble accessing the resources they need to have healthy, safe, and happy lives. So, we are asking everyone in our clinic these questions.

Please answer the following questions so that we can better understand any needs you have and provide help if you want it. This survey is optional.

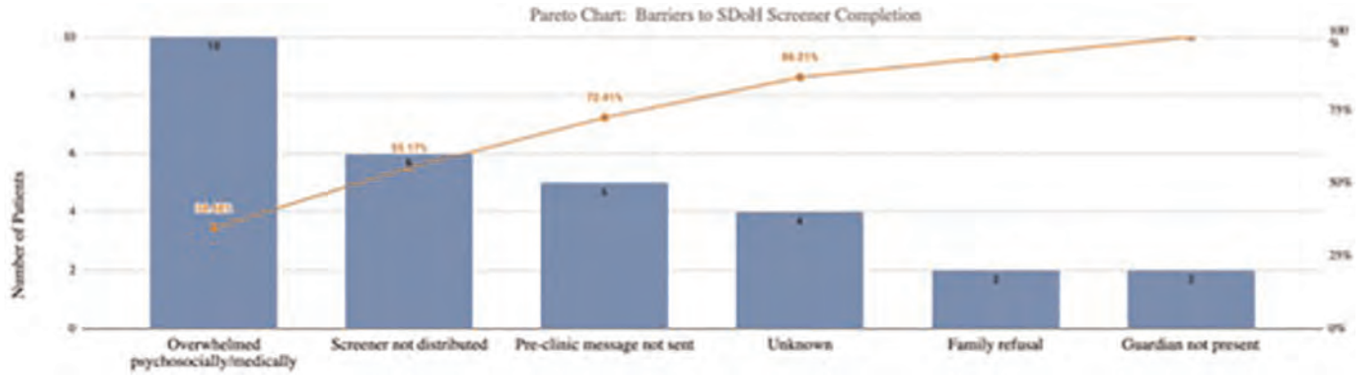
		Yes / No
	In the last 6 months, **have you worried whether food would run out** before you got money to buy more?	Y N
	In the last 6 months, has the **electric, gas, oil, or water company threatened to shut off your services** in your home?	Y N
	Are you worried that in the next few months, you **may not have stable housing?**	Y N
	Do problems with getting **childcare make it difficult for you to work or study?**	Y N
	Sometimes people find that their **income does not quite cover their living costs.** In the last 6 months, has this happened to you?	Y N
	In the last 6 months has **lack of transportation** kept you from medical appointments, meetings, work, or getting things for daily living?	Y N
	During the last month, **have you been actively looking for work?**	Y N
	Do you ever need help **reading or understanding medical information?**	Y N
	Do you have **concerns about your family's immigration status?***	Y N
	In the last year, **have you ever felt threatened in your home or been afraid** of your partner or ex-partner (or someone who cares for you)?**	Y N
	If you checked YES to any boxes above, **would you like help** with any of these needs? Or any other needs we have not asked about? _____	Y N

Person completing form: ☐ Patient ☐ Parent/guardian ☐ Other family member ☐ Other

This figure shows the screener used to screen for social determinants of health in the pediatric Rheumatology clinic.



This figure shows the rate of completion of social determinants of health screeners from August 2022 to December 2022. The initiation of different PDSA cycles are indicated.



This figure shows the rate of different barriers that prevented the completion of the social determinants of health screener.

Methods: An interprofessional team was created to proactively identify psychosocial barriers to successful transition from pediatric to adult care in an outpatient pediatric rheumatology clinic. Eligibility criteria included established patients, age 14 or older, with chronic rheumatic disease, present with a legal guardian. The SDoH screener, adapted from the Health Leads Screening Toolkit, was given to eligible patients every 6 months (Figure 1). The primary outcome was the percent of eligible patients who completed the SDoH screener, and the secondary outcome was the percent of positive screens who met in-person or via phone with the clinic's social worker. A daily secure message was sent to providers and medical assistants to confirm family suitability for receiving the SDoH screener. Upon checking-in, eligible patients' guardians were given the screener and a cover sheet explaining its purpose. Medical assistants collected the screeners and promptly shared results with the provider and social worker in the original text thread. Families with positive screens, defined as one or more "yes" responses, were asked if they would like to meet with the social worker. The SDoH screener was integrated into the clinic in August 2022. At the end of August, a meeting was held to reeducate staff about the SDoH screening process flow (Plan-Do-Study-Act (PDSA) cycle 1). In October 2022, the workflow was modified to include patients seen in the infusion center (PDSA cycle 2).

Results: Preliminary results were collected August through December, 2022. On average, 71% of eligible patients completed the SDoH screener each month (Figure 2). Of those completed (n = 59), 15% (n = 9) of patients screened positive. 89% of patients with a positive screen directly interfaced with social work at the time of their visit or via phone

shortly thereafter. Identified barriers to completion of the SDoH screener include families feeling psychosocially/medically overwhelmed, staff forgetting to disseminate the SDoH, and the morning message not being sent before family arrival (Figure 3).

Conclusion: Implementation of a standardized SDoH screener was successful through interdisciplinary teamwork and communication. This has led to identification of potential barriers to care and proactive integration of social work support for the adolescent rheumatology patients. Next steps include collecting patient-facing feedback regarding SDoH screening acceptability and collection procedures, analysis of the correlation between SDoH screener results and transition outcomes and implementation of further PDSA cycles to address barriers to integration of SDoH screeners into clinic workflow.

Disclosure: S. Campbell: None; **R. Peterson**: None; **S. Barrientos**: None; **E. Benett**: None; **C. Christenholz**: None.

Abstract Number: 134

Patient-Provider Communication in Pediatric Rheumatology

Julie Samuels, Emma Wojtal and Rebecca Trachtman, Icahn School of Medicine at Mount Sinai, New York, NY

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Quality, Health Services, and Education II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Communication is an essential part of medical care, especially in Pediatric Rheumatology where children have varying complex chronic diseases. However, there is paucity of data about effective patient-provider communication. Studies have shown that improvements are required in information accessibility and ensuring children feel safely engaged in healthcare conversations. Further evidence suggests that good communication leads to improved adherence and outcomes, as well as treatment satisfaction in children with juvenile idiopathic arthritis (JIA). In this study, we aim to evaluate communication quality in Pediatric Rheumatology clinical practice using the Communication Assessment Tool (CAT) and assess the correlation between the CAT and patient-specific factors.

Methods: We consecutively recruited 41 patients with follow-up appointments between 10/27/2022 and 12/22/2022. Fifteen patients declined participation. Parent proxies completed surveys for children younger than 9 years. Participants completed a demographic survey, CAT, Patient-Reported Outcomes Measurement Information System Pediatric Global Health Measure 7 (PROMIS), and Connor Davidson Resilience Scale 2 (CD-RISC 2). Raw PROMIS scores were converted to T-scores using standard software. Spearmans correlations were performed with statistical significance set to p 0.05.

Results: The majority of our cohort was female (68%) and had Medicaid insurance (66%). The average age was 12.98 years. The patients primary visit diagnoses included systemic rheumatic disease (48.8%), JIA (14.6%), Uveitis (9.8%), and other (26.8%). The mean CAT score was 4.78 and median was 5 [IQR 4.79, 5], indicating overall high satisfaction with communication quality. Average PROMIS was 46.76 ± 8.89 and CD-RISC 2 was 5.24 ± 2.0, both consistent with reported means for the general population. CAT was only correlated with gender ($r = -0.34$, $p = 0.03$), with males endorsing lower scores; while age was strongly negatively correlated with PROMIS ($r = -0.65$, p 0.001), with older children endorsing worse health-related quality of life (HRQOL) (Table 2).

Conclusion: The mean CAT score of 4.78 indicates overall high communication quality in our practice; however, the percentage of excellent scores (5) was 17.8% lower than the initial report, indicating that areas for improvement exist. There were no significant correlations between CAT and PROMIS, CD-RISC 2, or diagnosis, suggesting that communication

Table 1. Demographic Data. Demographic data for patients recruited in Pediatric Rheumatology.

Characteristic	N = 41
Age	
Mean years ± SD	12.98 ± 5.56
Median years (IQR)	15 (9,17)
<9 years	10 (24.4%)
≥ 9 years	31 (75.6%)
Gender	
Female	28 (68.3%)
Male	13 (31.7%)
Ethnicity	
Hispanic	21 (51.2%)
Not Hispanic	19 (46.3%)
Unknown / Not Reported	1 (2.4%)
Race	
Asian	4 (9.8%)
Black or African American	11 (26.8%)
White	12 (29.3%)
More Than One Race	8 (19.5%)
Unknown / Not Reported	3 (7.3%)
Other	3 (7.3%)
Insurance	
Private	14 (34.1%)
Medicaid	27 (65.9%)
Income	
Less than $20,000	12 (29.3%)
$20,000 - $60,000	13 (31.7%)
$60,000 - $100,000	4 (9.8%)
Greater than $100,000	5 (12.2%)
Not reported	7 (17.1%)
Clinic Day	
Tuesday	21 (51.2%)
Thursday	17 (41.5%)
Friday	3 (7.3%)
Diagnosis	
Juvenile idiopathic arthritis	6 (14.6%)
Uveitis	4 (9.8%)
Systemic rheumatic disease	20 (48.8%)
Other	11 (26.8%)

Table 2. Spearman Correlation Coefficients. Bolded R value signifies p<0.05. Gender is correlated with the Communication Assessment Tool, with males endorsing worse communication quality. PROMIS is negatively correlated with age, with older children endorsing worse quality of life.

Characteristic	Sample Size	Communication Assessment Tool R Value	PROMIS PGH 7 R Value
Age	41	-0.11	**-0.65**
Gender	41	**-0.34**	-0.15
Diagnosis	41	0.13	-0.06
Clinic Day	41	-0.12	-0.03
Income	41	0.00	0.25
Insurance	41	-0.08	-0.11
Provider	41	-0.11	0.13
Parent vs. Patient	41	0.18	**0.32**
PROMIS PGH7	41	0.22	n/a
CDRISC 2	41	0.03	**0.31**

quality is not associated with HRQOL, resilience, or presentation. Male patients endorsed worse communication quality; given that the majority of our providers are female, additional research is needed to determine the drivers of lower scores in males, and whether they may be due to different communication styles or intrinsic biases. Older patients reported worse HRQOL than younger children, and the reasons for this are unclear. This study is limited by a small sample size in a single center. Additional research is needed to further assess the drivers of high and low communication quality and improve communication in this population.

Disclosure: J. Samuels: None; **E. Wojtal**: None; **R. Trachtman**: None.

Abstract Number: 135

Assessing Medication Adherence in JIA: Pilot Phase Results from a Single-Center Quality Improvement Initiative

Dori Abel[1], Joyce Chang[2], Jon Burnham[3], Chen Kenyon[4] and Sabrina Gmuca[5], [1]Department of Pediatrics, Division of Rheumatology, Children's Hospital of Philadelphia; PolicyLab, Children's Hospital of Philadelphia, Philadelphia, PA, [2]Boston Children's Hospital, Boston, MA, [3]Department of Pediatrics, Division of Rheumatology, Children's Hospital of Philadelphia; Department of Pediatrics, University of Pennsylvania Perelman School of Medicine, Philadelphia, PA, [4]Department of Pediatrics, University of Pennsylvania Perelman School of Medicine; PolicyLab, Children's Hospital of Philadelphia, Philadelphia, PA; Leonard Davis Institute of Health Economics, University of Pennsylvania Perelman School of Medicine, Philadelphia, PA, [5]Department of Pediatrics, Division of Rheumatology, Children's Hospital of Philadelphia; Center for Pediatric Clinical Effectiveness, Children's Hospital of Philadelphia; PolicyLab, Children's Hospital of Philadelphia; Department of Pediatrics, University of Pennsylvania Perelman School of Medicine, Philadelphia, PA

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Quality, Health Services, and Education II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Suboptimal medication adherence is a widespread problem in JIA. There are several unique features to medication adherence in JIA, including that the medications used are 1) primarily injectable or intravenous with varying administration frequencies, 2) difficult to safely dispose of if unused, and 3) quite expensive. Our pediatric rheumatology practice is currently developing a standardized method for assessing and addressing medication adherence barriers in patients with JIA during clinical care. Through a pilot phase of a quality improvement (QI) initiative, we aimed to test the feasibility of the Barrier Assessment Tool (BAT), created by the Pediatric Rheumatology Care and Outcomes Improvement Network (PR-COIN), within our local clinic setting and to explore perceived barriers and facilitators of adherence among a small sample of patients with JIA and their caregivers to inform future QI and qualitative work, with the long-term goal of improving adherence and minimizing the burden of treatment.

Methods: This single-center, pilot phase, QI study leveraged outpatient visit-level data from a large pediatric rheumatology clinic between May and December 2022 to assess medication adherence in JIA. One provider piloted a brief interview script to prompt discussion about medication adherence during routine clinical visits among a convenience sample of children with JIA and their caregivers. The BAT was administered to caregivers at the same visit.

We retrospectively reviewed responses to adherence interviews and the BAT and manually abstracted clinical information. We reviewed the interview text documented within the visit note to describe responses and summarize commonly reported themes.

Table 1. Patient Characteristics and BAT/Interview responses (total N=10)

Characteristics	Value
Age	11.5 (7.75-12.75)
Biological sex	
Female	8 (80%)
Male	2 (20%)
Race	
White	10 (100%)
Insurance type	
Private	6 (60%)
Public	3 (30%)
Self-pay	1 (10%)
Medication and route	
Methotrexate PO monotherapy	1 (10%)
Methotrexate SQ monotherapy	3 (30%)
Adalimumab SQ monotherapy	1 (10%)
Methotrexate SQ + adalimumab SQ combined therapy	3 (30%)
Etanercept SQ monotherapy	2 (20%)
JIA subtype	
Oligoarticular	6 (60%)
Polyarticular, RF-negative	3 (30%)
Undifferentiated	1 (10%)
Co-morbid mental health diagnosis	
Anxiety	1 (10%)
cJADAS	0.5 (0-1.875)
Disease duration	
< 6 months	3 (30%)
6-12 months	2 (20%)
13-24 months	2 (20%)
> 4 years	3 (30%)
Any missed medication doses in past 12 weeks	
Yes	3 (30%)
No	7 (70%)
Interested in JIA support group	
Yes	3 (30%)
No	7 (70%)
Number of identified barriers on BAT	
0	2 (20%)
1	2 (20%)
2	1 (10%)
3	2 (20%)
4	1 (10%)
5	2 (20%)

Categorical values are reported as counts and percentages. Continuous variables are reported as medians and interquartile ranges (IQR).

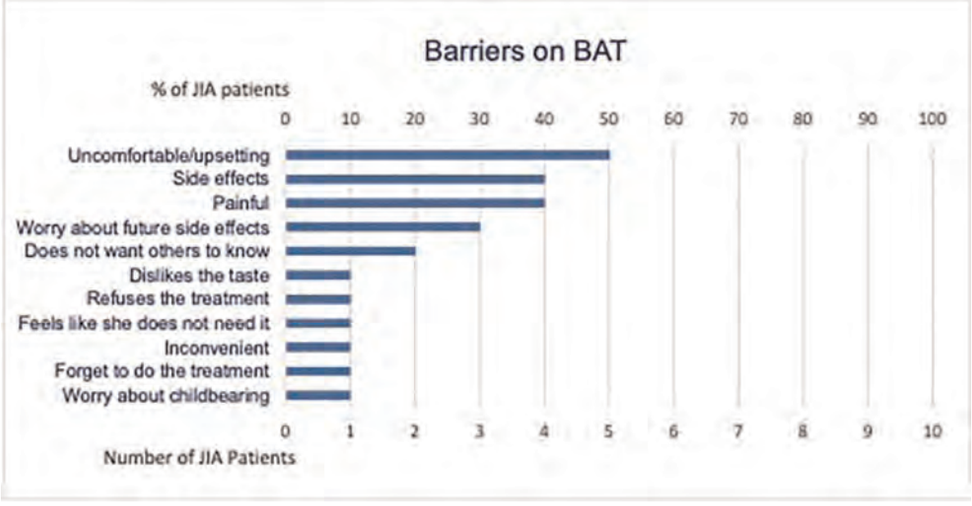


Figure 1. Barriers on BAT

Table 2. Moderators of Medication Adherence

	Barriers/Facilitators	Examples	N (%)
Barriers	Pain	Physical pain of the needle Burning sensation	5 (50%)
	Anxiety/fear	Needle anxiety Fear/anticipation prior to injection	5 (50%)
	Side effects	Nausea the day after injection Fatigue the day after injection General sick feeling the next day	4 (40%)
	Difficult to remember	No routine or reminder system Delaying the injection, then forgetting	3 (30%)
Facilitators	Belief in medication	Knowing that it helps prevent flares Knowing my disease will be better controlled Knowing that it helps my joint pains	6 (60%)
	Injection routine	Icing before Icing after Use of a phone for distraction Hold a stuffed animal Hold caregiver's hand Watch a TV show	6 (60%)
	Reward	Eating ice cream after injection Eating candy after injection	2 (20%)
	Access and affordability	Pharmaceutical company has good customer service Low copays	2 (20%)

Results: A total of 10 patients were included in this study (Table 1). Patients were primarily white females with oligoarticular JIA on subcutaneous medications. Only 30% reported recent missed doses. Most patients reported at least 1 barrier (Figure 1). Patients completed the BAT with ease and expressed the importance of discussing medication adherence.

Table 2 highlights emergent themes from the interviews regarding hypothesized moderators of medication adherence. Barriers included pain and anxiety related to injections, side effects, and forgetfulness. Facilitators included belief that the medication is helpful, injection routines, food rewards, and ease of access.

Conclusion: Our findings suggest that, despite few reports of missed doses, patients with JIA experience significant challenges associated with taking their medications and are interested in discussing them. Patients reported various factors impacting adherence and only some were interested in peer support, suggesting that interventions to address adherence and treatment burden may not be a one-size-fits-all approach.Given that self-reported adherence is known to overestimate true adherence, our findings should be validated with pharmacy fill data. Our study team aims to conduct a larger study using fill data to measure adherence combined with semi-structured interviews to further explore mechanisms influencing adherence. Future QI work will focus on assessing and addressing adherence barriers on a larger, division-wide scale.

Disclosure: D. Abel: None; **J. Chang**: GlaxoSmithKlein(GSK), 5; **J. Burnham**: None; **C. Kenyon**: IngenioRx, 12, Member of the Pharmacy and Therapeutics Committee; **S. Gmuca**: None.

Abstract Number: 136

Incidence and Disease Burden of Juvenile Idiopathic Arthritis and Rheumatoid Arthritis After Non-Pharmaceutical Interventions in the COVID-19 Era: A Nationwide Observational Study in Korea

Je Hee Shin[1], Jung Yoon Pyo[2], Minkyung Han[3], Myeongjee Lee[3], Sung Min Lim[1], Jee Yeon Baek[1], Ji Young Lee[1], Ji-Man Kang[1], InKyung Jung[3] and **Jong Gyun Ahn**[1], [1]Severance Children's Hospital, Yonsei University College of Medicine, Seoul, Republic of Korea, [2]Department of Internal Medicine, Yonsei University College of Medicine, Seoul, Republic of Korea, [3]Department of Biomedical Systems Informatics, Yonsei University College of Medicine, Seoul, Republic of Korea

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Quality, Health Services, and Education II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Several countries have implemented non-pharmaceutical interventions (NPIs) against the coronavirus disease 2019 (COVID-19) pandemic. We investigated the impact of NPIs on the incidence of juvenile idiopathic arthritis (JIA) and rheumatoid arthritis (RA) in Korea and that of the COVID-19 pandemic on healthcare services and medical expenses for patients with JIA and RA.

Methods: We included all cases of JIA and RA reported between January 2016 and February 2021 based on the National Health Insurance Service data. NPI period was defined as February 2020–February 2021. We evaluated the change in incidence trends for JIA and RA before and after NPI implementation using segmented regression analysis. Changes in health care utilization and medical costs for JIA and RA before and after NPI implementation were also investigated.

Results: A significant decreasing trend was observed over time in the pre-NPI period for both JIA and RA (-0.015 per 1,000,000, 95% confidence interval [CI] -0.021 to -0.009, $p < 0.001$ in JIA; -0.110 per 1,000,000, 95% CI -0.177 to -0.044, $p=0.002$ in RA). However, there was no significant change in the incidence trend between the NPI period and the pre-NPI period for both JIA and RA. A significant decrease was observed in the annual average number of hospitalizations, the mean days of hospitalization and the number of annual OPD visits for any cause per patient during the NPI period for RA, whereas there was no statistically significant change for JIA. Further, the prescription days per OPD visit significantly increased during the NPI period compared to those in the pre-NPI period for RA, conversely, they significantly decreased during the NPI period compared to those in the pre-NPI period for JIA (28.0±29.6 vs 23.0±29.0, $p < 0.001$). Although the total annual medical costs per patient for RA decreased during the NPI period, there was no significant difference in those for JIA compared to before NPI.

Conclusion: There was no change in the incidence of RA and JIA during the COVID pandemic period. In RA, there were changes in the medical utilization pattern and cost of treatment during the pandemic period, whereas no significant changes were observed in JIA.

Disclosure: J. Shin: None; **J. Pyo**: None; **M. Han**: None; **M. Lee**: None; **S. Lim**: None; **J. Baek**: None; **J. Lee**: None; **J. Kang**: None; **I. Jung**: None; **J. Ahn**: None.

Abstract Number: 137

Geographic Mapping of Adolescents with Rheumatic Disease: Racial and Ethnic Diversity by Texas County

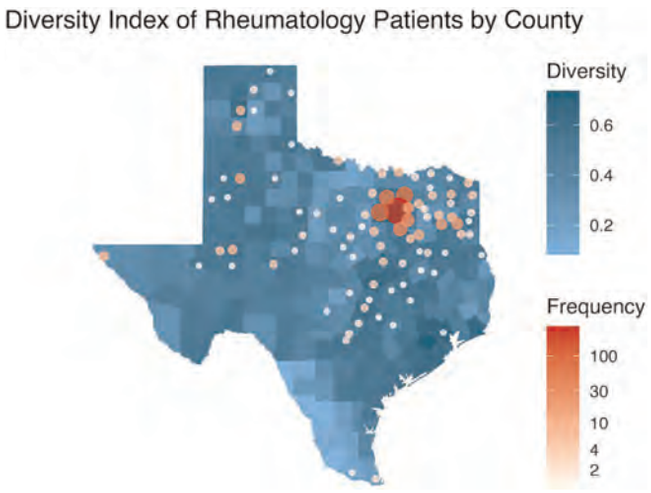
Kristina Ciaglia[1], Chan-hee Jo[2], Yuhan Ma[2], Tracey Wright[3] and Lorien Nassi[1], [1]University of Texas Southwestern, Dallas, TX, [2]Scottish Rite Hospital for Children, Dallas, TX, [3]University of Texas Southwestern, Scottish Rite Hospital for Children, Dallas, TX

SESSION INFORMATION
Session Date: Friday, March 31, 2023
Session Title: Posters: Quality, Health Services, and Education II
Session Type: Poster Session B
Session Time: 5:00PM–6:00PM

Background/Purpose: Rheumatic disease disproportionately impacts specific racial and ethnic groups frequently, resulting in health care inequities. Health care disparities are prevalent within certain geographic areas including the Southern United States, and Texas has been reported as having lower quality of care and worse racial and ethnic disparities.[1] The LUMINA cohort demonstrated that adult Texan Hispanics with lupus have increased disease activity and greater damage accrual compared to other groups.[2] Our objective was to estimate the geographic distribution of race and ethnicity in a cohort of adolescents with rheumatic disease followed at a single center to begin to explore risk factors for disparities in our patient population.

Methods: Demographic data of patients 14 years of age and older with a permanent Texas address was extracted from the EMR. Zip codes were translated into counties and mapped by geographic region and color coded by numeric frequency. Diversity index (DI) extracted from the 2013 Census data was used to measure the probability that two people chosen at random will be from different racial and ethnic groups. The DI ranges from 0 indicating that the population has the same racial and ethnic characteristics, to 1 indicating completely different backgrounds.[2] Using zip code and county the DI was reported geographically.

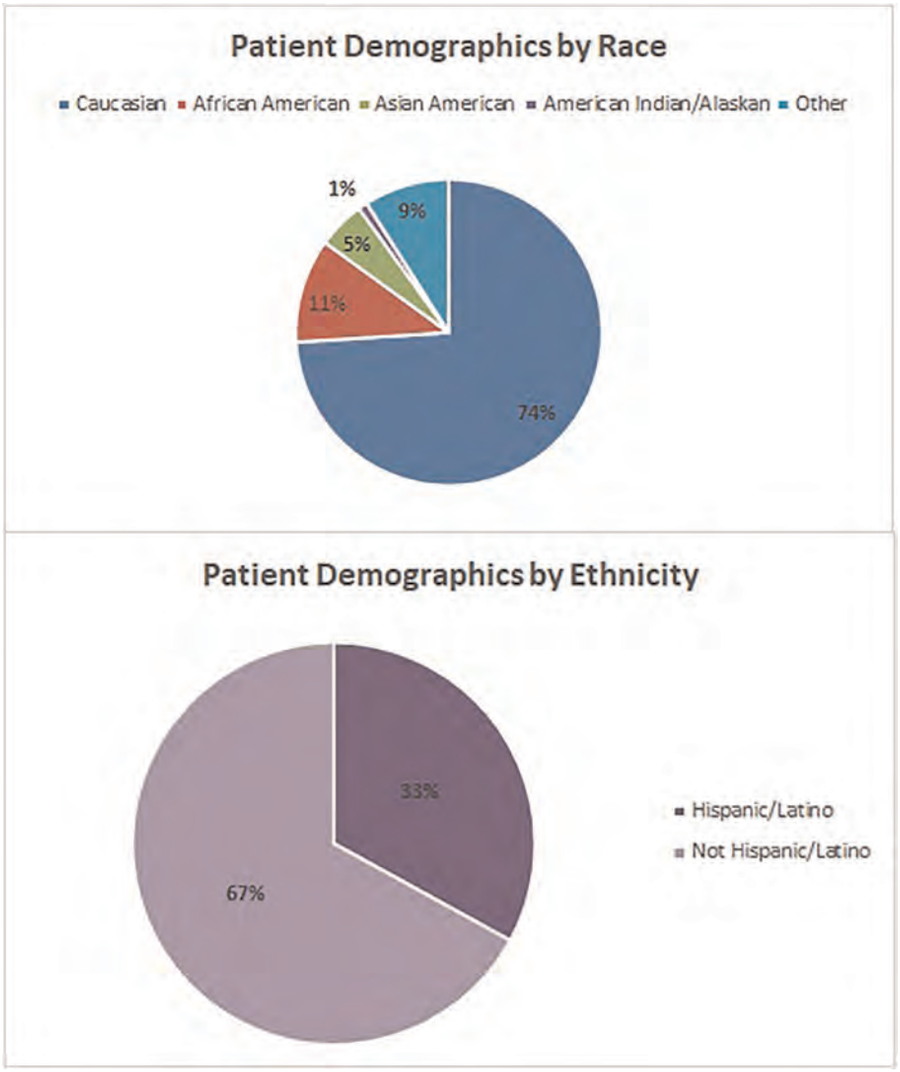
Results: A total of 834 unique patients were identified and mapped geographically. Forty-three patients were excluded as they lived out of state. Seventy-six percent identified as female. Seventy-four percent reported Caucasian race, and 11% African American. Thirty-three percent identified as Hispanic/Latino ethnicity. Fifty-eight percent had private commercial



The distribution of adolescent rheumatology patients across Texas by patient frequency and county diveristy index (DI)

Table dipicting Texas Counties with the least and most diversity, and most patient frequency in a single center rheumatology clinic

Texas County	Diversity Index	Number of Patients
Collin	0.578365	71
Dallas	0.69039	277
Denton	0.558831	57
Ellis	0.518114	42
Gregg	0.572375	20
Henderson	0.350927	11
Kaufman	0.490261	30
Lubbock	0.570089	12
Rockwall	0.437303	17
Tarrant	0.644473	86
Smith	0.558215	17
Fort Bend	0.736427	0
Starr	0.082592	0



Patient demographics by race and ethnicity

insurance, 34% government insurance, and 8% were uninsured. The county with the highest DI was Fort Bend, and the lowest DI in Starr County. Most (33%) patients reside in Dallas County. The farthest distance counties included Cameron at 478 miles and El Paso at 572 driving miles from clinic.

Conclusion: Patients in our center come from diverse racial and ethnic backgrounds. Some have the burden of driving a long distance to their pediatric rheumatologist. Ease of access to a pediatric rheumatologist is a health care systems factor that may contribute to disparities.[3] Provider awareness of patient diversity, in terms of both race/ethnicity and geography is essential to understanding inequities in health care. Our findings demonstrate a broad range of diversity across Texas and are the first step in evaluating barriers to care.

1. 2022 National Healthcare Quality and Disparities Report. AHRQ Pub. No. 22(23)-0030.

2. Gonzalez LA, Pons-Estel GJ, Toloza SMA, Ugarte-Gil MF, Alarcon GS. Understanding Risk Factors for Poor Outcomes in a Multiethnic Longitudinal Cohort: The LUMINA (Lupus in Minorities: Nature vs. Nurture) Experience (LUMINA LXXXII). *Rheum Dis Clin North Am*. Feb 2021;47(1):55-64. doi:10.1016/j.rdc.2020.09.002

3. Census US. Racial and Ethnic Diversity in the United States. 2023.4. Rubinstein TB, Knight AM. Disparities in Childhood-Onset Lupus. *Rheum Dis Clin North Am*. Nov 2020;46(4):661-672. doi:10.1016/j.rdc.2020.07.007

Disclosure: K. Ciaglia: None; **C. Jo**: None; **Y. Ma**: None; **T. Wright**: None; **L. Nassi**: None.

Author Index

Keyword Index