O070

Rheumatic diseases in Mexican children and their psychosocial and economic impact on caregivers

F. García Rodríguez[1], B. D. J. Fortuna Reyna [2], I. Peláez Ballestas[3], E. Faugier Fuentes [4], S. Mendieta Zerón [5], G. Reyes Cordero[6], S. Jiménez Hernández[7], A. V. Villarreal Treviño[1], J. Guadarrama Orozco[8], S. Rosiles de la Garza[9], M. De la O Cavazos[9], N. Rubio Pérez[7]
[1]Pediatric Rheumatology, Hospital Universitario "Dr.José E. González"; [2]Pediatric Rheumatology, Hospital Universitario "Dr. José E. González", Monterrey; [3]Rheumatology, Hospital General de México "Dr. Eduardo Liceaga"; [4]Pediatric Rheumatology, Hospital Infantil de México Federico Gómez, CDMX; [5]Pediatrics, Hospital Regional de Toluca ISSEMyM, Toluca; [6]Rheumatology , Hospital Infantil de Especialidades de Chihuahua, Chihuahua; [7]Pediatric Rheumatology, Hospital Universitario "Dr. José E. González", Monterrey; [8]Palliative Care, Hospital Infantil de México Federico Gómez, CDMX; [9]Pediatrics, Hospital Universitario "Dr. José E. González", Monterrey, Mexico
Correspondence: F. García Rodríguez
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Introduction: Pediatric rheumatic diseases (PRD) are a heterogeneous group of disorders. PRD patients and their caregivers face a number of challenges, these include the consequences of the PRD in patients and the impact on multiple dimensions of the caregiver's daily life. Our group developed and validated the CAREGI VERS questionnaire to measure the impact on caregivers of children with PRD.
Objectives: The objective of this study was to measure the economic, psychological and social impact that PRD has on the caregivers of Mexican children and the factors associated with these impacts.
Methods: This is a cross-sectional study in which primary caregivers were prospectively included between April and November 2019 in four public hospitals of specialized care. Descriptive statistics used used to the sociodemographic characteristics of the participants and the patients' clinics, a univariate analysis was performed with the interview responses of the CAREGIVERS questionnaire and the sociodemographic, clinical, and health system variables using the Chi square, Mann-Whitney U, and Kruskal-Wallis tests ($p < 0.05$).
Results: 200 participants were included, women (84.5%) with median age of 38 years; 54.5% cared for patients with JIA, 14% with JDM and 31.5% with JSLE. Most of the caregivers felt concern (42.5%) when learning about the diagnosis, which then was modified by tranquility (44%) when the current feeling was questioned; however, 40 expressed sadness when sharing the patient's PRD (20%) and 39 do not like to do so (19.5%).The main cause of concern is pain (41.5%), followed by difficulty in movement (28.5%) and covering the costs of treatment (25%). Social impact: In 99 caregivers (49.5%), the use of their time changed a lot upon learning the PRD. Social life varied according to the PRD, in JSLE it had a significant change (39.6%), but it did not change in JIA (44%) and it slightly changed in JDM (53.5%, $p < 0.01$).Financial impact: the family financial situation worsened upon diagnosis of the patient in most cases (JIA 63 [57.8%], JSLE 19 [69.8%] and JDM 44 [67.8%], $p = 0.27$). Almost two thirds had had to borrow money, more frequently in JSLE (48 [76.1%] vs JIA 62 [56.8%] and JDM 19 [67.8%], $p = 0.03$); 63 stopped buying medicines due to lack of money (31.5%) and 86 received additional financial support for the treatment (43%).The emotional impact increased in caregivers of male patients. Social dimension showed significant differences regarding PRD, healthcare system, time to reach the center, presence of disability, active disease, cutaneous and systemic manifestations and treatment.
Conclusion: This study highlights a series of lessons learned and the most important is the need to improve opportunities for support, especially regarding financial support, for caregivers of patients with PRD. The study has shown that social status can be devastating in the impact that PRD can have on families. We feel confident that, although all the participants are Mexican, the findings can be generalized to populations with similar characteristics in other regions.

Disclosure of Interest
None declared

O071

Barriers and facilitators to physical activity in Juvenile Idiopathic Arthritis (JIA): a scoping review

L. Larkin[1,2], K. O'Donoghue[1]
[1]School of Allied Health; [2]Health Research Institute, University of Limerick, Limerick, Ireland
Correspondence: L. Larkin
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Introduction: Physical activity is an important aspect in the management of JIA (Kuntze et al 2018). However physical activity levels are low in this population (Bos et al 2016). Limited research has been conducted to identify definitive barriers and facilitators to physical activity in children and adolescents who have JIA.
Objectives: The objective of this scoping review was to identify the common barriers and facilitators to physical activity in JIA.
Methods: Original studies, either quantitative or qualitative, including participants with a diagnosis of JIA, who were under 18 years of age were included. Two independent reviewers carried out a search of the literature and full text reviews of papers to determine eligibility for inclusion. The Critical Skills Appraisal Programme (CASP), Appraisal tool for Cross-Sectional Studies (AXIS) and Downs and Black critical appraisal tools were used to assess the quality of the included research articles.
Results:

Category	Quantitative studies (N)	Qualitative studies (N)
Barriers		
Physical barriers	N=13	N=4
Psychological barriers	N=7	N=5
Management barriers	N=5	N=3
Other barriers	N=7	N=0
Facilitators		
Physical facilitators	N=5	N=0
Psychological facilitators	N=6	N=6
Management facilitators	N=10	N=7
Other facilitators	N=4	N=0

Eighteen studies were included in the review. The included studies were of a variety of low, moderate and high quality. The synthesis of the data identified pain to be the most common barrier and the modification of physical activities to the need of the individual to be the most common facilitator to physical activity in JIA.
Conclusion: Identifying the most common barriers and facilitators to physical activity allows clinicians to apply better management strategies when treating an individual with JIA. Our findings demonstrate the need for further research in this area to assist increasing physical activity participation for children and adolescents who have JIA.

Disclosure of Interest
None declared

O072

Under detection of interstitial lung disease in Juvenile Systemic Sclerosis (JSSC) utilizing pulmonary function tests. Results from the juvenile scleroderma inception cohort.

I. Foeldvari[1], B. Hinrichs[2], K. Torok[3], M. J. Santos[3], O. Kasapcopur[3], A. Adrovic[3], V. Stanevicha[3], F. Sztajnbok[3], M. T. Terreri[3], A. P. Sakamoto[3], E. Alexeeva[3], J. Anton[3], M. Katsikas [3], V. Smith[3], T. Avcin [3], R. Cimaz[3], M. Kostik[3], T. Lehman[3], W.-A. Sifuentes-Giraldo[3], A. S. Appenzeller, Simone[3], M. Janarthanan [3], M. Moll[3], D. Nemcova [3], D. Schonenberg-Meinema[3], C. Battagliotti[3], L. Berntson[3], B. Bica[3], J. Brunner[3], P. Costa Reis[3], D. Eleftheriou[3], L. Harel[3], G. Horneff[3], T. Kallinich[3], D. Lazarevic[3], K. Minden[3], S. Nielsen[3], F. Nuruzzaman[3], A. Patwardhan[3], Y. Uziel [3], N. Helmus[1]
[1]Hamburg Center for Pediatric and Adolescent Rheumatology, Am Schoen Klinik Eilbek; [2]Kinderklinik Heidberg Pulmologie; [3]jSSc Collaborative Group, Hamburg, Germany
Correspondence: I. Foeldvari
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Introduction: Juvenile systemic sclerosis (jSSc) has a prevalence in around 3 in a million children. Pulmonary involvement occurs in approximately 40 % in the international juvenile systemic scleroderma cohort (JSScC). Traditionally in jSSc, pulmonary function testing (PFT) with FVC and DLCO are used for screening and computed tomography (HRCT) was more reserved for those with abnormal PFTs. More recently, it has become apparent that PFTs might not be sensitive enough for detecting interstitial lung disease (ILD) in children.
Objectives: To assess the sensitivity and specificity of FVC and DLCO assessment to detect ILD
Methods: The international juvenile systemic scleroderma cohort (JSScC) database was queried for available patients with recorded PFT parameters and HRCT performed to determine sensitivity of PFTs detecting disease process.
Results: Of 129 patients in the jSScC, 67 patients had both CT imaging and an FVC reading from PFTs for direct comparison. DLCO readings were also captured but not in as many patients with tandem HRCT (n =55 DCLO and HRCT scan). Therefore, initial analyses focused on the sensitivity, specificity and accuracy of the FVC value from the PFTs to capture the diagnosis of interstitial lung disease as determined by HRCT.
Overall, 49% of the patients had ILD determined by HRCT, with 60% of patients having normal FVC (>80%) with positive HRCT findings, and 24% of patients having normal DLCO (> 80%) with positive HRCT findings. Fourteen percent (n = 3/21) of patients with both FVC and DLCO values within the normal range had a positive HRCT finding.
Conclusion: The sensitivity of the FVC in the JSScC cohort in detecting ILD was only 39%. Relying on PFTs alone for screening for ILD in juvenile systemic sclerosis would have missed the detection of ILD in almost 2/3 of the sample cohort, supporting the use of HRCT for detection of ILD in children with SSc. In addition, the cut off utilized, of less than 80% of predicted FVC or DLCO could be too low for pediatric patients to exclude beginning ILD. This pilot data needs confirmation in a larger patient population.
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Disclosure of Interest
None declared

O073

Cross-cultural adaptation and validation of the Localised Scleroderma Quality of Life Instrument (LOSQI) in JLS: a multicentre study of the PRES scleroderma working party in collaboration with members of the carra scleroderma working group

C. Pain[1], H. Lythgoe[1], V. Leone[2], G. Martini[3], O. Kasapcopour[4], K. Torok[5], S. Li[6], K. Ardalan[7], C. Zigler[7], I. Foeldvari[8] on behalf of PRES WP2.0 scleroderma and PRINTO
[1]Alder Hey Children's NHS Foundation Trust, Liverpool; [2]Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom; [3]Padova University, Padova, Italy; [4]Cerrahpasa Medical School, Istanbul University, Cerrahpasa, Turkey; [5]Children's Hospital of Pittsburgh of the University of Pittsburgh Medical Center, , Pittsburgh; [6]Joseph M. Sanzari Children's Hospital; Hackensack Meridian School of Medicine, New Jersey; [7]Duke University School of Medicine, , Durham, United States; [8]Hamburg Center for Pediatric and Adolescent Rheumatology, Am Schoen Klinik Eilbek, Hamburg, Germany
Correspondence: C. Pain
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Introduction: ***Juvenile localised scleroderma (JLS)*** is characterised by chronic inflammation within the skin and tissues leading to fibrosis [1]. It is associated with significant complications including joint contractures, limb length discrepancy and facial atrophy that impact quality of life. Patient reported outcomes (PRO) are not well established within research settings and are not part of routine clinical care in many centres [2].
Several studies have measured health-related quality of life (HRQoL) in JLS, most commonly using the Children's Dermatology Life Quality Index[3-6]. This measure only captures impact of skin involvement and not HRQoL from extra-cutaneous manifestations. Psychometric analysis shows that it incompletely measures important concepts of HRQoL that are unique to JLS [7]. The Localised Scleroderma Quality of Life Instrument (LoSQI) has been developed in partnership with patients and families to capture aspects of disease which may not be well defined within generic HRQoL measures [8]. The initial development and validation process was iterative, patient-centred, and consistent with best practices in PRO development. Currently, the LoSQI is the only JLS-specific PRO, and the only PRO that includes both qualitative and quantitative validity evidence. It is currently being utilized within two large American scleroderma registries but will require cross-cultural adaptation for international use.
Objectives: to undertake cross-cultural adaptation and validation of the Localised Scleroderma Quality of Life Instrument (LoSQI) in juvenile localised scleroderma (JLS).
Methods: Workstream 1: cross-cultural adaptation of LoSQI via methods previously described by Guillemin et al, with pre-testing in selected study population. A single site in up to 35 PRINTO represented countries will take part.
Workstream 2: validation of the LoSQI via a multicentre prospective cohort study of 100 patients at 2 time points.
Results: This study was successful in obtaining funding from the **PRES 2025 / PRINTO Research Award** and is in study set up stage.
Conclusion: Collaboration between PRINTO centres, CARRA and PRES scleroderma working party members in partnership with patients and families will facilitate shared aims and this will be the first multinational study of a disease-specific patient-reported outcome (PRO) in JLS. To allow further validation work of outcome measures, this important step will allow cohorts from multiple countries to combine datasets and results with an overarching aim to embed PRO in routine clinical practice. This is invaluable for a rare disease population, where research is continuously limited by small samples sizes, large geographical dispersion of subjects, and lack of consensus in selection and use of outcome measures.

Disclosure of Interest
None declared

O074

Ischemic stroke in children with scleroderma en coupe de sabre

M. S. Pavlova[1], M. K. Osminina[1], L. G. Khachatryan[1], N. S. Podchernyaeva[1], V. M. Trepilets[2], O. V. Shpitonkova [1], V. A. Seraya[2], S. V. Trepilets[3]
[1]N.F.Filatov Clinical Institute of Child Health; [2]Sechenov First Moscow State Medical University; [3]Burdenko National Medical Research Center of Neurosurgery, Moscow, Russian Federation
Correspondence: M. S. Pavlova
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Introduction: Neurologic disturbances (ND) in children with localized scleroderma (LS) occur more frequent in linear scleroderma en coupe de sabre (ECDS). The frequency of Central nervous system involvement in pediatric craniofacial scleroderma is estimated to be 28–38%. Mostly epilepsy, headache, focal symptoms, neuropsychiatric disorders are described. Only several cases of stroke were recorded in adults with ECDS.
The origin of ND is still unclear. There is data for neurovasculitis hypothesis with endothelial cell injury, microthrombotic angiopathy. Other data suggests a prenatal malformation of one side of rostral neural tube resulting in hemiatrophy of facial tissue and underlying brain parenchyma.
Objectives: To analyze frequency of neurologic involvement ECDS in children, describe 3 cases ischemic stroke (IS).
Methods: Retrospective analysis of ND in ECDS childhood cases was done. All children carried out physical and neurologic examination, brain magnetic resonance imaging (MRI), electroencephalography (EEG), rheumatological observation (physical, instrumental and laboratory including homocystein serum level (Hcy), evaluation for genetic thrombophilia (GThr).
Results: We observed 115 children with ECDS, aged from 3 to 16 years, the mean age 12,4 years (M ±3,52), 63 girls and 52 boys (girls/boys = 1.2:1).