Notes from the PR~~E~~eS Juvenile Scleroderma Working Group meeting-
Friday 25th of September 2020 (11.-12.30 am central European time)

Chair: Ivan Foeldvari

Notes: Ozgur Kasapcopur, secretary

Participants: 95-100.

Short Introduction: Ivan Foledvari

Program:

I 11.05- 11.20, Valentina Leone (Department of Paediatric Rheumatology, Leeds, UK)**:** Raynaud's phenomenon: a UK survey against the PRES Scleroderma Working Group recommendations.
-Valentina firstly presented data from the literature regarding the recommendation for assessing children with Raynand phenomenon
-Afterward, Valentina presented data from the electronic survey regarding the pediatric rheumatology practice for pediatric patients with RS in UK and Ireland
-64 respondents, 60% unaware of PReS recommendations
-Conclusion: variations in practice; poor access to imaging
-Plan: prospective 5-years study aiming to identify patients who are at risk of progression to scleroderma (pediatric arm of the adult study supported by the Grant from the Leeds Care)
-For those interested to take part in RS survey and further projects:
Valetina.leone@nhs.net, catrionaanderson86@gmail.com

Q (Natasa Toplak): How do you deal with patients who have "cold hands"?
A (Valentina Leone): The RS in pediatric patients is not clearly defined. There is a need for careful follow up.
A (Ivan Foeldvari): There is a need for standardization of NFC changes in pediatric population. It would be useful to perform the survey regarding the values and measurements that ped. Rheumatologists are looking for when performing the NFC. NFC in pediatric scleroderma teaching session would be useful.

II 11.20-11.35, Ivan Foeldvari (Hamburg Centre of Pediatric and Adolescent Rheumatology, Hamburg, Germany): Update of the juvenile systemic scleroderma inception cohort.
-Ivan presented data from the ongoing cohort on juvenile systemic sclerosis which is the largest cohort of the sort, temporary including 150 patients
-Ivan invited more participants:
www.juvenile-scleroderma.com
-Contact:
foeldvari@t-online.de, inceptioncohort@kinderrheumatologie.de

Q (Clare Pain): It would be useful to separate patients with long-term disease and those with new disease onset.

Q (Liza McCann): Do the NFC progress even if the disease improves?
A (Ivan Foeldvari): We have to look at that point. Currently it seems that NFC improve with disease improvement.
Q: Is the HRCT necessary?

A (Ivan Foeldvari): The HRCT is a standard for ILD but we should look for reduction in radiation.
A (Clare Pain): Baseline CT is highly important. Further follow-up with pulmonary function tests and CT with reduced number of slides seem reasonable.
A (Ivan Foeldvari): Follow-up with FVC and DLCO measurements and 8-min walking distance is reasonable. HRCT could be repeated according to results. We have to prevent lung fibrosis.

Q: What about data regarding the arrythmia that seems more common among patients with limited disease form?
A (Ivan Foeldvari): It would be valuable to perform study on cardiac MRI among pediatric patients, to assess the possible cardiac fibrosis (already done in adults).

III 11.35 -11.50, I**van Foeldvari** (Hamburg Centre of Pediatric and Adolescent Rheumatology, Hamburg, Germany) : Establishing a "Minimal data set " in the care of children with localized Scleroderma
-Ivan Foeldvari gave a short introduction of the project
-Ivan Foeldvari pointed out the importance and the need for the standardization of the follow-up and treatment for patients with juvenile localized scleroderma between pediatric rheumatologists and pediatric dermatologists.

IV 11.50-12.05, **Clare Pain** (Alder Hey Children's NHS Foundation Trust, Liverpool, UK): Cross-cultural adaptation and validation of the LoSQI (Localised scleroderma Quality of life Instrument
-Clare Pain gave a talk on details regarding the study related to validation and adaptation of LoSQI
-Feasibility: of 638 contacted centers, 248 (39%) responded; 158/248 (64%) interested. In participating; estimated number of patients 1048-2297
-Aim to open Workstream: 01.03.2021; Workstream: 02.09.2022

V 12.05-12.20, **Suzanne Li,** (Joseph M. Sanzari Children's Hospital, Hackensack Meridian Health, Hackensack, NJ, USA): Morbidity in Juvenile Localized Scleroderma: Capturing Cutaneous and Extracutaneous involvement with the Total Morbidity Score.
-Suzanne Li gave a talk on a morbidity score for juvenile localized scleroderma
-Conclusion: TMS shows good correlation with PGA-damage
-Plan: performance in more patients; module for head and neck should be improved.
-Contact: sl07078@gmail.com

Q (Ivan Foeldvari): Can we use TMS in our studies?
A (Suzanne Li): The score could be used in studies but we still need more refinement of the head and neurological scoring module. It is possible that it will work better to have the head and neurological module separate from the other 4 modules of the TMS, and that patients with head involvement would then have 2 scores, one of the TMS (4 modules) and the other of the Head and neurological module.

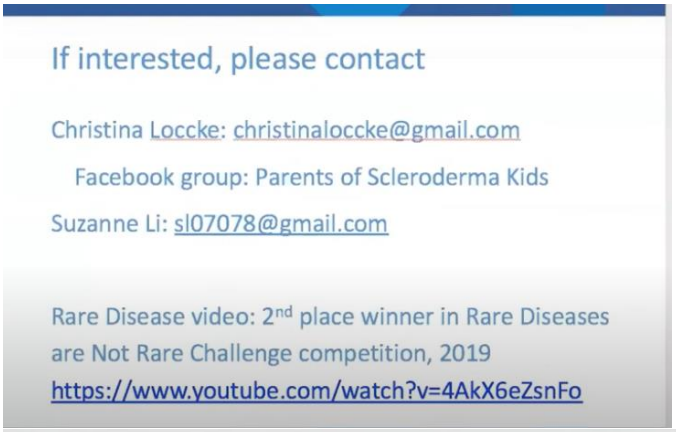
Q (Ivan Foeldvari): Did you ask patients to evaluate the damage?
A (Suzanne Li): No, this is the damage evaluation based on physicians'' assessment, so it is intended more to help the physicians track morbidity extent and severity.

Q: What is the minimum-maximum score for the damage?
A (Suzanne Li): The minimum score is 0. The maximum score is not clear, depends on the extend of damage; for a case vignette of a severe pansclerotic morphea patient, the total score was >60.

Q (Ivan Foeldvari): How do you evaluate the facial damage?
A (Suzanne Li): We score for level of facial disfigurement. Our CARRA localized scleroderma workgroup agreed upon 5 severity levels (mild to severe), and put photo examples of these levels in our scoring atlas; photos had to achieve $\geq$ 75% consensus agreement to be included. The TMS also scores for hair loss, dyspigmentation, tissue atrophy, and extent of skin damage features.

Q (Ivan Foeldvari): Is the atlas with pictures for evaluation available to everyone?
A (Suzanne Li): The atlas is copyrighted by CARRA and is available as a print version. We did not make it web-based because we found issues with the reproducibility of the color which made scoring erythema level less accurate. If you have a meeting, we could take pre-orders to cover costs and distribute at meeting.

VI 12.20-12.25, Christina Loccke (a parent of JLS child, Scleroderma Foundation, USA): F**amilies and Physicians Partnering to Develop a Pediatric Scleroderma Website.**
- Christina Locke gave details regarding the patients-physicians partnership and the development of pediatric scleroderma website.
-The website will fulfil the gap between pediatric scleroderma patients and physicians, provide accurate information about the disease and treatment to help counter some of the misinformation on the web, provide families with some resources, and facilitate helping patients and parents to connect with each other. .



Q (Ivan Foeldvari): It would be valuable for parents' representative of the PReS Scleroderma working group to get connected and make a collaboration.

VII 12.25-12.30, **Giorgia Martini** (University Children's Hospital, Padua, Italy): Update on the PRES Scleroderma Educational course.
- Giorgia Martini gave details on the educational course that was planned to be organized but it was postponed due to COVID-19 pandemic.
-The course was planned for 30 participants: young physicians particularly interested in juvenile scleroderma.
-The course would consist of two parts: theoretical and practical.
-Plan: to be organized in 2021.

VIIClare Pain: Transitional care during COVID-19 pandemic for patients with juvenile-onset inflammatory myopathies. (From the PReS JDM working party)
-Clare Pain give a short talk on proposed recommendations for transitional care for JDM patients during COVID-19 pandemic that are open for discussion.

Q (Ivan Foeldvari): There is a different approach to transition care among different countries.
A (Liza McCann): The transition is a process that should start at younger age before the patient has been transferred to adult physicians.

Closing remarks: Ivan Foeldvari