[FRI0537] UPDATE ON THE JUVENILE SYSTEMIC SCLEROSIS INCEPTION COHORT WWW.JUVENILE-SCLERODERMA.COM

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Background: Juvenile systemic sclerosis (jSSc) is a orphan autoimmune disease. Currently only retrospective data is existing without a standardized assessment regarding the organ involvement and evolvement of the disease. Our project is the first projects, where prospectively and with a standardized assessment data of early jSSc patients are collected.
Objectives: To learn about the evolvement of juvenile systemic sclerosis
Methods: Patients with less the 18 months of disease duration, after the first Non-Raynaud symptomatic, are prospectively assessed, using the proposed standardized patient assessment protocol.
Results: We report the characteristics of the patients at 0, at 6 and 12 months of follow up. We present date on 24 patients. The mean follow up of the patients in the cohort are 3.4 years. No patient died during the follow up. Eighteen of the 24 patients were female. The mean age of the onset of Raynaud symptomatic was 11.2 years, the youngest 4.2 years old. The mean age at the onset of the non-Raynaud symptomatic were 11.7 years. 20 of the 24 have diffuse subtype, 5 of them have an overlap symptomatic, two of them associated with diffuse subtype. ANA positive were 19, and 6 of them were anti-Scl 70 positive. None of them was anticentromere positive The mean modified Rodnan Skin Score was at timepoint 0, 6 and 12 month 19.4, 15.7 (n=19) and 15.6. (n=13). Raynaud's Phenomen occurred in 20/23 at time point 0 and 14 of 19 at time point 6 months and 10 of 15 at 12 months. 16 of 23 of them have capillary changes at time point 0, 9 of 18 in m6 and 7 of 14 in m12. 7 of them have already ulcerations at time point zero, 9 of 18 at month 6 and 4 of 10 at months 12. 13 of them have cardiopulmonary involvement, at time point zero already, 8 of them have interstitial lung disease. 4 of 19 have cardiopulmonary involvement in month 6 and 6 of 16 in month 12. Two of them have renal involvement at time point 0 and 3 at time point 6 and 12 months. 9 of 23 had gastrointestinal involvement, and 5 of them oesophageal involvement at time point zero, 3 from 18 at time point month 6 and 5 of 16 at time point 12 months. Twenty of 23 have musculoskeletal involvement 17 of 19 In month 6 and 15 of 16 at time point 12 months.
Conclusions: We present the data on the first 24 prospectively assessed patients with jSSc. The current recruitment data confirms that pediatric patients are different from the adult patients. Unfortunately despite the prospective data collection, we miss some data. We are only at the first phase of this project and hope to recruit up to 50 patients and follow them prospectively over the next 5 years at least.
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