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Is the Presentation and Severity Different of the Juvenile Diffuse and Limited Subtype Systemic Sclerosis? Results of Juvenile Scleroderma Inception Cohort

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SESSION INFORMATION

Session Date: Sunday, November 10, 2019

Session Title: Pediatric Rheumatology – ePoster I: Basic Science, Biomarkers, & Sclerodermic Fever

Session Type: Poster Session (Sunday)

Session Time: 9:00AM–11:00AM

Background/Purpose: Juvenile systemic scleroderma (jSSc) is an orphan disease with an estimated prevalence of 1 in 1 000 000 children. In the adult systemic scleroderma population there are large differences regarding organ pattern and severity between diffuse and limited subtypes.

Methods: We reviewed all patients of juvenile scleroderma inception cohort (jSScC) at the time of inclusion till 15th May 2019. The jSScC is a cohort, where patients, who fulfill the adult 2013 classification criteria , are age under 18 at the time of inclusion and developed the first non-Raynaud before the age of 16 years are included.

Results: 131 patients were included, 72.5% with diffuse subtype. 75% females in the diffuse (djSSc) and 71% in the limited subtype(ljSSc). 86% of patients were Caucasian. Mean age of onset of Raynauds was 9.7 years in the djSSc and 10.7 years in the ljSSc (p=0.8). Mean age of onset of the non-Raynauds was 9.9 years in the djSSc and 11.2 years in the ljSSc (p=0.7). Mean disease duration at time of inclusion was 3.4 years in the djSSc and 2.4 years in the ljSSc There was no significant difference in the ANA, anti-Scl-70 and anticentromere positivity. The mean modified skin score was significantly higher in the djSSc (17.3 compared 7.1, (p=0.3)). They were significantly more teleangiectasia in the djSSc group (39% compared to 19% (p=0.003)). Cardiac involvement was significantly higher in the ljSSc group (19% compared to 3% (p=0.005) There was no significant difference in the proportion of ILD, pulmonary hypertension, gastrointestinal involvement and renal involvement. No renal hypertension was observed. There was significantly more muscle weakness observed in the ljSSc group (38% compared to 17% (p=0.029)). There was no significant difference regarding number of joints with contractions. Physician rated disease activity (40 compared to 29, on 100 mm VAS scale (p=0.013)) and disease damage (37 compared 18, on a 100 mm VAS scale (p< 0.001)) was significantly higher in the djSSc. This significant difference was not found in rating of patients of disease damage and activity.

Conclusion: ljSSc and djSSc seems to be more similar than in adult patient with these subtypes, although physician rating of disease activity and damage found the djSSc more severe.

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