Pädiatrische Rheumatologie

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Patient and physician reported outcomes of juvenile systemic sclerosis patients significantly improve over 12 months observation period in the juvenile systemic scleroderma inception cohort. www.juvenile-scleroderma.com

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Einleitung:

Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1 000 000 children (1). The Juvenile Systemic Scleroderma Inception cohort (jSScC) is the largest cohort of jSSc patients in the world. The jSScC collects longitudinal data prospectively in jSSc, allowing the evaluation of the development of organ involvement and patients and physician reported outcomes in jSSc over time.

Methoden:

The jSScC cohort enrolls jSSc patients who developed the first non-Raynaud´s symptom before the age of 16 years and are under the age of 18 years at the time of inclusion (2, 3). We reviewed jSScC patient clinical data and patient and physician reported outcomes, who had 12 months follow up from the time of inclusion until 1st of December 2021.

Ergebnisse:

We could extract data of 113 patients. The female/male ratio was 3.5:1. Median age of onset of Raynaud´s was 10.1 years and the median age of onset of non-Raynaud´s was 10.8 years. Eighty-eight percent of the patients were treated with disease modifying anti-rheumatic drugs (DMARDs) at time of inclusion in the cohort (T0) and 93% after 12 months (T12). Median disease duration was 2.5 years at T0. Antibody profile stayed unchanged. Only 3 clinical parameters changed and improved significantly, the median modified Rodnan skin score improved from 13 to 8 (p=0.002), the number of patients with swollen joints decreased from 17% to 8% (p=0.043) and number of patients with joints