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Patients with juvenile systemic sclerosis have a distinct pattern of organ involvement. Results from the juvenile systemic sclerosis inception cohort.

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Background

Juvenile systemic sclerosis (jSSc) is a rare disease with a prevalence of around 3 in 1,000,000 children. To better capture the clinical manifestations of jSSc the juvenile systemic sclerosis inception cohort (jSScC) has been prospectively enrolling patients with predetermined clinical variables over the past 12 years. One of the goals is to study the demographic, clinical features, and physician and patient reported outcome differences between those with juvenile limited cutaneous (lc) compared to diffuse cutaneous (dc) disease subtypes, to determine if characteristics are similar or different between dc and lc jSSc.

Objectives

Evaluation of the baseline clinical characteristics of jSSc patients in the jSScC. Compare clinical phenotype between diffuse (dcjSSc) and limited cutaneous (lcjSSc) subtypes.

Methods

Demographic, physical examination, organ system evaluation, autoantibody profile, treatment, and patient and physician reported outcome variables were evaluated from the jSSc Inception cohort and summary statistics applied using chi-square test and Mann Whitney U-test comparing lcjSSc and dcjSSc subtypes.

Results:

At the time of data extraction, 187 jSSc patients were enrolled in the cohort, 80% were Caucasian and 80% female. Diffuse cutaneous jSSc subtype predominated (72%). Median Disease duration was 2.5 years (1 – 4.4). Median age at Raynaud´s was 10.4 years (7.2 – 13.1) and median age of first non-Raynaud´s was 10.9 (7.4 – 13.5). Significant differences were found between dcjSSc versus lcjSSc, regarding several clinical characteristics. Patients with diffuse cutaneous subtype had significantly higher modified Rodnan skin score ($p < 0.001$), presence of sclerodactyly ($p=0.003$), presence of Gottron's papules ($p=0.008$), presence of telangiectasia ($p=0.005$), history of digital tip ulceration ($p=0.001$). Cardiac involvement was significantly higher in limited cutaneous jSSc subtype ($p=0.001$). Diffuse cutaneous jSSc patients had significantly worse scores for Physician Global Assessment of disease activity (35 vs 20; $p < 0.001$) and disease damage (30 vs 15; $p < 0.001$).

Conclusion:

Results from this large international cohort of jSSc patients demonstrate significant differences between dcjSSc and lcjSSc patients. According to the general organ involvement and physician global scores, the dcjSSc patients had significantly more severe disease. These observations strengthen our previous findings of the unique organ pattern of pediatric patients.

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