



Rheumatologie GKJR - Autoimmunität / Kollagenosen

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Male juvenile systemic sclerosis patients have more severe disease: results from the international juvenile scleroderma inception cohort. www.juvenile-scleroderma.com

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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 male and female patients, to determine if characteristics are similar or different.

Objective

To compare organ involvement and disease severity between male and female patients with juvenile onset systemic sclerosis (jSSc).

Methods

Demographics, organ involvement, laboratory evaluation, patient reported outcomes and physician assessment variables were compared between male and female jSSc patients enrolled in the prospective international juvenile systemic sclerosis cohort (jSScC) at their baseline visit.

Results

175 jSSc patients were evaluated, 142 female and 33 male. Race, age of onset, disease duration, and disease subtypes (70% diffuse cutaneous) were similar between males and females. Active digital ulceration, very low body mass index, and tendon friction rubs were significantly more frequent in males. Physician global assessment of disease severity and digital ulcer activity was significantly higher in males. The composite pulmonary involvement was also more frequent in males, though not statistically significantly.

Conclusion

In this cohort, jSSc had a more severe course in males. This reflects the adult-onset SSc cohort data and parallels it in regards to increased digital ulcers, interstitial lung disease, and global severity. Differences from adult findings include no increased signal of pulmonary arterial hypertension or heart failure in male pediatric patients. While monitoring protocols of organ involvement in jSSc need to be identical for males and females, our findings suggest a higher index of suspicion of certain organ involvement in males.

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