Patient Consent
Not applicable (there are no patient data)

Disclosure of Interest
None Declared

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Scleroderma and related syndromes
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Is there a difference in the number of involved organ systems between Juvenile diffuse and limited subtype systemic sclerosis patients?
Ivan Foeldvari [1], Jens Klotsche[2], Kathryn Torok[3], Ozgur Kasapcopur[3], Amra Adrovic[3], Brian Feldman[3], Flavio Sztajnbok[3], Jordi Anton[3], Sindhu Johnson[3], Maria Teresa Terreri[3], Ana Paula Sakamoto[3], Raju Khubchandani[3], Valda Stanevicha[3], Gülcan Özomay Baykal[3], Dieneke Schonenberg-Meinema[3], Eslam Al-Abadi[3], Ekaterina Alexeeva[3], Maria Katsicas [3], Sujata Sawhney[3], Vanessa Smith[3], Simone Appenzeller[3], Tadey Avcin [3], Mikhail Kostik[3], Thomas Lehman[3], Suzanne Li[3], Hana Malcova[3], Edoardo Marrani[3], Clare Pain[3], Anjali Patwardhan[3], W.-Alberto Sifuentes-Giraldo[3], Natalia Vasquez-Canizares[3], Sima Abu Al-Saoud[3], Patricia Costa Reis[3], Stefanie Hajek[3], Mahesh Janarthanan [3], Monika Moll[3], Dana Nemcova [3], Siri Opsahl Hetlevik[3], Maria Jose Santos[3], Cristina Battagliotti[3], Lillemor Berntson[3], Bica, Blanca Bica, Blanca[3], Jürgen Brunner[3], Despina Eleftheriou[3], Liora Harel[3], Gerd Horneff[3], Daniela Kaiser[3], Tilmann Kallinich[3], Dragana Lazarevic[3], Kirsten Minden[2], Susan Nielsen[3], Farzana Nuruzzaman[3], Mihaela Sparchez[3], Yosef Uziel [3], Nicola Helmus[1]
[1]Hamburg Centre for Pediatric and Adolescence Rheumatology, Hamburg; [2]Deutsches Rhemaforschungszentrum, Berlin; [3]jSSc collaborative group, Hamburg, Germany
Correspondence: Ivan Foeldvari
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Introduction: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1 000 000 children. In adult patients diffuse subtype is associated with higher number of organ systems involvement. In a CARRA North American study, it was noted that 38% of jSSc patients had four or more organ systems involved. This topic has not yet been assessed in the currently largest juvenile scleroderma inception cohort (jSSic) cohort.
Objectives: To assess the number of organ systems involved in the diffuse and limited jSSc patients at the time of inclusion in the cohort.
Methods: The jSSic is a prospective cohort including patients, who fulfill the adult SSc criteria[1], with first non-Raynaud symptom before the age of 16 years and under 18 years of age at the time of inclusion. We reviewed the number of organ systems involved at the time of inclusion into the cohort in patients, who were included till 1st of April 2024. The categorization of the organ system involvement was skin, vascular, muscular, articular, pulmonary, cardiac, gastrointestinal, renal and nervous system. We compared the number of involved systems between diffuse and limited jSSc subtype.
Results: Until 1st of April 2024, 268 patients were enrolled and 188 of them had diffuse subtype. The median age at the onset of Raynaud´s was 10.4 years. The median age of the first non-Raynaud organ involvement was 10.9 years. The median disease duration was 2.5 years. 48% in the diffuse subset and 39% of the limited subset had in involvement of 4 or more organ systems involved. There was no significant difference between the cumulative number of organ systems, number of involved organs 1 to 7, between the jSSc subtypes in the Inception cohort.
Conclusion: In this currently largest jSSc cohort in the world, around 43 % the enrolled children have 4 or more organ systems involved, which highlights the overall severity of the disease. There was no significant difference in jSSc children skin subtypes, lcSSc or dcSSc regarding, the cumulative number of organ systems involved, number of involved organs 1 to 7, although as shown in our publications [2] the diffuse subtype presented more severe disease.

Patient Consent
Not applicable (there are no patient data)

Disclosure of Interest
None Declared

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Scleroderma and related syndromes
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Juvenile localized scleroderma: experience of a pediatric rheumatology unit
M Ines Nunes Marques, Inês Madureira, Cristina Henriques, Marta Conde, Margarida P. Ramos
Pediatric Rheumatology Unit , Hospital Dona Estefânia, ULS São José, Lisbon, Portugal
Correspondence: M Ines Nunes Marques
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Introduction: Juvenile localized scleroderma (LS), also called morphea, is a rare disease characterized by inflammation of the skin and subdermal tissues, triggering fibrosis that can lead leading to functional and aesthetic sequelae. The Pediatric Rheumatology European Society classifies LS into five subtypes: linear (most frequent), circumscribed, generalized, pansclerotic, and a mixed subtype which includes a combination of two or more subtypes.
Between 20-70% of LS patients have extracutaneous involvement (musculoskeletal, neurological, ocular and oral). Approximately 10% of these patients have an additional autoimmune (AI) disease.
Objectives: Characterization of demographics, clinical and complementary findings, treatment and outcome in children with LS, followed in a tertiary reference center for pediatric rheumatic diseases.
Methods: A single center retrospective analysis of children diagnosed with LS from 2008 to April 2024.
Results: Of the 18 patients, 16 were female, with a median age at diagnosis of 7 years (5-18y), and a median time to diagnosis of 2.3y (0.15-15y).
The most common form of LS was linear scleroderma (9/18; 6 with head and neck involvement, 2 head and limbs and 1 only the limbs), followed by circumscribed (5/18), mixed (3/18) and generalized morphea (1/18).
Half had family history of AI diseases and 4/18 had previous diagnosis of other AI diseases.
Regarding extracutaneous manifestations: neurological manifestations were present in 4 patients, all with Linear Head Scleroderma (2 preceding the soft-tissue lesions); 4/18 had limited articular mobility; and 2/18 had Raynaud Phenomenon.
At diagnosis, 12/17 had positive ANA (^{3}1:80), 2/17 had positive anti-Scl70 (without evidence of any systemic involvement) and 1/17 had