6	M	6/15	Negative	Uveitis, granulomatous colitis, pneumonitis, splenomegaly	512	MTX
7	F	10/10	Pending	Uveitis, granulomatous gastroenterocolitis and bronchitis	N/A	CS, ADA

Conclusion: Manifestations of sarcoidosis vary significantly across the paediatric age spectrum. While EOS is a known juvenile idiopathic arthritis mimic, nephritis and GIT inflammation may lead to a non-rheumatologic misdiagnosis. Apart from specific infections other childhood diseases may also present with granuloma formation: Crohn's disease, chronic granulomatous disease (and other primary immunodeficiencies), granulomatosis with polyangiitis. Our small series reflects disease heterogeneity and diagnostic difficulties that prolonged the diagnosis by years in 4/7 patients.

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AB0966 PROPOSAL OF OUTCOME MEASURES TO BE USED ON A 12-MONTH OPEN LABEL DRUG TRIAL IN JUVENILE SYSTEMIC SCLEROSIS. RESULTS OF THE 3RD CONSENSUS MEETING IN HAMBURG DECEMBER 2018

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Background: Juvenile systemic sclerosis (jSSc) is an orphan disease, associated with high morbidity and mortality. New treatment strategies are much needed. To develop an open label drug trial for the treatment of jSSc patients, it is necessary to clearly define how to evaluate outcomes in this disease, which are currently not existing. A group of experts in jSSc has met annually and worked to develop an index to evaluate outcomes in this disease.

Objectives: The aim of our third consensus meeting was to establish the domains and the items that should be assessed in a clinical drug trial in jSSc.

Methods: In the consensus meeting 26 jSSc international experts with various specialties participated (22 voted). In a nominal group technic, moderated by DEF, was used to develop the outcome measures. Agreement was defined if 80% or more of the participants approved an item.

Results: Domains and items suggested in the 2017 consensus meeting were reconsidered and selected or rejected during the 2018 meeting, as were additional domains/items (Table).

Domain	Item	voted for - yes/nominator
Global Disease Activity	Physician global of disease activity	22/22
	Change in HAQ/CHAQ- DI	22/22
	Scleroderma HAQ	22/22
Skin	Change in Modified Rodnan Skin Score	22/22
Raynaud Phenomenon	Scleroderma HAQ question regarding Raynaud	22/22
Digital ulcerations	DUCAS Score	22/22
Musculoskeletal System	Juvenile Idiopathic Arthritis Definition of active joint	22/22
Cardiac Involvement	Left ventricular ejection fraction	22/22
	Development of clinically significant arrhythmia as a sign of non-response	22/22
Pulmonary Involvement	FVC	22/22
	age- defined DLCO in all trials	20/22
Renal	New occurrence of renal crisis	22/22
Gastrointestinal Involvement	Body Mass Index	22/22
	Scleroderma HAQ- Gastrointestinal section	22/22
Global health/Health related Quality of Life (QOL)	QOL instrument should be used	18/18

Conclusion: We reached consensus on domains and items which should be assessed in an open label 1 year clinical jSSc trial. We also listed research items which should be assessed but should not currently be included as an outcome in such a trial.

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AB0967 IS THERE A DIFFERENCE IN PRESENTATION OF FEMALE AND MALE PATIENTS WITH JUVENILE SYSTEMIC SCLERODERMA. AN UPDATE FROM THE JUVENILE SYSTEMIC SCLERODERMA INCEPTION COHORT. WWW.JUVENILE-SCLERODERMA.COM

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Background: Juvenile systemic scleroderma (jSSc) is an orphan disease with a prevalence of 3 in 1 000 000 children (1). There are limited data published regarding the differences in clinical presentation of male and female patients with jSSc. The Juvenile Systemic Scleroderma Inception Cohort (JSSIC) is a multinational cohort with a prospective standardized assessment of the patients. The data regarding the difference in clinical characteristics at time of inclusion in the cohort are presented.

Objectives: Evaluation of the differences in clinical presentation of male and female jSSc patients at the time of inclusion in the JSSIC.

Methods: The JSSIC is a prospective multicentre registry of patients with jSSc, who fulfil the adult classification criteria (2), and presented the first non-Raynaud symptoms before 16 years old and were younger than 18 years old at the time of inclusion in the cohort. Patients characteristics at time of inclusion in the cohort were evaluated.

Results: As of 15th of December 2018 120 patients are included in JSSIC. The great majority are female (80%). There were more female patients with CK elevation (29% vs 22%) and more female patients with Gottron papulae (25% vs 12%). The mean modified skin score was higher in males (18.6 vs 13.9).

Sclerodactyly was more frequent in males (90% vs 76%). Active ulceration was present in 33% of males compared to 14% of females (p=0.026). FVC<80% occurred more often in males with 47% compared with 24% in females (p=0.018). Pulmonary hypertension was more