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

common in females with 7% compared to 4% in males. Urine sediment changes were more common in males (8% vs 4%). Gastrointestinal involvement was more common in females (37% vs 29%). Contractures occurred more often in males with 62% compared with 46% in females. Tendon friction rub was observed in 21% of males and 3% of females ($p=0.001$). Physician global scores of disease activity and damage were higher in males with 48 for both assessments compared to 36 and 30 in females.

Conclusion: Male patients with jSSc have a higher severity of disease, as it has been reported in adults.

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AB0968

INTEGRATED RHEUMATOLOGY AND OPHTHALMOLOGY CARE IN PEDIATRIC AUTOIMMUNE UVEITIS – OUTCOME AND EFFECTIVENESS

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Background: Uveitis is an ocular inflammatory disease that may lead to irreversible blindness if not treated early and properly. May be induced by infectious and non-infectious diseases (autoimmune diseases, especially juvenile idiopathic arthritis), or “masquerade” causes. Some uveitis are termed “idiopathic” although autoinflammatory evidence. Treatment effectiveness depends on the accuracy of the ophthalmologic evaluation and the appropriate management, especially when immunosuppressive medications are necessary. (1, 2)

Objectives: To describe the first year experience of the Integrated Ophthalmology and Pediatric Rheumatology Outpatient Clinic in the management of pediatric patients with autoimmune uveitis.

Methods: Retrospective chart review study of twenty-four patients followed up from June 2017 to October 2018. The patients clinical evolution at 0, 3, 6, 9 and 12 months was analyzed.

Results: In this group, 62% were female, and mean age at first appointment and at symptoms onset were 11.7 (2.9 to 17.5) and 7.3 (1.1 to 12.6) years, respectively. Anterior uveitis was present in 70.8% of the cases, and in 65.6% it was bilateral; cataract or glaucoma was observed in 20.5%. The identified diagnosis during follow-up were: idiopathic autoimmune uveitis in 45.8%, juvenile idiopathic arthritis in 41.6% (20.8% oligoarticular, 16.6% polyarticular, 4.1% associated with enthesitis) and juvenile systemic lupus erythematosus, Kawasaki Disease and Vogt-Koyanagi-Harada syndrome one case of each. Antinuclear antibodies were present in 41% and inflammatory markers in 28.7% patients. At first appointment, 20.5% presented ocular inflammatory activity and 53.3% had sequelae when examined in slit lamp or indirect ophthalmoscopy. Medications used were: topical corticosteroids (61.6%), topical mydriatic (49.2%), oral corticosteroid (41%), methotrexate (41%), etanercept (12.3%), adalimumab (28.7%) and periocular injections of triamcinolone, cyclosporin, tocilizumab, azathioprine (4.1% each). The comparison of the data at inclusion and during follow-up is reported in Table:

	At inclusion	3 months	6 months	9 months	12 months
Ocular inflammation(%)	20.5	24.6	12.3	12.3	12.3
Topical Corticosteroid(%)	61.6	32.8	16.4	8.2	20.5
Topical Mydriatic (%)	41	24.6	28.7	20.5	20.5
Oral Corticosteroid (%)	73.8	20.5	12.3	12.3	20.5
Methotrexate (%)	12.3	53.3	49.2	36.9	28.7
Etanercept (%)	12.3	4.1	4.1	4.1	4.1
Adalimumab (%)	20.5	20.5	16.4	16.4	16.4
Others (%)	12.3	4.1	4.1	4.1	4.1
No medication (%)	4.1	8.2	8.2	8.2	4.1

At the end of 12 months, only 12.3% of the patients still had active uveitis. We have also observed a reducing in 66.6% of topical corticosteroids and 72.2% of oral corticosteroids use during the treatment of uveitis over 12 months

Conclusion: Considering that before the inclusion, patients were followed independently in each clinic, there was optimization of the treatment and reduction in number of medical appointments after the creation of the integrated outpatient clinic. The interprofessional management teams should be the standard of care for pediatric patients with autoimmune uveitis.

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AB0969

COMPARISON AMONG ACR1997, SLICC AND THE NEW EULAR/ACR CLASSIFICATION CRITERIA IN CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: To date there are no specific classification criteria for childhood-onset systemic lupus erythematosus (cSLE).

Objectives: This study aims to compare the performance among the American College of Rheumatology (ACR) 1997, the Systemic Lupus International Collaborating Clinics criteria (SLICC) and the new European League Against Rheumatism (EULAR)/ACR criteria, in a cSLE cohort.

Methods: Data were retrospectively collected from medical charts of cSLE cases and controls with defined rheumatic diseases, both ANA positive, to establish each ACR1997, SLICC and EULAR/ACR criterion fulfilled, at first visit and 1-year-follow-up.

Results: 122 cSLE cases and 89 controls were included. At first visit, SLICC criteria had greater sensitivity than ACR 1997 (89.3% versus 70.5%, $p < 0.001$), but similar specificity (80.9% versus 83.2%, $p = 0.791$), however performance was not statistically different at 1-year-follow-up. SLICC performed better in specificity compared to EULAR/ACR score ≥ 10 at first visit (80.9% versus 67.4%, $p = 0.008$) and 1-year