known about the association between those diseases and rheumatic disorders during childhood, such as juvenile idiopathic arthritis (JIA).

Objectives: Our objective was to evaluate the correlation between the thyroid function and the clinical presentation in children with juvenile idiopathic arthritis.

Methods: Ninety-seven children (56 female, 41 male, median age 9.57 years, range 1.1–17.5 years) with juvenile idiopathic arthritis were evaluated. All patients underwent thyroid function tests (TSH, free T₄ and free T₃), antithyroglobulin (TgA) and antiperoxidase (TPOA) antibodies. All patients had a thyroid high-resolution sonography examination. Patients with nodules on US examination, low-TSH level and/or high thyroid hormone levels or positive TPOA and/or TgA values were referred to thyroid scintigraphy. The gained data was correlated with disease activity indices. Thyroid autoimmunity was evaluated by fluorescence enzymatic immunoassays of TgA and TPOA antibodies, considering TgA >50 IU/ml and TPOA >100 IU/ml as positive values.

Results: Fifty-one of the cases were classified as oligoarticular, thirty-one as seronegative polyarticular, two as seropositive polyarticular, eleven as systemic, and two as enthesitis related- juvenile idiopathic arthritis.

No cases of overt clinical and biochemical hypothyroidism were found among children with juvenile idiopathic arthritis. On opposite, mean thyroid free T₃ levels were higher in 36.4% of cases and free T₄ in 29.7% cases. Anti-TPO antibodies were found in 5 of 97 patients (5.15%) and anti-TG antibodies were found in 2 patients (2.06%). All patients with thyroid antibodies positives were females. The ultrasound examination of thyroidal gland revealed abnormalities in 41% cases, most of them cystic changes (33.7%) and hypo-echogenicity (28.57%). In those patients with thyroid nodules on US examination, the thyroid scintigraphy did not revealed pathological activity or other processes. Although, 2 patients presented mean thyroid volume above 2SDS according their age reference values. An increased vascular flow pattern on Doppler examination of thyroidal gland was found in 26% cases. Correlation and regression analysis showed low age at diagnosis and longer duration of the disease to be predictors for those thyroid disorders.

Conclusion: These data seem to suggest careful monitoring of thyroid function and thyroid autoantibodies just in children with juvenile idiopathic arthritis presenting clinical symptoms of thyroid involvement.

REFERENCES

- [1] Fröhlich E., Wahl R., Thyroid Autoimmunity: Role of anti-thyroid antibodies in thyroid and extra-thyroidal diseases, *Front. Immunol.* 8:521. doi: 10.3389/fimmu.2017.00521
- [2] Stagi S. et al., Thyroid function, autoimmune thyroiditis and coeliac disease in juvenile idiopathic arthritis, *Rheumatology* 2005;44:517–520, doi:10.1093/rheumatology/keh531

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7974

AB0964 DIAGNOSIS OF CROHN'S DISEASE IN PATIENTS WITH PRIMARY RHEUMATOLOGICAL MANIFESTATIONS

Evgeny Fedorov¹, Svetlana Salugina¹, Natalia Schigoleva², Maria Kaleda¹, Irina Nikishina¹, Elena Komelyagina¹, Olga Borodacheva¹, Anastasia Schigoleva³.
¹V.A.Nasonova Research Institute of Rheumatology, Moscow, Russian Federation;
²National Pediatric Clinical Hospital, Moscow, Russian Federation; ³Pirogov Russian National Research Medical University, Moscow, Russian Federation

Background: Sometimes Crohn's disease (CD) manifests initially with rheumatological symptoms. Therefore, there's an important for a rheumatologist to recognize this pathology correctly and timely.

Objectives: To present cases of CD, referred to V.A. Nasonova Research Institute of Rheumatology.

Methods: Case series of 7 pts (5 boys, 2 girls) with initial rheumatological condition who developed CD. All pts were subjected to standard rheumatological examination. CD diagnosis was suggested by a rheumatologist and confirmed in gastroenterology hospital.

Results: Mean age at CD onset was 12.7±3.98 yrs, varying from 5 to 17 yrs. In 4 pts gastrointestinal (GIT) symptoms manifested simultaneously with rheumatological, in 2 pts GIT symptoms were delayed for 4 and 5 years. The time-interval from the onset of first symptoms until establishing CD diagnosis varied from 2 months till 6 years, averaging to 96±38 months. The list of initial rheumatological diagnoses included the following entities: juvenile idiopathic arthritis (JIA) with oligoarticular onset – in 5 pts, and systemic onset JIA – in 2 pts. In all cases arthritis manifested initially as an oligo-involvement of a lower limb joints. High active sacroiliitis with deep bone oedema was detected by MRI in 5 pts. Fever

was present in 5 pts, uveitis – in 1, cutaneous psoriasis – in 1. Other documented symptoms included: weight loss (3), hepatosplenomegaly (1), lymphadenopathy (1), aphthous stomatitis (1), nodular erythema (1), erythematous rash (1). HLA B27-antigen positivity was established in 3 pts out of 5 examined. All pts had significant increase in CRP and ESR levels and WBC counts; (additionally 3 examined for calprotectin cases demonstrated significant increase of its' concentration (>1000 µg/g).

Conclusion: IBD, including CD, should not be missed and ruled out in all pts with oligoarticular and systemic JIA onset. Fecal calprotectin should be checked in suspected CD cases among individuals with rheumatologic symptoms. Special attention should be given to cases manifesting during puberty, accompanied by weight loss, fever, GIT symptoms, increased levels of acute phase inflammatory markers. MRI stir lesion may be as suspected of IBD especial CD.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.4689

AB0965 PAEDIATRIC SARCOIDOSIS: A PROFESSIONAL IMITATOR

Sarka Fingerhutová¹, Hana Malcova², Dana Nemcova¹, Radoslav Srp¹, Tomas Piskovsky^{1,3}, Pavla Doležalová¹. ¹1st Faculty of Medicine, Charles University and General University Hospital, Paediatric Rheumatology Unit, Department of Paediatrics and Adolescent Medicine, Prague, Czech Republic; ²Motol University Hospital, Paediatric Rheumatology Unit, Prague, Czech Republic; ³Faculty Hospital in Ostrava, Ostrava, Czech Republic

Background: Sterile granulomatous inflammation is a hallmark of disorders covered by the term „paediatric sarcoidosis“, a rare disease classified among autoinflammatory conditions. While the classical triad of arthritis, uveitis and dermatitis associated with the NOD2 gene mutation is typical for Early-onset sarcoidosis (EOS), mutation-negative disease has extremely variable phenotype in children and can mimic various other conditions.

Objectives: To illustrate variability of clinical presentation of sarcoidosis in a cohort of children from 2 collaborating paediatric rheumatology units

Methods: Retrospective case analysis.

Results: Seven Caucasian patients (2 boys) followed for a median of 1 (0.2-12) year were identified (Table). Patients 1 and 3 presented with typical rash (with biopsy-proven granuloma), polyarthritis and severe, treatment-resistant uveitis which has never been fully controlled. Patient 2 presented with recurrent self-limited febrile episodes and was investigated for periodic fever syndrome. Within 3 months she developed erythema nodosum-like rash, granulomatous uveitis and arthritis. In 4 patients with disease onset after 10 years of age two major organ systems were affected: renal and gastrointestinal (GIT). In patients 4 and 5 renal biopsy performed for insufficiency showed tubulointerstitial nephritis with granulomas. Patients 6 and 7 had been initially treated for “atypical inflammatory bowel disease” with granulomas on biopsy. In both of them finding of granulomatous uveitis eventually lead to the diagnosis of sarcoidosis. Additional symptoms are listed in the Table. Presence of granulomatous inflammation was confirmed in all patients. Chronic uveitis was present in 6/7 patients and was an important diagnostic clue in EOS as well as adult-type of sarcoidosis while respiratory tract was affected subclinically in 2 older patients only. Apart from combinations of clinical symptoms elevation of chitotriosidase significantly aided diagnostic process and when available, it also reflected disease activity. (1)

Pt – patient, F – female, M – male, Dx – diagnosis, CS – corticosteroids, MTX – Methotrexate, ADA – Adalimumab, MMF – Mycophenolate mophetil

Pt	Sex	Age (yrs) at Onset/ Dx	NOD2 gene mutation	Manifestations	Chitotriosidase N 4.4-89 nmol/ml/h	Therapy
1	F	0,25/4	R334Q	Rash, fever, uveitis, arthritis, meningitis	299	MTX, MMF, ADA, MTX
2	F	0,6/1	Negative	Rash, fever, uveitis, arthritis, panniculitis	433	ADA, CS, MTX
3	F	1/3	Variants S178S, V955I, N289S	Rash, fever, panuveitis, arthritis, liver granuloma	N/A	CS, MTX, ADA, Infliximab
4	M	4/15	Pending	Fever, arthralgia, lymphadenopathy, granulomatous nephritis	N/A	CS, MMF
5	F	13/13	Negative	Uveitis, granulomatous nephritis, hearing loss	604	CS, MTX, ADA

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.

REFERENCES

- [1] Popevic S. et al. Verifying Sarcoidosis Activity: Chitotriosidase versus ACE in Sarcoidosis – A Case-control Study. *J Med Biochem.* 2016 Nov 2;35 (4):390-400

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7849

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

Ivan Foeldvari¹, Kathryn Torok², Lusine Ambartsumyan², Jordi Anton², Christian Beyer², Michael Blakley², Tamas Constantin², Patricia Costa Reis², Megan Curran², Maurizio Cutolo², Francesco Del Galdo², Christopher Denton², Kim Fligelstone², Bernd Hinrichs², Antonia Höger², Francesca Ingegnoli², Ozgur Kasapcopur², Li Suzanne², Dana Nemcova², Catherine Orteu², Clarissa Pilkington², Vanessa Smith², Anne Stevens², Brandi Stevens², Allison Zheng², Dinesh Khanna², Daniel Furst². ¹Hamburger Zentrum für Kinder- und Jugendrheumatologie, Hamburg, Germany; ²Participant of the Hamburg Outcome measure consensus process, Hamburg, Germany

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.

Disclosure of Interests: Ivan Foeldvari Consultant for: Chugai, Novartis, Kathryn Torok: None declared, Lusine Ambartsumyan: None declared, Jordi Anton Grant/research support from: Grant/research support, consultant or speakers bureau from AbbVie, Alexion, Bristol-Myers Squibb, ChemoCentryx, Gebro, GlaxoSmithKline, Novartis, Novimmune, Pfizer, Roche, Sanofi and Sobi, Consultant for: Grant/research support, consultant or speakers bureau from AbbVie, Alexion, Bristol-Myers Squibb, ChemoCentryx, Gebro, GlaxoSmithKline, Novartis, Novimmune, Pfizer, Roche, Sanofi and Sobi, Speakers bureau: Grant/research support, consultant or speakers bureau from AbbVie, Alexion, Bristol-Myers Squibb, ChemoCentryx, Gebro, GlaxoSmithKline, Novartis, Novimmune, Pfizer, Roche, Sanofi and Sobi, Christian Beyer: None declared, Michael Blakley: None declared, Tamas Constantin: None declared, Patricia Costa Reis: None declared, Megan Curran: None declared, Maurizio Cutolo: None declared, Francesco Del Galdo: None declared, Christopher Denton Grant/research support from: GlaxoSmithKline, Inventiva, CSF Behring, Consultant for: Roche-Genentech, Actelion, GlaxoSmithKline, Sanofi Aventis, Inventiva, CSL Behring, Boehringer Ingelheim, Bayer, Kim Fligelstone: None declared, Bernd Hinrichs: None declared, Antonia Höger: None declared, Francesca Ingegnoli: None declared, Ozgur Kasapcopur: None declared, Suzanne Li: None declared, Dana Nemcova: None declared, Catherine Orteu: None declared, Clarissa Pilkington: None declared, Vanessa Smith: None declared, Anne Stevens: None declared, Brandi Stevens: None declared, Allison Zheng: None declared, Dinesh Khanna Shareholder of: Eicos Sciences, Inc, Grant/research support from: Bayer, BMS, Pfizer, Horizon, Consultant for: Actelion Acceleron, Arena, Bayer, BI, BMS, CSL Behring, Corbus, Cytari, GSK, Genentech/Roche, Galapagos, Employee of: Elcos Sciences, Inc, Daniel Furst Grant/research support from: F. Hoffmann-La Roche, Genentech

DOI: 10.1136/annrheumdis-2019-eular.3198

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

Ivan Foeldvari¹, Jens Klotsche², Ozgur Kasapcopur³, Amra Adrovic³, Kathryn Torok³, Valda Stanevich³, Flávio R. Sztajnbock³, Ana Paula Sakamoto³, Ekaterina Alexeeva³, Jordi Anton³, Maria Katsikas³, Vanessa Smith³, Tadej Avčin³, Rolando Cimaz³, Mikhail Kostik³, Thomas Lehman³, Walter Alberto Sifuentes-Giraldo³, Simone Appenzeller³, Mahesh Janarthanan³, Monika Moll³, Dana Nemcova³, Maria Jose Santos³, Amara Nassar³, Cristina Battagliotti³, Lillemor Berntson³, Blanca Bica³, Juergen Brunner³, Patricia Costa Reis³, Despina Eleftheriou³, Liora Harel³, Gerd Horneft³, Tilmann Kallinich³, Dragana Lazarevic³, Kirsten Minden³, Susan Nielsen³, Farzana Nuruzzaman³, Anjali Patwardhan³, Yosef Uziel³, Nicola Helms³. ¹Hamburger Zentrum für Kinder- und Jugendrheumatologie, Hamburg, Germany; ²Deutsches Rheuma-Forschungszentrum (DRFZ), Berlin, Germany; ³JSSC Collaborative Group, Hamburg, Germany

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