AB0954 RELATIONSHIP BETWEEN SERUM COMPLEMENT LEVELS AND RENAL PATHOLOGICAL CLASSIFICATION IN CHILDREN WITH SILENT LUPUS NEPHRITIS

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Background: Silent lupus nephritis is defined by some pathological evidence of lupus nephritis with normal urinary findings. It is therefore apparent only upon renal biopsy. Serum complement levels have been associated with overall lupus disease activity. Despite recent progress in understanding the pathogenesis of lupus nephritis, including complement, our understanding of silent lupus nephritis, especially in children, remains limited.
Objectives: To search whether serum complement levels are associated with renal pathological classification in children with silent lupus nephritis.
Methods: We determined serum C3 and C4 levels and International Society of Nephrology/Renal Pathology Society classification in 25 patients with paediatric silent lupus nephritis before initial therapy who were admitted to our hospital. Patients were classified as having silent lupus nephritis based on normal urinary findings at baseline renal biopsy in juvenile systemic lupus erythematosus.
Results: Serum C3 levels varied between International Society of Nephrology/Renal Pathology Society classes, with significantly lower levels for silent lupus nephritis patients in class III compared to class II, and for class II compared to class I. There was no significant difference in serum C4 levels between International Society of Nephrology/Renal Pathology Society classes in patients with silent lupus nephritis.
Conclusions: Our results suggest that serum C3 levels were associated with renal pathological classification in children with silent lupus nephritis. We propose that serum C3 levels would provide a useful tool for predicting latent severe nephritis in patients with juvenile systemic lupus erythematosus who have normal urinary findings before initial therapy.
Disclosure of Interest: None declared
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AB0955 NEW ONSET OF UVEITIS, PSORIASIS OR IBD AS PARADOXICAL EFFECTS OF BIOLOGICS IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS: SINGLE CENTER EXPERIENCE

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Background: Biological agents (BA) are high efficacy options for current therapy for patients (pts) with juvenile idiopathic arthritis (JIA). They are successfully used not only for the arthritis but also for uveitis, psoriasis and inflammatory bowel disease (IBD). However, paradoxical induction of these conditions in pts treated with BA is a well-established phenomenon.
Objectives: To evaluate the frequency of new onset of uveitis, psoriasis or IBD occurring under BA therapy in JIA pts, to establish clinical features, which may be associated with development of such effects.
Methods: Retrospective cohort study involved all JIA pts (740) who were treated with BA in our clinic from 2004 to 2016. All cases of new onset (*no*)-uveitis/psoriasis/IBD collected; clinical features of disease onset and course, activity level, JIA category, exposure to Methotrexate (MTX) and BA, presence of ANA, HLA B27 were studied.
Results: We identified 20 (2.7%) pts (11 female/9 male) with *no*-uveitis under BA, mostly during etanercept (ETA) therapy (18 cases from 285 ETA courses, 6.3%), 1/112 - in abatacept (ABA) and 1/233 - in adalimumab (ADA); 2.46 events/100 patient-year (PY) vs 0.31 in ABA, and 0.15 in ADA), ETA exposure was 14.9±9.9 months (mo). JIA subtypes were as follows: RF-neg polyarthritis 5 (25%), persistent oligoarthritis 3 (15%), extended oligoarthritis 10 (50%) ERA - 2 (10%). JIA started in this group at the age of 4.5±3.9 yrs.18/20 patients had high laboratory activity (CRP 55±25 mg/l) and severe arthritis before BA initiation. Most of pts (16/20) achieved 90–100% ACRpedi-response by the uveitis development. 1 pt was treated by ADA for the 71 mo *and* switched to ABA. in 1 pt *no*-uveitis was obseved under ABA. 12/20 pts were ANA-positive, 10/20 pts had HLAB27, 1 pt did not have ANA or HLAB27. Uveitis was occurred earlier in ANA plus B27 positive pts (mean exposure - 10.7 mo vs 27.4 in ANA+ and 21.6 in B27+ pts). 17/20 (85%) of pts received MTX. In all cases of *no*-uveitis BA was switched. 5 pts from 740 (0.7%) developed *no*-psoriasis under BA: INF - 2 cases (0.62/100PY), ADA - 2 (0.15/100PY), ABA -1 (0.31/100PY). JIA subtypes were presented by RF-negative polyarthritis (1), extended oligoarthritis – 3. All received MTX. 1/5 pts was ANA+, 2/5 - HLAB27+. Average age of disease onset was 9.8±7.8 years; BA exposure before psoriasis was 25±11.5 mo. Therapy was continued in 3/5 pts; switched from INF to ADA in 2. Only 1 case of *no*-IBD was occurred in female patient fulfilled to systemic-onset JIA criteria years. She was treated by INF for the 62 months and switched to ADA due to secondary inefficiency presented by active polyarthritis and sacroiliitis (HLAB27neg), but 2 months later severe gastrointestinal symptoms appeared and diagnosis of Crohn's disease was established by endoscopy.
Conclusions: Our study suggested that new onset of uveitis, psoriasis and IBD is rare adverse event during BA therapy in JiA. It seemes to be as delayed implication of disease natura. but not therapy complication. Uveitis observed mostly in pts receiving ETA, unlike the psoriasis mainly developed during TNF-monoclonal antibodies using. High activity aggressive manifestations at disease onset and good initial response to BA are typical features for all pts, who developed paradoxical effects under BA therapy.
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AB0956 INFLUENCE OF BIOLOGICAL THERAPY ON BONE MINERAL DENSITY IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile idiopathic arthritis (JIA) is one of the most common and most disabling rheumatic diseases in children. JIA is a systemic chronic inflammatory immunopathological disease, which leads to dysfunction of the joints, their deformation and limitations of life of the child, violates the child's growth and development. One of the complications of systemic JIA in children is osteoporosis. In the pathogenesis of osteoporosis important place is given to the use of glucocorticoids. In world Rheumatology successfully used drugs aimed at the major pro-inflammatory cytokines such as tumor necrosis factor (TNF), interleukin-1 (IL-1), interleukin-6 (IL-6), and others.
Objectives: The purpose of research - to determine the influence of biological therapy on bone mineral density in children with JIA.
Methods: 15 children with systemic JIA (mean age 11,9±3,4 years) were examined in the rheumatological department of 4th city clinical hospital of Minsk. Bone mineral density was assessed by 2-energy X-ray absorptiometry (DEXA) at two points of the skeleton. The level of mineralization of skeletal mineral content was estimated in the bone tissue (BMC), bone mineral density (BMD) and the parameter Z-score. Z-score was used to determine the incidence of osteopenia and osteoporosis in children surveyed. In accordance with the WHO criteria for normal bone mineral density was diagnosed with the Z-score> -1 SD, osteopenia - at Z-score <-1 SD, but> - 2,5 SD, osteoporosis - with Z-score <-2,5 SD. Statistical data processing carried out using an integrated application package "Statistics 6 for Windows".
Results: In a study of children osteopenia was diagnosed in 9 patients with systemic JIA (mean Z-score -2,3 SD) Osteoporosis was diagnosed in 6 patients with systemic JIA (mean Z-score -2,7 SD). All children received a mean dose of methotrexate 12.7 mg/m^2 of body surface area, an average dose of methylprednisolone 0.34 mg/kg body weight per day and treatment of IL-6 inhibitor - tocilizumab 8 mg/kg body weight every 2–4 weeks. Bone mineral density was measured prior to initiating therapy tocilizumab and after 2 years of therapy.
During tocilizumab therapy achieved remission of the disease, all children was canceled methylprednisolone. After re densitometry of 2 years after the beginning of therapy improvement noted tocilizumab Z-score in children with osteopenia with Z-score -2.3 SD to Z-score -1.4 SD, in children with osteoporosis a Z-score -2 7 SD to Z-score - 2,1 SD.
Conclusions: The results indicate a positive influence tocilizumab therapy to bone mineral density in children with JIA.
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AB0957 IS THERE A DIFFERENCE IN THE CLINICAL PRESENTATION OF JUVENILE SYSTEMIC SCLERODERMA PATIENTS ACCORDING THE AGE OF ONSET: RESULTS FROM THE JUVENILE SCLERODERMA INCEPTION COHORT WWW.JUVENILE-SCLERODERMA.COM

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Background: Juvenile systemic sclerosis (jSSc) is an orphan autoimmune disease. It was rarely looked at the differences between the clinical presentations of patients at different paediatric age groups. The juvenile scleroderma inception cohort (www.juvenile-scleroderma.com) is a prospective standardized register for patients with jSSc.
Objectives: Comparison of clinical characteristics of patients with different age range at the time of inclusion in the registry.
Methods: Patients with jSSc were included worldwide to the juvenile scleroderma inception cohort. We compared the demographics and clinical characteristics of

the patients at different age ranges. We created 3 cohorts with different age ranges at onset of disease. Patents aged less than 5 years (Group1), 5–10 years (Group2) and over 10 years (Group3) at the time of diagnosis of the first non-Raynaud involvement of jSSc.
Results: Up till now 88 patients were enrolled,14 patients (15%) in Group1, 22 (25%) in Group2 and 52 (59%) in Group3. Diffuse subtype occurred in 71% in Group1, in 82% in Group2 and in 65% in Group3. Most patients were Caucasian. Disease duration at time of inclusion into the cohort was 3.9 years in Group1, 4.9 years in Group2 and 2.2 years in Group3. ANA positivity was 57% in Group1, 77% in Group2 and 86% in Group3. Anti-scl 70 was around 30% in all groups. Anti-Centromere positivity was 7 to 10%. Mean modified skin score was 12.4 in Group1, 16.5 in Group2 and 15.9 in Group3. Raynaud Phenomenon occurred in 85 to 95% of the patients. History of active or inactive ulceration occurred in 57% in Group1, 62% in Group2 and 43% in Group3. Decreased FVC under 80% occurred in 43% in Group1, 32% in Group2 and 30% in Group3. Pulmonary hypertension occurred in 7% in Group1 and in 10% in Group3. No renal hypertension was observed. Urinary sedimentary changes occurred in 7% in Group1 and in 10% in Group3. Gastrointestinal involvement occurred in 21% in Group1, 45% in Group2 and 27% in Group3. Musculoskeletal involvement occurred in 58 to 64%. Patient global disease activity (VAS 0–100) was 42.8 to 47.9. Patient global disease damage (VAS 0–100) was 39.6- to 45.0. Physician global disease activity (VAS 0–100) was 35.4–40.0. Physician global disease damage (VAS 0–100) was 37.1 in Group1, 41.3 in Group2 and 27.7 in Group3.
Conclusions: It seems to be that patients, with onset of the disease in younger age have more severe disease as patients with disease onset after the age of 10 years. We need more patients in our cohort to gain more sufficient data to prove our preliminary observation.
Disclosure of Interest: None declared
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AB0958 PROPOSAL FOR A JUVENILE SYSTEMIC SCLEROSIS RESPONSE INDEX (JSSCRI): RESULT OF THE CONSENSUS MEETING IN HAMBURG. GERMANY 11TH OF DECEMBER 2016

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Background: Juvenile Systemic Sclerosis (jSSc) is an orphan disease. There is increasing interest in testing novel therapies in the management of fibrotic diseases. Therefore, it is very important to develop a Response Index for jSSc (JSScRI) to distinguish effective therapies from placebo. In 2014 at the 1st JSScRI Consensus Meeting in Hamburg, following two rounds of a Delphi process, a proposal for domains and items for a JSScRI were made. In 2016, the 2nd JSScRI Consensus Meeting was held in Hamburg, Germany.
Objectives: To conduct a Nominal Group Technique (NGT) and select potential core data set items that could change as outcome measures) that will be incorporated in the development of a JSScRI.
Methods: Before the 2nd JSScRI Consensus Meeting, the items from the 1st JSScRI Consensus Meeting (2014) were scored via Email, in a Delphi by the participants of the current meeting. Participants included 14 experts in adult and juvenile SSc and a patient partner. During the subsequent face to face NGT meeting, moderated by DEF, and the items were scored anonymously by the participants after a nominal group discussion. The domains and items were scored regarding their importance for 1 year clinical trial from 1 (not relevant at all) to 9 (most relevant). A priori, it was agreed by the participants that the goal of the NGT was to exclude items that: 1. Are not feasible and 2. do not represent a changeable outcome measure even though they may represent the impact of disease on quality of life, vocational or recreational activity. Items with a median score of <4 in which greater than 1/3 of participants scored 1–3 [despite the item having a median score >3] were excluded.

Table 1. Assessment of the Activity of the Musculoskeletal domain

	Whole Group	1–3	4–6	7–9
1) Swollen joints	7	0	1	12
2) Limited range	7.5	1	1	10
3a) MMT	8	0	1	11
3b) CMAS	7	4	1	7
4) Presence of tendon friction rub	7	1	2	10
5a) CK	8	1	1	11
5b) Aldolase	7	3	3	7

Results: Seventy-one items in 13 domains were scored. Six items were not scored as they were felt to not represent an outcome measure or were non-feasible and six items received a median score less then 4.
Table 1 provides an example of the musculoskeletal domain with the median scores for different outcome measures.
Conclusions: In a rigorous, NGT consensus meeting, some item reduction for the JSScRI was achieved. Items will be tested in a prospective way in the patients of the inception cohort of juvenile SSc (www.juvenile-scleroderma.com).
Disclosure of Interest: None declared
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AB0959 EVALUATE THE CARDIOVASCULAR RISK THROUGH CAROTID INTIMA-MEDIA THICKNESS IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS IN THE YOUNG ADULT AGE

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Background: The relationship between inflammation and atherosclerosis has been demonstrated, so it is important to identify early markers of the disease. Ultrasound carotid intima-media thickness (CIMT) measurement is a non-invasive, consistent, validated technique used as a marker to identify subclinical arteriosclerotic disease.
The long-term risk of cardiovascular disease in young adult patients with juvenile idiopathic arthritis (JIA) is unclear and there are no risk management guidelines for these patients.
Objectives: To assess whether there is an increase in CIMT in the young adult with a history of JIA and to relate CIMT with classic cardiovascular risk factors in these patients.
Methods: Observational and cross-sectional study. Follow-up patients from transitional care between 18 and 36 years old, with JIA diagnosis by ILAR classification. Filiations data, anthropometric variables and activity disease scores were collected.
We performed, prior informed consent, CIMT measurement by radiofrequency with Esaote MyLab 70XVG. Three measurements were performed on each carotid artery, according to the protocol of the American Society of Echocardiography.
Results: Of the 20 patients, 17 (85%) women and 3 (15%) men. Subtype distribution was 8 (40%) oligoarticular, 1 of them ANA negative; 8 (40%) polyarticular being 4 seropositive and 4 seronegative; 1 systemic (5%); 2 (10%) psoriatic arthropathy and 1 (5%) HLA B27 positive arthritis. 66.7% are with disease modifying drugs (26.7% synthetic and 40% biological), while 33.3% do not have specific treatment.
The main variables studied are described in the attached table (Table 1).
In two previous independent studies conducted by Falaschi and Huang, CIMT was found to be 0.54±0.03 mm and 0.54±0.06 mm in 26 and 38 patients, respectively. From these results a mean and standard deviation of 0.54±0.05 mm were obtained. Using this last result as a control group, we compared it with the results of our series (0.466±0.068), with a significant difference ($p<0.001$).
In the statistical analysis, using the Rho Spearman, a significant correlation of the CIMT with the time of evolution of the disease in years ($r=0.579$) and systolic blood pressure ($r=0.621$) was observed at the level of 0.01 and with C Reactive Proteine ($r=0.524$) and BMI ($r=0.471$) at the 0.05 level.

	N	Minimum	Maximum	Media	Typical deviation
Age	20	18	36	24,75	5,350
Diagnosis age	20	1	16	9,70	5,048
Evolution time (years)	20	5	27	15,00	6,720
BMI (Kg/cm^2)	19	15,5	38,5	24,505	6,4347
Abdominal perimeter	20	58	126	81,33	19,392
Waist-hip index	20	44	135	90,40	26,009
Systolic blood pressure	20	96	130	114,05	7,258
Diastolic blood pressure	20	60	99	73,80	12,271
Left CIMT	20	394	634	464,15	54,788
Right CIMT	20	337	600	467,45	80,275
HAQ	17	,00	2,25	,3529	,59986
VAS pain (mm)	18	0	8	3,17	2,618
CRP (mg/L)	17	,3	57,7	9,419	14,6040
DAS28	16	,97	3,97	1,8856	,88204

Conclusions: The carotid intima-media thickness of patients with JIA were lower than the controls previously described in the literature, so we will complement this study with our population controls.
In addition to classic cardiovascular risk factors such as systolic blood pressure and BMI, there is correlation with the evolution time in years of the disease and CRP, so that in transitional care programs we must the activity of the disease and insist on identification, control and follow-up of the classics factors associated with cardiovascular risk.
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