Objectives: To assess AIDAI score and evaluate correlation between AIDAI and disease/response characteristics over 16 weeks (wks) of CAN treatment in CLUSTER.

Methods: CLUSTER study design and results have been presented.² AIDAI was calculated as the sum of 12 items¹ for 30 consecutive days. AIDAI score was calculated if the first score was recorded ≥ 29 days before baseline (BL). Missing items were imputed beyond last evaluable measurement by LOCF. Proportion of pts with inactive disease (ID; AIDAI score <9) was calculated at Wk 16. Correlation analysis of AIDAI with C-reactive protein (CRP), serum amyloid A (SAA), physician global assessment (PGA) and sheehan disability score (SDS), and child health questionnaire–psychological/physical (CHQ–PSC/PCS) and short form 12–physical/mental (SF12–PCS/MCS) component summaries were performed at BL and Wk 16, with significance set at $p < 0.05$.

Results: Median AIDAI scores decreased over time (Fig 1). Proportion of pts with ID at Wk 16 was 52% in crFMF, 40% in HIDS/MKD and 46% in TRAPS cohorts. AIDAI at Wk 16 correlated significantly with: SDS in all 3 cohorts; PGA in HIDS/MKD and TRAPS; SF12–MCS in crFMF and HIDS/MKD (Table 1). CRP and SAA did not correlate with AIDAI.

Table 1. Correlation between AIDAI and disease activity/response variables at Week 16

	Correlation coefficient (95% CI)		
	crFMF (N=63)	HIDS/MKD (N=72)	TRAPS (N=46)
CRP	-0.12 (-0.36; 0.14)	0.23 (-0.01; 0.45)	0.12 (-0.19; 0.42)
SAA	-0.01 (-0.27; 0.25)	-0.05 (-0.30; 0.21)	0.06 (-0.26; 0.37)
PGA	0.23 (-0.02; 0.46)	0.35[§] (0.12; 0.55)	0.73[*] (-0.54; 0.85)
CHQ – PSCS	-0.18 (-0.56; 0.26)	-0.25 (-0.55; 0.11)	-0.33 (-0.72; 0.22)
CHQ – PCS	-0.33 (-0.66; 0.11)	-0.46[§] (-0.70; -0.14)	-0.48 (-0.80; 0.04)
SF12 – PCS	-0.26 (-0.57; 0.11)	-0.23 (-0.68; 0.35)	-0.52[*] (-0.81; -0.03)
SF12 – MCS	-0.45[†] (-0.70; -0.10)	-0.55[†] (-0.84; -0.03)	0.09 (-0.43; 0.56)
SDS	0.47[†] (0.22; 0.67)	0.37[§] (0.10; 0.59)	0.41[†] (0.06; 0.67)

* $p < 0.0001$; † $p < 0.001$; § $p < 0.01$; ‡ $p < 0.05$.

Fig 1. Median AIDAI scores over 16 weeks

AIDAI score was calculated as the sum of all 12 daily items (YES=1 or NO=0) during 30 days. Missing AIDAI assessments between first and last AIDAI assessments were imputed as NO. Missing AIDAI assessments after last AIDAI assessment were imputed as NO. LOCF, last observation carried forward.

Conclusions: CAN demonstrated rapid and sustained disease control assessed with AIDAI over 16 wks. Approximately half of crFMF and TRAPS pts, and 40% of HIDS/MKD patients had inactive disease after 16 wks of treatment. AIDAI improvements at Wk 16 correlated with patient and physician driven evaluations (PGA, SF12–MCS and SDS). CRP and SAA are indicators of response to treatment, rather than a disease activity parameter.

References:

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THU0510 UKRAINIAN REGISTRY OF JIA PATIENTS RECEIVING BIOLOGICS: IMPLEMENTATION INTO CLINICAL PRACTICE AND RECENT DATA

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Background: The situation with a treatment of juvenile idiopathic arthritis (JIA) is complicated in Ukraine. Mainly due to financial reason biological therapy is hardly accessible for Ukrainian patients. Nevertheless, some patients with JIA receive biologics within governmental programme. Clinical data of these patients were scattered and not systematized until 2014. The Task Force of Ukrainian Association of rheumatologists, by analogy to other European registries, has developed a registry of JIA patients receiving biologics. Ukrainian register of JIA patients is an observational, prospective, non-interventional clinical study.

Objectives: The main aim of Ukrainian register of JIA patients is an assessment of long-term safety, efficacy and cost of biological treatment of JIA. in the routine clinical practice.

Methods: 33 clinical sites from different regions of Ukraine are participating. Inclusion criteria: i) diagnosis of JIA; ii)age ≤ 2 years old, iii)planned start of biological therapy due to JIA; iv)negative screening for tuberculosis; v)provided informed consent. Patients have undergone standard clinical assessment every 3 months. Disease activity is measured using JADAS27 in all age groups. ANA, HLAB27, RF and anti-CCP detection are highly recommended. Uveitis and other comorbid conditions should also be fixed.

Results: 339 patients were enrolled into the study during 3 years. 64% of patients are girls. Mean age–10.98±4.41 years, with mean disease duration –5.81±3.48 years. The duration of the period between diagnosis and biologics start was 54.31±40.28 months. Comorbid conditions were found in 41.03% patients. In 14.65% of the patients uveitis was diagnosed. Most common JIA subtypes in patients receiving biologics are pJIA with negative RF (45%), sJIA (20%), enthesitis-associated JIA (11%) and persistent oligoarthritis (11%). 67.3% of enrolled patients received adalimumab (ADA); 27.9%>tocilizumab (TOZ) and 4.8%>etanercept, respectively. During observational period biologics was discontinued in 19.8% of patients due to different reasons: adverse events were observed in 6.7% (ADA) and 16.7% (TOZ), insufficient efficiency of 23.3% (ADA) and 33.3% (TOZ), remission - 6.7% (ADA); drug absence - 63.3% (ADA) and 50% (TOZ), respectively. Comparative analysis of ADA and TOZ efficacy was performed in the 144 patients with pJIA with negative RF. Administration either ADA or TOZ resulted in statistically significant reducing of disease activity according to JADAS27. In ADA group after 3 months of administration JADAS27 decreased from 16.3±10.3 to 10.0±7.8 ($p < 0.00001$). In TOZ group after 3 months of administration JADAS27 reduced from 22.2±12.2 to 13.1±9.1 ($p = 0.0012$). The functional disability of the patients also statistically significant decrease in both treatment group starting from 3 months of administration: 1.1±0.8 to 0.7±0.7 ($p = 0.0081$) in ADA and 1.6±1.0 to 1.0±0.7 in TOZ, respectively.

Conclusions: Ukrainian national registry of JIA patients provides real-life long-term data concerning safety, efficacy, outcomes and comparative analysis of biologics in Ukrainian population of JIA patients. Data collection continues and the data received expected to be a background for clinical decision-making in future.

Disclosure of Interest: None declared

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THU0511 TOCILIZUMAB IS A PROMISING TREATMENT OPTION FOR THERAPY RESISTENT JUVENILE LOCALISED SCLERODERMA PATIENTS

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Background: Juvenile localised scleroderma (jLS) is an orphan disease. Most patient respond to treatment of methotrexate or mycophenolate. In case of nonresponse or partial response, based on the promising tocilizumab (TOC) data of adult systemic sclerosis studies, TOC seems to be a promising option. There is no publication regarding the effectiveness of tocilizumab in jLS.

Objectives: To assess the effectivity of TOC in jLS patients, who had nonresponse or partial response on conventional therapy.

Methods: Participants of the pediatric rheumatology email board were asked, if they follow patients with jLS, who are treated with TOC. Clinical characteristics and the response to TOC was assessed.

Results: Six centers responded to the survey from the email board, with around 800 participatns, and reported 11 patients. The mean age of the patients at disease onset was 5.5 years. Disease duration at time of the initiation of TOC was 53.5 months (range 9 to 109). 5 patients had linear subtype, 3 of them with facial involvement, 2 of them Parry Romberg and one of them coup de sabre. Three had generalized subtype, 2 mixed subtype and 1 limited subtype/morphea. Before starting TOC patients received 10/11 Methotrexate, 7/11 Mycophenolate, 1 abatacept and 1 anti-TNF therapy. Reason to start TOC was in 9 patients increase in Localised Scleroderma Activity Index [1] (mLoSSI). In two patients increased extracutaneous activity was the indication, in one increased activity of arthritis and in the other increased activity of the central nervous system involvement.

The mean duration of tocilizumab therapy was 14.75 months. 2 patients received s.c. according to the poly JIA dosing and all other i.v. There were different i.v. doses applied, 5 of them 8mg/kg every 4 weeks, one of them 8 mg /kg every three weeks, 1 every two weeks and 1 patient received 10 mg /kg every 3 weeks. 3/11 received TOC as monotherapy. 8/11 as combination therapy, 6 of them with Methotrexate and one of each with Mycophenolate or Tacrolimus. Therapy success was reflected by a decreased mLoSSI in 8/11 patients and in 6 patients by a decrease in the Localized Scleroderma Skin Damage Index [1] (LoSDI). No new lesion occurred during the treatment and in the patients with Parry Romberg subtype (n=2) no increase in the facial atrophy occurred. In 8/8 patients physician global (VAS 0–100) decreased and in 8/8 the patients global disease activity (VAS 0–100) decreased. In 3/3 patients, where it was applicable, the number of active joints decreased, in one patient the limb discrepancy decreased. The mean modified Rodnan skin score assessed in 8 patients decreased from the mean value of 9.6 to 5.5.

Conclusions: In this small cohort of patients TOC seems to be a promising rescue medication in methotrexate/mycophenolate nonresponsive patients. A prospective controlled study would be important to prove the seen effect in a controlled way.

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THU0512 FLUORESCENCE OPTICAL IMAGING IN JUVENILE PATIENTS WITH AND WITHOUT INFLAMMATORY PEDIATRIC RHEUMATIC JOINT DISEASES

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Background: Imaging techniques play an important role in making a diagnosis and in the evaluation of treatment effectiveness as well as in the outcome assessment of juvenile idiopathic arthritis (JIA). Fluorescence optical imaging (FOI) has been shown to visualize inflammation in arthritis of wrist and finger joints. FOI is a simple and cost-effectively imaging technique that is well tolerated by the patients.

Objectives: Firstly, to determine the association and agreement of FOI with ultrasonography (US) and physician's assessment of swollen and active joints. Secondly, to estimate the predictive power of FOI to distinguish between patients with and without inflammatory pediatric rheumatic joint diseases.

Methods: A total of 95 patients were enrolled in three pediatric rheumatology centers in Berlin, Germany. FOI and US (in greyscale (GS) and power Doppler (PD)) were performed in each patient. The FOI software automatically generated the PrimaVista mode (PVM). Furthermore, three phases (P1, P2, P3) were defined dependent on the signal intensity in the fingertips. Each joint was scored semiquantitatively (0=no signal up to 3=strong signal, more than 50% of affected joint area) in each of the three phases and PVM. US was additionally graded by a semiquantitative score of each joint for synovitis (synovial thickening and joint effusions) in GS and hyperperfusion in PD mode. The joints were defined as active if the FOI or US reached a score of at least 1, respectively. We report the results on 27 patients in this interim analysis.

Results: The mean disease duration was 3.5 years (SD=3.2), the mean cJADAS-10 was 11.0 (SD=12.3), the mean number of active joints in the hand was 3.4 (SD=5.8). Half of the patients had polyarthritis (51.8%) and one third had a non-inflammatory rheumatic disease. A total of 810 joints in 27 patients could be analyzed. Among those, 140 (17.3%) had a positive US synovitis score, 87 (10.7%) a positive US power Doppler signal, 93 (11.5%) a clinically active joint and 133 (16.4%) a positive FOI PVM. Taking the US synovitis score as reference, the FOI PVM had a sensitivity of 40%, a specificity of 86% and an overall agreement of 79%. Taking the active joint count as reference, the FOI PVM had a sensitivity of 46%, a specificity of 88% and an overall agreement of 83%. The area under curve was 0.91 for US power Doppler, 0.84 for US GS synovitis, 0.76 and 0.93 for FOI PVM and P2 for the ability to distinguish between patients with and without inflammatory rheumatic diseases. FOI and US scores correlated highly with the cJADAS-10 and the physicians global. In contrast, the patient-reported outcomes pain and fatigue did not show any correlation with FOI and US scores.

Conclusions: FOI and US had a comparable predictive power to distinguish between patients with and without inflammatory rheumatic diseases in pediatric/juvenile patients. The agreement between active joint count, US and FOI was high. FOI may provide a cost-effective method to evaluate inflammation in finger and hand joints.

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THU0513 NEONATAL MANIFESTATIONS OF IMMUNE-MEDIATED RHEUMATIC DISEASES: A RETROSPECTIVE LONGITUDINAL STUDY IN A TERTIARY HOSPITAL

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Background: Autoimmune rheumatic diseases such as systemic lupus erythematosus (SLE), antiphospholipid antibody syndrome (APS) and Sjögren's syndrome (SS) are part of a clinical spectrum eligible to affect women in child-bearing ages, affecting neonatal outcomes. Cardiac, cutaneous, haematological, hepatic complications and, more rarely, pulmonary complications have been described.

Objectives: This project aims to describe the occurrence of neonatal lupus manifestations and possible associated clinical factors among women with immune-mediated rheumatic diseases.

Methods: A retrospective longitudinal study was performed including pregnant women with immune-mediated rheumatic diseases seen in a multidisciplinary group for autoimmune diseases during pregnancy between January 2010 and December 2015. Clinical and demographic data as well as and pregnancy outcomes and neonatal manifestations were collected through consultation of clinical files. Patients with and without neonatal lupus were compared using Mann-Whitney, chi-square and Fisher tests (SPSS 24.0). Significance level was set as <0.05.

Results: We included 151 gestations from a total of 140 women with a mean age of 32.5±4.4 years; 4 gestations were twin pregnancies. Within these 151 gestations, 54 (35.8%) women had SLE, 17 (11.3%) had Sjögren's syndrome, 17 (11.3%) had rheumatoid arthritis, 41 had APS (27.2%), 11 (7.3%) had Behçet's disease, 4 (2.6%) had systemic sclerosis, 8 (5.3%) had mixed connective tissue disease and 16 (10.6%) had other immune-mediated diseases. 35 (23.2%) had anti-SSA/La antibodies, 18 (11.9%) had anti-SSB antibodies, 6 (4.0%) had anti-URNP antibodies and 43 (28.5%) had anti-nuclear antibodies. During follow-up, 142 (94.0%) babies were born and 7 (4.6%) abortions and 2 (1.3%) foetal losses occurred. 6 (4.2%) neonates were born with neonatal lupus and 1 (0.7%) died in uterus with a complete heart block. Out of the 6 babies with manifestations, 4 (66.7%) were cardiac, 2 (33.3%) were cutaneous, 1 (16.7%) was hepatic, 2 (33.3%) were haematological and 1 (16.7%) was pulmonary. Neonatal lupus manifestations occurred more frequently in mothers with SS (23.5% vs 2.2%; p=0.003), anti-SSA/Ro (20% vs 0%; p<0.001), and anti-SSB/La (27.7% vs 1.5%; p<0.001).

Conclusions: Our study proved a link between immune-mediated rheumatic diseases and specific neonatal outcomes.

Disclosure of Interest: None declared

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THU0514 PREDICTIVE VALUE OF SUBCLINICAL SYNOVITIS DETECTED BY DOPPLER ULTRASOUND IN RELATION TO FLARE IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS TREATED WITH BIOLOGIC THERAPY AFTER TAPERING BIOLOGIC THERAPY

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Background: Anti-TNF therapy is effective and safe in JIA. Changes in anti-TNF doses are common when remission is achieved¹. Subclinical synovitis on Doppler mode (PD) detected by ultrasound can predict flares in adult RA, but it is not yet clear in JIA².

Objectives: The aim of this study is to evaluate the predictive value of subclinical synovitis detected by PD-US in relation to flares in patients with JIA on remission under anti-TNF when therapy is tapered. The preliminary results were presented at the EULAR congress 2015 in Rome (FRI0520).

Methods: Observational, prospective and multicenter study. We included JIA patients on remission at least 6 months with anti-TNF, ETN and ADA, in whom anti-TNF was tapered due to clinical decision. ETN was tapered by increasing the injection 3 days and ADA by increasing a week. Patients were clinically assessed every 3 months and also with PD-US at baseline. Bilateral US assessment included joints and tendons. Adult synovitis definitions and semiquantitative scoring system were used, no synovitis definitions are available for JIA. We collected demographics (date of birth, JIA subcategory, previous and current treatments). Flare was defined as clinical signs and/or symptoms of arthritis that required increase of systemic therapy.

Results: We included 57 patients, with 19 patients (33.33%) having a flare