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–PSCS/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 participants, 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.