A. Alshevskaya³, A. Moskalev³, I. Kriulin^{1,2}. ¹Sechenov First Moscow State Medical University (Sechenov University), Moscow, Russian Federation; ²National Medical Research Center of Children's Health, Moscow, Russian Federation; ³Biostatistics and Clinical Trials Center, Novosibirsk, Russian Federation

Background: Early-onset form of systemic juvenile idiopathic arthritis (sJIA) often presents severe disease course. Choosing the optimal therapy option as first-line treatment is necessary for rapid improvement of patients' quality of life and prevention of further radiologic progression.

Objectives: To evaluate the long-term effectiveness and safety of tocilizumab (TOC) in sJIA patients depending on the duration of the disease treated in the National Medical Research Center of Children's health, Moscow, Russia.

Methods: The study was conducted as a subanalysis of the prospective cohort study to evaluate the efficacy of biologics in children with sJIA. Analysis included sJIA patients younger than 4 years of age at the moment of TOC initiation. Patients were divided into 2 groups: with disease duration shorter than 6 month (ShorterDD group, n=35) and more than 6 month (LongerDD group, n= 19). Treatment efficacy was evaluated according to the dynamics of clinical and laboratory signs using the ACRPedi criteria. The Wallace criteria were used to evaluate whether or not remission had been achieved. Treatment safety was evaluated according to the data presented in the Adverse Event Reports.

Results: TOC was first biologics in 34/35 (97.1%) patients in ShorterDD group and 18/19 (94.7%) patients in LongerDD group. Groups were comparable in terms of disease activity at TOC initiation with 100% of patients presented active systemic features. 31/35 (88.6%) patients in ShorterDD group and 17/19 (89.5%) patients in LongerDD group have median 3 (IQR 1: 6) and 5 (IQR 3: 7.5) active joints, respectively (p=0.119). JADAS-71 level was 17.14 ± 6.25 ShorterDD group and 17.36 ± 5.45 in LongerDD group (p=0.895).

TOC showed high efficacy after first months of treatment with only 6/35 (17.1%) patients in ShorterDD group and 7/19 (36.8%) in LongerDD group remained with active systemic features (p=0.181). JADAS-71 level decreased to 0 points 26/35 patients (74.3%) in ShorterDD group and in 11/19 patients (57.9%) LongerDD group (p=0.237). After 3 month of treatment, WID was achieved by 27/35 patients (77.1%) in ShorterDD group and by 9/19 patients (47.4%) LongerDD group (p=0.038). ACR Pedi 50/70/90 was achieved by 88.6%/85.7%/80% of patients in ShorterDD group and by 84.2%/73.7%/68.4% of patients in LongerDD group after 1 months of treatment and in 77.1%/74.3%/74.3% and 84.2%/78.9%/68.4%, respectively, after 3 months of treatment.

Conclusion: Initiation of tocilizumab treatment in sJIA patients under 4 years of age is highly effective. However, early treatment within first 6 month after disease onset had advantages in speed of reaching an inactive disease as soon as after 3 months of therapy.

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SAT0493

CLINICAL PROFILE OF JIA PATIENTS WITH THE CERVICAL SPINE INVOLVEMENT: A SINGLE CENTER RETROSPECTIVE CONTINUOUS STUDY.

R. Raupov¹, L. Sorokina^{2,3}, M. Dubko¹, L. Snegireva¹, T. Likhacheva¹, A. Santimov¹, E. Gaidar¹, E. Isupova¹, I. Chikova¹, T. Kornishina¹, V. Masalova¹, M. Kostik^{1,3}. ¹Saint Petersburg State Pediatric Medical University, Saint Petersburg, Russian Federation; ²Leningrad Regional Children's Hospital, Saint Petersburg, Russian Federation; ³Almazov National Medical Research Centre, Saint Petersburg, Russian Federation

Background: JIA is the most common chronic condition in pediatric rheumatology. The cervical spine (CS) involvement is associated with severe disease activity and disability and has been recognized as a factor of a poor prognosis. Data about the CS involvement is contradictory due to silent CS involvement in some patients.

Objectives: the aim of our study was to provide a clinical profile of the patients with the CS involvement.

Methods: 753 patients for last 10 years with JIA were analyzed. Patients were divided depending on the CS involvement, which was confirmed by clinical (pain, LOM) and radiological features (effusion in the CS joints). We evaluated active joints and routine tests, such as CRP, ESR, ANA-positivity and HLA B27

Results: The CS involvement was in 101 patients (13.4%). The data are in the table. The CS involvement was more frequently associated with joints of upper body, such as TMJ (23.7% vs 2.9%, p=0.000001), shoulder (29.7% vs 2.9%, p=0.000001), elbow (34.2% vs 12.2%, p=0.000001), wrist (61.4% vs 21.8%, p=0.000001), MCP (43.6% vs 18.4%, p=0.000001), PIP (52.5% vs 21.3%, p=0.000001), DIP (23.8% vs 7.1%, p=0.000001) and hip (44.6% vs 16.6%, p=0.000001), and ankle (60.4% vs 40.2%, p=0.0001) from lower body.

Parameters	CS, yes (n=101)	CS, no (n=652)	p
Female, n (%)	69 (68.3)	388 (59.5)	0.092
ANA-positivity, n (%)	22/57 (38.6)	190/403 (47.2)	0.226
HLA B27-positivity, n (%)	12/33 (36.4)	88/275 (32.0)	0.613
Onset age, years	5.3 (2.7-10.1)	6.1 (3.0- 10.4)	0.241
ESR, mm/h	12.0 (5.0-31.0)	7.0 (3.0- 18.0)	0.0006
CRP, mg/l	3.9 (0.0- 20.0)	1.1 (0.0-9.2)	0.002
Active joints, n (%)	16.0 (9.0-28.0)	5.0 (3.0-10.0)	0.000000
Time before remission, years	2.9 (1.5-5.1)	2.2 (1.4-4.6)	0.046
Oligoarthritis	5 (5.0)	199 (30.5)	0.0000001
Polyarthritis	48 (48.0)	217 (33.3)	
Psoriatic arthritis	7 (7.0)	33 (5.1)	
Enthesitis-related arthritis	22 (21.8)	164 (25.2)	
Systemic arthritis	19 (18.9)	39 (6.0)	
Uveitis, n (%)	9/76 (11.9)	107/444 (24.1)	0.018
Oral glucocorticosteroids, n (%)	37 (36.7)	115/651 (17.7)	0.00001
Biologic, n (%)	68 (67.3)	283 (43.4)	0.000007
Remission, n (%)	57 (56.4)	428 (65.6)	0.072
Flare, n (%)	10 (9.9)	128/651 (19.7)	0.018

Conclusion: The main risk factors of CS involvement in JIA were polyarthritic and systemic arthritis, high inflammatory activity and involvement of joints of upper body. Patients with CS involvement required more often biologics.

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SAT0494

RITUXIMAB IN REFRACTORY PEDIATRIC RHEUMATIC DISEASES: FOCUS FOR THE SAFETY IN REAL CLINICAL PRACTICE

M. Kaleda¹, I. Nikishina¹, E. Nikolaeva¹, S. Arsenyeva¹, V. A. Nasonova Research Institute of Rheumatology, Pediatric Department, Moscow, Russian Federation

Background: Rituximab (RTX) is now approved only for pediatric patients (pts) 2 years of age and older with granulomatosis with polyangiitis or microscopic polyangiitis, but it has been used with success to treat another rheumatic diseases (RD) in children despite the status "of label".

Objectives: to analyze the safety of RTX in children with various RD who did not respond to conventional therapy.

Methods: In our retrospective study safety data was analyzed for all pts, who received at least one infusion of RTX. The dose of RTX was established as 375 mg/m² of body surface area, administered by intravenous infusion once weekly for 1 to 4 weeks, depending on the CD19 lymphocyte count.

Results: 81 patients with RD, who received RTX, were included: 38 (46.9%) with systemic lupus erythematosus (SLE), 16 (19.75%) – JIA, polyarthritis (2 pts - RF negative, 14 - RF positive), 9 (11.1%) - systemic JIA (sJIA), 6 (7.4%) – systemic sclerosis (SSc), 5 (6.2%) – primary Sjogren's syndrome (pSS), 2 (2.5%) – juvenile dermatomyositis (JDM), 4 (4.9%) – mixed connective tissue disease (MCTD) and 1 with livedoid vasculitis (LV). Most were female – 69 (85.2%). The median age at onset – 11.6 years [interquartile range (IQR) 7.9; 14.3], median age of the starting therapy – 15.2 [IQR 12.5; 16.85] and median disease duration – 2.8 [IQR 1.0; 4.6]. 53 pts (65.4%) reported more than one course of RTX, maximum – 10. The median time between each course was 182 days [IQR 156–315]. The RTX was effective in 95% pts, ineffective in 5% (2 pts with sJIA, 2 pts with SLE and macrophage activation syndrome (MAS)). Adverse events (AE) were recorded in 23 (28.4%) pts, included upper respiratory tract infections – 7 (8.6%), urinary tract infections – 2 (2.5%), short-term infusion reactions that did not require discontinuation of therapy – 2 (2.5%), clinically insignificant neutropenia (grade I-II) – 4 (4.9%), decrease of IgG level was detected in 14 (17.5%) pts (median 5.5 g/l [IQR 4.0; 6.9]). The infection rate in pts with a low IgG level was 35.7%, in pts with neutropenia wasn't recorded. Serious AE were recorded in 16 (19.7%) pts: sepsis – 4, pneumonia – 3, herpes zoster – 1, serious infusion reactions – 2, serious postinfusion reactions within 3 to 10 days – 4 (3 – MAS, 1 – hemorrhagic vasculitis), death – 2 pts with SLE and MAS (therapy of RTX was inefficiency). In general, various AE were registered in 55.6% of pts with sJIA, in 52.6% of pts with SLE, 50% of pts with SSc and JDM, and 80% of pts with pSS. Discontinuation of therapy due to SAE was observed in 15 pts (18.5%).