PTX3 is known to have an interesting role in regulation of innate immunity and it is a key player in regulation of many inflammatory reactions [2].

Objectives: This study aimed to measure serum and synovial fluid (SF) levels of PTX3 in juvenile idiopathic arthritis (JIA) patients and to correlate them with different clinical, laboratory and musculoskeletal ultrasound parameters of disease activity.

Methods: We measured PTX3 in the serum (n=57) and SF samples (n=18) from 57 JIA patients and in the serum from twenty healthy control. Disease activity was calculated using the Juvenile Arthritis Disease Activity Score in 27 joints (JDAS27) and musculoskeletal ultrasound examination (MSUS) was performed using grey scale (GS) and power Doppler (PD) 10-joint score (bilateral knee, ankle, wrist, elbow and the 2nd metacarpophalangeal (MCP) joints)[3]; Erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) levels, serum ferritin, rheumatoid factor (RF) titre were measured and the Juvenile arthritis multidimensional assessment report (JAMAR) was documented.

Results: Serum PTX3 level was significantly higher in JIA patients (3.59 ± 2.38 ng/mL) compared to serum level in the healthy controls (1.5 ± 0.9 ng/mL) ($p < 0.001$). There was no significant difference between SF PTX3 level (4.73 ± 2.62 ng/mL) and its level in paired serum samples (4.23 ± 2.93 ng/mL) ($p = 0.59$). JIA patients with systemic onset subtype (n=13) had higher PTX3 serum levels (5.57 ± 2.52 ng/mL) compared to serum level in the oligoarticular (n=25) and polyarticular (n=19) subtypes (2.88 ± 2.13 ng/mL and 3.18 ± 1.19 ng/mL respectively) ($p = 0.001$ and 0.005 respectively). In JIA patients, serum PTX3 level significantly correlated with JDAS27 ($r = 0.52$, $p < 0.05$), CRP titres ($r = 0.46$, $p < 0.05$) and serum ferritin ($r = 0.48$, $p < 0.05$). Both serum and SF PTX3 levels significantly correlated with PD score ($r = 0.51$ and 0.48 respectively, $p < 0.05$). SF PTX3 ($p = 0.001$) was shown to be superior to serum PTX3 ($p = 0.02$), ESR ($p = 0.07$) and comparable to CRP ($p = 0.001$) at predicting PD synovitis score.

Conclusion: JIA patients have significantly increased serum and synovial fluid levels of Pentraxin 3 that remarkably correlated with the JDAS27 and MSUS parameters of inflammations suggesting that it could be a useful marker to reflect JIA disease activity.

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AB0984

INTERSTITIAL LUNG DISEASE IN CHINESE JIA PATIENTS

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Background: Rheumatic diseases and/or medications (for example methotrexate) used to treat the underlying condition are associated with the development of interstitial lung disease (ILD). Development of ILD in subjects with juvenile idiopathic arthritis (JIA) is assumed to be rare, however, severe respiratory complications are increasingly reported in JIA patients.

Objectives: To describe the prevalence of respiratory diagnoses and ILD in a group of Chinese JIA patients at a tertiary center.

Methods: Retrospective chart review of JIA patients attending rheumatology clinic during 2012 to 2019

Results: Fifty-eight cases of JIA were identified. Thirty-three (57%) received DMARDs, majority were on methotrexate (MTx, 24, 73%) and

sulphasalazine (SSZ, 14, 42%). Eleven (19%) were currently or previously on biologics (adalimumab 5, etanercept 5, anakinra 1, previous exposure: infliximab 1, etanercept and anakinra 1).

Respiratory diagnosis was made in 11 (19%) patients. There were 2 (3.4%) cases of ILD diagnosed during the study period (see below). The rest were asthmatics. Among them, 9 were on DMARDs [MTx 8(89%), SSZ 4(44%), leflunomide 2(LEF), hydroxychloroquine 1(HCQ)] or biologics [Etanercept 3(33%), adalimumab 1, tocilizumab and anakinra 1(sequential use)]. Four subjects were on combination DMARDs [MTx&SSZ in 2, 1 each for MTx+LEF+HCQ+SSZ (sequential combination)& MTx+LEF]. Three received combination DMARDs and biologics (1 each for MTx+adalimumab, MTx+etanercept & MTx+anakinra).

Cases of ILD

One was a SJIA patient diagnosed at 2 years old. She failed methotrexate, etanercept & tocilizumab (infusion reaction). She was on low dose prednisolone, methotrexate and anakinra when she presented to us with acute right heart failure requiring ICU care 3 years after diagnosis. Advanced clubbing was noted (fig 1). HRCT thorax on presentation showed severe interstitial lung disease with marked fibrosis and traction bronchiectasis (fig 2). She was diagnosed acute on chronic cor pulmonale and pulmonary hypertension secondary to ILD.

The other was an oligoarticular JIA patient, diagnosed at 2 years old. She was refractory to intra-articular steroid injections and combination DMARDs including MTx, SSZ, LEF & HCQ. Arthritis was controlled by Etanercept. At 12 years of age, she started to have chest pain. Lung function test showed restrictive lung pattern. HRCT thorax showed pulmonary fibrosis.

Conclusion: From this Chinese cohort we identified 2 cases of ILD. They did not present with any symptoms until advanced stage or when complications set in. Both were very young at diagnosis and were difficult to control, as evidenced by the use of multiple DMARDs and biologics including methotrexate, IL6 and IL1 blockade. The causes of ILD are believed to be multifactorial. For severe case, prognosis is poor¹. Although rare, methotrexate induced pneumonitis do occur.

Symptoms of ILD may not be obvious until late stage. Clinical examination alone is not sensitive enough to detect disease progression. Asthma is common, as in this cohort, and may further complicated the assessment. It is recommended in the recent SHARE initiative on JSLE to screen for lung disease². We believe it should be performed in JIA patients as well. 6 minute walk test is a noninvasive way to assess cardiopulmonary functional status and is validated in children³. Patients with unexplained chest symptoms or impaired lung function should be referred to pulmonologist for further assessment.

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Figure 1

Figure 2

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AB0985 EVALUATION OF SERUM CALPROTECTIN (MRP-8/MRP-14) LEVEL IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Evaluation of inflammatory activity is an important element in the management of patients with juvenile idiopathic arthritis (JIA), for which C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) are traditionally used. However, they might be uninformative in case of subclinical inflammation. The serum level of calprotectin MRP-8/MRP-14 (sCal) correlates well with arthritis activity, as it is produced by activated cells directly in synovia.

Objectives: We evaluate the level of sCal in patients with JIA depending on the type of therapy in order to assess comprehensively the disease activity for further treatment correction.

Methods: 74 patients with JIA were examined, 18 of them had oligoarticular disease subtype, 39 – polyarticular, 17 – systemic. The mean age was 11.3 ± 0.4 years; the disease duration was 5.2 ± 0.4 years. Among them, there were 49 (66%) females and 25 (34%) males. All patients were divided into 2 groups depending on the therapy type. Group I consisted of 33 children treated with methotrexate, while 11 of them were in a state of clinical remission. Group II included 41 children treated with biologic DMARDs (adalimumab, etanercept, tocilizumab), while 14 of them achieved clinical remission. All children had normal levels of CRP and ESR. Quantitative indicators distribution is given as a median [5th; 95th percentile], the calculations were carried out using the Mann-Whitney U test.

Results: Level of sCal in the active disease stage in children of Group I was 8,750 ng/ml [3,700; 17,100], while sCal level in Group II was 2,900 ng/ml [1,200; 24,900]; sCal level in children of Group I which achieved clinical remission – 3,400 ng/ml [1,200; 6,000], and the same indicator in Group II – 1,000 ng/ml [100; 2,800]. sCal level was significantly higher in the group of patients who did not receive biologic DMARDs, both in the active stage of disease ($p = 0.000006$, $U = 71.5$) and in the stage of clinical remission ($p = 0.00034$, $U = 11$). sCal level is 5,800 ng/ml less in patients with active stage of disease and 2,400 ng/ml less in patients with clinical remission, both treated with biologic DMARDs. In addition, the level of sCal is 5.5 times higher in our patients (3,300 ng/ml) compared with healthy children (600 ng/ml) ($p = 0.015$). The moderate positive correlation of sCal and JADAS-27 activity index (r-Spearman's = 0.58) was credibly established.

Conclusion: The level of sCal can reflect the degree of inflammatory activity in JIA, it is significantly higher in the group of patients who did not receive biologic DMARDs in the treatment regimen, both in the active disease stage ($p = 0.000006$, $U = 71.5$) and in the stage of clinical remission ($p = 0.00034$, $U = 11$), which indicates the effectiveness of biologic DMARDs in the treatment of JIA. We assume that it would be appropriate to estimate the serum calprotectin level in the comprehensive

analysis of clinical status in JIA patients for the further correction of therapy.

Disclosure of Interests: None declared

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AB0986 VITAMIN D LEVEL AND BONE MINERAL DENSITY IN PATIENTS WITH JUVENILE RHEUMATOID ARTHRITIS

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Background: An impaired bone metabolism is observed in patients with juvenile idiopathic arthritis (JIA), associated both with the activation of pro-inflammatory cytokines and use of steroid medications. The majority of children with JIA have low vitamin D level, which may complicate the disease course. We regard the monitoring of bone mineral density (BMD) in patients with juvenile rheumatoid arthritis (JRA) should be complemented with an additional assessment of calcidiol (25(OH)D) serum level as an indicator of BMD and a criterion of successful therapy.

Objectives: To evaluate the bone densitometry data and calcidiol level in patients with JRA in order to estimate the osteopenic syndrome and the advisability of prescribing the vitamin D additional doses.

Methods: The calcidiol level and BMD data in 65 patients with JIA were assessed (41 girls and 24 boys). All children were divided into 2 groups, depending on the therapy type. Group I consisted of patients who received methotrexate ($n = 37$), Group II – patients who received the biological DMARDs ($n = 28$), namely tocilizumab (6 patients), adalimumab (20 patients) and etanercept (2 patients). At the time of the study, 14 children of the Group II were prescribed the biological DMARDs only, while the 12 patients were in the state of clinical remission unlike the 7 patients of Group I. It should be mentioned that the majority of children in the Group II had high disease activity degrees before the start of treatment with biological DMARDs. In order to show the correlation between BMD and disease activity, cJADAS-27 score result for the last 6 months was estimated. The data were processed using Pearson's chi-squared test and Spearman's Rank correlation coefficient.

Results: The study revealed, 60 children (92% of all patients) showed vitamin D insufficiency, that mainly manifested by decreased calcidiol level (from 21 to 29 ng/ml), only 8 children (12%) showed calcidiol level deficiency (<20 ng/ml). A significant difference was found in the BMD results depending on the therapy type ($\chi^2 = 10.05$; $p < 0.01$) using Pearson's chi-squared test. As a result, children who received biological DMARDs, demonstrated significantly better results according to BMD assessment data. A direct association of moderate strength was found (the correlation coefficient (r-Spearman's) is 0.39). The strong negative association (r-Spearman's was -0.72) was observed between BMD assessment data and cJADAS-27 score result, which confirmed our statement – high disease activity affects the bone tissue mineralization. There was a weak negative association between 25(OH)D level and cJADAS-27 score result (r-Spearman's was -0.15).

Conclusion: There was a paucity of vitamin D level in 92% of patients with JRA, in 60% secondary osteopenic syndrome. It was revealed that patients, who receive biological DMARDs in integrated treatment, demonstrate significantly better results according to BMD assessment data. Calcidiol level has a moderate effect on BMD, while BMD strongly depends on disease activity degree according to the Spearman's Rank correlation coefficient.

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AB0987 CHOICE AND SWITCH BETWEEN BIOLOGICAL AGENTS IN NON-SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS (JIA)

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Background: Over the past few decades, biologic therapy has significantly improved the prognosis in children with JIA. The issue of optimal and personalised biologics prescription and the problem of switching between drugs are relevant questions of modern paediatric rheumatology.