Conclusion: Children with JIA have adiposity similar to healthy controls. Inflammatory activity measured by JDAS is associated with fat mass but not to anthropometric measurements such as body mass index (BMI).

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POS1329

A COHORT STUDY ON THE RISK OF DEVELOPING OBSTRUCTIVE SLEEP APNEA, TEMPOROMANDIBULAR JOINT DISORDERS AND CRANIOFACIAL DEFORMITIES IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

K. S. K. MA¹, L. T. Wang². ¹National Taiwan University, Taipei, Taiwan, ROC, Department of Life Science, Taipei, Taiwan, Republic of China; ²National Taiwan University Hospital, Department of Obstetrics & Gynecology, Taipei, Taiwan, Republic of China

Background: Juvenile Idiopathic Arthritis (JIA), an autoimmune disease, has been proposed to be comorbid with obstructive sleep apnea (OSA).

Objectives: We aimed at identifying the relationship between JIA and OSA.

Methods: We performed a cohort study including JIA and OSA patients from 1999 to 2013. A total of 2791 patients diagnosed with OSA after JIA onset were recruited, which 11,164 eligible individuals without JIA history were selected as matched-controls. A Cox proportional hazard model was developed to estimate the risk of OSA in JIA patients. A cumulative probability model was adopted to assess the time-dependent effect of JIA on OSA development, implying the casual link of the association. To identify whether JIA patients have higher risks for developing temporomandibular joint (TMJ) disorders, craniofacial anomalies and deformities than non-JIA individuals, subgroup analyses was conducted. Finally, Ingenuity Systems Pathway Analysis (IPA) was conducted to identify underlying mechanisms of the above disease correlation among peripheral blood mononuclear cells (PBMCs) from rheumatic factor (RF)-positive and RF-negative JIA patients, and subcutaneous fat tissues from OSA patients, using p-value visualization for RNA-seq analyses.

Results: The Cox proportional hazard model showed that JIA patients were more likely to have OSA than non-JIA individuals (adjusted hazard ratio =1.949, 95% CI =1.264–3.005). The incidence of developing OSA was particularly high among patients who developed JIA aged 18-30 years old (aHR= 2.034, 95% CI=1.305-3.169) and males (aHR=1.82, 95% CI=1.121-2.954). The risk of developing OSA increased within 0-36 months (aHR = 2.216, 95% CI = 1.001 – 4.907) and over 60 months (aHR = 2.558, 95% CI = 1.346 – 4.860) of follow-up duration after JIA onset. Subgroup analyses showed that JIA patients were more likely to have TMJ disorders (relative risk = 2.047, 95% CI = 1.446-2.898) and to receive treatment for craniofacial deformities (RR = 1.722, 95% CI = 1.38-2.148) than non-JIA controls. IPA analyses suggested that the underlying mechanisms involved activation of antigen presentation pathway followed by antigen presentation to CD4+ and CD8+ T lymphocytes, as well as B cell development.

Conclusion: Our findings identified high risks of developing OSA, TMJ disorders, and craniofacial deformities following JIA onset, which the underlying mechanisms may involve both cellular and humoral immunity.

Disclosure of Interests: None declared

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POS1330

SAFETY OF ANAKINRA IN PATIENTS WITH CRYOPYRIN ASSOCIATED PERIODIC SYNDROMES (CAPS) USING A GRADUATED PRE-FILLED SYRINGE

R. Papa^{1,2}, G. Giancane^{2,3}, H. J. Lachmann⁴, P. Brogan⁵, E. Legger⁶, D. Lindqvist⁷, S. Cederholm⁷, F. Bagnasco⁸, N. Ruperto⁹, M. Gattorno¹ on behalf of Paediatric Rheumatology International Trials Organisation (PRINTO) and Eurofever registry. ¹IRCCS Istituto Giannina Gaslini, UOSD Centro Malattie Autoinfiammatorie e Immunodeficienze, Genoa, Italy; ²Università degli Studi di Genova, Dipartimento di Neuroscienze, Riabilitazione, Oftalmologia, Genetica e Scienze Materno-Infantili (DiNOGMI), Genoa, Italy; ³IRCCS Istituto Giannina Gaslini, Clinica Pediatrica e Reumatologia, Genoa, Italy; ⁴Royal Free Campus, National Amyloidosis Centre, London, United Kingdom; ⁵UCL Institute of Child Health, Great Ormond Street Hospital, Infection, Inflammation and Rheumatology Unit, London, United Kingdom; ⁶Beatrix Kinderkliniek,

University Medical Center, Department of Pediatric Rheumatology, Groningen, Netherlands; ⁷Sobi, Stockholm, Sweden; ⁸IRCCS Istituto Giannina Gaslini, Servizio di Epidemiologia e Biostatistica, Genoa, Italy; ⁹IRCCS Istituto Giannina Gaslini, Clinica Pediatrica e Reumatologia, PRINTo, Genoa, Italy

Background: Cryopyrin associated periodic syndromes (CAPS) are a group of ultra-rare autoinflammatory diseases caused by mutations in the *NLRP3* gene, leading to overproduction of IL-1 β . CAPS includes the following subdiagnoses: familial cold autoinflammatory syndrome (FCAS), Muckle-Wells syndrome (MWS) and chronic infantile neurological cutaneous articular syndrome/neonatal-onset multisystem inflammatory disease (CINCA/NOMID), where FCAS is the mildest form of CAPS and CINCA/NOMID the most severe. A graduated pre-filled syringe of the IL-1 receptor antagonist anakinra was introduced to meet the need of smaller and varying doses when treating children with CAPS.

Objectives: To evaluate the safety of anakinra in CAPS patients using the novel pre-filled syringe.

Methods: Investigators managing patients with CAPS treated with anakinra were identified via the Eurofever registry. Follow-up data of at least 3 years for each patient were prospectively collected and analyzed (EUPAS6366). The primary study endpoints were the occurrence of adverse events (AEs) with focus on serious infections, malignancies, injection site reactions, allergic reactions and medication errors, including re-use of the syringe. Secondary endpoints included dose at specific time points, discontinuations, and switch to other IL-1 blocking treatment.

Results: 12 patients with CAPS were included in the study, 8 with MWS, 2 with FCAS and 2 with CINCA/NOMID. The majority were male (75%) and white (83.3%). At baseline, all but one patient were already using the graduated pre-filled syringe with anakinra. 5 patients required a median dose of 2 to 3 mg/kg/day and 6 patients below 2 mg/kg/day. During a total of 26.1 patient years of treatment, there were 7 AEs with rate of 26.8 (95% CI 4.2-169.6) per 100 patient years. All AEs were infections from 1 patient with CINCA/NOMID and were considered unrelated to anakinra by the investigator. Two of the AEs were considered serious as hospitalization was required (1 tonsillitis and 1 urinary tract infection). No AEs were severe, 6 of the AEs were of moderate severity and 1 was of mild severity. In total, 6 patients discontinued anakinra permanently, 2 during the first year of treatment, 3 during the second year and 1 after the third year of follow-up. The reasons for discontinuation included switch to canakinumab (4 patients), switch to canakinumab and inefficacy (1 patient) and non-compliance (1 patient). The remaining 5 patients continued anakinra until the end of the study. 1 patient discontinued anakinra temporarily during the second year of treatment due to non-compliance. No deaths, malignancies, injection site reactions, allergic reactions or medication errors, including re-use of the pre-filled syringe, were observed.

Table 1. Study results

Patients	12 (100)
Gender, M:F	9:3 (75:25)
Ethnicity, white:black	10:2 (83.3:16.7)
Age at baseline, median (range), years	25.3 (1.4-54.9)
Age at disease onset, median (range), years	2.5 (0-29.5)
Age at diagnosis, median (range), years	24.4 (0.6-54.6)
Disease duration, median (range), years	15.5 (1.4-51.3)
Patients already using anakinra at baseline	11 (91.7)
Patients with history of other IL-1 blocking treatments at baseline	3 (25)
Anakinra dose at baseline, median (range), mg/kg/day	1.7 (1.1-2.5)
Anakinra dose during follow-up years 1:2:3>3, median (range), mg/kg/day	1.6:1.6:2.6:1.4 (1.1-3.9:1.4-4.1:7.3:1-3.2)
Patients with AEs	1 (8.3)
Patients with permanent discontinuation of anakinra	6 (50)
Patients with continuation of anakinra until the end of the study	5 (41.7)
Patients with temporary discontinuation of anakinra	1 (8.3)
Total duration of anakinra exposure, median (range), years	1.3 (0.5-7)
Any AE	7 (100)
Urinary tract infection	1 (14.3)
Upper respiratory tract infection	1 (14.3)
Tonsillitis	5 (71.4)

Results are shown as number (%) unless stated otherwise.

Conclusion: The results of the present study confirm the safety profile of anakinra treatment in CAPS patients using the graduated pre-filled syringe. No new safety findings were identified.

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of: Sobi employee and holder of Sobi shares, Speakers bureau: Sobi employee and holder of Sobi shares, Paid instructor for: Sobi employee and holder of Sobi shares, Consultant of: Sobi employee and holder of Sobi shares, Grant/ research support from: Sobi employee and holder of Sobi shares, Employee of: Sobi employee and holder of Sobi shares, Francesca Bagnasco: None declared, Nicolino Ruperto: None declared, Marco Gattorno: None declared
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POS1331

LONG-TERM OUTCOME IN CHILDREN WITH JUVENILE DERMATOMYOSITIS FROM SOUTHERN INDIA

P. R. Chickermane¹, M. Kuriakose², A. Tiwari², A. Shahul², S. Balan². ¹Amrita Institute of Medical Sciences, Clinical Immunology and Rheumatology, Kochi, India; ²Amrita Institute of Medical Sciences, Clinical Immunology and Rheumatology, Kochi, India

Background: There is paucity of data regarding long-term outcome and cumulative damage in children with juvenile dermatomyositis (JDM) from the Indian subcontinent.

Objectives: To assess the long-term outcome and cumulative damage in children with JDM receiving treatment at a tertiary hospital in southern India.

Methods: Retrospective review of records and cross-sectional assessment of outcome and damage in 29 patients with JDM at a tertiary hospital in Kochi, India. The disease course was categorized as monocyclic, polycyclic and chronic progressive. Cumulative damage was assessed using the IMACS myositis damage index (MDI).

Results: Twenty-nine patients (male-16) diagnosed with definite or probable juvenile dermatomyositis based on the Bohan and Peter criteria and having a minimum follow-up period of 3 years each were enrolled. Of these, 20 children were diagnosed and initiated on treatment at our institute (inceptional) and 9 were diagnosed elsewhere and referred to our centre for further management (non-inceptional). The mean age at disease onset was 7.01 ± 3.34 years (range 1.0 to 13.5 years). The median interval from onset to diagnosis was 3 months (range- 3 weeks to 8.75 years). Delayed diagnosis defined as interval from onset to diagnosis exceeding 6 months was noted in case of 8 children. Among patients in the non-inceptional group, six were considered to have not received standard of care treatment prior to referral to our centre. Standard of care treatment was defined as initiation of a treatment regimen comprising of a combination of glucocorticoids with an immunosuppressive agent within 4 weeks of diagnosis. A total of 11 children had a delayed diagnosis and/or had not received standard of care treatment prior to referral.

At our centre, all patients received oral steroids and subcutaneous methotrexate as standard therapy. Pulse steroids were used to induce remission in 12 patients and as a rescue for relapses in 2 patients. Intravenous immunoglobulin was used in 10 children with severe myositis, oropharyngeal weakness, refractory cutaneous disease including calcinosis and concomitant infection, where affordable with good results. Hydroxychloroquine was added in 15 patients with dominant cutaneous manifestations. Mycophenolate mofetil, azathioprine and tacrolimus were used in patients refractory or intolerant to methotrexate. Cyclophosphamide and rituximab were used in 4 patients each with refractory disease and extra-muscular manifestations such as interstitial lung disease. Seven patients with refractory calcinosis received pamidronate infusions.

The total follow-up duration was 121.06 patient-years. A monocyclic course was observed in 11/29 (37.9%), chronic progressive course in 16/29 (55.2%) and polycyclic course in 2/29 (6.9 %). Nearly half of the patients (14/29) had damage in at least one organ using the MDI (MDI ≥ 1). 38% of the patients (11/29) had at least one sign of cutaneous damage, the most common being calcinosis (n=11) and lipodystrophy (n=4). This was followed by skeletal damage (n=6, joint contractures-5, osteoporosis with fracture-1), muscle and pulmonary (pulmonary

fibrosis) damage in 3 patients each, endocrine damage in 2 patients (diabetes mellitus-1, growth failure and delayed development of secondary sexual characteristics-1) and chronic infection in one patient. All patients with damage in at least one organ had a chronic progressive course, indicating damage accrual. Delayed diagnosis and lack of standard of care treatment prior to referral (i.e., longer duration of untreated/ sub-optimally treated disease) were associated with an increased risk of cumulative damage ($p < 0.05$).

Conclusion: Nearly half of the patients had damage in at least one organ using the MDI. Cutaneous damage was the most common, followed by skeletal, muscle, pulmonary and endocrine damage. Longer duration of untreated/ sub-optimally treated disease significantly increases the risk of cumulative damage, highlighting the need for an early diagnosis and referral to pediatric rheumatology services.

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POS1332

ETANERCEPT-ASSOCIATED NEW ONSET UVEITIS IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS UNDER BIOLOGICAL THERAPY: SINGLE CENTER EXPERIENCE

I. Nikishina¹, S. Arsenyeva¹, M. Kaleda¹, O. Kostareva¹, A. Shapovalenko¹, E. Denisova², A. Panova^{1,2}. ¹V.A. Nasonova Research Institute of Rheumatology, Pediatric Department, Moscow, Russian Federation; ²Helmholtz Moscow Research Institute of Eye Diseases, Pediatric Department, Moscow, Russian Federation

Background: Biological agents (BA), especially TNF inhibitors, 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 JIA-associated uveitis, however, development of uveitis *de novo* in pts treated with BA is a well-established paradoxical phenomenon.

Objectives: to evaluate the frequency of new onset (*no*-) uveitis, 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 (1136) who were treated with BA in our clinic from 2004 to 2020. All cases of *no*-uveitis were collected for the describing of their clinical features in disease onset and course, activity level, JIA category, exposure to Methotrexate (MTX) and BA, presence of ANA, HLA B27. **Results:** among of 1136 pts treated with different BA we identified 36 (3.3%) pts (19 female/17 male) with *no*-uveitis under BA. Mostly during etanercept (ETA) therapy (34 cases from 488 ETA courses, 7%), 1/166 - in abatacept (ABA) and 1/372 - in adalimumab (ADA). 30 pts (83%) with *no*-uveitis developed it on the 1st line of BA treatment (29 ETA vs 1 ADA). 4pts (11%) developed *no*-uveitis on 2nd line (3 ETA vs 1 ABA). 2 pts (6%) on third line (all ETA, both pts had also psoriasis). There are no cases of *no*-uveitis under other BA. Frequency of *no*-uveitis was much higher in ETA group. ETA exposure was 26.8 ± 28.8 months (mo). It means there are no "safe" period of therapy from paradoxical phenomenon of *no*-uveitis. JIA subtypes were as follows: RF-neg polyarthritis 9 (25%), persistent oligoarthritis 3 (8%), extended oligoarthritis 21 (59%), enthesitis-related arthritis (ERA) - 3 (8%). Average age at JIA onset was 4.6 ± 3.9 yrs. 20/36 patients had high laboratory activity (CRP 54 ± 23 mg/l; ESR 41 ± 19 mm/h) and severe arthritis before BA initiation. However most of pts (25/36) achieved 90-100% ACRpedi-response by the uveitis development. 23/36 pts were ANA-positive, 17/36 pts had HLAB27, including 7 pts who had the both features. Uveitis was occurred earlier in ANA plus HLAB27-positive pts (mean exposure - 15.3 mo) than in only ANA-positive or HLAB27-positive pts (27.7 mo and 27.6 mo accordingly). 29/36 (81%) of pts received methotrexate (MTX) in mean dosage $11.5 \text{ mg/m}^2/\text{week}$. There are no differences in time of uveitis development depending of MTX. In all cases of *no*-uveitis BA was switched.

Conclusion: Our study suggested that new onset of uveitis is rare adverse event during BA therapy in JIA. Uveitis can develop despite the excellent effect of therapy on joint manifestation. The most typical development of *no*-uveitis is under ETA therapy, especially in the predisposed cases (certain variants of JIA, ANF positivity, HLAB27 presence) and in patients with very high disease activity at the time of the start of biological therapy.

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POS1333

ANKLE ARTHRITIS IN JUVENILE IDIOPATHIC ARTHRITIS: DON'T MISS TENOSYNOVITIS. CLINICAL AND SONOGRAPHIC FEATURES OF ANKLE TENOSYNOVITIS IN JIA

S. Pastore¹, S. Della Paolera¹, A. Zabotti^{2,3}, A. Tommasini^{1,4}, A. Taddio^{1,4}. ¹IRCCS Burlo Garofolo, Pediatric Department, Trieste, Italy; ²Azienda Sanitaria