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

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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

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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 (1.1-3.9:1.4-4.1:7.3:7.1-3.2)
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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