THU0589

PHYSICAL ACTIVITY IN CHILDREN AND ADOLESCENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

M. Sellami, A. Faza, S. Miladi, K. Ouenniche, S. Kassab, S. Chekili, K. Ben Abdelghani, A. Laatar. *Rheumatology, Mongi Slim Hospital – La Marsa, Tunis, Tunisia*

Background: Juvenile Idiopathic Arthritis (JIA) is the most common rheumatic disease of childhood which may cause physical dysfunction and inactive sedentary lifestyle. Physical activity (PA), known to maintain optimal metabolic function and normal development could be impaired during JIA.

Objectives: The aim of this study was to assess PA in children and adolescents with JIA compared with a healthy population and to analyse associations between PA, functional ability and disease activity.

Methods: This is a cross-sectional study including patients with JIA (ILAR criteria) and age and gender-matched healthy Tunisian schoolchildren. Disease activity was evaluated by Juvenile Arthritis Disease Activity Score (JADAS) and functional ability by the Childhood Health Assessment Questionnaire (CHAQ). PA was assessed by children's physical activity questionnaire (CPAQ) for children and for adolescents (APAQ) and expressed as physical activity level (PAL). Moderate to vigorous PA (MVPA) (hours/day) and sedentary time (hours/day) were determined.

Results: A total of 55 patients (38 boys and 17 girls) with JIA and 60 healthy schoolchildren were included. No significant differences for age, height, weight, and body mass index between JIA patients and controls were observed. A quarter of children lived in poor socio-economic conditions. Subtypes of JIA were: oligoarthritis (52.7%), rheumatoid factor-negative (16.3%) or positive (10.9%) polyarthritis, systemic JIA (9%), and enthesitis-related arthritis (7.2%). The mean disease duration was 3.2 years.²⁻⁶ Twelve patients had active disease (21.8%), 25 patients had inactive disease (45.4%) and 18 patients were in remission (32.7%). The mean average of CHAQ was 0.98. Seventy six percent of the JIA group spent most of the time (average of 15 hours/week) on the two lowest categories: sleeping and sitting compared with the control group (average of 8 hours/week). Only 11 children with JIA (20%) played sport regularly with an average of 40 minutes/week which was significantly lower compared with controls (120 minutes/week; $p=0.01$). Children with JIA spent less time in MVPA compared with the control group (0.5 ± 0.6 versus 1.9 ± 0.9 hours/day; $p<0.01$). Only 34.5% of JIA patients met public health recommendations to perform ≥ 1 hour daily MVPA compared with 66.6% in the control group ($p<0.01$). No associations were found between PA level, disease duration and disease activity. However, lower PAL in children with JIA was significantly associated with young age ($p=0.03$), worse well-being ($p=0.01$) and CHAQ >1 ($p<0.001$). A higher CHAQ score was associated with less time spent in MVPA ($p<0.001$) and poor willingness to practice PA ($p=0.001$). Sedentary time was associated with higher body mass index ($p=0.02$).

Conclusions: In our study, children and adolescents with JIA were less physically active than the healthy peers and less active than recommended for general health. This population needs more attention in achieving normal PA.

Disclosure of Interest: None declared

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THU0590

THE ROLE OF STAT4 RS7574865 G/T AND IFR5 RS2004640 G/T GENE POLYMORPHISM IN JUVENILE IDIOPATHIC ARTHRITIS SUSCEPTIBILITY AMONG RUSSIAN PAEDIATRIC POPULATION

M. Krylov¹, E. Fedorov¹, S. Salugina¹, N. Kononova², D. Varlamov². ¹V.A. Nasonova Research Institute of Rheumatology, ²All-Russian Research Institute of Agricultural Biotechnology, Moscow, Russian Federation

Background: Juvenile idiopathic arthritis (JIA) is the common chronic condition in paediatric population aged under 16 y.o. JIA is an autoimmune disorder with the potential contribution from genetic factors and environmental exposures. Signal transduction and activation of transcription 4 (STAT4) gene and interferon regulatory factor 5 (IRF5) are the common set of susceptibility genes in many autoimmune disorders.

Objectives: To study STAT4 rs7574865 G/T and IFR5 rs2004640G/T gene polymorphisms impact on JIA susceptibility among Russian children.

Methods: The study included 177 subjects, 66 JIA cases and 111 healthy volunteers (the control group). Mean age of JIA patients was 11.7 ± 4.2 y (3–17 y), mean disease duration was 4.8 ± 3.8 y. All JIA patients were on the record at V. A. Nasonova Research Institute of Rheumatology, meeting ILAR-2001 criteria for JIA diagnosis. JIA group was comprised of 30 (45%) patients with oligoarthritis (oligo-JIA), 20 (30%) patients with polyarthritis (poly-JIA) and 16 (24%) patients with systemic onset arthritis (SoJIA). Oligo-JIA subgroup included 20 (67%) HLA-B27-seropositive patients (JIA-B27), and 10 (34%) patients with anterior uveitis (JIA-uveitis). The variables used for patients classification were: sex (44 girls and 22 boys); disease duration: ≤ 4 years - 34 (51%) patients and >4 years - 32 (49%) patients; the number of joints involved: ≤ 4 - 29 (44.0%) and >4 - 37 (56.0%);

positive antibody titers to nuclear antigens (ANA) - 26 (39%) patients. STAT4 and IRF5 genes polymorphism was studied using real time PCR.

Results: There was no significant difference between JIA and healthy volunteers in the incidence of various genotypes and alleles in the studied range of polymorphisms. The only significant difference was found between the rates of T STAT4 allele in oligo-JIA group and T allele in the control group (38% and 20.4%, respectively, $p=0.006$). Similar differences with the controls in T allele rates were observed in JIA-B27 and JIA-uveitis subgroups (35% and 45% vs 20.4% $p=0.044$ and $p=0.021$, respectively). Therefore, carriers of T STAT4 allele demonstrate high JIA susceptibility as compared to healthy subjects (OR 2.43 95% CI (1.23–4.70), $p=0.007$). The risk in JIA-B27 and JIA-uveitis was (OR 2.10 95% CI (0.38–4.60), $p=0.070$ and OR 3.19 95% CI (1.09–9.06), $p=0.021$, respectively). There was no specific association between studied gene polymorphisms and increased JIA susceptibility depending on sex, disease duration, number of joints involved and ANA status.

Conclusions: Obtained data confirm the specific role of STAT4 rs7574865 G/T polymorphism as the susceptibility factor to oligo-JIA associated with uveitis, HLA-B27 in Russian paediatric population.

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THU0591

SERUM 25 OH VITAMIN D IN PAEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS: IN RELATION TO DISEASE ACTIVITY, DURATION AND BONE MINERAL DENSITY

M.H. Abu-Zaid¹, H.H. Abdelnabi², A.M. El-Barbary¹. ¹Rheumatology and Rehabilitation; ²Pediatric, Faculty of Medicine, Tanta University, Egypt., Tanta, Egypt

Background: Vitamin D has immunomodulatory effects and is commonly deficient in Paediatric SLE (pSLE) so associated with the disease activity and low bone density^{1,2}

Objectives: To assess serum level of 25(OH)vitamin D in paediatric lupus patients and correlate it with disease activity, duration and bone density

Methods: A retrospective cohort study conducted on 90 patients with pSLE classified into three groups (30 patients in each) according to disease activity and duration. Group I (Initial active patients), group II (relapsing active patients) and group III (inactive patients), compared with 60 healthy children as controls (group IV). Disease and drugs duration, present steroid dose, medications' history and SLEADI for disease activity were evaluated. CBC, serum C3, C4, 24 hour urinary proteins, anti-dsDNA, Ca, P, PTH, 25(OH) D levels and lumbar spine BMD Z score were measured. Vitamin D levels were correlated with clinical, laboratory and radiological parameters.

Results: There were significant differences in mean 25(OH) D concentration between the patients' groups (I= 15.37 ± 5.72 , II= 17.70 ± 4.88 , III= 26.98 ± 6.59 ng/ml) and controls= 35.90 ± 9.66 ng/ml ($p<0.05$), with significant difference between active groups (I, II) and inactive group III ($p<0.05$). Lumbar spine Z-score was significantly different between groups (I: -0.39 ± 0.81 , II: -1.58 ± 1.12 , III: -0.99 ± 0.89 , IV: 0.58 ± 0.71). There were significant negative correlations between serum 25(OH) D and SLEADI ($r = -0.545$, $p = 0.001$), steroid dose ($r = -0.561$, $p = 0.001$), anti-dsDNA ($r = -0.685$, $p = 0.006$), 24 hr-proteinuria ($r = -0.738$, $p = 0.001$) and PTH ($r = -0.335$, $p = 0.001$), significant positive correlations between 25(OH) D and C3 ($r = 0.617$, $p = 0.001$), C4 ($r = 0.544$, $p = 0.001$) and serum Ca ($r = 0.424$, $p = 0.001$) with non-significant correlations between 25(OH) D and disease duration, steroids' duration, serum P and Z scores ($p>0.05$).

Abstract THU0591 – Table 1. different correlations in this study

Pearson Correlation	25(OH) Vit D	
	(r)	(P value)
SLEADI	-0.545	0.001*
Steroid dose	-0.561	0.001*
Anti-dsDNA	-0.685	0.006*
24 hr-proteinuria	-0.738	0.001*
PTH	-0.335	0.001*
C3	0.617	0.001*
C4	0.544	0.001*
Ca	0.424	0.001*
Disease duration	-0.215	0.080
Steroid duration	-0.089	0.363
Z-score	0.084	0.431
P	0.154	0.092

Conclusions: Vitamin D deficiency is common in pSLE and is considered a risk factor for the disease occurrence as it is correlated significantly with the disease activity not duration.

REFERENCES:

- [1] Vojinovic J, Cimaz R. Pediatric Vitamin D update for the pediatric rheumatologists. *Rheumatol* 2015;13:18.
- [2] Stagi S, Cavalli L, Bertini F, et al. Vitamin D levels in children, adolescents, and young adults with juvenile-onset systemic lupus erythematosus: a cross-sectional study *Lupus* 2014; 23:1059–1065.

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THU0592

PUBERTAL DELAY DESPITE INTENSIVE TREATMENT OF JUVENILE IDIOPATHIC ARTHRITIS: RESULTS OF A LONGITUDINAL STUDY

P. Van Pelt^{1,2}, A. Hokken-Koelega³, R. Dolhain⁴, A. Kruize⁵, H. Bijlsma⁵, N. Wulfrat². ¹Rheumatology and paediatric rheumatology, Erasmus Medical Centre Rotterdam, Rotterdam; ²Paediatric immunology and rheumatology, University Medical Centre Utrecht, Utrecht; ³Paediatric endocrinology; ⁴Rheumatology, Erasmus Medical Centre Rotterdam, Rotterdam; ⁵Rheumatology, University Medical Centre Utrecht, Utrecht, Netherlands

Background: Delayed puberty and reduced adult height have been reported in JIA before the era of biologicals. Long-term consequences of delayed puberty are among others growth disturbances, low bone mineral mass and decreased fertility. Treatment with anti-TNF restores growth, but data on puberty are unknown

Objectives: We evaluated onset and course of puberty and growth, in JIA-patients who are treated intensively, including the possibility of biologicals, and identified variables related with puberty and growth

Methods: In a longitudinal JIA-cohort, all consecutive patients (10–21 years) were followed for three years. Annual examinations were performed regarding demographic and disease-related items as well as Tanner pubertal stages and anthropometric measurements. Median ages at reaching each stage were estimated by Kaplan-Meier curves. Parametric tests were used to determine differences between patients and healthy controls, non-parametric tests between patient-groups. Univariate analyses and mixed models were used to identify associated variables

Results: 138 patients were included (66% girls). Median disease-duration was 7.8 years (IQR 3.7–10.5), median JADAS-27 3.7 (IQR 1.3–8.0), DAS-28 2.16.^{1,5–2,8} Puberty onset was 1.2 years delayed in girls ($p < 0.01$), in boys 0.6 years (ns). The progression was also delayed: end-stage (Tanner-5) in girls was 3.3 years delayed ($p < 0.01$), in boys 1.7 years ($p < 0.01$). A positive association was found for longer disease-duration and a lower BMI. Biological-use was not associated. Both for girls and boys, standardised height was not different at the onset and at the end of puberty

Abstract THU0592 – Figure 1 Vertical. lines indicate median age of healthy population of Tanner stages

Conclusions: In contrast to normalised longitudinal growth, we found in JIA-patients in disease remission or with low disease activity a delayed onset and progression of puberty despite intensive treatment including biologicals. Eventually, puberty was completed and normal adult height was reached. The effects on bone mass and fertility will have to be evaluated in cohort studies

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THU0593

MULTICENTRIC OSTEOLYSIS WITH NODULOSIS AND ARTHROPATHY (MONA): REPORT OF THE FIRST LEBANESE FAMILY

R. Dagher¹, E. Saliba², M. Rizkallah², M.C. Fadous Khalife¹, A. Megarbane³. ¹pediatrics, Notre Dame de Secours University Hospital, Byblos; ²orthopedics, Hotel Dieu de France University Hospital, Beirut, Lebanon; ³medical genetics, Institut Jérôme Lejeune, Paris, France

Background: “Multicentric Osteolysis with Nodulosis and Arthropathy” (MONA) also known as Winchester-Torg syndrome is a rare chronic skeleton disorder caused by matrix metalloproteinase 2 (MMP2) deficiency. It is characterised by facial dysmorphism, subcutaneous fibrocollagenous nodules, carpal and tarsal osteolysis and interphalangeal joint erosions. Short stature and Osteopenia are frequent and heart defects have been described. As children first present with joint pain, swelling and stiffness, MONA is often misdiagnosed as Juvenile Idiopathic Arthritis (JIA).

Objectives: We report the first Lebanese family with 3 siblings presenting MONA, two of which were diagnosed at first as JIA.

Methods: The proband is the eldest boy born of consanguineous parents. At birth, a ventricular septal defect (VSD) was noted. At the age of 5 bone erosions and nodules in his hands and feet, and cuneiform vertebrae of unknown cause appeared. He was diagnosed with JIA. His pain partially improved with methotrexate and TNF agonist treatment; steroid injections were performed in wrists and permitted a gain in range of motion. Secondly, Osteopenia was detected.

The middle boy was examined at the age of 7; he had wrist arthritis and metacarpal tenosynovitis. He was known to have psoriasis. He was treated first with steroid injections then with NSAID and methotrexate with a good response but his condition slowly progressed to deviated fingers.

The youngest boy suffered from foot pain and Kohler disease was diagnosed at the age of 4. Soon after he developed global stiffness of the foot with erosions and nodules. He had failure to thrive and a VSD was detected.

Results: This family history along with the progressive coarsening of face features in the 3 siblings raised the possibility of a genetic disorder. Exome sequencing for skeletal dysplasia in the eldest boy detected a mutation in the MMP2 gene (NM_001127891:exon2:c.8A>G:p.Y3C). The same mutation was found in the 2 other siblings.

Conclusions: These cases are to add to the 44 individuals coming from 27 different families, with molecularly proven MONA reported in the medical literature. This diagnosis should be raised in front of patients having resistant articular erosions with hand and feet nodules, despite anti-rheumatic drugs. Till date, no specific therapy is available and management is only supportive. In this described family, steroid injections were efficient when children presented with swelling and stiffness of joints and seemed to slow progression to erosion.

REFERENCES:

- [1] Bhavani GS, Shah H, Shukla A, Gupta N, Gowrishankar K, Rao AP, et al. Clinical and mutation profile of multicentric osteolysis nodulosis and arthropathy. *Am J Med Genet A* 2016 Feb;170A(2):410–7.