AB1041

PREVALENCE OF JUVENILE IDIOPATHIC ARTHRITIS (JIA) SUBGROUPS AND JIA-ASSOCIATED UVEITIS AMONG JIA PATIENTS ADMITTED TO REFERRAL PEDIATRIC RHEUMATOLOGY CLINICS IN TURKEY: A RETROSPECTIVE STUDY, JUPITER

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Background: Juvenile Idiopathic Arthritis (JIA) is a chronic childhood arthritis with onset before age of 16 and has a significant degree of morbidity that negatively affects quality of life. Uveitis, which is defined as the inflammation of the iris, ciliary body and choroid, is the most common cause of morbidity of JIA. This study was planned to collect data from a Turkish cohort to provide the initial national prevalence data of patients with JIA.

Objectives: The objective of this study was to determine the frequency of JIA subtypes in Turkey. We also aimed to assess the frequency and characteristics of eye involvement in JIA.

Methods: This national, non-interventional, multicenter, observational study was conducted in a retrospective manner in four study centers which were main referral pediatric rheumatology clinics across Turkey. Data on patient demography, medical history, JIA disease characteristics, laboratory data, cases of JIA-associated uveitis, JIA treatment history and data on other comorbidities were collected from a cohort of 500 patients.

Results: Oligoarthritis (n=194, 38.8%) was the most common JIA disease characteristic in this study cohort. The frequency of the subgroups was as follows: Enthesitis-Related Arthritis (ERA) in 23.2% (n=116), polyarthritis in 15.6% (n=78), systemic arthritis in 12.2% (n=61), psoriatic arthritis in 5.2% (n=26), idiopathic arthritis in 2.8% (n=14) and polyarthritis (RF+) in 2.2% (n=11) of patients were identified. The most frequently prescribed treatment for JIA was methotrexate (n=384, 76.8%). A total of 85 comorbidities were reported, and the most frequently reported comorbidity was Familial Mediterranean Fever (FMF) (n=63, 12.6%).

The number of patients with JIA-associated uveitis diagnosis was 34 (6.8%), and the mean duration of uveitis was 3.2 (±2.3) years. The mean duration between the initial JIA diagnosis and diagnosis of uveitis was 1.8 (±1.9) years. Among 34 patients with uveitis, 45 eye involvements were identified; left eye, right eye and both eyes were affected in 5, 8 and 16 patients, respectively. Five patients (14.7%) had uveitis-related complications that required surgical intervention.

Conclusion: The main difference from the European Caucasian population is the lower frequency of oligoarticular JIA and higher frequency of ERA in Turkish JIA patients. Uveitis was also somewhat lower than expected. Geographic and ethnic factors, that may affect these differences, need further investigation.

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AB1042

THE EFFECT OF INTRA-ARTICULAR STEROID INJECTION ON THE CARTILAGE THICKNESS IN JUVENILE IDIOPATHIC ARTHRITIS

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Background: Intra-articular corticosteroid injection is a choice in the treatment of juvenile idiopathic arthritis (JIA), especially in large and few joints involvement. Intra-articular steroid injection provides relief in active synovitis but its effects on cartilage are controversial.

Objectives: The aim of this study is to evaluate the effect of intra-articular steroid injection on distal femoral cartilage thickness in patients with JIA by ultrasonography.

Methods: Distal femoral cartilage thicknesses were measured before the procedure and 6 months later by ultrasonography in the patients that were injected an intra-articular steroid to the knee.

Results: The mean age of the patients was 12.1±4.8 years. Nine of patients were girls and 2 were boys. The mean disease duration was 5.1±4.2 years. Ten patients had oligoarticular JIA and one patient had enthesitis-related arthritis. The number of patients injected to the right knee was 3, the left knee was 5 and both the knee was 3. The femoral cartilage thickness before the procedure was 3.0 mm [min-max 2.0-3.65 mm], 6 months after the procedure was 2.95 mm [min-max 2.0-3.55 mm] and there was no statistical difference (p>0.05).

Conclusion: In this study, no effect of intra-articular steroid treatment on cartilage was observed. However, the number of patients is lack. Therefore, we planned a comprehensive study with more patients.

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AB1043

CLINICAL OUTCOMES OF ANTI-NUCLEAR ANTIBODY POSITIVENESS IN THE 0-18 AGE GROUP: SINGLE-CENTER REAL-WORLD EXPERIENCE

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Background: Anti-nuclear antibody (ANA) assay is important in the diagnosis of autoimmune diseases. However, it has a poor positive predictive value and is detected positive in 10% of general population. In this study, the children that ANA assay was performed in a tertiary pediatric clinic, was evaluated and was aimed to investigate the clinical outcomes in positives.

Objectives: In this study, the children that ANA assay was performed in a tertiary pediatric clinic, was evaluated and was aimed to investigate the clinical outcomes in positives.

Methods: 0-18 age group patients that ANA assay was requested in the various clinics of Erciyes University were included. The patients with positive ANA assay and access to hospital records were retrospectively examined. Patients with and without autoimmune disease were identified as group 1 and group 2, respectively.

Results: The number of patients who required ANA assay in pediatric clinics was 3812. There was a positive ANA assay in 1010 of these patients. The medical records of 909 patients were reached. There were 345 (38%) patients in group 1 and 564 (62%) in group 2. In group 1, female gender was higher compared to group 2 (p <0.05).

Musculoskeletal, hematologic, enterohepatic, and ocular findings as initial findings were significantly high in group 1 ($p < 0.05$). Pediatric Rheumatology, general pediatrics and pediatric neurology are the three clinics which the assay was requested most common. The patients who were examined in pediatric rheumatology, pediatric hematology-oncology, pediatric gastroenterology departments had a higher incidence of autoimmune disease ($p < 0.05$). In patients with autoimmune disease, $\geq 1/1000$ ANA titer and homogenous staining were frequent ($p < 0.05$).

Conclusion: In our center, which is a tertiary health center, the autoimmune disease was not detected in most of the patients whose ANA assay was positive. The proportion of patients with autoimmune disease was low in most of the commonly requested departments. The ANA assay should be requested after the patient's clinical findings are evaluated in detail.

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AB1044

CYTOKINE PROFILE OF MACROPHAGE ACTIVATION SYNDROME ASSOCIATED WITH KAWASAKI DISEASE

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Background: Macrophage activation syndrome (MAS) is a severe, potentially life-threatening complication of childhood systemic inflammatory disorders. MAS occurs most often in children with systemic juvenile idiopathic arthritis and less commonly in children with Kawasaki disease (KD).

Objectives: Our study aimed to assess the kinetics of cytokine release and compare the accuracy of serum biomarkers for diagnosis of MAS, including neopterin, IL-18, IL-6 and soluble TNF receptor type I (sTNFR-I) and sTNFR-II levels, we analysed these levels in patients with KD, including those with MAS, and compared them to the clinical features of KD and MAS.

Methods: Serum neopterin, interleukin (IL)-18, IL-6 and soluble tumour necrosis factor receptor type I (sTNFR-I) and sTNFR-II levels were determined using enzyme-linked immunosorbent assay in 78 patients with KD, including five with MAS. Results were compared to the clinical features of MAS.

Results: Serum neopterin, IL-18, sTNFR-II levels and sTNFR-II/I ratio were significantly elevated in KD patients with MAS compared to those in the acute phase. Receiver operating characteristic curve analysis revealed areas under the curve and cutoff values of neopterin, IL-18, sTNFR-II levels and sTNFR-II/I ratio were 0.9750/30.0 nmol/L, 0.9813/1165 ng/mL, 0.9969/16,600 pg/mL and 0.9875/4.475, respectively. Serum sTNFR-II levels correlated positively with disease activity.

Conclusion: These findings indicate that interferon (IFN)- γ and tumour necrosis factor- α (TNF- α) are closely associated with the pathogenesis of MAS associated with KD. Serum sTNFR-II levels might be a useful marker to diagnose the transition to MAS.

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AB1045

MULTICENTRIC CARPOTARSAL OSTEOLYSIS (MCTO) IN PRACTICE OF PEDIATRIC RHEUMATOLOGIST: DIFFERENTIAL DIAGNOSIS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Rare genetic pathologies involving the musculoskeletal system can be erroneously misdiagnosed as juvenile idiopathic arthritis (JIA).

Idiopathic multicentric carpotarsal osteolysis (MCTO) is a congenital disease, linked to MAFB-gene mutation, described in 2012. MCTO is characterized by progressing osteolysis, mostly of carpal and tarsal bones, leading to articular deformities and functional impairment, with or without nephropathy. The incidence has not yet been established. And there is no available treatment as of today

Objectives: To share the experience of MCTO identification in pediatric rheumatologist practice at federal center.

Methods: Totally 2 MCTO cases were identified in boys 6 and 13 years old during the period 2009 – 2018. Standard rheumatological examination was performed. Genetic testing (Sanger sequencing) in 1 patient identified MAFB-gene mutation.

Results: Both patients had pain and swelling in wrist and ankle joints, flexion contractures in elbow joints, and gait abnormalities. Disease duration at the time of MCTO verification was 4 and 11 years. Both patients went through erroneous polyarticular JIA diagnoses. During the follow up patients' ESR and CRP were normal, HLA B27 - negative, ANA and RF - negative. No visceral pathology, including kidneys, was found. Therapy included NSAIDs, glucocorticosteroids – in one case, MTX and Tocilizumab gave no effect in the second patient. Radiographic findings were severe osteolysis of wrist and feet bones. JIA diagnosis was ruled out and MCTO suspected, and later confirmed in both patients by a geneticist. Heterozygous MAFB gene c.206C>T (p.Ser69Leu) mutation was detected in one patient.

Conclusion: Specific phenotypical features and patient's articular status, osteolysis of the carpal and tarsal bones, absence of laboratory activity signs and of response to antirheumatic therapy is a sound motive to continue diagnostic elaboration in order not to miss rare genetically-linked conditions of the musculoskeletal system.

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AB1046

PHYSICAL ACTIVITY ASSESSMENT IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS COMPARED TO CONTROLS

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Background: Physical activity (PA), known to maintain optimal metabolic function and normal development could be impaired during Juvenile Idiopathic Arthritis (JIA).

Objectives: The aim of our study was to assess PA in children and adolescents with JIA compared to healthy peers using the physical activity questionnaire for children (cPAQ) and adolescents (aPAQ).

Methods: This is a cross-sectional study of measured level of PA in children and adolescents with JIA, compared to age and gender-matched healthy Tunisian schoolchildren. PA was estimated by cPAQ and aPAQ filled by the patient group and the reference group. If the child is unable or unsure to answer the questions we have helped with the parents' response. The PAQ scores 2 as "low activity," >2 and ≤ 3 as "moderate activity," and >3 as "high to vigorous activity".

Results: A total of 55 patients (38 boys and 17 girls) with JIA and 60 healthy control schoolchildren were included. No significant difference in demographic background was found between the two groups. The mean age was 8.5 ± 4.12 years in the JIA group and 9.2 ± 3.51 years in the control group. Thirty-one patients (53%) had persistent oligoarticular JIA, 15 (27%) had polyarticular JIA, 5 patients (9%) had systemic JIA, and 4 (7%) had enthesitis-related arthritis. The median disease duration was 3.2 ± 2.8 years. The mean cPAQ was 2.101 ± 0.722 in the JIA group and 4.112 ± 0.644 in the control group ($p=0.0001$). Children and adolescents with JIA had a significantly lower levels of PA compared with their healthy peers as assessed by cPAQ/aPAQ ($p=0.012$). The time spent in each cPAQ/aPAQ item was significantly lower in the JIA group compared to the healthy group: sports activities (3.4 ± 0.5 versus 4.7 ± 1.4 hours/week, $p=0.002$), leisure time activities (2.2 ± 0.3 versus 6.2 ± 1.3 hours/week, $p=0.001$), activities at school (1.1 ± 0.3 versus 2.1 ± 0.5 hours/week, $p=0.001$), and after school activities (0.5 ± 0.5 versus 2.5 ± 0.8 hours/week, $p=0.001$). Seventy six percent of the JIA group spent the day on the two lowest PA categories: sleeping and sitting, which was significantly higher compared with the reference group ($p=0.001$ and $p=0.055$, respectively).

Conclusion: In our study, children and adolescents with JIA were less physically active than the healthy peers as assessed by the PAQ. More objective methods are needed to better evaluate and quantify the PA.

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