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COMORBIDITIES IN FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF) is a periodic fever syndrome, characterized by recurrent episodes of fever and serosal inflammation accompanied with high acute phase reactants. The analysis of possible comorbidities is important to understand the impact of these conditions on clinical care and whether they share a common etiological pathway.

Objectives: We aimed to evaluate the comorbidities associated with FMF patients in a large genetically diagnosed cohort.

Methods: We retrospectively evaluated the medical records of FMF patients who were followed up at Department of Pediatric Rheumatology in Hacettepe University between 2000 and 2015. This study was approved by the Research Ethics Committee and was conducted in accordance with the Declaration of Helsinki. The diagnosis of FMF was made according to Tel Hashomer diagnosis criteria for patients who applied prior to April 2009 and to the Turkish FMF pediatric diagnosis criteria after April 2009. The FMF patients who had homozygous or compound heterozygous mutations were included in the study. Comorbidities associated with FMF were divided into three groups; associated with increased inflammation, associated with FMF and incidental.

Results: A total of 1999 patients were enrolled in the study. Of all 1999 FMF patients, 636 were children (31.8%), 1029 were males (51.4%), with a mean age of 31.60±16.01 years. The mean follow up time was 4.50 ±3.99 years (median:3.84 range from 0.21-29.4 years). 880 of 1999 (44%) FMF patients had homozygous MEFV gene mutation, the most common mutation was M694V homozygous. The remaining were compound heterozygous. 656 patients (32.8%) had one or more than one comorbidity associated with FMF. Ankylosing spondylitis was the most common comorbidity associated with increased inflammation while the most common comorbidity in FMF related comorbidities was renal amyloidosis. The frequency of ankylosing spondylitis, henoch schonlein purpura, juvenile idiopathic arthritis, polyarteritis nodosa (PAN), multiple sclerosis (MS) and Behçet's disease were increased in patients with FMF when compared to those in the literature. Systemic lupus erythematosus was observed less frequently in the patients with FMF than in the population. While the increase in the frequency of MS was 3.3 times, the frequency of PAN was increased 110 times.

Conclusion: This study shows that FMF is a hereditary disease associated with significant comorbidity. We also confirm that inflammatory and rheumatic diseases are more common in FMF.

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VITAMIN D STATUS IN CHILDREN WITH JIA DEPENDING ON THE SEASON

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Background: Plenty of studies demonstrated the relationship between vitamin D and Juvenile idiopathic arthritis (JIA) disease activity [1,2]. Others reported no relationship between 25(OH)D and disease activity [3,4]. The main source of vitamin D is endogenous vitamin D synthesis induced by sunlight exposure.

Objectives: The aim of this study was to examine the status of vitamin D in children with JIA depending on the peculiarities of the disease and the season.

Methods: 92 patients with JIA were examined. The median age of them was 10.5±1.7 years, from 1.8 to 17.6 years (55 female, 37 male). The serum level of vitamin D was measured through blood test by chemiluminescence method. The relationship between the level of vitamin D and disease activity was analyzed based on juvenile arthritis disease activity score (JADAS27).

Results: The average level of vitamin D was 22,75±1,97ng/ml (corresponded to an insufficient level). It was not found relationship between the frequency of vitamin D deficiency and gender. Vitamin D status changed throughout the year from lowest value 19,52±1,61 ng/ml (in May) till the greatest value 29,62±2,49 ng/ml (in September). Significantly higher level of vitamin D was in September compared to most months (January, p=0,04; February, p=0,04; March, p=0,01; April, p=0,02; May, p=0,01; October, p=0,03; November, p=0,03; December, p=0,01).

The geographical location of Kharkiv (Ukraine) is at 50° latitude. It was proved that UVB radiation above the 33° latitude is not intense enough for the synthesis of vitamin D during the whole year [5].

At the same time there was no significant relationship between the low level of vitamin D in serum and disease activity.

Conclusion: A decrease of vitamin D status were observed throughout the year. Despite the fact that in September was the highest level of vitamin D, the normal concentration was not reached. Seasons should be taken into account, but patients with JIA need supplementation of vitamin D all around the year.

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EXPLORING UVEITIS IN EARLY ONSET ANA POSITIVE JIA

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Background: Juvenile idiopathic arthritis (JIA) is the most frequent childhood systemic disease affiliated with uveitis, where uveitis occurs in 10-13% of the patients, frequently causing long lasting consequences when unrecognized, untimely or incorrectly treated.

Objectives: To explore correlations between age, gender, ANA, RF titer in patients with JIA, occurrence of ocular manifestations and its complications. To examine similarities and differences between patients with different subtypes of JIA and uveitis.

Methods: The retrospective study included 31 children treated for JIA and uveitis in the period 2009-2017 at the Department of Paediatrics, UHC Zagreb. The SUN working group classification and grading system were used to evaluate ocular manifestations. Data analysis was executed using R programming language.

Results: We followed 31 patients (81% female) suffering from JIA with ocular manifestations. Median age at JIA onset in girls was 2.5 (1-14) years and in boys 8.6 (1-14.5) years, while girls had median age 4.25 (1-14) years at first ocular manifestation and boys 8.25 (4 -13.5) years. All patients were RF negative. 61% of patients was ANA positive, out of