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THU0618 HYPOPHOSPHATASIA IN FRENCH TERTIARY CARE HOSPITALS

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Background: Hypophosphatasia is a rare heritable metabolic disorder. Its prevalence is estimated at 1: 1 00 000. Its diagnosis can only be established after genetic confirmation. A low serum total alkaline phosphatase (ALP) level is the hallmark for the diagnosis of hypophosphatasia. Its prevalence is 0.05% in the general population and may be associated with symptoms similar to those of adult forms of hypophosphatasia: excess of joint pathology (chondrocalcinosis, osteoarthritis), periarticular disorders (calcifications, tendinopathies enthesopathies), and disorders of bone mineralization (risk of fracture).

Objectives: The aim of this study was to assess the recognition of persistent low ALP in 3 tertiary care hospitals in France.

Methods: All of the ALP assays of 3 tertiary care hospitals measured in 2013 were reviewed. Persistent hypophosphatasemia was defined by at least one assay <30 IU/L without any assay >40 IU/L. Selected records were analysed to eliminate secondary causes of hypophosphatasemia. A telephone questionnaire was conducted with included patients from the rheumatology and internal medicine departments.

Results: In 2013, 288,851 PAL assays were performed in 1 24 044 patients. Excluding emergency and intensive care unit services, 216,817 PAL assays were performed in 83 657 patients. 716 patients had a value ≤30 IU/L. Of these, 174 had 1 single dosage, 542 multiple dosages, of which 186 never had value >40. 31 patients were excluded due to secondary hypophosphatasemia: severe caloric restriction (n=10), massive surgery (n=6), cancer/hemopathy (n=8), acute pathology -sepsis/voluntary drug intoxication (n=4), high-dose corticosteroid therapy (n=3)). 155 patients were selected; the prevalence of hypophosphatasemia in hospitals is therefore 0.124%. Hypophosphatasemia was noticed in the summary discharge in 1,3%. 4 patients received bisphosphonates despite low PAL (before treatment) and 2 patients had a fracture under treatment. Of the 155, 38 were followed in the rheumatology and internal medicine departments and 33 answered a standardised telephone questionnaire (78% women, average age 43.8 years). 11 patients reported a history of fracture, 2 patients had a history of rickets in childhood, and 1 had known hypophosphatasia in the family. 9 patients had tooth enamel disorders, 7 had gingival recession, 3 had spontaneous tooth loss, and 1 had lost their deciduous teeth by the age of 3 years.

Conclusions: The prevalence of hypophosphatasemia is higher in hospitals than in the general population. This biological anomaly is almost never recorded in the files. However, the existence of hypophosphatasemia should be systematically reported as it is a contraindication to anti-resorptive therapy because of the risk of atypical femoral fracture.

Disclosure of Interest: None declared

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THU0619 THE FREQUENCY OF EXON-10 MUTATIONS IN MEFV GENE IN "PROBABLE" DIAGNOSED FMF PATIENTS ACCORDING TO TEL HASHOMER CRITERIA

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Background: Familial Mediterranean Fever (FMF) is characterised with recurrent inflammatory attacks with serosal inflammation. The clinical findings of FMF is seen in a large spectrum. Tel Hashomer criteria are widely used for classifying FMF. According this criteria set FMF is classified as 'definite' and 'probable' disease.

Objectives: We aimed in this study to investigate the frequency of exon-10 MEFV mutations in 'probable' FMF patients according to Tel Hashomer criteria.

Methods: The study group consisted of 117 patients (79 male, 38 female, median age:31±11, ¹²⁻⁶⁹ which is classified as 'probable' FMF according to Tel-Hashomer criteria. The 12 frequently seen mutations in Turkey analysed in all blood samples and compared with the previous reported 'definite' FMF data from Turkey.

Results: We found in probable FMF group single mutation in 36 patients (%30,8), two mutations in 56 patients (%47,9), 3 mutations in 5 patients (%4,3) and no

mutations in 20 patients (%17,1). The distribution of exon 10 mutations showed single exon-10 mutation in 46 patients (%39,3) and two exon-10 mutations only in 23 patients (%19,7). The detailed distribution of MEFV mutations in 'probable' FMF group is shown in table 1.

Abstract THU0619 – Table 1. The Distribution of MEFV gene mutations in 'probable' FMF group

	Homozygout	Heterozygout	Negative
M694V	5 (4,3%)	42 (35,9%)	70 (59,8%)
M680I	2 (1,7%)	18 (15,5%)	82,8 (96%)
V726A	1 (0,9%)	12 (10,3%)	104 (88,9%)
M694I	-	3 (2,6%)	114 (97,4%)
R202Q	2 (1,7%)	35 (29,9%)	80 (68,4%)
E148Q	1 (0,9%)	15 (12,8%)	101 (86,3%)
P369S	-	3 (2,6%)	114 (97,4%)
R761H	1 (0,9%)	6 (5,1%)	110 (94%)
K695R	-	2 (1,7%)	115 (98,3%)
A744S	-	2 (1,7%)	115 (98,3%)

Conclusions: We found decreased frequency of exon 10 mutations in the MEFV gene in "probable FMF" group according to Tel-Hashomer criteria in comparison previous reported MEFV mutations data. The distribution of non-exon 10 mutations were similar in the definite FMF group. It is needed more clinical studies with large patient group for the clinical significance of non-exon 10 mutations in "probable" FMF patients.

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THU0620 ON DEMAND USE OF ANAKINRA FOR THE ATTACKS OF FAMILIAL MEDITERRANEAN FEVER (FMF)

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Background: IL-1 blocking agents have been shown to be effective in the prevention of attacks in colchicine-resistant FMF (crFMF) patients by their regular use. However, their high cost, side effects and treatment noncompliance limit their use which might be overwhelmed by on-demand use of them which has not been reported in FMF patients. Herein, we evaluated the efficacy of on demand use of anakinra in crFMF patients.

Methods: Data were derived from Gazi FMF cohort which was established in the year 2010. From that date patients with FMF who were diagnosed according to the Tel Hashomer criteria were registered. Co-morbidities, detailed attack characteristics, type, duration, severity, treatments, laboratory parameters and impact of FMF on their life in terms of quality of life and work productivity were recorded either by FMF diary or a mobile phone application (FMF AIDD free to download from AppStore and Android market). A retrospective cohort analysis was made from records of patients who have ever been treated with IL-1 inhibitors.

Results: A total of 60 patients were treated with anakinra in our cohort and 15 patients were identified who were received on demand anakinra protocol. Rationale for on demand use of anakinra was prominent prodrome or trigger for attacks and patients' personal claim. Six patients were switched from regular use and 9 were directly started as on demand use. All were using background colchicine in maximum tolerated doses. None of patients had evidence of persistently elevated acute phase reactants or proteinuria. The median duration of on demand anakinra use was 6 (min 3- max 36) months. Pre- and post- on demand anakinra periods were compared (table 1). Patient reported attack severity (p=0.002), duration (p=0.001), absenteeism (p=0.001) and presenteeism (p=0.002) were significantly