

## Clinical features of familial mediterranean fever in fmf clinic of Bouali Hospital in Ardabil

### Abstract

Introduction: FMF is an inflammatory and hereditary periodic disorder. The symptoms can occur in the first decade of life in more than 80%. With regard to high prevalence of this disorder in this area, this study conducted to introduce FMF as a common disorder with significant characteristic in this area, on the basis of Data outline in our web.

### Materials and Methods :

This is a descriptive study. 422 patients with FMF diagnosis according to the Tel Hashomer criteria were entered. Finally 403 patients enrolled in the study and data of all patients as a web designed questionnaire analysed by SPSS v20 software by using simple descriptive statistics .

### Results :

In this study 175(43.4%) patients were female and 228(56.6%) patients male. , the mean age of patients was 21.03 years. The frequency of symptoms were abdominal pain in 93.3%, fever in 88.1%, weakness in 70% , myalgia in 29.3% , arthralgia in 25.6% , constipation in 23.8% , respiratory discomfort in 21.6% , chest pain in 17.9% , diarrhea in 17.1% , arthritis in 7.7% of patients were seen. Abdominal pain was the main complaint in ( 49.6%) the average duration of pain was  $43.3 \pm 34.5$  hours and the average attack-free period was  $36.5 \pm 29.6$  days. 15.1% of patients had positive family history and 12.7% had history of appendectomy.

In 52.3% delay. in diagnosis was more than three years.

Genetic analysis have been done in 239 patients, 21.33% had no mutations, 39.7% were compound heterozygous genotype, 25.52% heterozygous and 13.38% had compound homozygous mutations . The most common mutations were M694V/V726A (10.46%) and the most common Alleles were M694V(20.9%) and V726A(12.7%). In male genotype M694V-V726A ( 12.7% ) was the most common mutation and in female M694V/M694V (10.4%) was the common mutation. Among the patients with abdominal pain M694V/V726A(12.5%) was more common. Genotypes M680I/V726A(13.9%) , M694/V726A and M694V/R761H(16.7%) , M694V/M694V(33.3%) were common mutations in patients with fever , chest pain and joint symptoms respectively.

**Conclusion:** This study showed the common age for manifestation of this disease is first decade. M694V is the most common mutation and M694V-V726A is the common compound heterozygous. It is concluded that clinical criteria are the best way in FMF diagnosis.

**Keywords:** familial Mediterranean fever, MEFV gene, autoinflammatory syndromes