Pediatric Rheumatology

Poster presentation

Familial Mediterranean Fever (FMF) before the age of one year

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Background

Familial Mediterranean Fever is an autosomic recessive autoinflammatory disease reaching populations of Mediterranean countries. It is mainly a paediatric affection, with a usual presentation at the age of 4 years. However, some cases may start within the first year of life. Only few is known, on their presentation, course and outcome.

Objectives

To identify FMF symptoms in patients before the age of one year. To compare these patients to those with later onset of FMF (after the age of one year).

Population and methods

Retrospective chart review of genetically confirmed FMF patients, comparing clinical symptoms, ethnic origin, response to colchicine and MEFV gene mutations, in the two age groups of patients.

Results

We identified 446 patients divided as such: 37 in the early-onset group (A) and 409 in the other group (B). A statistically significant difference was noted between the 2 groups (A vs B) for the ethnic origin (Sefaradic Jews $p < 0.001$), male preponderance ($p < 0.015$), number of crises/month ($p < 0.015$), response to colchicine treatment, and type of mutations in codon 694 and 680 (for M694V, $p < 0.04$).

Conclusion

This study shows a 10% prevalence rate of FMF before the age of 1 year, and highlights the difficulty to make early diagnosis. The early appearance of the first symptoms may be predictive of disease severity, as these patients have more severe phenotype and carry mutations known as the most severe.