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Anti-IL1 therapy in patients with refractory FMF living inGermany

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Introduction

About 10-20% of patients with familial Mediterranean fever (FMF) show an inadequate response to colchicine. Patients with colchicine-resistant FMF with or without AA-Amyloidosis can be treated with Interleukin-1 (IL-1)-inhibiting drugs.

Objective

We report our experience in adult patients with colchicine-resistant FMF who were treated with anakinra or canakinumab.

Patients and methods

Demographic data, clinical and laboratory parameters, MEFV mutations, patient reported outcomes and physician global health were analyzed in 15 patients treated with anakinra or canakinumab.

Results

Within our cohort of 160 adult patients with FMF, we identified 15 patients (4 female and 11 male) who were treated with anakinra (n=13) or canakinumab (n=2). Twelve of 15 patients (80%) were of turkish-armenian ancestry. The median FMF severity score was 8 (range 5-14). Patients carrying two high-penetrance MEFV mutations (M694V or M680I) had a severity score of 9 (8/15=53%). Patients with a single high penetrance mutation had a severity score of 11 (3/15=20%). Four patients (4/15=27%) had no MEFV mutations and the FMF severity score was 7.5 (p=0.2). FMF-related AA amyloidosis was diagnosed in 6 patients (40%) and the median FMF severity score was 10 compared to a severity score of 7 in 9 patients without amyloidosis (60%) (p=0.3). Anakinra was used continuously in 13 patients and in 2 patients only during attacks. The number of FMF attacks was significantly reduced by anti-IL1 treatment (p=0.0024). The patient reported health and the physician reported global health were both improved significantly (p

Conclusion

IL-1-blocking therapies are well tolerated and effective in patients with colchicine-resistant FMF. Blocking IL-1 reduced the number and severity of FMF attacks.

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