Treatment

Specialized care and regular monitoring of the disease are necessary.

The objectives of treatment are:

- **To reduce or even eliminate attacks.**
- **To improve quality of life.**
- **To normalize blood markers of inflammation (CRP, SAA).**
- **To avoid long-term complications, especially renal failure linked to inflammatory amyloidosis (AA).**

The standard treatment is ***colchicine***, which is started at 1 mg/day. The daily dose will be increased if necessary to suppress attacks and normalize **CRP levels between attacks.** The maximum daily dose is 2.5 mg/day. Treatment is lifelong and effective in more than 90% of cases.

If attacks or blood inflammation persist despite proper treatment with *colchicine*, anti-interleukin-1 biotherapy may be discussed after consultation with an FMF expert reference center, particularly CEREMAIA.

Factors that trigger attacks must be identified. Stress management techniques and regular gentle physical activity are often beneficial in preventing attacks.

CeRéMAIA Tenon

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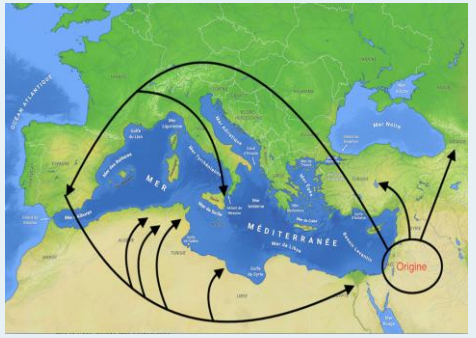
Familial Mediterranean Fever "FMF"



Hôpital Tenon AP-HP

European Reference Network
for rare or low prevalence complex diseases
Network
Immunodeficiency, Autoinflammatory and Autoimmune Diseases (ERN RITA)
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Mediterranean fever Familial

Definition

A rare genetic disorder affecting patients from the Mediterranean region.

Formerly known as: periodic disease.

Epidemiology

It is estimated that there are 100,000 FMF patients worldwide and 5,000 to 10,000 in France. It affects both men and women equally.

Genetics

Familial Mediterranean fever (FMF) is an autosomal recessive genetic disorder. Patients with FMF have two mutations in the MEFV (MEditerraneanFeVer) gene.

The *MEFV* gene encodes the"pyrin," protein which is involved in the production of inflammatory molecules such as interleukin 1, CRP, and SAA in the blood.

Mutations in the *MEFV* gene are common in Armenia, Turkey, and Sephardic and Arab populations originating from North African countries.

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Clinical

The first symptoms of the disease appear very early in life: generally before the age of 5 in 75% of cases and before the age of 1 in 10% of cases.

FMF progresses in painful attacks that last from 24 to 72 hours. Usually, there are no symptoms between attacks. The interval between attacks varies.

During attacks, the fever is often high (38.5°C to 39°C, but can reach 40°C) and may be preceded by a feeling of fatigue or malaise. Other signs may be associated with this fever. The most common is intense abdominal pain, with an abdomen that may resemble appendicitis.

Other symptoms include chest, knee, or ankle pain and red swelling of the ankles called pseudoerysipelas.

Some patients may experience muscle pain, particularly in the legs.

Attacks are often identical for the same individual but may differ within a family.
Sometimes, other inflammatory diseases are associated with it, such as psoriasis, spondyloarthritis, and hidradenitis suppurativa

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Diagnosis

FMF is suspected based on clinical and biological criteria and then confirmed by genetic testing to detect mutations in the MEFV gene, mainly in exon 10.

Progression

Nowadays, if FMF is diagnosed in a child and effective treatment is started, their life expectancy is similar to that of the general French population.

If diagnosis and treatment are delayed, chronic inflammation can affect the kidneys (inflammatory amyloidosis), more rarely the hip joint, and very rarely the liver.

It is necessary to monitor inflammation and kidney function with blood tests twice a year and to perform an annual urine test to screen for the onset of renal amyloidosis.

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