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Familial Fever in Children

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What is Familial Mediterranean Fever?

Familial Mediterranean fever (FMF) also known as 'periodic peritonitis,' is an autoinflammatory genetic disorder that causes recurrent fevers and serosal inflammation of the abdomen, lungs, and joints leading to severe pain. The first attack frequently occurs in childhood, and it usually begins before the age of 20 years. All attacks develop over 2 to 4 hours and last anywhere from 6 hours to 4 days.



Epidemiology of FMF

FMF is commonly seen in people of Mediterranean and Middle Eastern descent.

The carrier rate of FMF is around 1 in 5 healthy individuals.

Recent studies showed that prevalence of FMF to be around 1 in 1000 children.



How to be Diagnosed?

Clinical Findings

- Fever, recurrent abdominal pain, Joint pain, chest pain

Family History

- A family member with confirmed diagnosis of FMF

Genetic Testing

- Genetic Mutation is confirmed

Ethnic Origin

- Individuals of Mediterranean descent



Why it is Difficult to be Diagnosed?

Insufficient knowledge & understanding of this disease are driven by

- The mimicking manifestation of other inflammatory conditions.
- The diversity of symptoms, which varies from one individual to another and within the same family.


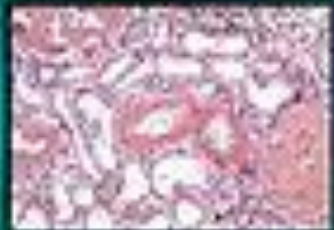


This imposes delayed or missed diagnosis, unbeneficial treatment risks, and progression of serious complications



Why FMF is Considered a Serious Condition?

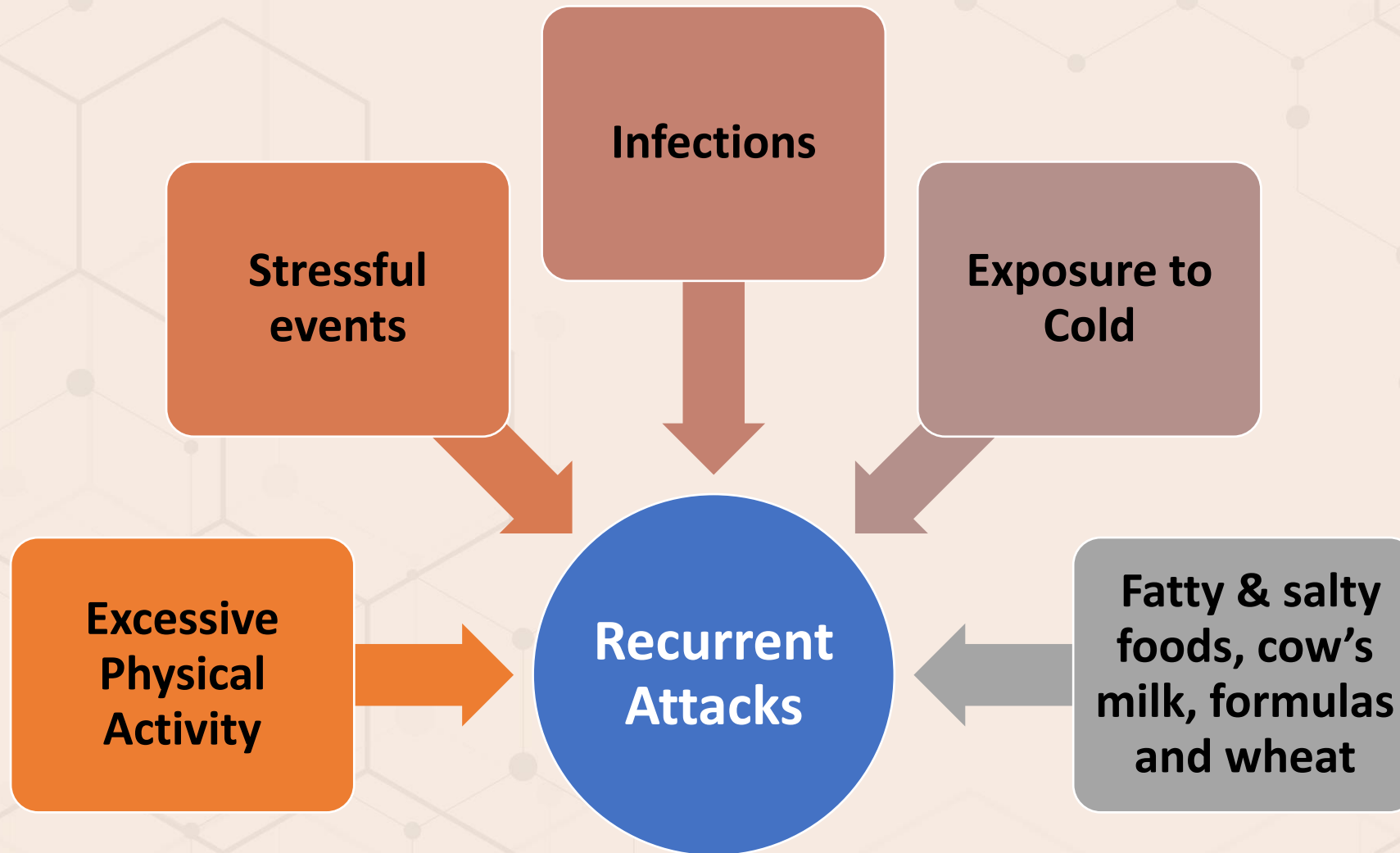
Amyloidosis is considered the most serious complication of FMF, in which amyloid builds up and accumulates in multiple tissues causing organ damage. This often starts in the kidney with nephrotic syndrome and progresses into renal failure.

Primary Amyloidosis



- Plasma cell dyscrasia
- Monoclonal light-chains
- Spontaneous fibril formation
- Widespread tissue deposition
 - Heart
 - Kidney (glomeruli)
 - Nerves (sensorimotor, autonomic)
 - Tongue
 - Vascular (bleeding)
 - Gut

Triggers of Recurrent Attacks



How to Control the Disease?

*It is essential to start a lifelong daily treatment with **Colchicine***

Colchicine has two aims

- Reduce frequency of the attacks
- Restrain the formation and progression of amyloidosis.



Our Role as Pediatric Nurses

*Although, FMF in children is fluctuating between **Overdiagnosis** and **Misdiagnosis** pediatric nurses have a crucial role in containment of disease process. Role of pediatric nurses focusses on the following*

- ☐ Symptom Management
- ☐ Lifestyle Modifications
- ☐ Education and support



Our Role as Pediatric Nurses

□ Symptom Management:

- Colchicine administration
- Pain relief measures, as applying warm compresses, administering analgesics, and encouraging rest.



Our Role as Pediatric Nurses

□ Lifestyle Modifications:

- Encouraging the child to maintain a healthy lifestyle, including a balanced diet, regular physical activity, and adequate rest.
- Identifying and avoiding potential triggers for FMF attacks, such as certain foods, stress, or environmental factors.
- Promoting strategies to manage stress and improve overall well-being, such as relaxation techniques

Our Role as Pediatric Nurses

□ Educational Support:

- Educating the child and family about the long-term nature of FMF and the importance of adherence to the treatment plan.
- Providing resources and information about FMF
- Emphasizing importance of follow up visits and periodical laboratory tests.

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