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Tumour Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) or Familial Hibernian Fever

Version of 2016

2. DIAGNOSIS AND TREATMENT

2.1 How is it diagnosed?

An expert physician will suspect TRAPS on the basis of clinical symptoms identified during a physical examination and a family medical history.
Several blood analyses are useful for detecting inflammation during the attacks. The diagnosis is confirmed only by genetic analysis providing evidence of mutations.
Differential diagnoses are other conditions presenting with recurrent fever, including infections, malignancies and other inflammatory chronic diseases, including other autoinflammatory diseases, such as Familial Mediterranean Fever (FMF) and Mevalonate Kinase Deficiency (MKD).

2.2 Which examinations are needed?

Laboratory tests are important in diagnosing TRAPS. Tests such as erythrocyte sedimentation rate (ESR), CRP, serum Amyloid A protein (SAA), whole blood count and fibrinogen are important during an attack to assess the extent of inflammation. These tests are repeated after the child becomes symptom-free to observe if the results are back to or near normal.
A sample of urine is also tested for the presence of protein and red blood cells. There may be temporary changes during attacks. Patients with amyloidosis will exhibit persistent levels of protein in urine tests.

Molecular analysis of the TNFRI gene is performed in specialised genetic laboratories.

2.3 What are the treatments?

To date, no treatment exists to prevent or cure the disease. Non-steroidal anti-inflammatory drugs (NSAIDs such as ibuprofen, naproxen or indomethacin) help to relieve symptoms. High dose corticosteroids are often effective but sustained use may lead to serious side effects. Specific blockade of the inflammatory cytokine TNF with the soluble TNF receptor (etanercept) has been shown to be an effective treatment in some patients for the prevention of fever attacks. Conversely, the use of monoclonal antibodies against TNF has been associated with an exacerbation of the disease. Recently a good response to a drug blocking another cytokine (IL-1) has been reported in some children affected with TRAPS.

2.4 What are the side effects of drug therapy?

Side effects depend on the drug that is used. NSAIDs can give rise to headaches, stomach ulcers and kidney damage. Corticosteroids and biologic agents (TNF and IL-1 blockers) increase susceptibility to infections. In addition, corticosteroids may cause a wide variety of side effects.

2.5 How long should treatments last?

Due to the rather small number of patients treated with anti-TNF and anti-IL-1, it is not entirely clear whether it is better to treat each new fever attack as it occurs or to treat continuously and if so, for how long.

2.6 What about unconventional or complementary therapies?

There are no published reports of effective complementary remedies.

2.7 What kind of periodic check-ups are necessary?

Patients being treated should have blood and urine tests at least every 2-3 months.

2.8 How long will the disease last?

TRAPS is a life-long disease, although fever attacks may decrease in intensity with age and a more chronic and fluctuating course may be observed. Unfortunately, this evolution does not prevent the possible development of amyloidosis.

2.9 Is it possible to recover completely?

No, because TRAPS is a genetic disease.