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Periodic Fever With Apthous Pharyngitis Adenitis (PFAPA)

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2. DIAGNOSIS AND TREATMENT

2.1 How is it diagnosed?

There are no laboratory tests or imaging procedures specific for diagnosing PFAPA. The disease will be diagnosed based on the combination of physical examination and laboratory tests. Before the diagnosis is confirmed, it is mandatory to exclude all other diseases that may present with similar symptoms.

2.2 What type of laboratory exams are needed?

The values of tests such the erythrocyte sedimentation rate (ESR) or the C-reactive protein (CRP) levels in the blood are raised during attacks.

2.3 Can it be treated or cured?

There is no specific treatment to cure PFAPA syndrome. The aim of treatment is to control symptoms during the episodes of fever. In a large proportion of cases, the symptoms will decrease with time or spontaneously disappear.

2.4 What are the treatments?

Symptoms do not usually respond completely to paracetamol or non-steroidal anti-inflammatory drugs but they may provide some relief. A single dose of prednisone, given when symptoms first appear, has been

shown to shorten the length of an attack. However, the interval between the episodes may also be shortened with this treatment and the next febrile episode may recur earlier than expected. In some patients, a tonsillectomy may be considered, especially when the quality of life of the child and the family is significantly affected.

2.5 What is the prognosis (predicted outcome and course) of the disease?

The disease may last for a few years. With time, the intervals between the febrile attacks will increase and the symptoms will resolve spontaneously in some patients.

2.6 Is it possible to recover completely?

Over the long term, PFAPA will spontaneously disappear or become less severe, usually before adulthood. Patients with PFAPA do not develop damage. The growth and development of a child are usually not affected by this disease.