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Periodic Fever With Apthous Pharyngitis Adenitis (PFAPA)

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1. WHAT IS PFAPA

1.1 What is it?

PFAPA stands for Periodic Fever Adenitis Pharyngitis Aphthosis. This is the medical term for recurrent attacks of fever, swelling of the lymph nodes in the neck, sore throat and mouth ulcers. PFAPA affects children in early childhood, usually with onset before five years of age. This disease has a chronic course but is a benign disease with a tendency toward improvement over time. This disease was first recognised in 1987 and called Marshall’s syndrome at that time.

1.2 How common is it?

The frequency of PFAPA is not known but the disease appears to be more common than generally appreciated.

1.3 What are the causes of the disease?

The cause of the disease is unknown. During periods of fever, the immune system is activated. This activation leads to an inflammatory response with fever and inflammation of the mouth or throat. This inflammation is self-limiting as there are no signs of inflammation between two episodes. There is no infectious agent present during attacks.

1.4 Is it inherited?

Familial cases have been described but no genetic cause has been found to date.

1.5 Is it infectious?

It is not an infectious disease and is not contagious. However, infections may trigger attacks in affected individuals.

1.6 What are the main symptoms?

The main symptom is a recurrent fever, accompanied by a sore throat, mouth ulcers or enlarged cervical lymph nodes (an important part of the immune system). The episodes of fever start abruptly and last for three to six days. During episodes, the child looks very ill and has at least one of the three above-mentioned symptoms. The episodes of fever are recurring every 3-6 weeks, sometimes at very regular intervals. Between episodes, the child is well and activity is normal. The child appears to be perfectly healthy between attacks and there is no impact on development.

1.7 Is the disease the same in every child?

The main features described above are found in all affected children. However, some children may have a milder form of the disease, while others may have additional symptoms, such as malaise, joint pain, abdominal pain, headache, vomiting or diarrhoea.

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.

3. EVERYDAY LIFE

3.1 How might the disease affect the child and the family's daily life?

Quality of life can be affected by recurrent episodes of fever. There can often be considerable delay before the correct diagnosis is made, which may give rise to parental anxiety and sometimes to unnecessary investigations.

3.2 What about school?

Regular fever flares may affect school attendance. It is essential to continue education in children with chronic diseases. There are a few factors that may cause problems for school attendance and it is therefore important to explain the child's possible needs to teachers. Parents and teachers should do whatever they can to allow the child participate in school activities in a normal way, in order not only for the child to be successful academically but also to be accepted and appreciated by both peers and adults. Future integration in the professional world is essential for the young patient and is one of the aims of the global care of chronically ill patients.

3.3 What about sports?

Playing sports is an essential aspect of the everyday life of any child. One of the aims of therapy is to allow children to conduct a normal life as much as possible and to consider themselves not different from their peers.

3.4 What about diet?

There is no specific dietary advice. In general, the child should observe a balanced, normal diet for his/her age. A healthy, well-balanced diet with sufficient protein, calcium and vitamins is recommended for a growing child.

3.5 Can climate influence the course of the disease?

No, it cannot.

3.6 Can the child be vaccinated?

Yes, the child can be and should be vaccinated; however, the treating physician should be informed before administering live attenuated vaccine, to give proper advice on a case-by-case basis.

3.7 What about sexual life, pregnancy, birth control?

So far, no information on this aspect in patients is available in the literature. As a general rule, like for other autoinflammatory diseases, it is better to plan a pregnancy in order to adapt treatment in advance due to the possible side effect of anti-inflammatory drugs on a foetus.