Pædiatric Rheumatology
InterNational Trials
Organisation

Periodic fever with Aphtous Pharyngitis Adenitis (PFAPA)

What is it?

The patient suffers from recurrent attacks of fever and affects children in early childhood, two to four years). This disease has a chronic course, but is a benign disease with a tendency toward improvement over time. This disease was recognised for the first time in 1987 and called Marschalls' syndrome at that time.

How common is it?

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

What are the causes of the disease?

The exact cause of the disease is currently 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-limited as there are no signs of inflammation to be found between two episodes. There is no infectious agent present during attacks.

Is it inherited?

Familial cases have been described, but no genetic cause has been found so far.

Is it contagious?

Infectious agents may play a role in the PFAPA syndrome, but it is not an infectious disease and is not contagious.

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 complains about at least one of the three above-mentioned symptoms. The episodes of fever are recurring every few weeks. Between episodes, the child is asymptomatic and his activity is normal. There is no consequence at all on the development of the child, who looks perfectly healthy between attacks.

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, or may present additional symptoms, like malaise, joint pain, abdominal pain, headache, vomiting, diarrhoea or cough.

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

What type of laboratory exams are needed?
Values of tests, like the erythrocyte sedimentation rate (ESR) or the C-reactive protein (CRP) levels in the blood, are raised during attacks.

Can it be treated or cured?
There is no specific treatment to cure PFAPA syndrome. The aim of the treatment will be to control symptoms during the episodes of fever. In a large proportion of cases, the disease will spontaneously disappeared with time.

What are the treatments?
Symptoms do not usually respond to paracetamol, or non-steroidal anti-inflammatory drugs. 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 can be considered.

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.

Is it possible to recover completely?
Over the long term PFAPA will spontaneously disappear, usually before adulthood. Patients with PFAPA do not develop damage. The growth and development of the child are usually not affected by this disease