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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.