

Periodic fever adenitis pharyngitis aphthous ulcer (PFAPA) syndrome

Reviewed by SickKids Staff | Last updated: June 5th 2019

Information about the symptoms, causes, diagnosis and treatment of periodic fever adenitis pharyngitis aphthous ulcer (PFAPA) syndrome.

Key points

- PFAPA is a disease that causes regular attacks of fever.
- The attacks are usually accompanied by swollen neck glands, sore throat and/or sores in the mouth.
- PFAPA can be treated with medications or removal of the tonsils.
- PFAPA will go away in almost all children by the early teen years.

What is periodic fever adenitis pharyngitis aphthous ulcer (PFAPA) syndrome?

Periodic fever adenitis pharyngitis aphthous ulcer (PFAPA) syndrome causes sudden attacks of high fever. These attacks occur on a regular basis. Children with PFAPA can also have swollen neck glands, sore throat and/or sores in the mouth. Fever that occurs on a regular basis is called periodic fever. Swollen glands are called adenitis. Sore throat is called pharyngitis. Sores in the mouth are called aphthous ulcers.



Sometimes children do not complain of these other symptoms. A doctor should check whether these symptoms are present with the fever.

Symptoms of PFAPA

The major symptom of PFAPA is fever. The episodes of fever occur every three to six weeks, while the fever itself usually lasts about three to five days. Children may complain of pain in the

throat and they may find it hard to swallow because of this pain. They may also complain of pain in the mouth because of mouth ulcers. The glands in the neck may be enlarged. Children seem quite ill during attacks. However, they recover completely between attacks.

The attacks eventually stop by late childhood. There are no long-term effects. Children with PFAPA grow and develop normally.

Causes of PFAPA

The cause of PFAPA is not yet known. For reasons that are not yet understood, the body's inflammation system becomes active, leading to the symptoms of PFAPA.

PFAPA is not caused by an infection that we know about. Children are not contagious when they have a fever from PFAPA. PFAPA is not hereditary. It is not common for more than one child in a family to have it.

Diagnosis of PFAPA

It takes time to diagnose PFAPA. Usually infection is suspected at first. Antibiotics are often prescribed with the first few attacks. After a while, the frequency of the attacks and the similarity of the episodes make the physician and the parents suspect that infection is not the cause.

There are no specific tests to diagnose PFAPA. However, many tests are often done to exclude other causes of fevers. Blood tests should be done both when the child has fever and when the child is well. In a child who has PFAPA, the test results should come back to normal between attacks.

Treatment of PFAPA

Although PFAPA does not have any long-term consequences, most children (and families) are quite disturbed by the frequency of episodes and by how badly the children feel during the attacks. Antibiotics do not work at all. The fevers are only partly controlled with medications like [acetaminophen](#) or [ibuprofen](#). The fever responds better to ibuprofen than acetaminophen. Fever medication does not shorten the length of the attacks.

There is no medication that can cure PFAPA. Steroids like prednisone given at the start of an attack can shorten the length of the attack quite a bit. Sometime two doses, 24 hours apart, are

needed. However, the attacks may happen a little more often if steroids have been used. Seeing how children do with steroid treatment does help confirm the diagnosis of PFAPA.

A newer medicine, anakinra, given once per day by injection into the skin is also very effective. It is only given during the episode until the fever stops.

Cimetidine, a drug given twice a day, may work in some patients. The same is true for colchicine, a medication used in other forms of periodic fever.

[Montelukast](#), a drug for [asthma](#) given once a day, may also work in some children.

Another treatment that has been successful is removal of tonsils and adenoids. It is not clear why this works.

In some children, attacks may return after several years.

How you can help your child with PFAPA

Typical episodes of PFAPA should not be treated with antibiotics. However, children with PFAPA may also get usual childhood infections. If you are concerned that a fever episode is not PFAPA, always contact your child's doctor.

Notify your child's school about the diagnosis. Children with PFAPA are not contagious. They should not be excluded from school.

Children with PFAPA should be encouraged to carry on with normal activities.

Follow-up care at the rheumatology clinic

Your child may be referred to a specialty clinic such as rheumatology for follow-up care. Usually, a rheumatology clinic will have a team of doctors and nurses who have a lot of experience treating children with PFAPA.

Other members of the rheumatology team include:

- a nurse with expertise in inflammation-related illnesses and the impact of these diseases on patients and families and knowledge of different treatments
- a social worker and a child life specialist to help with emotional and behavioural problems that might result from the disease
- a dietitian to help with your child's diet and nutrition

Preparing for visits to the rheumatology clinic

Your child may need a blood test during a clinic visit. This will help doctors monitor the disease. Blood tests also help check for side effects of the medicine your child may be taking. If your child needs to prepare for clinic visits in any other way, a member of the rheumatology team will tell you before the visit.

After you have met the rheumatology team, you will know more about how to care for your child and how to plan for future clinic visits.

Please visit [AboutKidsHealth.ca](https://www.aboutkidshealth.ca) for more child health information.

©2023 The Hospital for Sick Children