



Periodic Fever, Aphthous Stomatitis, Pharyngitis, Adenitis Syndrome (Juvenile)

Fast Facts

- PFAPA usually resolves spontaneously during the second decade of life. However, the disorder may continue into adulthood, although episodes usually become less frequent.
- Steroids given at the start of an episode usually end it, but the result may be more frequent episodes.
- Removing the tonsils in children with many episodes may cure the disease.
- Episodes may affect the quality of life of the child and the family and result in many missed days of school. There is no danger to the long-term health of the child.



Periodic Fever, Aphthous Stomatitis, Pharyngitis, Cervical Adenitis (PFAPA) is a syndrome that consists of recurrent episodes of fever, sore throat, mouth sores and swelling of the glands in the neck. The frequency of PFAPA is not known, but the disease appears to be more common than originally thought, and may be the most common recurrent fever (autoinflammatory) syndrome that does not come from an infection. Both males and females and all ethnic groups can develop PFAPA. PFAPA usually starts in early childhood, between the ages of two to five years. Occasionally PFAPA may develop at an older age, including rare cases in adults.

It is not yet known what causes this disorder. No gene defect has yet to be found in PFAPA, although sometimes more than one family member has the disease and a history of a tonsillectomy (surgical removal of the tonsils) may be reported among family members. No infection has been found in PFAPA, and it is not a contagious disease. It is clear that the inflammatory process is active during episodes, but it is not clear why this happens.

⊕ What is Periodic Fever, Aphthous Stomatitis, Pharyngitis, Cervical Adenitis (PFAPA)?

This syndrome includes recurrent episodes of fever with aphthous stomatitis (mouth sores) and pharyngitis (sore throat with redness). Occasionally, there also may be exudate (white patches on the tonsils) and usually the lymph nodes in the neck are enlarged (adenitis). Episodes of fever start suddenly and last for three - seven days.

Fevers occur routinely every few weeks (usually between three -six weeks); often, families know the exact day when an episode will start. Some children have other symptoms like joint pain, rash, abdominal pain, headache, vomiting or diarrhea. Children are completely well between episodes.

The disease may last for several years but usually will resolve by itself in the second decade of life. However, in nearly 15% of patients episodes (although less frequent) may continue to occur during adulthood. Over time, the time between the episodes will increase. Children with PFAPA continue to grow and develop normally.

+ How is Periodic Fever, Aphthous Stomatitis, Pharyngitis, Cervical Adenitis diagnosed?

There are no laboratory tests specific for diagnosing PFAPA. The disease is diagnosed based on symptoms and physical examination. White blood cell counts and markers of inflammation including the sedimentation rate and the C-reactive protein, all of which can be measured with a blood test, increase during episodes. It is important to exclude all other diseases that may present with similar symptoms (especially a Streptococcus infection) before confirming the diagnosis. The dramatic response to treatment with steroids also helps diagnose PFAPA. In cases without a classic presentation it may be necessary to exclude other causes of recurrent fever (see other patient sheets on this topic), especially if symptoms start in the first year of life and markers of inflammation are increased also between episodes.

+ How is Periodic Fever, Aphthous Stomatitis, Pharyngitis, Cervical treated?

The aim of the treatment will be to control symptoms during the episodes of fever, to shorten the duration of the episodes, and to prevent episodes from occurring. The fever usually does not respond well to acetaminophen (Tylenol) or nonsteroidal anti-inflammatory drugs like ibuprofen (Advil or Motrin). A single dose of steroids (usually prednisone or prednisolone), given when the symptoms first start, dramatically shortens, and often even ends, the episode. However, the time between episodes also may be shortened with this treatment, and the next episode may occur earlier than expected.

Medications like cimetidine and colchicine, when used regularly, may prevent future episodes in about a third (cimetidine) to half (colchicine) of the children. Several studies have found that a tonsillectomy (removing the tonsils by surgery) cures PFAPA in the majority of patients (more than 80%) but the role and timing of surgery in treating PFAPA has still not been fully clarified. Alternatives to steroids should be considered in patients needing steroids more frequently than once every three - four weeks.

Additional Information

The American College of Rheumatology compiled this list to give you a starting point for your own additional research. The ACR does not endorse or maintain these websites, and is not responsible for any information or claims provided on them. It is always best to talk with your rheumatologist for more information and before making any decisions about your care.

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