PATIENT FACT SHEET

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

CONDITION DESCRIPTION

Periodic Fever, Aphthous Stomatitis, Pharyngitis, Cervical Adenitis, or PFAPA, is a syndrome that usually affects children. PFAPA usually starts between ages two and five. Both males and females of any ethnic group can get PFAPA.

PFAPA is an autoinflammatory disease, but its cause is unknown at this time. It is not triggered by infection and is not contagious. In about 15% of patients, PFAPA persists into adulthood with occasional episodes. Episodes usually become less frequent over time.

There is no gene linked to PFAPA, but it sometimes runs in families. Inflammation is involved in PFAPA, but it's unclear why this happens. Children with PFAPA grow and develop normally.

SIGNS/ SYMPTOMS

PFAPA causes recurrent fevers with mouth sores (stomatitis) and red, sore throat (pharyngitis). Children may have white patches on their tonsils and swollen lymph glands in the neck (adenitis). Other possible signs are joint pain, rash, vomiting, diarrhea, headache or stomach pain.

Fevers start suddenly and last from 3-7 days. Episodes may happen every few weeks with no illness in between. A doctor will diagnose PFAPA with a physical examination to check for symptoms. There is no laboratory test for PFAPA, but two common markers of inflammation that show up on blood tests (C-reactive protein and sedimentation rate) rise during these fevers.

Diagnosis may first rule out other causes for the fevers and spike in inflammatory markers, such as Streptococcus infection, especially if symptoms start in the child's first year of life. Prompt reaction to steroid treatment may help confirm PFAPA.

COMMON TREATMENTS

Goals of PFAPA treatment are to ease symptoms, shorten the duration of fevers and prevent recurrence. These fevers usually don't respond well to over-the-counter treatments like acetaminophen (Tylenol) or ibuprofen (Advil, Motrin).

When symptoms first appear, one dose of steroids like prednisone or prednisolone usually shortens or ends the episode. Steroids may shorten time between episodes, however.

Children who need steroids more often than every three to four weeks may need to try other treatments. Tonsillectomy (surgery to remove tonsils) cures PFAPA in 80% or more children, but the best timing for this treatment is unclear. Other medications like cimetidine and colchicine may prevent future PFAPA episodes in many children.

CARE/ MANAGEMENT TIPS

Parents should let their child's doctor know if there is a family history of tonsillectomies, as this could be a sign of past, undiagnosed PFAPA in relatives. This syndrome is more common than doctors once thought.

While PFAPA may cause children to miss days of school, the syndrome isn't a long-term danger to the physical health of the child.

Children may have dramatic response to steroids if they are treated as soon as symptoms appear. Do not delay setting up a pediatrician appointment if the child shows signs of PFAPA. Seek care from a pediatric rheumatologist for treatment and ongoing management.

Updated March 2017 by James Udell, MD, and reviewed by the American College of Rheumatology Communications and Marketing Committee. This information is provided for general education only. Individuals should consult a qualified health care provider for professional medical advice, diagnosis and treatment of a medical or health condition.