Prevalence of Periodic Fever, Aphthous Stomatitis, Pharyngitis and Cervical Adenitis Syndrome (PFAPA): Eastern Wisconsin

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Case Description
- 12 y/o male with multiple handicaps (probably incidental/incunomcapable)
- Began having episodes of aphthous stomatitis with fever and pharyngitis &/or cervical adenopathy & non-specific symptoms
- Every 3 to 5 weeks
- At first were diagnosed as aphthous stomatitis PLUS pharyngitis or otitis media or sinusitis. Antibiotics given.
- After about 6 bouts, Dx: PFAPA Syndrome
- RAS can be due to: vitamin & mineral deficiencies, esp B, Fe, folic acid, Zn; sprue, gluten sensitivity; inflam bowel disease; methotrexate; neutropenia

PFAPA Syndrome
- Etiology: Unknown
- Epidemiology: Onset 2 – 5 y/o, Male>Female
- Fever: 38.9 – 41.1 deg C X 3-4 days
- Repeats q 26 – 30 days (on average)
- Diagnosis: Clinical. Three or more episodes; otherwise normal before onset & in between
- Treatment: Try single dose of prednisone (2mg/kg) at onset of fever. May give 4 day taper if recurrence of sx’s that episode.
- Future plans for other recurrences in some children
- The disorder may be more heterogeneous than previously recognized (eg. older aged children, range of symptoms)
- Thought to be rare, the true prevalence rate is unknown

Other observations
- Antibiotics were prescribed in 4% of all AS cases without other diagnosis.
- Anecdotally, primary care clinicians exhibited significant confusion in their documentation of mouth sores: AS vs. Herpes vs. Herpangina.
- e.g., wrong terms or two different terms used to describe the lesions in the same chart note.

Conclusions
- PFAPA Syndrome indeed appears to be rare in a community population.
- The recurrent nature of AS appears to be poorly documented, but does not contribute significantly to inappropriate antibiotic prescriptions.

Method
- Retrospective electronic chart review of children with surnames A-J seen in a large eastern Wisconsin integrated health system, 2002-2007, with diagnosis of stomatitis (ICD-9 codes 528.0, 528.2)
- AS syndromes were enumerated and demographic, clinical and treatment histories were compared utilizing chi-squared test for categorical variables and Mann-Whitney test for non-normal continuous variables

Results
- Review of 949 records revealed 353 cases of apparent AS
- 47 (13%) were documented to be recurrent (at least two episodes in one year), significantly lower than 50% as reported in current literature
- There was a trend toward recurrent cases (RAS) being more commonly male (60% versus 40% non-recurrent), p=0.08
- Otherwise RAS cases did not differ from non-recurrent cases by age, race or specialty of diagnosing clinician; or presence of other diagnosis.
- Antibiotics were prescribed in 4% of all AS cases without other diagnosis.
- Herpes vs. Herpangina.
- Anecdotally, primary care clinicians exhibited significant confusion in their documentation of mouth sores: AS vs. Herpes vs. Herpangina.
- e.g., wrong terms or two different terms used to describe the lesions in the same chart note.

Conclusions
- PFAPA Syndrome indeed appears to be rare in a community population.
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