

Poster presentation

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## Characteristics of a PFAPA cohort in a single European centre

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### Background

PFAPA is characterized by periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis. Children are healthy between fever attacks. First manifestation is usually before 5 years of age. The aetiology is unknown. There are few diagnostic criteria which help to make the clinical diagnosis. Laboratory parameters are non-specific.

### Methods

Retrospective case analysis.

### Results

Over the years 2004 – 2007 17 boys and 15 girls were diagnosed with PFAPA out of 87 patients referred to the fever clinic (36.8%). Median age at onset was 24 months (4–56), interval between attacks 4 weeks (2–12) and fever duration 3.5 days (1.5–7). Fever was generally above 39°C, associated with pharyngitis in 23 cases (71.9%), cervical adenitis in 24 (75%) and aphthous stomatitis in 11 (34.4%). Other symptoms: abdominal pain, arthralgia (each in 7 cases), vomiting and headache (each in 4 cases). All children had elevated inflammatory parameters during attack (median: CRP 64.5 g/l, ESR 32/h) with subsequent normalization. The single prednisone dose of 1 mg/kg administered at the onset of an episode helped to reduce symptoms in 16/18 children. In 2 cases tonsillectomy led to the resolution of symptoms. After the median follow-up of 8.7 months (3.1–35.5) 4/32 patients (12.5%) have been in the full remission.

### Conclusion

PFAPA appears to be a relatively common cause of recurrent fever in early childhood. The diagnosis was made after clinical exclusion of hereditary fevers and other systemic diseases or immune deficiencies. Our current diagnostic algorithm, therapy and follow-up scheme need further prospective evaluation.