

Poster presentation

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## Periodic fever syndrome with aphthous stomatitis, pharyngitis and cervical adenopathy treated with ketotifen – a case report

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### Background

The pathogenesis of PFAPA syndrome is unknown. According to Stojanov *et al.* cytokine profile in PFAPA syndrome suggests Th1 mediated inflammatory process resulting in continuous inflammation and reduced Th2 anti-inflammatory response. Interferon  $\gamma$  might be responsible for suppressing the production of IL-4 and IL-10. Hung *et al.* showed suppressive effects of ketotifen on the expression of Th1 and Th2 related chemokines of human monocytes.

### Case report

We report a case of a four year old girl with PFAPA syndrome. Attacks of fever started in the first year of life and recur every 2–4 weeks. The genetic testing for FMF, HIDS and TRAPS was negative. No immunodeficiency or autoimmune disease was proven. She responded well to oral corticosteroid treatment but intervals between attacks became shorter. Given the recent finding of possible suppressive effect of ketotifen on Th1 and Th2- related chemokines of monocytes and favourable safety profile of this medication, the patient was prescribed ketotifen 1 mg twice per day for 8 months. During this period we observed significant prolongation of interval between fever attacks. She had only 3 attacks in 8 months. During the period of treatment with ketotifen she was without febrile attacks for 3 months, the longest period ever. After cessation of therapy with ketotifen attacks recur with previous frequency.

### Conclusion

Our case report suggests possible beneficial effect of ketotifen on the frequency of fever attacks in patients with PFAPA syndrome. Ketotifen may be considered as an alternative medication in patients with PFAPA syndrome and recurrent fever attacks.