Pediatric Rheumatology

Poster presentation

PFAPA syndrome: is it a family history?

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Background

PFAPA syndrome is a recurrent febrile disease characterized by periodic fever, aphtous stomatitis, pharyngitis and cervical adenitis. Since first description no clear etiology has been found, no genetic origin was underlined and no familial tendency was reported until now. To better understand this disease, we created a web-based international registry (8 countries and 14 centers).

Aim

To investigate the familial tendency to present PFAPA or another rheumatologic disease.

Patients and methods

In 2 of the participating centers (Lausanne-Geneva, Switzerland; Bordeaux, France), we questioned all parents during a phone call interview to complete the family history. We used the same questionnaire for a control group from a general pediatric consultation. We asked for positive family history of recurrent fevers, PFAPA and rheumatologic diseases. Patients and controls are matched for age and sex.

Results

We recruited 84 patients with PFAPA and 47 control children. Family history for recurrent fever was positive in 37/84 (44%, p = 0.00), always negative in the control group. 9/84 (10%, p = 0.02) PFAPA patients had a family member with PFAPA, none in the control group. The family history for rheumatologic diseases (arthritis, polyarthritis) 14/84 (17%, p = 0.0122) is also more frequently positive in the PFAPA group than in the control 1/47 (2%).

Conclusion

These data show that history of recurrent fever and PFAPA is found more often in patients with PFAPA than in the general pediatric population. They suggest a familial susceptibility and a potential genetic origin for the PFAPA syndrome. This opens a wider spectrum for future research.