



<https://printo.it/pediatric-rheumatology/TW/intro>

International Children's Rheumatology Alliance, **PFAPA**

2023

### 3. Overview

#### 3.1 Definition

PFAPA is a clinical entity characterized by recurrent, self-limiting febrile attacks, periodicity, and absence of identifiable infectious, neoplastic, or autoimmune aetiology.

#### 3.2 Epidemiology

PFAPA is a rare condition with a prevalence of approximately 1:10,000 children. It is most commonly diagnosed between 3 and 6 years of age, with a male-to-female ratio of approximately 1:1.

#### 3.2 Clinical presentation

The clinical presentation of PFAPA is characterized by recurrent, self-limiting febrile attacks, periodicity, and absence of identifiable infectious, neoplastic, or autoimmune aetiology.

#### 3.4 Diagnosis

Diagnosis is based on clinical criteria: recurrent febrile attacks, periodicity, and absence of identifiable infectious, neoplastic, or autoimmune aetiology.

#### 3.5 Treatment

The mainstay of treatment is low-dose corticosteroids, which rapidly reduce the duration and severity of febrile attacks.

#### 3.6 Prognosis

The prognosis is generally good, with most children achieving long-term remission. However, some children may have a more prolonged course.

#### 3.7 Differential diagnosis

