

PFAPA SYNDROME IN CHILDREN

Zespół PFAPA u dzieci

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SUMMARY

PFAPA is a rarely described, autoinflammatory disease whose symptoms include periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis in young children. The disease usually has a benign and self-limiting course. The pathogenesis of this disease has not been recognized yet. The disease more frequently concerns boys at the age from 2 to 5. Patients are asymptomatic between episodes and present normal growth. The aim of the following paper is to present the major issues concerning PFAPA syndrome.

Key words: PFAPA, nursing care, child.

STRESZCZENIE

Zespół PFAPA jest rzadkim, autozapalnym zespołem charakteryzującym się występowaniem epizodów gorączkowych, z towarzyszącym aftowym zapaleniem jamy ustnej, zapaleniem gardła i powiększeniem węzłów chłonnych szyjnych. Zespół zwykle ma łagodny i samoograniczający przebieg. Patogeneza zespołu pozostaje nieznana. Choroba dotyczy najczęściej chłopców w wieku od 2. do 5. roku życia. Rozwój dzieci z PFAPA pozostaje prawidłowy, pomiędzy epizodami gorączki pacjenci pozostają bezobjawowi. Celem pracy jest zaprezentowanie głównych zagadnień dotyczących zespołu PFAPA.

Słowa kluczowe: PFAPA, opieka pielęgniarska, dziecko.

BACKGROUND

Fever occurring in children has always been a source of anxiety of parents and the most frequent reason of the pediatric appointments. PFAPA syndrome, which stands for periodic fever, aphthous stomatitis, pharyngitis, cervical adenitis, was first described by Marshall in 1987 [1]. It may be indicated by periodic fevers occurring in children under 5 years of age and generally resolves by adolescence. The fevers, usually no lower than 39°C, occur approximately every 14–50 days, associated with at least one of three symptoms: pharyngitis, aphthous stomatitis and cervical adenitis [2–4]. Additional symptoms include: chills, stomach-aches, headaches, enlarged spleen and liver, weakness, joint pains, nausea, vomiting and diarrhoea [3, 4]. Other alarming symptoms are not diagnosed between the recurring fevers, children feel well and they develop properly. The illness is typical to stay for a couple of years and it wears off over time [3]. Belongs to the group of recurring febrile diseases which also include Familial Mediterranean fever, HIES (Hyperimmunoglobulinemia E syndrome), tumor necrosis factor receptor-associated periodic syndrome and the cyclic neutropenia.

The pathogenesis of this disease has not been fully recognized. No genetic relationship has been documented although the family history may indicate the occurrence of the disease [5, 6]. The presence of variants in inflammasome related genes, in *NLRP3* and *MEFV*, may suggest a potential role of these genes in PFAPA pathogenesis [7]. PFAPA is currently assigned to the group of autoinflammatory diseases [4].

No significant examination or diagnostic tests to identify PFAPA are currently available. The diagnosis bases mainly on the profound observation of a clinical picture, the fulfillment of the recognition criteria (Table 1) and the elimination of occurring other conditions with periodic fever [4].

Table 1. PFAPA – diagnostic criteria [1, 3]

1. Regular recurring fevers beginning at the age lower than 5 years old.
2. At least one of the following symptoms:
 - aphthous stomatitis,
 - pharyngitis,
 - adenopathy syndrome.
3. Eliminating the cyclical neutropenia.
4. Asymptomatic interval between episodes.
5. Normal growth and development.

Acute-phase reactants (C-reactive protein, ESR) are elevated during a febrile episode. Blood culture as well as oropharyngeal and tracheal are negative [2, 3, 8]. The following symptoms have to appear to diagnose the syndrome: at least 3 fever episodes lasting for 2 up to 7 days and recurring regularly every 14–50 days, aphthous stomatitis, pharyngitis and adenitis accompanied by a proper child development and remission periods of the symptoms between the disease episodes [4].

Children diagnosed with PFAPA show significantly lower concentration of procalcitonin than children suffering from bacterial infections. In contrast, the CRP strongly increases in children suffering from PFAPA compared to the patients with the viral infections [9].

Non-steroidal anti-inflammatory agents and anti-pyretics have shown poor results in resolving symptoms of PFAPA syndrome. According to many authors, corticosteroids, seem to be most effective in controlling the fever episodes. They might not, however, prevent the disease recurrences, but only target the episode's symptoms [2, 3, 8]. Data from Goldsmith [8] confirm the effectiveness of 20–40 mg/kg daily, orally dose of cimetidine, which helps to eliminate the symptoms in 30–40% of patients. The positive effects of using the 6-month-long cimetidine treatment along with its immunomodulatory influence has also been discussed by Krzywicka et al. [2]. A 150 mg per day dose significantly reduces the frequency of fever and the coexisting symptoms; moreover, it can even result in the fully successful treatment [2]. In case of the fever recurrences, Wolska-Kuśnierz et al., find it necessary to apply a 1 mg/kg dose of prednisone [4]. The last method that is recommended by many authors is operation [10, 11]. When other treatment methods have already been used with no effective results, tonsillectomy and adenoidectomy should be performed. The surgery can be performed after individual examination of a particular case. Tonsillectomy in PFAPA syndrome remains controversial.

PFAPA syndrome is distinguished from the cyclical neutropenia, JIA (juvenile idiopathic arthritis), HIDS (hyperimmunoglobulinemia D with recurrent fever), TRAPS (Twin reversed arterial perfusion sequence – also called TRAP), MWS (Muckle-Wells syndrome), and FMF (Familial Mediterranean fever) [2, 3, 8]. It is also necessary to eliminate urinary tract infections and all the congenital defects which lead to infective endocarditis as well as to perform the dental examination including the possible cavity treatment [4].

NURSING CARE

It is an essential part of nursing care to understand the process of temperature control and take appropriate steps to identify if a child's fever is a sign of severe illness. Caring for the child with PFAPA syndrome a nurse must be guided by scientific knowledge about the concept of fever, thermoregulation and methods of measuring body temperature, combined with assessment of the individual patient. Considering the fact that a patient is a child, parents should be treated with care as well. Anger or anxiety might negatively influence the wellbeing of the child.

Due to the fact that a nurse is responsible for taking test materials, it is essential to keep a good relation between her and the child's parents. It is obligatory to explain the aim of the performed activities to the parents and to a child as well. EMLA patches, may be used to mitigate the fear and the pain during the blood collection [12, 13].

During hospitalization, the nurse should observe a child and notice any occurring symptoms. The primary purpose for intervening when a child has a fever is to increase the child's comfort. The nurse measures temperature and notes the result on the medical records. In case of convulsions, arrhythmia or breath condition, the nurse should make a proper note and inform a doctor immediately. To lower the temperature, cold compresses should be put on a head and the patient needs to be watered properly to avoid dehydration resulting from fever. Tepid sponging to reduce symptoms of fever have been questioned, suggests that sponging may cause discomfort as the body will attempt

to compensate by shivering. Changing the bedding by excessive sweating increases the child's comfort. Removal of clothing can help in reducing body temperature. The child should be encouraged to drink fluids to replace those lost through sweating. Maintaining the proper humidity in the room and lips moisturizing are also of high importance [14]. A nurse should constantly observe the patient's mouth for potential aphthae. In case of noticing any lesions in the mouth, the doctor should be informed and symptomatic treatment plan needs to be set. In a child suffering from aphthae's pain, the ailments can be healed with dental floss, gels or sprays. The nurse should also educate the patients about the diet, which is recommended to be light, mild and non-acid. The products should be served fragmented or semi-fluid and tepid. The tooth paste should not contain either SLS or SDS. Such a diet is also recommended in case of pharyngitis because it is not supposed to irritate mucous tissue in the mouth [14]. Additionally a nurse should measure and record heart rate, respiratory rate and capillary refill time (CRT) as part of the routine assessment of a child with a fever. If the child shows signs of circulatory problems such as an abnormal heart rate or a prolonged capillary refill time, a nurse should measure child's blood pressure. All the above mentioned symptoms and other individual ones significantly and negatively influence the child's life quality; therefore, they should be cured in the possibly most efficient way. Where possible, parents or carers of child in hospital should assist with nursing care.

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