



The Place of PFAPA Among the Indications of Tonsillectomy in Children

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Tonsillar and adenoid tissue are among the most important defense systems in human organisms. Adenotonsillar hypertrophy is one of the most common diseases that can occur during childhood; thus, tonsillectomy/adenotonsillectomy is the most frequently performed operation at this time of life (1,2). The main reason this operation is performed so frequently is that there are several indications for it. While tonsillar and adenoid tissue are a very small lymphoid tissue at birth, it may undergo hypertrophy due to the development of the immune system and frequent infections, which may cause obstruction of the airway (3). There is a wide spectrum of indications for adeno/tonsillectomy, ranging from chronic upper respiratory tract infections to sleep apnea (4). Although tonsillectomy is controversial in the treatment of PFAPA (periodic fever, aphthous stomatitis, pharyngitis, and cervical adenitis), it is still performed as a treatment. But until now, in the literature PFAPA has not been mentioned among the absolute or relative indications of tonsillectomy (4,5). Therefore, in this study we aimed to investigate the place of PFAPA among the tonsillectomy indications.

Our computer records of 328 children who underwent tonsillectomy between June 2009 and May 2016 were retrospectively examined in the department of otorhinolaryngology at Fatih University, Turkey. Demographic characteristics of patients and causes of tonsillectomy/adenotonsillectomy were recorded for this study. Besides, for this study it has received permission from the hospital ethics committee. For the statistical analysis the program SPSS was used.

In the study, 147 female (44.8%) and 181 (55.2%) male children were included with a mean age of 6.5 ± 1.7 years. Preoperative diagnosis of the children were episodes of recurrent tonsillitis 164 (50%), tonsillar hypertrophy, adenoid hypertrophy and sleep apnea 73 (22.2%), tonsillar hypertrophy and chronic otitis media (purulent/serous) 59 (17.9 %) and PFAPA 32 (9.7%).

There is a wide spectrum of indications for

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adenotonsillectomy, ranging from chronic upper respiratory tract infections to sleep apnea (4). Many conditions are relative rather than absolute indications, and indications may vary depending on the nature and severity of the underlying problem. There is no standard template for establishing a diagnosis for tonsillectomy/adenotonsillectomy. Hypertrophy and infections of the tonsil and adenoid must be assessed individually for each child, and the decision of operation should be made after evaluating the advantages and disadvantages.

PFAPA is the most common periodic fever condition in children. The true prevalence and etiology exactly remain unknown although one study estimated the incidence to be 2.3 per 10000 children (6). The diagnosis is set according to clinical criteria (Box 1) (7).

The disease is usually seen in children under 5 years old, although recently it was described also in adults (8). The clinical manifestations are defined as a high fever recurring at intervals of 3 to 8 weeks and lasting for 3 to 8 days accompanied by aphthous stomatitis, pharyngitis/

Box 1. Diagnostic criteria for Marshall's/PFAPA syndrome (7)

Regularly recurring fevers with an early age of onset (<5 years of age)
Symptoms in the absence of upper respiratory tract infection with at least one of the following clinical signs:

- Aphthous stomatitis
- Cervical lymphadenitis
- Pharyngitis exclusion of cyclic neutropenia

Completely asymptomatic interval between episodes normal growth and development



tonsillitis, and cervical adenitis. The mean fever duration is reported as 5.1 ± 1.2 days and the mean frequency of episodes as 5.50 ± 1.2 weeks. Current treatment for PFAPA includes corticosteroids given at the onset of an episode, daily cimetidine or colchicine, and tonsillectomy (9). In a meta-analysis study, tonsillectomy was shown to be the most effective treatment method in prevention of long-term episodes of PFAPA (10). Kyvsgaard et al found that fever episodes decreased in 70% of the cases in 20 pediatric patients (11). In a series of 102 PFAPA patients, Licameli et al reported improvement in 99 patients (12). Garavello et al. analysed 15 studies on this subject and concluded that surgical treatment would be an option in PFAPA patients (13). There are also some studies that showed that surgery was not effective (14-16). In our study, only one of the ten patients who underwent tonsillectomy -have had been PFAPA. Therefore, we may say that PFAPA should be its place among the least relative indications of tonsillectomy. But Optimal treatment in PFAPA continues to be debated.

Ethical Issues

Not applicable.

Conflict of Interests

None to be declared.

Financial Support

None to be declared.

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