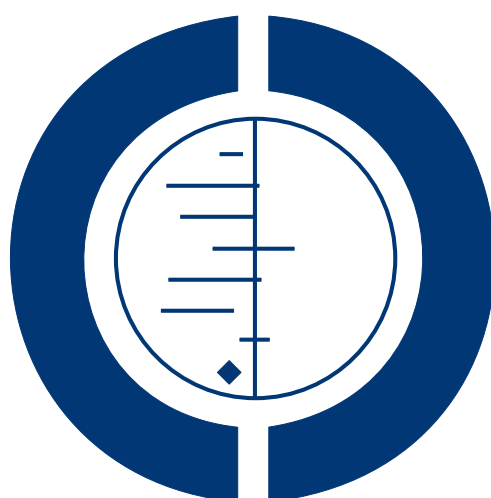


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[Intervention Review]

Tonsillectomy for periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis syndrome (PFAPA)

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ABSTRACT

Background

PFAPA syndrome (periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis syndrome) is a rare clinical syndrome of unknown cause usually identified in children.

Objectives

To assess the efficacy of tonsillectomy (with or without adenoidectomy) in children with PFAPA.

Search strategy

We searched the Cochrane Central Register of Controlled Trials (CENTRAL) (*The Cochrane Library*, 2010 Issue 1); MEDLINE (PubMed); EMBASE; CINAHL; *mRCT* (*meta*Register of clinical trials, including ClinicalTrials.gov); NRR (National Research Register); LILACS; KoreaMed; IndMed; PakMediNet; China Knowledge Network; CAB Abstracts; Web of Science; BIOSIS Previews; ICTRP (International Clinical Trials Registry Platform) and Google. The date of the last search was 21 January 2010.

Selection criteria

Randomised studies comparing adeno-/tonsillectomy with non-surgical treatment.

Data collection and analysis

Two authors independently assessed trial quality and extracted data.

Main results

Two trials involving 67 children were included. One high quality study demonstrated a dramatic benefit of adenotonsillectomy in children with PFAPA diagnosed according to rigid, standard criteria with a relative 'risk' (RR) of symptom resolution after 18 months of 12.63 (95% CI 1.81 to 87.98) and a lower rate of episodes per patient-month (rate ratio 0.07; 95% CI 0.04 to 0.13). A less methodologically rigorous study enrolled some children with PFAPA, but probably included others with alternative types of recurrent pharyngitis, and performed tonsillectomy alone. This also demonstrated a significant benefit for surgery at six months: RR 1.93 (95% CI 1.11 to 3.36); rate ratio episodes per patient-month 0.10 (95% CI 0.04 to 0.28). The pooled relative risk of symptom resolution was 3.25 (95% CI 1.78 to 5.92) and the resulting number needed to treat (NNT) 2 (95% CI 1 to 3).

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Authors' conclusions

The trials included in this review reported follow up at 18 and six months respectively but it is well-established that children with PFAPA recover spontaneously and treatment can be administered to try and reduce the severity of individual episodes. Therefore, the parents and carers of children with PFAPA must weigh the risks and consequences of surgery (hospitalisation, a predictable period of time postoperatively away from school/nursery, the risks of surgery) against the alternative of a finite period of recurrent episodes of disease at predictable intervals, potentially requiring time off school and the regular use of medication. It is uncertain whether adenoidectomy combined with tonsillectomy adds any additional benefit to tonsillectomy alone.

PLAIN LANGUAGE SUMMARY

Tonsillectomy for periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis syndrome (PFAPA) (a rare type of recurrent throat infection)

PFAPA syndrome is a rare cause of regular, repeated episodes of fever, sore throat and swollen neck glands in children. It is a diagnosis that needs confirmation by paediatricians working in centres with expertise in this condition and most children who suffer from a lot of sore throats and tonsillitis will not have it. Those children that do have it have been shown to benefit from having their tonsils removed in two recent trials. The benefit seems to be large; many children had no further episodes after their tonsils were removed. The key issue for parents is that they need to balance the risks and benefits of surgery (tonsillectomy) with the alternative: waiting until their child grows out of the problem and treating each regular episode with medication.