

CARRA PFAPA Consensus Treatment Plan Workgroup. Consensus treatment plans for periodic fever, aphthous stomatitis, pharyngitis and adenitis syndrome (PFAPA): a framework to evaluate treatment responses from the childhood arthritis and rheumatology research alliance (CARRA) PFAPA work group.

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Background: Periodic fever, aphthous stomatitis, pharyngitis, and cervical adenitis (PFAPA) syndrome is the most common periodic fever syndrome in children. There is considerable heterogeneity in management strategies and a lack of evidence-based treatment guidelines. Consensus treatment plans (CTPs) are standardized treatment regimens that are derived based upon best available evidence and current treatment practices that are a way to enable comparative effectiveness studies to identify optimal therapy and are less costly to execute than randomized, double blind placebo controlled trials. The purpose of this project was to develop CTPs and response criteria for PFAPA.

Methods: The CARRA PFAPA Working Group is composed of pediatric rheumatologists, infectious disease specialists, allergists/immunologists and otolaryngologists. An extensive literature review was conducted followed by a survey to assess physician practice patterns. This was followed by virtual and in-person meetings between 2014 and 2018. Nominal group technique (NGT) was employed to develop CTPs, as well as inclusion criteria for entry into future treatment studies, and response criteria. **Consensus required 80% agreement.**

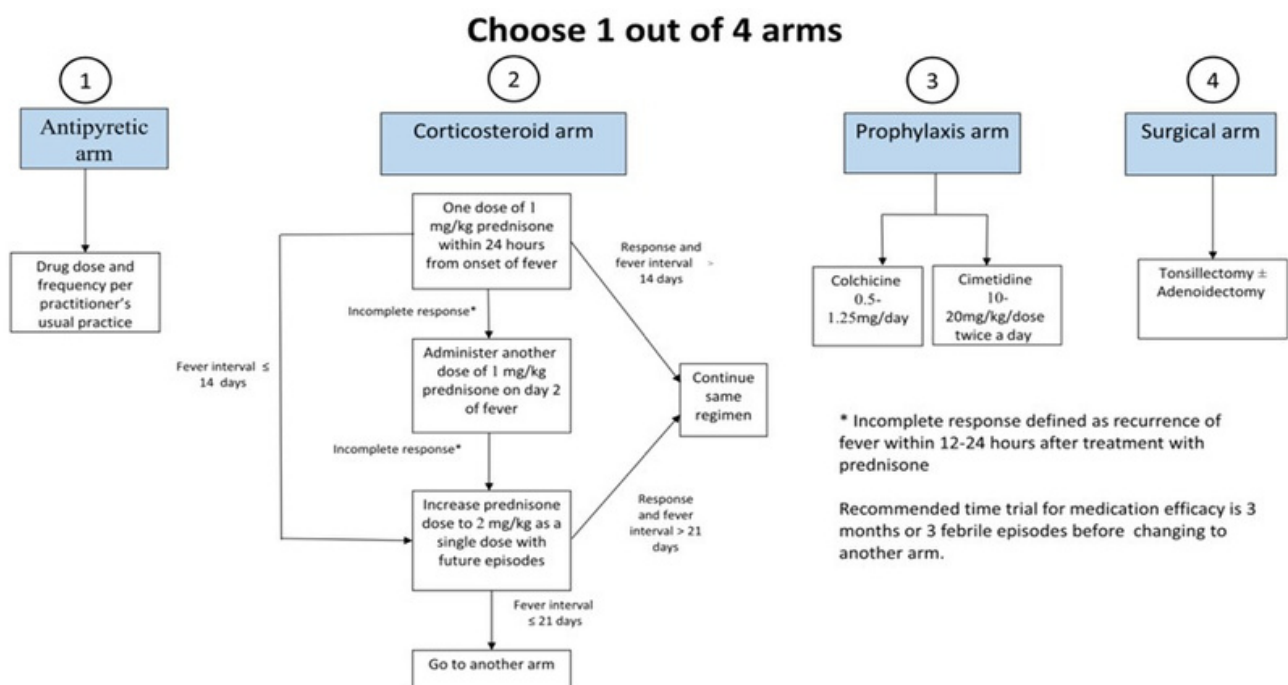
ACADEMIC P.E.A.R.L.S

Pediatric Evidence And Research Learning Snippet



Consensus treatment for periodic fever, aphthous stomatitis, pharyngitis and adenitis syndrome (PFAPA)

Results: The PFAPA working group developed CTPs resulting in 4 different treatment arms: 1. Antipyretic, 2. Abortive (corticosteroids), 3. Prophylaxis (colchicine or cimetidine) and 4. Surgical (tonsillectomy). Consensus was obtained among CARRA members for those defining patient characteristics who qualify for participation in the CTP PFAPA study.



Conclusion: The goal is for the CTPs developed to lead to future comparative effectiveness studies that will generate evidence-driven therapeutic guidelines for this periodic inflammatory disease.

EXPERT COMMENT

“PFAPA is an under recognised condition in office practice. It is not uncommon for children to have undergone a wide array of investigations and multiple courses of antimicrobials before a diagnosis of PFAPA is made. Episodes of fever lasting for 3-6 days with recurrence every 3-8 weeks and associated symptoms such as aphthous stomatitis, cervical adenitis and pharyngitis are suggestive of PFAPA. It usually has a benign and self-limiting course.”

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With warm regards,

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