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Autoinflammatory Diseases with Periodic Fevers

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Abstract

Purpose of Review

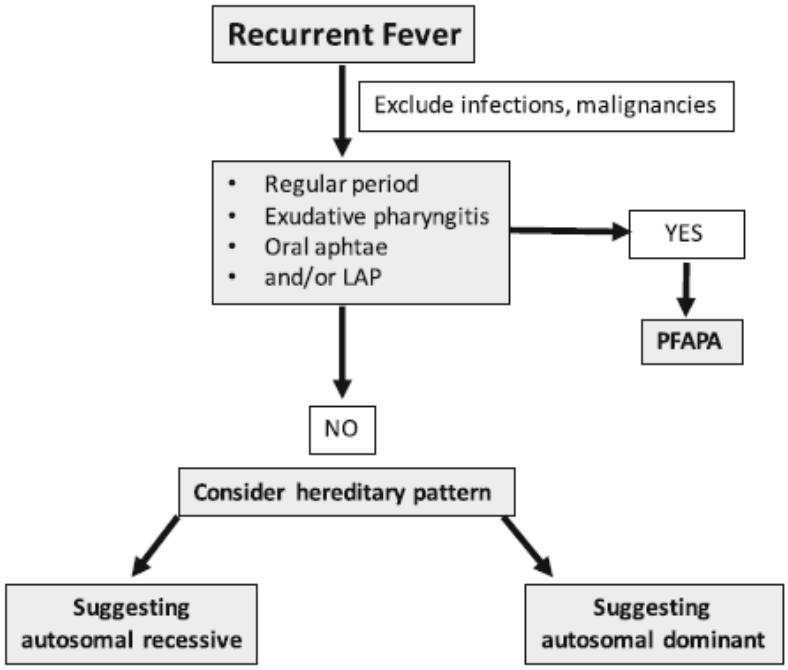
One purpose of this review was to raise awareness for the new autoinflammatory syndromes. These diseases are increasingly recognized and are in the differential diagnosis of many disease states. We also aimed to review the latest recommendations for the diagnosis, management, and treatment of these patients.

Recent Findings

Familial Mediterranean fever (FMF), cryopyrin-associated periodic syndrome (CAPS), tumor necrosis factor receptor-associated periodic fever syndrome (TRAPS), and hyperimmunoglobulinemia D and periodic fever syndrome/mevalonate kinase deficiency (HIDS/MVKD) are the more common autoinflammatory diseases that are characterized by periodic fevers and attacks of inflammation. Recently much collaborative work has been done to understand the characteristics of these patients and to develop recommendations to guide the physicians in the care of these patients. These recent recommendations will be summarized for all four diseases.

Summary

FMF is the most common periodic fever disease. We need to further understand the pathogenesis and the role of single mutations in the disease. Recently, the management and treatment of the disease have been nicely reviewed. CAPS is another interesting disease associated with severe complications. Anti-interleukin-1 (anti-IL-1) treatment provides cure for these patients. TRAPS is characterized by the longest delay in diagnosis; thus, both pediatricians and internists should be aware of the characteristic features and the follow-up of these patients. HIDS/MVKD is another autoinflammatory diseases characterized with fever attacks. The spectrum of disease manifestation is rather large in this disease, and we need further research on biomarkers for the optimal management of these patients.



	HIDS	FMF	CAPS	TRAPS
Onset	<1 yrs (0.1-1.4 yrs)	2.7 yrs (1.1-5.3 yrs)	<1 yrs (0-3 yrs)	4.3 yrs (0-63 yrs)
Attack duration	4 days (3-6 days)	1-3 days	<2 days – continous	>7-14 days (median 10.8 days, 33% less than 7 days)
Trigger	Vaccination, stress, infection	Stress, menstruation, infection	Cold	Stress, menstruation, fatigue, infection, vaccination
Cutaneous	Maculopapular rash	Erysipelas-like erythema	Urticeria-like rash	Migratory rash over myalgia
Musculoskeletal	Arthralgia, myalgia	Monoarthritis	Artralgia, arthritis, abnormal bony growth (NOMID)	Severe myalgia, arthralgia
Abdominal	Severe abdominal pain, diarrhea, vomiting, Splenomegaly	Severe abdominal pain	-	Abdominal pain
Eye	-	-	Conjunctivitis, episcleritis, uveitis, optic disc edema	Periorbital edema, pain, conjunctivitis
Spesificities	Cervical LAP, aphtous ulcers, headche, mental retardation (in severe form)	Erysipelas-like eryhtema, severe serositis (abdominal pain, pleurisy, pericarditis), arhtritis	Urticeria-like rash, chronic aseptic meningitis, sensorineural hearing loss	Migratory myalgia and rash, periorbital edema
Gene	MVK	MEFV	NLRP3	TNFRSF1A
Treatment	Anti-TNF, Anti-IL1	Colchicine, Anti-IL1	Anti-IL1	Anti-TNF, Anti-IL1