

Characteristics of the Main Autoinflammatory Syndromes												
Disease	Onset Age	Typical Ethnicity	Common Triggers	Episode Duration	Interval between Episodes	Serositis	Skin	Musculoskeletal	Eyes	Mucous Membranes	Reticuloendothelial	Neurologic
Familial Mediterranean fever (FMF)	Mostly first decade	Sephardic Jewish Armenian, Arab Moslem Turks, Italian	None, exercise, stress Menstrual period	Hours to 3 days	Variable	Peritonitis, pleuritis pericarditis, scrotum	Erysipelas-like, vasculitis	Acute monoarthritis, 5-10% chronic arthritis, exercise-related myalgia prolonged febrile myalgia	No	No	Splenomegaly, adenopathy	Headaches
Hyperimmunoglobulin D syndrome (HIDS)	First year of life	Dutch, Western Europe	Vaccines, infections	3-7 days	3-6 weeks	Abdominal pain, vomiting, diarrhea	Maculo-papular rash, morbilliform	Arthralgia	No	Aphthous, vaginal sores	Cervical adenopathy	No
Tumor necrosis factor receptor associated periodic syndrome (FRAPS)	Mostly first decade Can start in adults	Any, mainly Scottish, Irish (Hibernian)	Exercise, stress Minor trauma	Days to 6-8 weeks	Variable, usually >8 weeks	Peritonitis, pleuritis pericarditis	Painful, migratory, erythema, vasculitis	Myalgia, fasciitis, arthralgia, arthritis	Periorbital edema	Aphthous	Splenomegaly, adenopathy	Focal
Familial cold autoinflammatory syndrome (FCAS)	First year of life	United States	Cold	<24 hours	Variable	No	Urticaria Like	Arthralgia	Conjunctivitis	No	No	Headaches
Muckle-Wells syndrome (MWS)	Any age	Western Europe	Any	1-3 days	Variable	No	Urticaria Like	Arthralgia	Conjunctivitis, episcleritis, uveitis	No	No	Hearing loss, headaches
Neonatal-onset multisystem inflammatory disorder (NOMID)	Birth, infancy	Any	None	Continuous	None	No	Urticaria Like	Epiphyseal overgrowth with deformities, cartilage defect, arthritis	Conjunctivitis, uveitis, papillitis	No	Adenopathy	Chronic meningitis, hearing loss, mental retardation, headaches
Pyogenic sterile arthritis, pyoderma gangrenosum, acne syndrome (PAPA)	Mostly first decade	Western Europe	Minor trauma	Variable	Variable	No	Pyoderma gangrenosum, acne	Destructive 'pyogenic' large joint arthritis	No	No	No	No
Periodic fever, aphthous stomatitis, pharyngitis, adenitis (PFAPA)	Toddler (1 -4 yrs)	Any	None known	5-7 days	3-4 weeks, truly periodic	No	No	Arthralgia	No	Aphthous, pharyngitis	Cervical	No lymphadenopathy