	Cryopyrin-Associated Periodic Syndromes (CAPS)			Other Autoinflammatory Periodic Fever Syndromes (PFS)		
	NOMID/CINCA	MWS	FCAS/FCU	FMF (Familial Mediterranean Fever)	TRAPS (TNF Receptor Assoc. PFS)	HIDS (Hyper IgD PFS)
GENES & INHERITANCE	*NLRP3/CIAS1/NALP3* Autosomal Dominant Spontaneous mutations, few familial cases.[1]	*NLRP3/CIAS1/NALP3* Autosomal Dominant Spontaneous mutations, some familial groups.[1]	*NLRP3/CIAS1/NALP3* Autosomal Dominant Large familial groups, some spontaneous mutations.[1]	*MEFV* Autosomal Recessive Most common inherited periodic fever syndrome.[1]	*TNFRSF1A* Autosomal Dominant Spontaneous mutations, some familial groups.[1]	*MVK* Autosomal recessive
ETHNICITY	Any–present in all races.[1]	Affects all races, but many of European decent.[1]	Affects all races, but most are of European decent.[1]	Turk, Armenian, Arab, Sephardic Jew, or Italian.[1]	Any–present in many diverse races.[1]	Mostly of Dutch descent, or Northern European.[1]
FREQUENCY OF THE MUTATION IN THE WORLD	Estimated frequency 0.001= 1:1 million=possibly 7,000+ w/ CAPS mutation in world.[5]	(see NOMID) 1:1 million, maybe more due to some family groups.[5]	1:1 million, or more. In USA 300+ diagnosed–most in large family groups.[2,5]	In specific ethnic groups, carrier frequency of *MEFV* variants is up to 1:5 people.[1]	Unknown–over 1000 patients worldwide. (from TRAPS info at http://ghr.nlm.nih.gov	Unknown >200 known patients so far on HIDS registry (www.hids.net).
TIMING OF SYMPTOMS OR ATTACKS (FLARES)	Continuous w/ increased symptoms during flares.[1]	Often lasts 2-3 days, random onset–some pts. flares triggered by cold temperature.[1]	12-24 hours–Onset 1-3 hr. after exposure to cold or cooling temperatures.[1]	12-72 hours[1,9]	Days to weeks. Average flare is 3 weeks.[1,9]	3-7 days, recurrent bouts every 2-12 weeks.[1,9] Some flares occur after vaccines.[9]
AGE OF ONSET	Neonatal/early infancy. Rash, symptoms, abnormal labs often at birth.[1,6]	Infancy, but a few present w/ symptoms later in childhood or adolescence.[1]	Infancy[1]	Infancy, to under 20 years of age for the first symptoms.[9]	Most first attacks by 3 yrs, almost all begin by 20 yrs of age, a few start later in life.[9]	>90% present with symptoms in infancy.[9]
SYSTEMIC FINDINGS						
SKIN/CUTANEOUS	Ever present[1] Urticaria-like rash w/increased neutrophils at the eccrine coils. Rash increases w/ flares.[4]	Urticaria-like rash w/ increased neutrophils at the eccrine coils.[4] Almost daily rash–increases w/ flares.[1]	Cold induced Urticaria-like rash w/ increased neutrophils at the eccrine coils.[4] Many w/ daily rash.[1]	Erysipeloid erythema on the ankle–foot–below knee region, lasts 2-3 days during flares of symptoms.[1]	Migrating rash w/ deep pain under rash areas. Severe pain follows the rash path from the trunk out to limbs.[9]	Diffuse maculopapular rash. Some w/ petechiae or purpura present. A few w/ apthous ulcers.[1,9]
NEUROLOGIC	Headaches, fever, chronic aseptic meningitis, high CNS pressure. Many w/ mental &/or cognitive impairments, papilledema common.[6]	Some have headaches with fever & flares. Uncommon to have many other CNS symptoms.[1]	Some have headaches with fever after cold exposure. Unknown if there are notable CNS affects at this time.[1]	Fevers. Acute aseptic meningitis is rare and can occur during flares, but is never chronic.[1]	Fevers lasting >3 days at over 38º C w/ flares. Some have headaches w/ flares of symptoms.[1,9]	Headaches & fevers w/ flares of symptoms are common.[1,9] More severe neurological symptoms are rarely present in HIDS.[9]
AUDITORY	Many have increased sensorineural hearing loss, from infancy/childhood.[1,6]	Many have increased sensorineural hearing loss, starting in adolescence.[1]	Some pts have mild hearing loss–not currently known if it's from CAPS inflammation.[1]	Uncommon–not believed to be caused by FMF disorder.[1]	Uncommon–not believed to be caused by TRAPS.[1]	Uncommon–not believed to be caused by HIDS.[1,9]
OPHTHALMIC	Papilledema, uveitis, iritis, conjunctivitis. Some w/ corneal haze or vision loss.[6]	Conjunctivitis (non-infectious) episcleritis during flares, or corneal haze.[1]	Conjunctivitis (non-infectious) during flares.[1]	Very rare to uncommon.[1]	Conjunctivitis, & periorbital edema during flares.[1,9]	Very rare to uncommon.[9]
CARDIO-PULMONARY	Some cases of pericardial effusions, or pericarditis. [1]	Rare[1]	Not noted[1]	45% have pleuritis, painful respiration,w/ flares. Some w/ pericarditis.[1]	Common, including pleurisy[1]	Rare[1]
ABDOMINAL	Nausea & vomiting, pain w/ flares, or high CNS pressure.[6]	Some have abdominal pain w/ flares or other GI issues.[1]	Uncommon[1]	Sterile peritonitis, pain, &/or constipation w/flares.[1]	Peritonitis, diarrhea, & constipation w/ flares.[1]	Extreme pain, vomiting & diarrhea w/ flares.[1,9]
LYMPHATIC	Some pts. w/enlarged liver and/or spleen, many have large lymph nodes.[1]	Rarely noted[1]	Not noted[1]	Enlarged spleen is common, some have enlarged lymph nodes.[1]	Enlarged spleen common, some have enlarged lymph nodes.[1]	Enlarged cervical lymph nodes common in children.[1]
JOINTS/BONES MUSCLES & CARTILAGE	Joint pain, knee valgus or varus. Some w/ frontal bossing, saddleback nose, contractures clubbing [1] <50% of pts. knees have bony overgrowth.	Arthralgias, stiffness & swelling with flares.[1]	Arthralgias, stiffness & swelling with flares.[1]	Mono/Polyarthritis, oligoarthritis & clubbing common. Ankle arthralgias common. Severe arthritis of the hip or ankle is rare.[1]	Intermittent or chronic arthritis in large joints,w/ muscle pain & swelling.[1]	Arthralgias common, symmetric polyarthritis frequently noted.[1]
VASCULITIS	Vasculitis rarely develops.[1]	Not noted[1]	Not noted[1]	HSP, polyarteritis nodosa.[1]	HSP, lymphocytic vasculitis.[1]	Cutaneous vasculitis common, HSP is rare.[1]
AMYLOIDOSIS	Elevated SAA Amyloidosis in <2% pts.[1,6]	Elevated SAA >25 % w/amyloidosis.[1,9]	Elevated SAA Amyloidosis in some pts.[1,9]	Common >50% in untreated pts., depends on genotype.[9]	10-20% occurrence–higher risk w/ cysteine mutation.[9]	<10%, not common.[9]
ABNORMAL LABS	Chronically high: ESR, CRP, SAA, anemia, granulocyte leukocytosis.[1,6]	High: ESR, CRP, SAA. Leukocytosis, w/ flares.[1]	High: ESR, CRP, SAA. Leukocytosis w/ flares.[1]	High: ESR,CRP, SAA between flares. Fibrinogen, Leukocytosis present w/ flares.[1]	High: ESR, CRP, SAA. Elevated PMNs, polyclonal gammopathy, leukocytosis.[1]	High: ESR,CRP, SAA w/ flares. High IgD w/ IgA in 80% pts. Mevalonate aciduria noted.[1]

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