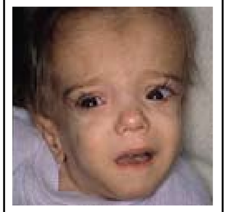
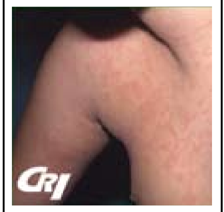
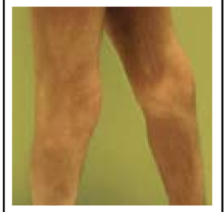
Comparison Chart of Systemic Autoinflammatory Diseases (SAID)

	Cryopyrin-Associated Periodic Syndromes (CAPS)				Pyrin
	Familial Cold Autoinflammatory Syndrome*	Muckle-Wells Syndrome*	Neonatal-Onset Multisystem Autoinflammatory Disease—aka Chronic Infantile Neurological Cutaneous Articular Syndrome (CINCA)*	Schnitzler Syndrome	Familial Mediterranean Fever*
ACRONYM	FCAS	MWS	NOMID/CINCA	SCHNITZLER	FMF
GENE	*NLRP3*	*NLRP3*	*NLRP3*	Currently unknown. (Some pts. w/somatic *NLRP3* mutations.)[76]	*MEFV*
INHERITANCE	Autosomal Dominant. Large familial groups, some spontaneous mutations.[1]	Autosomal Dominant. Spontaneous mutations, some familial groups.[1]	Autosomal Dominant. Spontaneous mutations, few familial cases.[1]	Unknown.	Autosomal Recessive. Some cases are gene-dosage-dependent autosomal dominant.[10]
ETHNICITY	Affects all races, but many are of European descent.[1]	Affects all races, but many are of European descent.[1]	Any–present in all races.[1]	Affects all races, but most cases are in Europe.[13] More men than women are affected.	Turk, Armenian, Arab, Sephardic Jew, Italian.[1] Most common inherited periodic fever syndrome.
FREQUENCY IN THE WORLD	1:1 million, or more. In USA 300+ diagnosed–most cases are from large family groups.[2,5]	1:1 million, maybe more. Some large family groups.[5] Frequency of CAPS in France is 1:360,000.[55]	Estimated frequency 1:1 million, mostly due to spontaneous genetic mutations.[5]	Unknown. Over 150 known cases, mostly in Europe.[13]	In specific ethnic groups, the carrier frequency of *MEFV* variants is up to 1:5 people.[1]
TIMING OF SYMPTOMS OR ATTACKS	12-24 hours, or longer. Onset of fever & flares is often 1-3 hours after exposure to cold or cooling temperatures.[1]	Often lasts 2-3 days. Random onset–flares of fever & symptoms are often triggered by cold or cooling temperature.[1]	Continuous w/increased symptoms & fever during flares.[1] Chronic inflammation noted between flares.	12-36 hours. Rash is present first. Intermittent fevers, that often occur separately from the rash.[13]	12-72 hours.[1,9] Recurrent fever & flares can occur weekly, or only a few times a year.
AGE OF ONSET	Infancy, but a few present w/symptoms later in childhood or adolescence.[1]	Infancy, but a few present w/symptoms later in childhood or adolescence.[1]	Neonatal/early infancy. Rash, symptoms, & abnormal labs are often present at birth.[1,6]	Most cases start in middle age, over 35-50 yrs. Youngest pt. was 13 yrs old. Symptoms start w/the rash.[13]	Infancy, to under 20 years of age for the first symptoms.[9]
SYSTEMIC FINDINGS:					
SKIN/ CUTANEOUS	Cold induced urticaria-like rash w/increased neutrophils at the eccrine coils.[4] Almost daily rash that increases w/flares.[1]	Urticaria-like rash w/increased neutrophils at the eccrine coils.[4] Most w/daily rash that increases w/flares.[1]	Ever-present[1] Urticaria-like rash w/increased neutrophils at the eccrine coils. Rash increases w/flares.[4]	Maculopapular rash, & plaques (sometimes itchy) on the chest & limbs. Dermis has neutrophillic infiltrate. Dermographism.[13]	Erysipeloid erythema on the ankle–foot–below knee region–lasts 2-3 days during flares of symptoms.[1]
NEUROLOGIC	Some have headaches, fatigue w/fever after cold exposure. Unknown if there are notable CNS affects at this time.[1]	Some have headaches, fatigue w/fever & flares. Uncommon to have many other CNS symptoms.[1] A few pts. have MWS/NOMID crossover of symptoms.	Headaches, fever, fatigue, chronic aseptic meningitis, & high CNS pressure (ICP). Many with mental &/or cognitive impairments. Papilledema is common.[6]	Intermittent fevers can rise > 40°C. Chills are uncommon. Fatigue & headaches are common w/fevers. Temperature changes, stress & exercise can trigger flares.[13]	Fevers. Acute aseptic meningitis is rare & can occur during flares, but is never chronic.[1] Other neurological involvement is very rarely seen in FMF.
AUDITORY	Some pts have mild hearing loss–not currently known if it's from CAPS inflammation.[1]	Many have increased sensorineural hearing loss, starting in adolescence.[1]	Many have increased sensorineural hearing loss, from infancy/childhood.[1,6]	Uncommon.[13]	Uncommon–not believed to be caused by a FMF disorder.[1]
OPHTHALMIC	Conjunctivitis (non-infectious) during flares.[1]	Conjunctivitis (non-infectious) during flares,[1] or corneal haze.[26] MWS/NOMID crossover pts. may have more eye involvement.	Papilledema, uveitis, iritis, conjunctivitis. Some w/retinal scarring, corneal haze or vision loss.[6,26]	Not noted.[13]	Very rare to uncommon.[1]
CARDIO-PULMONARY	Not noted.[1]	Rare.[1]	Some have clubbing of fingers. Some cases of pericardial effusions, or pericarditis.[1]	Not noted.[13]	45% have pleuritis, painful respiration, w/flares. Some w/pericarditis.[1]
ABDOMINAL	Uncommon.[1]	Some have abdominal pain w/flares or other gastrointestinal issues.[1]	Nausea, vomiting & abdominal pain w/flares, or w/high CNS pressure.[6]	GI symptoms are uncommon. Enlarged liver &/or spleen is common.[13]	Sterile peritonitis, pain, and/or constipation with flares.[1]
LYMPHATIC	Not noted.[1]	Rarely noted.[1]	Some pts. with enlarged liver and/or spleen, many have enlarged lymph nodes.[1]	<20% w/lymphoma, IgM myeloma, or Waldenströms. >45% w/enlarged lymph nodes.[13]	Enlarged spleen is common, some have enlarged lymph nodes.[1]
JOINTS/BONES MUSCLES & CARTILAGE	Arthralgias, stiffness & swelling with flares.[1]	Arthralgias, recurrent arthritis, stiffness & swelling with flares.[1]	Joint pain, knee valgus or varus. Some w/frontal bossing, saddleback nose, contractures, clubbing.[1] <50% of patients knees have bony overgrowth. Short stature, growth delays failure to thrive, arthritis, & osteopenia noted.[1,26]	80% have muscle, bone &/or joint pain; arthritis. Bone pain is most common in the iliac and tibia. <40% have bone lesions. Some w/osteocondensation & sclerotic bone marrow involvement in the legs.[13]	Mono/Polyarthritis, oligoarthritis & clubbing are common. Ankle arthralgias are common. Severe arthritis of the hip or ankle is rare.[1]
VASCULITIS	Not noted.[1]	Not noted.[1]	Vasculitis rarely develops.[1]	Vasculitis noted in 20% of patients.[13]	HSP, polyarteritis nodosa.[1]
AMYLOIDOSIS	Elevated serum amyloid (SAA). Secondary amyloidosis in some patients.[1,9]	Elevated SAA. >25 % w/secondary amyloidosis.[1,9]	Elevated SAA. Secondary amyloidosis in <2% pts.[1,6]	A few patients have developed secondary amyloidosis.[13]	Common >50% in untreated patients, it depends on genotype.[9]
ABNORMAL LABS	High: ESR, CRP, SAA. Leukocytosis with flares.[1]	High: ESR, CRP, SAA. Leukocytosis,with flares.[1]	Chronically high: ESR, CRP, SAA, anemia, granulocyte leukocytosis.[1,6]	Monoclonal IgM &/or IgG gammopathy. High: ESR, CRP. Leukocytosis. Complement normal to elevated. 50% w/inflammatory anemia.[13]	High: ESR, CRP, SAA between flares. Fibrinogen, Leukocytosis present with flares.[1]
	The rash can vary in size, & intensity in all forms of CAPS. (NOMID Alliance pt. image)	The CAPS rash is often more pronounced during flares. (NOMID Alliance pt. image)	NOMID pt. w/rash, frontal bossing, & saddleback nose. (NOMID Alliance pt. image)	Schnitzler syndrome: Urticarial skin rash on the arm. (cri-net.com Schnitzler image 6)	FMF: Erysipelas-like erythema around the ankle. (Arthritis Research & Therapy 2009 11:212)

	Protein Folding	Mevalonate Kinase Deficiencies		Inflammatory Bone Diseases	
	Tumour Necrosis Factor (TNF)-Associated Periodic Sydrome—aka Familial Hibernian Fever*	Hyperimmunoglobulinemia D with Periodic Fever Syndrome (HIDS)*	Mevalonate Aciduria (MA) (Mevalonate Kinase Deficiencies, such as HIDS & MA are also referred to as MKD)	Deficiency of Interleukin-1ß (IL-1ß) Receptor Antagonist (DIRA)—aka Osteomyelitis, Sterile Multifocal w/Periostitis Pustulosis	Majeed Syndrome—aka Chronic Recurrent Multifocal Osteomyelitis, Congenital Dyserythropoietic Anemia, & Neutrophilic Dermatosis
ACRONYM	TRAPS	HIDS	MA	DIRA/OMPP	MAJEED
GENE	*TNFRSF1A*	*MVK*	*MVK*	*IL1RN*	*LPIN2*
INHERITANCE	Autosomal Dominant. Spontaneous mutations, some familial groups.[1]	Autosomal recessive. Some cases w/only one mutation found.[33]	Autosomal recessive.	Autosomal recessive.	Autosomal recessive.
ETHNICITY	Affects all races. 2nd most common inherited SAID (after FMF.)[1]	Mostly of Dutch descent, or Northern European.[1]	Mostly of Dutch descent, or Northern European.[1]	Carriers in 0.2% population of Newfoundland & 1.3% in Puerto Rico. Also Dutch Brazilian & Lebanese pts.[16]	Currently, the only documented cases are of Middle Eastern ancestry.[18]
FREQUENCY IN THE WORLD	Unknown. TRAPS affects 0.01:10,000 people in the European Union.[51] >1000 pts. worldwide.[52]	Unknown, but very rare. >200-300 known patients worldwide, (>300, when suspected cases are also included.)[12]	Unknown, but very rare. <100 known patients worldwide.[11]	Unknown, but very rare. In some regions of Aricibo, Puerto Rico w/more DIRA carriers, DIRA may occur 1:6300.[16]	Unknown, but very rare. Very few documented cases at this time.[18,53]
TIMING OF SYMPTOMS OR ATTACKS	Days to weeks. Average flare is 3 weeks.[1,9]	3-7 days. Recurrent bouts of fever & flares every 2-12 weeks.[1,9] Some flares occur after vaccines.[9]	4-5 days. Recurrent flares & fever every 2-3 weeks. Patients have chronic inflammation noted between flares.[11]	Continuous inflammation from birth/fetal development. Untreated DIRA can lead to death in infancy–childhood.[16]	Flares last for a few days, w/1-4 exacerbations a month of high fevers, severe pain, & joint swelling.[18,53]
AGE OF ONSET	Most first attacks by 3 yrs, & almost all begin by 20 yrs. of age; a few start later in life.[9]	>90% present w/symptoms in infancy.[9]	Most present w/symptoms at birth, or in early infancy. Most have facial features noted at birth.[11]	Most have symptoms at birth, or as a neonate: pustular rash, bone pain, swollen joints, & oral ulcers.[16]	Most present w/symptoms in infancy to early childhood, between 3 weeks to 2 years of age.[18]
SYSTEMIC FINDINGS:					
SKIN/ CUTANEOUS	Migrating rash w/deep pain under rash areas. Severe pain follows the rash path from the trunk out to the limbs.[9]	Diffuse maculopapular rash. Some w/petechiae or purpura present. A few w/apthous ulcers.[1,9]	Diffuse maculopapular or morbilliform rash. Some w/petechiae or purpura present. A few w/apthous ulcers.[1,9,11]	Epidermal neutrophilic pustules at hair folicles. Oral ulcers, pathergy, hyperkeratosis, acanthosis; high neutrophil infiltrate of dermis.[16,26]	Most patients have inflammatory dermatosis, Sweet's syndrome, pustular skin lesions, psoriasis. Intra-epidermal neutrophils.[18,53]
NEUROLOGIC	Fevers lasting >3 days at over 38°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]	Fevers w/flares. Microcephaly, dolichocephaly, mental retardation, developmental delays, cerebellar ataxia, cerebellar atrophy & epilepsy often develop over time.[11]	High fevers are not common, or noted in the neonatal period. Neurological complications are not common. A few cases of cerebral vasculitis noted.[16,26]	High fevers last for a few days w/flares & severe pain. Other neurological symptoms are not noted. Growth delays in height, & chronic pain are common.[18,53]
AUDITORY	Uncommon–not believed to be caused by TRAPS.[1]	Uncommon–not believed to be caused by HIDS.[1,9]	Uncommon–not believed to be caused by MA.[1,9,11]	Not noted.[15,16]	Not noted.[18,53]
OPHTHALMIC	Conjunctivitis, & periorbital edema during flares.[1,9]	Very rare to uncommon.[9]	Uveitis, central cataracts, blue sclerae & tapetoretinal degeneration are often present, even in less severe cases.[11]	Eye issues are rare. Non-infectious conjunctivitis can be caused by DIRA.[15,16]	Not noted.[18,53]
CARDIO-PULMONARY	Common, including pleurisy.[1]	Rare.[1] Some pts. have developed severe respiratory infections. Higher risk for issues w/*S. pneumoniae* infections.[78]	Rare.[1,11]	Some w/resp. distress. 1 case: Pulmonary hemosiderosis & progressive interstitial fibrosis.[15,16,17]	Not noted.[18,53]
ABDOMINAL	Peritonitis, diarrhea, & constipation w/flares.[1]	Extreme pain, vomiting & diarrhea w/flares.[1,9] Some w/enlarged liver/spleen, other GI issues.[78]	Enlarged liver &/or spleen. Cholestatic liver disease. Pain, vomiting & diarrhea w/flares.[1,9,11]	Rarely have GI issues. Mouth ulcers, stomatitis, & failure to thrive are common.[16]	Enlarged liver & cholestatic jaundice in the neonatal period, but it is transient.[18,53]
LYMPHATIC	Enlarged spleen is common; some have enlarged lymph nodes.[1]	Enlarged cervical lymph nodes w/flares.[1] Few w/enlarged spleen.[78]	Enlarged spleen, &/or lymph nodes are common.[1,11]	Enlarged liver and/or spleen is common. Risk of organ failure if untreated.[16]	Neonates: enlarged liver & neutropenia; anemia is common–can be severe.[18]
JOINTS/BONES MUSCLES & CARTILAGE	Intermittent or chronic arthritis in large joints w/muscle pain & swelling.[1]	Arthralgias common, symmetric polyarthritis frequently noted.[1]	Congenital defects are often noted: microcephaly, dolichocephaly, wide irregular fontanels, low set and posteriorly rotated ears, downslanted palpebral fissures. Hypotonia, myopathy, & failure to thrive are common.[11]	Joint swelling, severe bone pain. Bone biopsy shows no infection. Common: Balloon-like widening of the anterior rib ends, periosteal elevation along multiple long bones, multifocal osteolytic lesions. Other bones affected.[16]	Periarticular tender soft tissue swelling. Bone biopsy shows no infection. Early-onset Chronic Recurrent Multifocal Osteomyelitis (CRMO), periarticular tender soft tissue swelling, short stature, delayed bone age, contractures.[18]
VASCULITIS	HSP, lymphocytic vasculitis.[1]	Cutaneous vasculitis common, HSP is rare.[1]	Not noted.[11]	A few w/localized or cerebral vasculitis.[16]	Not noted.[18,53]
AMYLOIDOSIS	10-20% occurrence–higher risk w/cysteine mutation.[9]	<5-10%–uncommon.[9]	Not noted-unknown.[9,11]	Not noted.[15,16,17]	Not noted.[18,53]
ABNORMAL LABS	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 during flares.[1]	Anemia, leukocytosis, thrombocytopenia. High: ESR, CRP, SAA, CK, IgD, IgA. IgE; chronically high Mevalonate aciduria.[1,11]	High: ESR, CRP, leukocytosis, chronic anemia.[16,56]	Congenital dyserythropoietic anemia (CDA). High ESR. WBC can be normal, or elevated–neutropenia in infancy. Cultures negative.[18]
	TRAPS rash on the chest of a child. (Swiss Med Wkly. 2012;142:w13602)	HIDS: Maculopapular rash on the hand. (cri-net.com Auto Inflammatory Diseases image 23)	 MA: 21 month old pt. w/ facial dysmorphism. (Orphanet Journal of Rare Diseases 2006 1:13)	DIRA: Generalized pustulosis. (NOMID Alliance pt. image)	Sweet's Syndrome is often present w/Majeed. (pbase.com/leobarco/image/69914524)

		Pyogenic Diseases			Granulomatous
	Chronic Recurrent Multifocal Osteomyelitis—aka Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis Syndrome	Deficiency of Interleukin-36-Receptor Antagonist (DITRA)—aka Generalized Pustular Psoriasis (GPP)	Familial Psoriasis (PSORS2)—aka CARD14-Mediated Pustular Psoriasis	Pyogenic Sterile Arthritis, Pyoderma Gangrenosum, & Acne Syndrome	Juvenile Systemic Granulomatosis—aka Blau syndrome, Pediatric Granulomatous Arthritis (PGA), Early Onset Sarcoidosis, or Jabs Syndrome
ACRONYM	CRMO/SAPHO	DITRA/PSORP	CAMPS/PSORS2	PAPA	BLAU/PGA/EOS
GENE	Currently unknown.	*IL36RN*	*CARD14*	*PSTPIP1*	*NOD2*
INHERITANCE	Currently unknown.	Autosomal recessive.	Autosomal Dominant. Spontaneous mutations, some familial groups.[23]	Autosomal Dominant. Spontaneous mutations, some familial groups.[29,30]	Autosomal Dominant.
ETHNICITY	Affects all races, but the majority of patients have European ancestry; more female pts. than males.[21,22]	May affect all races. Pts. w/Caucasian, Spanish, Asian, African, Algerian & Tunisian ancestry.[58,75,77]	Most w/European or Asian ancestry. Pts. in US, EU, Canada (Newfoundland), Haiti, & Taiwan.[23]	Currently, the only documented cases are from Europe, New Zealand & the USA.[30]	Affects all races.
FREQUENCY IN THE WORLD	Unknown, but rare.	Unknown but rare. 1% of Sfax, Tunisians are carriers, w/ a 0.52% chance of having the disease in this population.[58]	Unknown, but rare.	Unknown, but rare.	Unknown, but rare.
TIMING OF SYMPTOMS OR ATTACKS	At least 6 months w/chronic or relapsing symptoms. Often 7-25 yrs. of symptoms. Many bone lesions heal completely.[19,22]	Flares last days–weeks. Some w/chronic symptoms. Most flare w/infections, stress, medication changes, during pregnancy or menstruation.[58]	Continuous chronic pustular or plaque psoriasis, triggered by inflammatory stimuli. Some cases w/psoriatic arthritis.[23,24]	Early-onset, destructive, recurrent inflammation of the joints, skin & muscle. Flares often occur after mild injury, or injections.[29]	Intermittent-persistent daily fevers, rash & arthritis.
AGE OF ONSET	Mostly affects children–some adult onset. Peak incidence of flares is around 10 years of age.[22]	Variable age of onset. Many have symptoms starting in childhood. Some have symptoms beginning in adulthood.[58]	Variable age of onset from infancy–childhood to adulthood w/pustular psoriasis.[23,24]	First symptoms of arthritis develop by 1-10 yrs old, & skin lesions develop during adolescence.[29,32]	Rash often develops by 4 months of age, fevers and other symptoms present by 4 yrs. of age.[34]
SYSTEMIC FINDINGS:					
SKIN/ CUTANEOUS	Some patients have acne, &/or pustulosis on the palms &/or soles of their extremities (w/ SAPHO). 23% w/psoriasis.[19,22,54]	Recurrent, generalized pustular psoriasis & high fevers after erythematous rash. Some w/acral pustules & nail damage, or chronic plaques.[58,61]	Generalized pustular psoriasis (can be severe), &/or plaque psoriasis. Sometimes nails are affected w/ psoriasis.[23, 24]	Pathergy. Pyoderma gangrenosum ulcerative lesions, &/or severe cystic acne. Affected tissues w/high neutrophil infiltration.[29]	First symptom: scaly plaques. The rash often starts on the face, then on the torso. Biopsies w/ non-caseating granulomatous dermatitis.[34]
NEUROLOGIC	Fevers affect a number of patients during flares of CRMO. Other neurological symptoms are not noted. Some w/impaired bone growth, or overall impaired growth.[19,22,54]	Sudden onset high fever >40°C w/chills. Some pts. have a headache w/the onset of the rash & fever, plus muscle weakness & elevated heart rate.[58,61]	Not seen.[23, 24]	Fevers can accompany flares of joint inflammation and pain. Other neurological symptoms are not noted.[31]	Intermittent-persistent daily fevers. Some have cranial neuropathies. 80% have vision damage & joint deformities if untreated. Some cases have peripheral nerves affected.[34]
AUDITORY	Not noted.[19,21,22,54]	Not noted.[58,61,62]	Not seen.[23, 24]	Not noted.[29,30,31]	Not noted.[34]
OPHTHALMIC	Some cases of uveitis.[19]	Not noted.[58,61,62]	Not seen.[23, 24]	Not noted.[29,30,31]	Uveitis (some w/blindness) 50% w/cataracts, 1:3 pts. get 2° glaucoma, inflamed conjunctiva, lacrimal glands, retina & optic nerves.[34]
CARDIO-PULMONARY	Not common–some patients also have ANCA+ Vasculitis that can affect the lungs.[18,54]	Elevated heart rate. Electrolyte imbalances during fever & onset of pustular rash; Risk for cardiac arrest, & septicemia.[58,61]	Not noted.[23, 24]	Not noted.[29,30,31]	Some have atrial hypertension &/or pericarditis. Some cases with lung involvement.[34,35]
ABDOMINAL	Some patients also have inflammatory bowel diseases.[19]	Nausea during flares. At risk for loss of appetite.[58,60] Infant case w/failure to thrive, diarrhea.[63]	Not noted.[23,24]	Some patients also have irritable bowel syndrome.[29]	Enlarged liver &/or spleen. Some w/ GI pain, higher risk for kidney &/ or liver issues.[34,35,36]
LYMPHATIC	Some cases of ANCA+ Vasculitis that can affect the kidneys.[19]	Risk for renal and liver impairment & systemic infection w/severe flares.[58,61]	Not seen.[23, 24]	Not noted.[29,30,31]	Enlarged liver &/or spleen, enlarged lymph nodes.[34,35,36]
JOINTS/BONES MUSCLES & CARTILAGE	Joint swelling, limp, severe bone pain over affected bones (mostly long bones). 2-18 bone lesions are commonly found. Earlier age of onset & many bone lesions=more severe disease. Bone biopsy/cultures show no infection.[19,22]	Muscle weakness during fevers & flares. Risk for inflammatory arthritis.[59,61]	Intermittent joint pain, psoriatic arthritis. 30% of affected patients in one European family w/ PSORS2 also had psoriatic arthritis.[24]	Episodic inflammatory arthritis, often to one joint at a time that doesn't resolve on it's own. Intermittent sterile pauciarticular, peripheral erosive arthritis. Joint damage & destruction can often develop from the arthritis.[29,30,31,32,55]	Symmetrical chronic polyarthritis or oligoarthritis of the wrists, knees, ankles w/ a boggy appearance is usually caused by an exuberant tenosynovitis.[34,35,36]
VASCULITIS	Some w/Takayasu arteritis, or ANCA+ Vasculitis.[54]	Not noted.[59,60]	Not seen.[23, 24]	Not noted.[29]	Some w/vasculitis, leukocytoclastic vasculitis.[34]
AMYLOIDOSIS	Not noted.[19,22,54]	Not noted.[59,60]	Not noted.[23, 24]	Not noted.[29]	Not noted.[34]
ABNORMAL LABS	Whole body MRI can reveal multifocal bone lesions.[20] Normal or elevated WBC, ESR, CRP.[19,22,54]	High during flares (most pts.): ESR, CRP, neutrophils, lactate levels, Low: plasma albumin, calcium, zinc. Risk for infections w/flares.[59,60]	Mildly elevated WBC, CRP & ESR rarely elevated–only during flares of symptoms.[56]	Cultures of bone & skin are negative. Purulent synovial fluid full of neutrophils. High w/flares: CRP, ESR, WBC.[29,30,32]	High CRP & ESR, ACE, immunoglobulins. Anemia, leukopenia, eosinophilia, hematuria, proteinuria, pyuria, abnormal LFTs (LF).[34,36]
	Palmar pustulosis SAPHO (CRMO). (cri-net.com SAPHO Syndrome image 29: Spondylarthropathies)	3 yr. old w/generalized pustular psoriasis. (dermis.net/dermisroot/en/32493/image.htm)	Pustular psoriasis–seen w/ CAMPS. (dermatlas.med.jhmi.edu/image/pustular_psoriasis_1_061124)	PAPA: Pyoderma gangrenosum. (dermatlas.med.jhmi.edu/image/pyoderma_gangrenosum_1_020918)	Blau syndrome: Reddish-brown papules. (Dermatology Online Journal 15 (12): 5)

	Monarch-1	Proteasome	Idiopathic		Macrophage Activation Diseases
	NLRP12-Associated Periodic Fever Syndrome—aka Familal Cold Autoinflammatory Syndrome 2, or Guadaloupe Periodic Fever	Chronic Atypical Neutrophilic Dermatosis w/ Lipodystrophy & Elevated Temperature—aka Nakajo-Nishimura Syndrome	Behçets Disease	Periodic Fever, Aphthous Stomatitis, Pharyngitis, & Cervical Adenitis (PFAPA)—aka Marshall Syndrome	Systemic-Onset Juvenile Idiopathic Arthritis—aka Still's, Systemic Juvenile Idiopathic Arthritis
ACRONYM	NLRP12/FCAS2	CANDLE/PRAAS	BEHÇETS/BD	PFAPA	soJIA/sJIA
GENE	*NLRP12*	*PSMB8; also PSMB4, PSMB9, PSMA3, POMP*[59]	*ERAP1 (with HLA-B51);* also variants near: *CCR1, KLRC4, STAT4*[42]	Currently unknown.	Currently unknown. *HLA-DRB1* in some pts. w/European ancestry[57]
INHERITANCE	Autosomal Dominant. Spontaneous mutations, some familial groups.[29,30]	Autosomal recessive.	Complex.	Currently unknown.	Complex.
ETHNICITY	Unknown. Cases in Guadeloupe, US, Martinique, France, Italy, & Armenia.[38,39,75]	Unknown. Caucasian, Hispanic, Japanese pts. & one case in South Africa.[27,60]	Rare in the USA. More common in the Middle East, Asia & Japan. (Silk Road Route.)[42,43]	Affects all races.[40]	Affects all races. soJIA accounts for 10% of all JIA.[45]
FREQUENCY IN THE WORLD	Unknown, but rare.	Unknown, but rare.	Prevalence is 80-370:100,000 people in Turkey, 10:100,000 in Japan & 0.6:100,000 in Yorkshire, UK.[3]	Unknown. Most common non-infectious recurrent fever disorder.[40]	Uncommon. 0.4–0.9 cases per 100,000 people, per year.[46]
TIMING OF SYMPTOMS OR ATTACKS	1-3, to up to 7-15 days of fevers 39–40°C, rash & pain. Onset after exposure to cold or cooling temperatures.[38,39]	Frequent fevers w/disease flares. Inflammatory flares & symptoms are often present before the age of 6 months.[26,27]	Mouth ulcers are present in almost all patients during flares, for around 10 days w/inflammation in the eye, & arthritis.[42,43]	Periodic fevers & symptoms lasting 3-6 days, recurring every 21-28 days. Pts. are symptom-free between flares.[40]	Fevers often > 39°C 1-2 times/day for >2 weeks, most often occuring in the evening w/arthralgia, rash & other symptoms.[45,46]
AGE OF ONSET	Neonatal/early infancy. Rash, fevers, symptoms, may be present at birth.[38,39]	Onset at birth or in infancy. Progressive damage from chronic inflammation noted as the child grows.[26,27]	Most show symptoms in early adulthood (20's-30's) but the onset can be in childhood, or any age.[42,43]	Early childhood, usually between 2-5 years of age. A few adult-onset cases. Many teens outgrow it.[40]	Onset before the age of 16–most often by 2 years of age, or between 0-5 yrs. of age.[46]
SYSTEMIC FINDINGS:					
SKIN/ CUTANEOUS	Present during flares: Cold-induced urticarial or malar rash[39] noted in some pts. Some with buccal aphthosis.[38,39]	Annular cutaneous plaques w/residual purpura. Lipodsystrophy: first on face & around joints. Lips swell w/flares. Purple-red eyelids.[26,27]	Pathergy. Pseudofolliculitis, erythema nodosum-like &/or acneiform nodules. 98% w/mouth ulcers, & 65% have genital ulcers.[43]	Some have a rash with flares. Aphthous stomatitis, & pharygitis w/ exudate, (but no infection) is a classic finding.[40,41]	Rash: Fleeting, evanescent, migratory, bright salmon-pink, morbilliform, macular rash often presents w/onset of fevers.[45,46]
NEUROLOGIC	Fevers 39–40°C myalgia, headaches w/flares. Other neurological symptoms are not noted.[31]	Aseptic meningitis & systemic inflammation. Growth delays–low height & weight. Developmental delays.[26,27]	20-40% have Neuro-Behçets w/headaches, aseptic meningitis or meningoencephalitis, seizures, hemiplegia, or cranial nerve palsies. Cerebral venous thrombosis w/high ICP noted.[43]	High fevers for 3-6 days, w/chills & malaise. Some patients have headaches w/flares. Other neurological symptoms are not noted.[41]	High fevers >39°C 1-2 times/day for >2 weeks. Other neurological symptoms are rare. A few cases w/seizures, meningismus, irritability & decreased level of consciousness.[46]
AUDITORY	Many have increased sensorineural hearing loss.[38,39]	Some have frequent otitis &/or recurrent sinusitis.[27]	Not noted.[42,43]	Not noted.[40,41]	Not noted to be from soJIA.[45,46]
OPHTHALMIC	Not noted.[38,39]	Nodular episcleritis (inflammation on the eye.) Conjunctivitis. Keratitis. Periorbital edema. Purple-red eyelids.[26,27]	Frequent anterior &/or posterior uveitis. Cataract, retinal vasculitis <30% risk for blindness. Papilledema w/CNS involvement.[43]	Not noted.[40,41]	Uveitis can be a complication from soJIA.[46]
CARDIO-PULMONARY	Not noted.[38,39]	Clubbing of the fingers &/or toes. At risk for cardiac arrythmias & dilated cardiomyopathy.[26,27]	Myocarditis, endocarditis w/aortic or mitral insufficiency, arterial aneurysm, pulmonary embolism.[43]	Flares of fevers, stomatitis & pharygitis are not associated w/respiratory illness.[40,41]	Serositis (especially pericarditis) is often seen. Pleuritis, pleural effusions can occur. Risk for MAS.[46]
ABDOMINAL	Some patients have abdominal pain w/flares.[39]	Loose bowels w/flares. Enlarged liver & abdomen. Delayed or slow growth.[26,27]	Ulcers from mouth to anus. Nausea, abdominal pain, anorexia, diarrhea, (may be bloody).[43]	Abdominal pain, diarrhea are often present w/ flares.[40,41]	Peritonitis rarely occurs. 50% have an enlarged spleen, some w/an enlarged liver.[46]
LYMPHATIC	Some patients with adenopathy.[39]	Enlarged liver, with elevated liver enzymes; enlarged lymph nodes.[26,27]	Some w/enlarged liver &/or spleen; enlarged lymph nodes.[43]	Cervical adenopathy during flares.[40,41]	Many w/generalized lymphadenopathy. Some w/mesenteric adenitis.[45,46]
JOINTS/BONES MUSCLES & CARTILAGE	Myalgia, arthralgia, fatigue & malaise w/flares. Permanent bone or joint damage not noted.[39]	Joint Contractures, muscle atrophy, panniculitis induced lipodystrophy, myositis, fatigue and malaise. Inflammed nose & ear cartilage (chondritis). Growth delays–low height & weight.[26,27]	45% have arthralgias &/or arthritis–often the knees &/or ankles, but other joints can be affected. May be the first sign of Behçets. X ray is normal but synovium often has high neutrophils or mononuclear cells & a vasculitis process.[43]	Arthralgias, fatigue and malaise. No permanent joint or bone issues noted, and patients are symptom-free between PFAPA flares.[40,41]	Arthralgias may come before the arthritis. 88% have polyarticular or oligoarticular arthritis, most often in the wrists, knees, &/or ankles. Some w/cervical spine, hip, temporomandibular joint arthritis or synovial cysts.[45,46]
VASCULITIS	Not noted.[38,39]	Not noted.[26,27]	Extensive vasculits. 30% w/venous thrombosis.[43]	Not noted.[40,41]	Not noted.[45,46]
AMYLOIDOSIS	Not noted.[39]	Not noted.[26,27]	Not noted.[42,43]	Not noted.[40,41]	Amyloidosis occurs in 7.4% of pts. in the USA, and 16% in Turkey.[46]
ABNORMAL LABS	Elevated CRP may be noted during flares. But some pts. do not have elevated CRP w/ flares.[39]	Hypochromic or normocytic anemia. High CRP, ESR, triglycerides. Some w/elevated platelets, TSH, &/or LDL.[26,27]	Leukocytosis common. Normal–rarely elevated ESR or CRP. Some cryoglobulinemia, elevated factor VIII, fibrinolysis.[43]	High: ESR, CRP, WBC during flares–normal levels when not flaring.[40,41]	High: ESR, CRP, WBC, SAA, ferritin, aldolase. Elevated LFT's. Leukocytosis, thrombocytosis Anemia.[45,46]
	Urticarial rash w/flares after cold exposure. (NOMID Alliance pt. image)	CANDLE: Child w/facial lipodystrophy, rash & swollen lips. (NOMID Alliance pt. image)	Behçet's Disease: Oral ulcers. (dermis.net/dermisroot/en/17101/image.htm Behçet's disease)	PFAPA: Aphthous stomatitis. (drpaulose.com; blog.timesunion.com/mdtobe Mystery Monday 125)	soJIA: Salmon-pink rash on abdomen. (Joint Bone Spine, Volume 74, Issue 5, Pages 500-503)

			PLCG2-associated		*SLC29A3* related	*ADA2 deficiency*
	Adult-Onset Stills Disease—aka Adult Still's, Wissler-Fanconi Syndrome	(Primary) Familial Hemophagocytic Lymphohistiocytosis—aka Familial Erythrophagocytic Lymphohistiocytosis	PLCG2-associated Antibody Deficiency & Immune Dysregulation, (PLAID)—aka Familial Atypical Cold Urticaria (FACU) or FCAS3	Autoinflammation & PLCG2-associated Antibody Deficiency & Immune Dysregulation (APLAID)	SLC29A3 Spectrum Disorder—aka H. syndrome; Pigmented Hypertrichosis w/IDDM; Faisalabad Histiocytosis & Sinus Histiocytosis w/Massive Lymphadenopathy	Deficiency of Adenosine Deaminase 2 (DADA2)—aka Fever w/Early Onset Stroke (FEOS)
ACRONYM	AOSD	1° HLH/ FHL	PLAID/FCAS3	APLAID	SLC29A3	DADA2
GENE	Currently unknown.	*PRF1, STX11, STXBP2, MUNC13-4 , RAB27A X link: SH2D1A, BIRC4*	Heterozygous genomic deletions within the *PLCG2* gene[64]	*PLCG2* mutation	*SLC29A3*	*CECR1*
INHERITANCE	Currently unknown.	Autosomal recessive, but if X-linked: inheritance is dominant.	Autosomal Dominant.	Autosomal Dominant.	Autosomal recessive.	Autosomal recessive.
ETHNICITY	Rare. Affects all races.[44]	Affects all races. 80% of African Americans, & 20% of pts. w/European decent have *PRF1* mutations.[47,48]	Unknown. Most reported cases w/European ancestry.	Unknown.	Unknown. Many pts. w/ Middle Eastern ancestry. Some from India, Pakistan, Spain, Bulgaria.[70]	Unknown.
FREQUENCY IN THE WORLD	France: estimated that 0.16:100,000 people have AOSD. AOSD affects more women than men.[44]	1° HLH affects 1:50,000 people worldwide.[48]	Unknown but rare.	Unknown but rare.	Unknown but rare.	Unknown but rare.
TIMING OF SYMPTOMS OR ATTACKS	High fevers > 39°C that last for <4 hours, recurring more than once a week, w/ a maculopapular rash & arthralgia.[44]	Fevers often > 39°C 1-2 times/day for >2 weeks, most often occurring in the evening w/arthralgia, rash & other symptoms.[47,48]	Onset <5 minutes after exposure to cold air (evaporative cooling).[64] Frequent sinus/lung infections. Concurrent autoimmune diseases.[64,65]	Recurrent skin lesions, chronic inflammation, & progressive eye complications from this disease.[66,67,68]	Fever 39°C w/flares that last 7-10 days w/joint & abdominal pain; pericarditis & sometimes diarrhea. Flares occur once every 2-3 months.[70]	Intermittent, recurrent fevers, livedo reticularis rash, vasculopathy, & high risk for early-onset lacunar stroke.[73,74]
AGE OF ONSET	First onset of symptoms occurs between 16-35 yrs. of age. Affects all ages.[44]	Onset <1yr: often by 6 months–early childhood. Some in utero or late childhood. A few adult-onset cases.[47,48,49]	Onset in infancy-under 6 months of age. Lifelong symptoms, but some find the symptoms less severe in adulthood.[64,65]	Onset in infancy w/recurrent skin lesions, arthralgia, eye inflammation, & infections. Some w/ intestinal symptoms.[68]	Onset in infancy-starts w/recurrent fevers & flares. Chronic & progressive systemic symptoms develop.[69,70,71]	Onset of symptoms in infancy–early childhood. w/recurrent fevers, livedo reticularis rash, & vasculopathy.[73,74]
SYSTEMIC FINDINGS:						
SKIN/ CUTANEOUS	Evanescent, salmon-pink, mildly pruritic maculopapular rash on the proximal limbs & trunk.[44]	40% w/transient maculopapular, nodular or purpuric skin rashes during bouts of high fever. Jaundice.[47,48]	Cold urticaria, erythema & itching post cold exposure (air, wet skin, cold food). Some w/angioedema; chronic granulomata. Ice cube test negative.[64]	Erythematous plaques & vesiculopustular blistering rash that intensifies w/heat & sun exposure. Cellulitis often develops w/rashes.[66,67]	Hyperpigmentation w/hypertrichosis.[69,70,71,72] Some w/notable varicose veins on the legs.[69]	Livedo reticularis rash, few w/polyarteritis nodosa. Diffuse vasculopathy, w/impaired endothelial integrity & endothelial cellular activation.[73,74]
NEUROLOGIC	>95% have high, spiking fevers, fatigue and myalgia w/flares. Other neurological symptoms are very rarely seen.[44]	High fevers. Increased CSF protein. High ICP. Multifocal inflammation of the gray & white matter, intracranial bleeding, generalized atrophy or brain edema, seizures &/or coma.[47]	Not noted.[64,65] No fevers noted w/cold-induced urticaria.[64,65]	Not noted.[66,67,68] No fevers noted as a part of this disease (but fevers may be present w/infections).[66,67,68]	Fever 39°C w/flares lasting over a week.[69,70] Psychomotor delays, dysmorphic facial features noted.[69,70]	Recurrent fevers & early-onset lacunar strokes. Possible adult stroke risk. Brain biopsies: diffuse vasculopathy, w/impaired endothelial integrity & endothelial cellular activation.[73,74]
AUDITORY	Not noted.[44]	Not noted.[47,48,49]	Not noted.[64,65]	Not noted.[66]	Sensorineural hearing loss, from early infancy/ childhood.[69,70]	Unknown.[73,74]
OPHTHALMIC	Not noted.[44]	Blindness due to CNS inflammation.[48]	Uncommon.[66]	Many develop corneal erosions, blisters, ulcerations, intraocular hypertension, &/or cataracts.[66,67,68]	Uveitis. Blindness can occur from anterior uveitis & glaucoma. Ptosis, eyelid swelling from histiocytic deposits.[69,70,71]	Unknown.[73,74] Strokes have the potential to cause blindness.
CARDIO-PULMONARY	<25% have pleuritis, pericarditis (a few w/tamponade.) Some myocarditis, pleural effusions, ARDS.[44]	High risk for respiratory infections triggering fevers, systemic inflammation & MAS. Edema.[49]	44% w/recurrent sinus &/or respiratory infections, >50% w/allergies, asthma &/or autoimmune diseases.[65]	Mild humoral immune deficiency w/increased frequency of sinus &/or respiratory infections or interstitial pneumonia.[66,67]	Pericarditis w/flares. Cardiac defects noted: ASD, VSD, PDA, mitral valve prolapse, cardiomegaly & others.[69,70]	Unknown.[73,74]
ABDOMINAL	50-75% w/enlarged liver, abnormal LFTs. 43% w/enlarged spleen. Renal disease is rare.[44]	Liver disease is common. High risk of death from multi-organ failure in 2+ months if untreated.[49]	Not noted. Some have concurrent autoimmune diseases that may involve other organs.[65]	Some w/bouts of abdominal pain, bloody diarrhea, enterocolitis, or ulcerative colitis.[67,68]	Diabetes Mellitus. Enlarged liver/spleen. Abd. pain, diarrhea, failure to thrive. Hypogonadism.[69,70]	Enlarged liver & spleen; diffuse vasculopathy noted in the liver.[73,74]
LYMPHATIC	Lymphadenopathy is common. Many w/enlarged liver &/or spleen.[44]	Lymphoma. Hemophagocytosis–spleen/lymph nodes. Enlarged liver &/ or spleen.[49]	Not noted. Some need IVIG for low immunoglobulins & frequent infections. Few w/CVID.[65]	Not noted.[67,68]	Lymphadenopathy. Rosai-Dorfman sinus histiocytosis w/massive lymphadenopathy.[69,70,71]	Not noted.[73,74]
JOINTS/BONES MUSCLES & CARTILAGE	Myalgias, arthralgias &/ or arthritis are common. Wrist changes after 6 months. 41% develop intercarpal and carpometacarpal joint space narrowing a few yrs. after onset of AOSD–25% then develop pericapitate ankylosis.[44]	Hemophagocytosis in the bone marrow. Delayed closure of the bones of the skull in infants, bulging fontanel often noted. Neck stiffness, abnormal muscle tone, impaired muscle coordination, paralysis.[48,49]	Not noted.[64,65] Some have concurrent autoimmune diseases that may involve the joints, such as inflammatory arthritis or undifferenciated connective tissue diseases.[65]	Not noted.[67,68]	Short stature. Arthralgias. Dysmorphic facial features: Triangular face, rotated ears, macrocrania, exophthalmia. Pectus excavatum, wide-set nipples, widened ribs, long bone changes, short, square hands, sacrococcygeal dimple, contractures.[69,70]	Not noted.[73,74]
VASCULITIS	Not noted.[44]	Not noted.[47,48,49]	Not noted.[64,65]	Not noted.[67,68]	Not noted. Some w/varicose veins on legs.[69]	Diffuse vasculopathy in the skin, liver & brain.[73,74]
AMYLOIDOSIS	Very rare.[44]	Not noted.[47,48,49]	Not noted.[64,65]	Not noted.[67,68]	Not noted.[69,70,71,72]	Not noted.[73,74]
ABNORMAL LABS	High: ESR, CRP, LFTs, ferritin. Low glycosylated ferritin. Leukocytosis, anemia common w/flares. Prolonged PTT (DIC risk.)[44]	High: ESR, CRP, triglycerides, LFTs, soluble CD25, ferritin. Low: platelets, fibrinogen. Low NK cell cytotoxic function, neutropenia, anemia.[49]	High IgE. Low serum IgA, IgG, IgM. Decreased circulating CD19+ B cells, IgG+ & IgA+ memory B cells, NK cells. >60% +ANA. WBC normal.[64,65]	Low circulating IgA, IgM antibodies, decreased class-switched memory B cells & NK T cells. ANA negative.[67,68]	Chronically elevated, but increase during flares: CRP, ESR, WBC. High during flares: IgG, Ig A. Pts. can become very anemic.[69,70,71,72]	High: CRP, ESR w/flares. Cytopenia. Blood: 10-fold decrease in ADA2. Low ADA2-specific adenosine deaminase activity: blood & CD14+ monocytes.[73,74]
	 AOSD: Salmon trunk eruption on a young adult pt. (cri-net.com AOSD image 1)	Generalized purpuric macules in Familial HLH. (Arch Dermatol. 2002;138(9):1208-1212)	Cold-induced urticarial rash. (NOMID Alliance pt. image)	APLAID vesiculopustular rash. (sciencedirect.com/science/article/pii/S0002929712004181)	 Pt. w/hyperpigmentation. (onlinelibrary.wiley.com/doi/10.1111/pde.12085/full#pde12085-fig-0001)	Livedo reticularis type rash is seen on DADA2 pts.

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List of abbreviations:

ACE: Angiotensin-converting enzyme (lab test)
ADA2: Adenosine deaminase 2
ANCA+ Vasculitis: Granulomatosis w/polyangiitis (GPA); Wegener's
ARDS: Acute Respiratory Distress Syndrome
CD14+ monocytes: Cluster of differentiation 14 positive monocytes
CD19: B-lymphocyte antigen CD19-aka Cluster of Differentiation 19
CD25: Soluble interleukin-2-receptor
CNS: Central Nervous System (involving the brain, spinal cord)
CRP: C-reactive protein (lab test);
DIC: Disseminated intravascular coagulation
ESR: Erythrocyte sedimentation rate (lab test); Westergren ESR
GI: Gastrointestinal (organs in the abdomen)
HSP: Henoch–Schönlein purpura, anaphylactoid purpura
ICP: Intracranial pressure
IDDM: Insulin-Dependent Diabetes Mellitus
LFTs: Liver function tests (lab test): AST, ALT, GGT, ALK Phos, Bilirubin
NK cells: Natural killer cells
PMNs: Polymorphonuclear leukocytes (on lab tests w/ WBC count)
pt.: abbreviation for the word "patient"
PTT: Partial thromboplastin time (lab test)
SAA: Serum amyloid A protein (lab test)
TSH: Thyroid-stimulating hormone (lab test); thyrotropin
w/: abbreviation for the word "with"
WBC: White Blood Count (lab test)

***These noted diseases are also referred to as "Classic" Hereditary Periodic Fever Syndromes (CAPS, FMF, HIDS, & TRAPS)**

All Cited References & Full Image Credits are Listed on the Back Side of this Chart.

This chart is an educational reference to increase awareness about autoinflammatory diseases.

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The NOMID Alliance is a 501(c)(3) non-profit organization dedicated to improving awareness, care and treatment for patients with CAPS or other autoinflammatory diseases.

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