Online supplement to An International Delphi Survey for the Definition of New Classification Criteria for Familial Mediterranean Fever, Mevalonate Kinase Deficiency, TNF Receptor–associated Periodic Fever Syndromes, and Cryopyrin-associated Periodic Syndrome, *The Journal of Rheumatology,* doi:10.3899/jrheum.180056

Supplementary Table 1. Variables obtained for each disease at the end of the first Delphi survey.

MKD	
Duration of attacks 5-7 days	Characteristic of fever episodes
Duration of attacks 3-6 days	
Duration of attacks 3-7 days	
Duration of attacks 4-6 days	
Fever	
Irregular periodicity	
Recurrence every 2- 8 weeks	
Recurrence every 2-6 weeks	
Recurrence every 4 weeks	
Self-limiting episodes	
Consanguinity	History
Disease onset < 1 year	
Disease onset <2 years	
Disease onset <3 years	
Disease onset <5 years	
Early disease onset	
Exclusion of infection	
Incomplete/no response to steroids	
Positive family history	
Presence of autoimmunity	
Presence of triggering factors (immunization, infection, minor trauma, surgery)	
Response to steroids	
Response to treatment	
Abdominal pain	Signs and Symptoms
Amyloidosis	
Aphtosis	
Arthralgia	
Arthritis	
Aseptic furuncles	
Ataxia	
Bad general condition during episodes	
Bipolar aphtosis	
Cerebellar syndrome	
Cervical lymphadenopathy	
Chest pain	
CNS abnormality/epilepsy	

Colitis	
Complete well-being between episodes	
Conjunctivitis	
Diarrhea	
Early onset Inflammatory bowel disease/ colitis	
Encephalopathy	
Episcleritis	
Erythema marginatum like rash	
Erythema nodosum	
Eye involvement	
Fatigue	
Fever chills	
Gastrointestinal manifestation	
Genital ulcers	
Growth retardation	
Headache	
Hemocolitis	
Hepatomegaly	
Joint manifestation	
Lymphadenopathy (often painful)	
Machrophagic activation syndrome	
Maculopapular rash	
Muscle weakness	
Musculoskeletal pain	
Myalgia	
Nausea	
Odynophagia	
Oral sores	
Pericarditis	
Peritonitis	
Pharingotonsillitis	
Polymorphous rash	
Psoriatic like rash	
Psychomotor delay	
Purpuric lesions/petechiae	
Renal involvement	
Renal involvement	
Renal tubular acidosis	
Sensorineural hearing loss	
Serositis	
Skin manifestation	

Skin Rash	
Splenomegaly	
Strong local reaction to vaccination and perfusion	
Urticarial rash	
Uveitis	
Vertigo	
Vomiting	
Anemia	Laboratory tests
Coagulation tests	
Complete Cells Blood count	
Evaluation of IgG subclasses	
Evaluation of liver function	
Evaluation renal function	
Genetic exclusion of others Autoinflammatory diseases	
Increase of acute phase reactants and serum amyloid A during fever episodes	
Increase of acute phase reactants and serum amyloid A during fever episodes and well being	
increased IgA level	
Increased IgD levels	
Increased urinary mevalonic acid during episodes	
Level of autoantibodies	
Level of cholesterol	
Level of Immunoglobulines	
Level of procalcitonin	
Level of proteinuria	
Lumbar puncture	
Mevalonate kinase activity	
Normalization of inflammatory markers during well being	
Positive genetic test for *MVK* gene	
Throat swab	
Thrombocytopenia	
Urine analysis	
Urine culture	
Abdominal ultrasound	Other investigation
Cardiac ultrasound	
Chest ultrasound	
CT/MR scan	
Joint ultrasound	

Physical growth assessment	
X ray	

FMF	
Classic recurrent fever pattern	Characteristic of fever episodes
Duration of attacks 1-2 days	
Duration of attacks 1-3 days	
Duration of attacks 1-5 days	
Duration of attacks few hours to 3-4 days	
Fever duration less than 3 days	
Fever duration less than 4 days	
Presence of prodromal symptoms	
Recurrence every 2-4 weeks	
Regular periodicity	
Self-limiting episodes	
Age at onset	History
At least 3 attacks (x year)	
Consanguinity	
Early age at onset	
Ethnicity (turkish, arabs, armenian, kurdis jeweish)	
Exclusion of infection	
Positive family history	
Presence of triggering factors	
Response to colchicine	
Unproductive laparotomy	
Well-being between episodes	
Abdominal pain	Signs and Symptoms
Absence of adenopathy	
Absence of diarrhea	
Absence of recurrent aphtosis	
Absence of vomiting	
Acute scrotum	
Amyloidosis	
Aphtosis	
Arthralgia	
Arthritis	
Bone pain	
Chest pain	

Conjunctivitis	
Constipation	
Diarrhea	
Epididymitis	
Erysipeloid rash	
Fatigue	
Fever chills	
Flank pain	
Headache	
Hepatomegaly	
Lymphadenitis	
Myalgia	
Nausea	
Non amyloid nephropathy	
Normal growth/development	
Pain under the feet during exercise	
Pericarditis	
Periorbital edema	
Peritonitis	
Pharingotonsillitis	
Pleuritis	
Post exertional myalgia	
Retrosternal pain	
Satisfaction of Livneh criteria	
Serositis	
Skin rash	
Splenomegaly	
Testicular pain	
Testicular swelling	
Uveitis	
Vomiting	
Coagulation tests	Laboratory tests
Complete cells blood count	
Dosage of calprotectine	
Evaluation of hematuria	
Evaluation of liver function	
Evaluation of microalbuminuria	
Evaluation of neutrophilia	
Evaluation of proteinuria	
Evaluation of renal function	
Evaluation of serum lipid profile	

Evauation of ionogramme	
Hemoculture	
Increase of acute phase reactants and serum amyloid A during fever episodes	
Increase of acute phase reactants and serum amyloid A during fever episodes and well being	
Level of alkaline phosphatase	
Level of autoantibodies	
Level of carbamide	
Level of complement factors	
Level of fibrinogen	
Level of haptoglobine	
Level of IgD and/or IgA during fever	
Level of lupus anticoagulant	
Level of procalcitonine	
Level of S100 protein	
Level of serum immunoglobulin	
Level of serum proteins	
Level of tiroid hormons	
Negative genetic test for other monogenic Autoinflammatory diseases	
Positive genetic analysis for *MEFV* gene	
Urinary albumin/creatinine ratio	
Urine analysis	
Abdominal ultrasound	Other investigation
Cardiac ultrasound	
Chest X ray	
Fundoscopy	
Musculoskeletal ultrasound	
Physical growth assessment	
Renal, subcutaneous fat or rectal biopsy	
Slit lamp examination	

TRAPS	
Duration of attacks 1-3 weeks	Characteristic of fever episodes
Duration of attacks 2-3 weeks	
fever	
Fever lasting more than 10 days	

Fever lasting more than 5 days	
Fever lasting more than 7 days	
Fever of any duration in young children	
Irregular long lasting fever episodes	
Recurrent episodes of fever	
Recurrent prolonged episodes of fever	
Consanguinity	History
Early age at onset	
Onset after 1st decade of life	
Onset after 2nd decade of life	
Positive family history	
Presence of triggering factors	
Responce to therapy	
Response to anti-TNF therapy	
Response to steroids	
School attendance, social and extra-curricular activities	
Spontaneaous remission of episodes	
Well being between flares	
Abdominal pain	Signs and Symptoms
Absence of abdominal pain	
Absence of aphtosis	
Amyloidosis	
Aphtosis	
Arthralgias	
Arthritis	
Aseptic meningitis	
Auricular swelling	
Cervical lymphadenitis	
Chest pain	
Conjunctivitis	
Constipation	
Diarrhea	
Eye inflammation	
Eye involvement	
Eye involvment	
Fever chills	
Flank pain	
Headache	
Hepatomegaly	
Localized intense myalgia	
Lymphadenitis	

Macular rash	
Migratory rash	
Monocytic fasciitis	
Mucosal inflammation	
Muscular involvement	
Musculoskeletal pain	
Myalgia	
Myositis	
Nausea	
Painful maculopapular rash	
Pathergy	
Periorbital edema	
Periorbital pain	
Periorbital rash	
Peritonitis	
Pharingitis	
Pharingotonsillitis	
Pleuritis	
Pseudo-cellulitis	
Recurrent pericarditis	
Retrosternal pain	
Serositis	
Skin involvment	
Skin rash	
Splenomegaly	
Testicular pain	
Urticarial like rash	
Uveitis	
Vomiting	
Complete blood cells count	Laboratory tests
Evaluation of liver function	
Evaluation of microalbuminuria	
Evaluation of proteinuria	
Evaluation of renal function	
Increase of acute phase reactants and serum amyloid A during fever episodes	
Increase of acute phase reactants and serum amyloid A during fever episodes and well being	
Level of autoantibodies	
Level of complement	
Level of IgG	
Level of procalcitonine	

Level of soluble TNF receptor	
Patient not affected if carrier of R92Q e P46L unless typical clinical picture	
Positive genetic analysis for *TNFRSF1A* gene	
Throat swab	
Urine analysis	
Urinary albumine/creatinine ratio	
Urine cultures	
Abdominal ultrasound	Other investigation
Joint assessment	
Ophthalmologic evaluation	
Physical growth assessement	
Renal, subcutanous fat or rectal biopsy	
X ray	

CAPS	
Fever	Characteristic of fever episodes
Fever duration > 3 days	
Irregular intercritic periods	
Recurrent fever	
Absence of autoimmunity	History
Age at onset <1 month	
Age at onset <1 year	
Caucasian ethnicity	
Chronic disease course	
Consaguinity	
Episodes triggered by cold exposure	
Positive family history	
Presence of triggering factors	
Response to anti histaminic therapy	
Response to IL-1Beta blockade	
Response to steroids	
Abdominal pain	Signs and Symptoms
Absence of oral apthosis	
Amyloidosis	
Arthralgia	
Arthritis	
Band erythema over the knuckles	
Cartilage overgrowth	

Chest pain	
Chronic meningitis	
Chronic urticaria	
Conjunctivitis	
Diarrhea	
Edema of the extremities	
Episcleritis	
Erysipeloid rashof the ankle	
Eye involvement	
Fatigue	
Fever chills	
Frontal bossing	
Fundus oculi abnormalities	
Growth retardation	
Headache	
Hearing loss	
Hepatomegaly	
Intracranial hypertension	
Iritis	
Irritability during attack	
Joint contractures	
Limb pain	
Lymphadenopathy	
Malaise	
Myalgia	
Nail clubbing	
Nanism	
Nausea	
Neurocognitive impairment	
Neurologic involvement	
Optic disc changes	
Optical nerve athrophy	
Oral ulcers	
Osteitis	
Osteo-arthropathy	
Papilledema	
Papillitis	
Peculiar facies/dysmorphism	
Peculiar musculoskeletal features	
Pericarditis	
Peritonitis	

Pharingitis	
Renal involvement	
Seizures	
Solitary pretibial lesion similar to erythema nodosum	
Splenomegaly	
Urticarial rash	
Uveitis	
Visual loss	
Vomiting	
Anemia	
Coagulation tests	Laboratory tests
Complete cells blood count	
Dosage of calprotectine	
Elevated liver enzymes	
Evaluation of liver function	
Evaluation of microalbuminuria	
Evaluation of proteinuria	
Evaluation of renal function	
Exclusion of M protein (in adults)	
Gene sequencing for other monogenic Autoinflammatory diseases	
Increase of acute phase reactants and serum amyloid A during fever episodes	
Increase of acute phase reactants and serum amyloid A during fever episodes and well being	
Leucocytosis	
Level of complement factors	
Level of procalcitonine	
Level of serum Immunoglobulines	
Levels of autoantibodies	
Not necessary genetic confirmation	
Positive lumbar punction	
Positive *NLRP12* genetic test	
Positive *NLRP3* genetic test	
Urinanalysis	
Urinary albumine/creatinine ratio	
Abdominal ultrasound	Other investigation
Acoustic evoked potentials	
Bone x-ray	
Joint ultrasound	
Ophthalmologic evaluation	
Physical growth assessment	

Positive central nervouse system MRI	
Positive inner ear MRI	
Skin biopsy	
Subcutaneous fat biopsy	
Visual evoked potentials	

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Supplementary Table 2. Variables obtained after the second Delphi survey, divided into 5 categories.

Classic recurrent fever pattern	Characteristic of fever episode
Duration of attacks 5-7 days	
Duration of attacks 1-2 days	
Duration of attacks 1-3 days	
Duration of attacks 1-3 weeks	
Duration of attacks 1-5 days	
Duration of attacks 2-3 weeks	
Duration of attacks 3-6 days	
Duration of attacks 3-7 days	
Duration of attacks 4-6 days	
Duration of attacks few hours to 3-4 days	
Fever	
Fever duration > 3 days	
Fever duration less than 3 days	
Fever duration less than 4 days	
Fever lasting more than 10 days	
Fever lasting more than 5 days	
Fever lasting more than 7 days	
Fever of any duration in young children	
Irregular intercritic periods	
Irregular long lasting fever episodes	
Irregular periodicity	
Presence of prodromal symptoms	
Recurrence every 2- 8 weeks	
Recurrence every 2-4 weeks	
Recurrence every 2-6 weeks	
Recurrence every 4 weeks	
Recurrent episodes of fever	
Recurrent prolonged episodes of fever	
Regular periodicity	
Self-limiting episodes	
Well-being between flares	History
Unproductive laparotomy	
Spontaneous remission of episodes	
School attendance, social and extra-curricular activities	
Response to steroids	
Response to IL-1Beta blockade	
Response to colchicine	

Response to anti-TNF therapy	
Response to anti histaminic therapy	
Response to treatment	
Response to therapy	
Presence of triggering factors (immunization, infection, minor trauma, surgery)	
Presence of autoimmunity	
Positive family history	
Onset after 2nd decade of life	
Onset after 1st decade of life	
Incomplete/no response to steroids	
Exclusion of infection	
Ethnicity (turkish, arabs, armenian, kurdis jeweish)	
Episodes triggered by cold exposure	
Early disease onset	
Disease onset <5 years	
Disease onset <3 years	
Disease onset <2 years	
Disease onset < 1 year	
Consanguinity	
Chronic disease course	
Caucasian ethnicity	
At least 3 attacks (x year)	
Age at onset <1 year	
Age at onset <1 month	
Age at onset	
Absence of autoimmunity	
Abdominal pain	Signs and Symptoms
Absence of abdominal pain	
Absence of adenopathy	
Absence of diarrhea	
Absence of recurrent aphtosis	
Absence of vomiting	
Acute scrotum	
Amyloidosis	
Anemia	
Aphtosis	
Arthralgia	
Arthritis	
Aseptic furuncles	
Aseptic meningitis	

Ataxia	
Auricular swelling	
Bad general condition during episodes	
Band erythema over the knuckles	
Bipolar aphtosis	
Bone pain	
Cartilage overgrowth	
Cerebellar syndrome	
Cervical lymphadenitis	
Cervical lymphadenopathy	
Chest pain	
Chronic meningitis	
Chronic urticaria	
CNS abnormality/epilepsy	
Colitis	
Complete well-being between episodes	
Conjunctivitis	
Constipation	
Diarrhea	
Early onset Inflammatory bowel disease/ colitis	
Edema of the extremities	
Encephalopathy	
Epididymitis	
Episcleritis	
Erysipeloid rash	
Erysipeloid rash of the ankle	
Erythema marginatum like rash	
Erythema nodosum	
Eye inflammation	
Eye involvement	
Fatigue	
Fever chills	
Flank pain	
Frontal bossing	
Fundus oculi abnormalities	
Gastrointestinal manifestation	
Genital ulcers	
Growth retardation	
Headache	
Hearing loss	
Hemocolitis	

Hepatomegaly	
Intracranial hypertension	
Iritis	
Irritability during attack	
Joint contractures	
Joint manifestation	
Limb pain	
Localized intense myalgia	
Lymphadenopathy	
Lymphadenopathy (often painful)	
Machrophagic activation syndrome	
Macular rash	
Maculopapular rash	
Malaise	
Migratory rash	
Monocytic fasciitis	
Mucosal inflammation	
Muscle weakness	
Muscular involvement	
Musculoskeletal pain	
Myalgia	
Myositis	
Nail clubbing	
Nanism	
Nausea	
Neurocognitive impairment	
Neurologic involvement	
Non amyloid nephropathy	
Normal growth/development	
Odynophagia	
Optic disc changes	
Optical nerve atrophy	
Oral sores	
Oral ulcers	
Osteitis	
Osteo arthropathy	
Pain under the feet during exercise	
Painful maculopapular rash	
Papilledema	
Papillitis	
Pathergy	

Peculiar facies/dysmorphism	
Peculiar musculoskeletal features	
Pericarditis	
Periorbital edema	
Periorbital pain	
Periorbital rash	
Peritonitis	
Pharingitis	
Pharingotonsillitis	
Pleuritis	
Polymorphous rash	
Post exertional myalgia	
Pseudo-cellulitis	
Psoriatic like rash	
Psychomotor delay	
Purpuric lesions/petechiae	
Recurrent pericarditis	
Renal involvement	
Renal tubular acidosis	
Retrosternal pain	
Satisfaction of Livneh criteria	
Seizures	
Sensorineural hearing loss	
Serositis	
Skin manifestation	
Skin rash	
Solitary pretibial lesion similar to erythema nodosum	
Splenomegaly	
Strong local reaction to vaccination and perfusion	
Testicular pain	
Testicular swelling	
Urticarial rash	
Uveitis	
Vertigo	
Visual loss	
Vomiting	
Anemia	Laboratory tests
Coagulation tests	
Complete cells blood count	
Dosage of calprotectine	
Elevated liver enzymes	

Evaluation of hematuria	
Evaluation of IgG subclasses	
Evaluation of liver function	
Evaluation of microalbuminuria	
Evaluation of neutrophilia	
Evaluation of proteinuria	
Evaluation of renal function	
Evaluation of serum lipid profile	
Evauation of ionogramme	
Exclusion of M protein (in adults)	
Gene sequencing for other monogenic Autoinflammatory diseases	
Genetic exclusion of others Autoinflammatory diseases	
Hemoculture	
Increase of acute phase reactants and serum amyloid A during fever episodes	
Increase of acute phase reactants and serum amyloid A during fever episodes and well being	
increased IgA level	
Increased IgD levels	
Increased urinary mevalonic acid during episodes	
Leukocytosis	
Level of alkaline phosphatase	
Level of autoantibodies	
Level of carbamide	
Level of cholesterol	
Level of complement factors	
Level of fibrinogen	
Level of haptoglobine	
Level of IgD and/or IgA during fever	
Level of IgG	
Level of lupus anticoagulant	
Level of procalcitonine	
Level of proteinuria	
Level of S100 protein	
Level of serum immunoglobulin	
Level of serum proteins	
Level of soluble TNF receptor	
Level of tiroid hormons	
Lumbar puncture	
Mevalonate kinase activity	

Negative genetic test for other monogenic Autoinflammatory diseases	
Normalization of inflammatory markers during well being	
Not necessary genetic confirmation	
Patient not affected if carrier of R92Q e P46L unless typical clinical picture	
Positive genetic analysis for *MEFV* gene	
Positive genetic analysis for *TNFRSF1A* gene	
Positive genetic test for *MVK* gene	
Positive lumbar punction	
Positive *NLRP12* genetic test	
Positive *NLRP3* genetic test	
Throat swab	
Thrombocytopenia	
Urinary albumin/creatinine ratio	
Urine analysis	
Urine cultures	
Abdominal ultrasound	Other tests
Acoustic evoked potentials	
Bone x-ray	
Cardiac ultrasound	
Chest ultrasound	
Chest X ray	
CT/MR scan	
Fundoscopy	
Joint assessment	
Joint ultrasound	
Muscoloskeletal ultrasound	
Ophthalmologic evaluation	
Physical growth assessment	
Positive central nervouse system MRI	
Positive inner ear MRI	
Renal, subcutaneous fat or rectal biopsy	
Skin biopsy	
Slit lamp examination	
Subcutaneous fat biopsy	
Visual evoked potentials	
X ray	

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Supplementary Table 3. Variables included in the top quartile considering the total score obtained at the end of the second Delphi survey in the 2 groups of clinicians (European vs American).

FMF							
EUROFEVER/PRINTO				**CARRA/ PRCSG**			
		Rank	**Medium rank**			**Rank**	**Medium rank**
1	Positive genetic analysis for *MEFV* gene	515	7.4	**1**	Positive genetic analysis for *MEFV* gene	111	7.9
2	Response to colchicine	502	6.8	**2**	Duration of attacks 1-3 days	71	7.9
3	Increase of acute phase reactants and serum amyloid A during fever episodes	400	6.2	**3**	Increase of acute phase reactants and serum amyloid A during fever episodes	71	5.9
4	Abdominal pain	398	5.7	**4**	Classic recurrent fever pattern	69	9.9
5	Ethnia (turkish, arabs, armenian, kurdis jeweish)	360	6.5	**5**	Abdominal pain	68	4.9
6	Classic recurrent fever pattern	309	7.9	**6**	Positive family history	67	5.2
7	Duration of attacks 1-3 days	242	7.3	**7**	Ethnia (turkish, arabs, armenian, kurdis jeweish)	66	6
8	Positive family history	186	4.9	**8**	Response to colchicine	66	5.1
9	Serositis	175	6.0	**9**	Serositis	45	6.4
10	Erysipeloid rash	173	4.3	**10**	Self limiting episodes	29	7
11	Self limiting episodes	152	5.1	**11**	Duration of attacks few hours to 3-4 days	26	8.7
12	Chest pain	144	4.5	**12**	Erysipeloid rash	25	3.6
13	Duration of attacks few hours to 3-4 days	140	7.4	**13**	Arthritis	22	3.7
14	Well being between episodes	108	3.7	**14**	Arthralgias	19	3.2
15	Arthritis	105	4.0	**15**	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	17	5.6
16	Amyloidosis	89	5.6	**16**	Well being between episodes	16	5.3
17	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	83	5.9	**17**	Amyloidosis	15	3.75
18	Arthralgias	61	3.4	**18**	Chest pain	8	2.7

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MKD							
EUROFEVER/PRINTO				CARRA/ PRCSG			
		Rank	Medium rank			Rank	Medium rank
1	Positive genetic test for *MVK* gene	528	8.7	**1**	Positive genetic test for *MVK* gene	109	9.1
2	Increased urinary mevalonic acid during episodes	279	6.6	**2**	Increased urinary mevalonic acid during episodes	73	6.1
3	Disease onset < 1 year	279	8.2	**3**	Fever	65	8.1
4	Duration of attacks 3-7 days	265	6.6	**4**	Duration of attacks 3-7 days	63	7
5	Fever	262	7.7	**5**	Increase of acute phase reactants and serum amyloid A during fever episodes	55	5.5
6	Increase of acute phase reactants and serum amyloid A during fever episodes	221	5.8	**6**	Increased IgD levels	52	6.5
7	Increased IgD levels	200	5.4	**7**	Presence of triggering factors (immunization, infection, minor trauma, surgery)	46	5.8
8	Presence of triggering factors (immunization, infection, minor trauma, surgery)	172	6.4	**8**	Disease onset < 1 year	38	7.6
9	Abdominal pain	151	4.1	**9**	Abdominal pain	33	4.1
10	Lymphadenopathy (often painful)	124	5	**10**	Lymphadenopathy (often painful)	26	4.3
11	Cervical lymphadenopathy	120	4.3	**11**	Early disease onset	24	4.8
12	Early disease onset	115	5.8	**12**	Irregular periodicity	23	5.8
13	Gastrointestinal manifestation	104	5.5	**13**	Mevalonate kinase activity	22	7.3
14	Diarrhea	91	4.3	**14**	Skin Rash	20	5.0
15	Maculopapular rash	76	4.5	**15**	Cervical lymphadenopathy	17	5.7
16	Aphtosis	71	3.7	**16**	Disease onset <2 years	15	7.5
17	Skin Rash	63	3.7	**17**	Diarrhea	15	3
18	Self limiting episodes	62	5.2	**18**	Self limiting episodes	10	3.3
19	Mevalonate kinase activity	58	6.4	**19**	Gastrointestinal manifestation	9	4.5
20	Disease onset <2 years	58	5.8	**20**	Maculopapular rash	9	4.5
21	Irregular periodicity	50	3.3	**21**	Aphtosis	9	4.5

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TRAPS							
EUROFEVER/PRINTO				CARRA/ PRCSG			
		Rank	Medium rank			Rank	Medium rank
1	Positive genetic analysis for *TNFRSF1A* gene	513	8.4	**1**	Positive genetic analysis for *TNFRSF1A* gene	124	9.5
2	Increase of acute phase reactants and serum amyloid A during fever episodes	265	6.6	**2**	Periorbital edema	61	5.1
3	Recurrent prolonged episodes of fever	254	8.5	**3**	Irregular long lasting fever episodes	61	7.6
4	Periorbital edema	225	5.2	**4**	Positive family history	60	5,0
5	Positive family history	222	6.2	**5**	Abdominal pain	60	5,0
6	Irregular long lasting fever episodes	193	8.4	**6**	Increase of acute phase reactants and serum amyloid A during fever episodes	58	5.8
7	Abdominal pain	191	4.2	**7**	Recurrent prolonged episodes of fever	47	9.4
8	Fever lasting more than 7 days	150	7.5	**8**	Fever lasting more than 5 days	43	8.6
9	Myalgia	146	4.7	**9**	Migratory rash	34	4.9
10	Monocytic fasciitis	112	7	**10**	Skin rash	33	3.7
11	Localized intense myalgia	105	5.8	**11**	Myalgia	30	3.8
12	Duration of attacks 1-3 weeks	104	8.7	**12**	Localized intense myalgia	30	6,0
13	Arthralgias	98	3.9	**13**	Recurrent episodes of fever	27	9,0
14	Skin rash	96	4.2	**14**	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	23	7.7
15	Migratory rash	94	4.7	**15**	Arthralgias	21	3,0
16	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	91	6.5	**16**	Conjunctivitis	21	4.2
17	Painful maculopapular rash	82	6.8	**17**	Fever lasting more than 7 days	19	6.3
18	Conjunctivitis	65	4.3	**18**	Duration of attacks 1-3 weeks	17	5.7
19	Recurrent episodes of fever	55	7.9	**19**	Monocytic fasciitis	2	2,0
20	Fever lasting more than 5 days	43	8.6	**20**	Painful maculopapular rash	/	/

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CAPS							
EUROFEVER/PRINTO				CARRA/ PRCSG			
		Rank	Medium rank			Rank	Medium rank
1	Positive NLRP3 genetic test	430	7.7	**1**	Positive NLRP3 genetic test	124	8.3
2	Urticarial rash	377	7.3	**2**	Urticarial rash	102	7.3
3	Response to IL-1Beta blockade	377	6.7	**3**	Recurrent fever	102	7.3
4	Recurrent fever	332	8.1	**4**	Response to IL-1Beta blockade	92	7.1
5	Increase of acute phase reactants and serum amyloid A during fever episodes	279	6.5	**5**	Episodes triggered by cold exposure	83	5.9
6	Hearing loss	260	5.4	**6**	Positive family history	71	6.5
7	Episodes triggered by cold exposure	208	5.8	**7**	Hearing loss	54	6.0
8	Chronic urticaria	145	6.0	**8**	Increase of acute phase reactants and serum amyloid A during fever episodes	52	5.8
9	Fever	143	7.5	**9**	Age at onset <1 year	50	5.6
10	Age at onset <1 year	135	7.1	**10**	Chronic disease course	43	5.4
11	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	134	5.6	**11**	Chronic meningitis	42	4.2
12	Chronic meningitis	129	4.4	**12**	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	32	8.0
13	Chronic disease course	122	5.8	**13**	Chronic urticaria	32	6.4
14	Eye involvement	99	5.5	**14**	Positive NLRP12 genetic test	32	6.4
15	Neurologic involvement	81	5.4	**15**	Neurologic involvement	21	7.0
16	Positive family history	80	3.5	**16**	Fever	14	4.7
17	Conjunctivitis	72	4.0	**17**	Conjunctivitis	13	6.5
18	Osteo-arthropathy	69	5.3	**18**	Age at onset <1 month	10	
19	Cartilage overgrowth	63	4.8	**19**	Arthralgia	7	1.8
20	Arthralgia	63	3.2	**20**	Eye involvement	5	5.0
21	Positive NLRP12 genetic test	62	6.9	**21**	Osteo-arthropathy	5	5.0
22	Age at onset <1 month	56	5.6	**22**	Cartilage overgrowth	3	3.0

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Supplementary Table 4.

A. Differences in the ranks between Eurofever/PRINTO and American experts for FMF.

		Eurofever/PRINTO				**CARRA**			
		Rank	Medium rank		%	Rank	Medium rank		%
1	Positive genetic analysis for MEFV gene	515	7.4	910	56,6	111	7.9	180	61,7
2	Response to colchicine	502	6.8	910	55,2	66	5.1	180	36,7
3	Increase of acute phase reactants and serum amyloid A during fever episodes	400	6.2	910	44,0	71	5.9	180	39,4
4	Abdominal pain	398	5.7	910	43,7	68	4.9	180	37,8
5	Ethnicity (turkish, arabs, armenian, kurdis jeweish)	360	6.5	910	39,6	66	6	180	36,7
6	Classic recurrent fever pattern	309	7.9	910	34,0	69	9.9	180	38,3
7	Duration of attacks 1-3 days	242	7.3	910	26,6	71	7.9	180	39,4
8	Positive family history	186	4.9	910	20,4	67	5.2	180	37,2
9	Serositis	175	6.0	910	19,2	45	6.4	180	25,0
10	Erysipeloid rash	173	4.3	910	19,0	25	3.6	180	13,9
11	Self limiting episodes	152	5.1	910	16,7	29	7	180	16,1
12	Duration of attacks few hours to 3-4 days	140	7.4	910	15,4	26	8.7	180	14,4
13	Chest pain	144	4.5	910	15,8	8	2.7	180	4,4
14	Arthritis	105	4.0	910	11,5	22	3.7	180	12,2
15	Well being between episodes	108	3.7	910	11,9	16	5.3	180	8,9
16	Amyloidosis	89	5.6	910	9,8	15	3.75	180	8,3
17	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	83	5.9	910	9,1	17	5.6	180	9,4
18	Arthralgias	61	3.4	910	6,7	19	3.2	180	10,6

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Supplementary Table 4 B. Differences in the ranks between Eurofever/PRINTO and American experts for MKD.

		PRINTO				**CARRA**			
		Rank	Medium rank		%	Rank	Medium rank		%
1	Positive genetic test for MVK gene	528	8.7	770	68,6	109	9.1	170	64,1
2	Increased urinary mevalonic acid during episodes	279	6.6	770	36,2	73	6.1	170	42,9
3	Duration of attacks 3-7 days	265	6.6	770	34,4	63	7	170	37,1
4	Fever	262	7.7	770	34,0	65	8.1	170	38,2
5	Disease onset < 1 year	279	8.2	770	36,2	38	7.6	170	22,4
6	Increase of acute phase reactants and serum amyloid A during fever episodes	221	5.8	770	28,7	55	5.5	170	32,4
7	Increased IgD levels	200	5.4	770	26,0	52	6.5	170	30,6
8	Presence of triggering factors (immunization, infection, minor trauma, surgery)	172	6.4	770	22,3	46	5.8	170	27,1
9	Abdominal pain	151	4.1	770	19,6	33	4.1	170	19,4
10	Lymphadenopathy (often painful)	124	5	770	16,1	26	4.3	170	15,3
11	Early disease onset	115	5.8	770	14,9	24	4.8	170	14,1
12	Cervical lymphadenopathy	120	4.3	770	15,6	17	5.7	170	10,0
13	Gastrointestinal manifestation	104	5.5	770	13,5	9	4.5	170	5,3
14	Diarrhea	91	4.3	770	11,8	15	3	170	8,8
15	Maculopapular rash	76	4.5	770	9,9	9	4.5	170	5,3
16	Skin Rash	63	3.7	770	8,2	20	5.0	170	11,8
17	Mevalonate kinase activity	58	6.4	770	7,5	22	7.3	170	12,9
18	Aphtosis	71	3.7	770	9,2	9	4.5	170	5,3
19	Disease onset <2 years	58	5.8	770	7,5	15	7.5	170	8,8
20	Irregular periodicity	50	3.3	770	6,5	23	5.8	170	13,5
21	Self limiting episodes	62	5.2	770	8,1	10	3.3	170	5,9

Supplementary Table 4 C. Differences in the ranks between Eurofever/PRINTO and American experts for TRAPS.

		PRINTO					**CARRA**		
		Rank	Medium rank		%	Rank	Medium rank		%
1	Positive genetic analysis for TNFRSF1A gene	513	8.4	750	68,4	124	9.5	180	68,9
2	Increase of acute phase reactants and serum amyloid A during fever episodes	265	6.6	750	35,3	58	5.8	180	32,2
3	Recurrent prolonged episodes of fever	254	8.5	750	33,9	47	9.4	180	26,1
4	Periorbital edema	225	5.2	750	30,0	61	5.1	180	33,9
5	Positive family history	222	6.2	750	29,6	60	5,0	180	33,3
6	Irregular long lasting fever episodes	193	8.4	750	25,7	61	7.6	180	33,9
7	Abdominal pain	191	4.2	750	25,5	60	5,0	180	33,3
8	Myalgia	146	4.7	750	19,5	30	3.8	180	16,7
9	Fever lasting more than 7 days	150	7.5	750	20,0	19	6.3	180	10,6
10	Localized intense myalgia	105	5.8	750	14,0	30	6,0	180	16,7
11	Skin rash	96	4.2	750	12,8	33	3.7	180	18,3
12	Migratory rash	94	4.7	750	12,5	34	4.9	180	18,9
13	Duration of attacks 1-3 weeks	104	8.7	750	13,9	17	5.7	180	9,4
14	Arthralgias	98	3.9	750	13,1	21	3,0	180	11,7
15	Monocytic fasciitis	112	7	750	14,9	2	2,0	180	1,1
16	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	91	6.5	750	12,1	23	7.7	180	12,8
17	Fever lasting more than 5 days	43	8.6	750	5,7	43	8.6	180	23,9
18	Conjunctivitis	65	4.3	750	8,7	21	4.2	180	11,7
19	Recurrent episodes of fever	55	7.9	750	7,3	27	9,0	180	15,0
20	Painful maculopapular rash	82	6.8	750	10,9	/	/	180	/

Supplementary Table 4 D. Differences in the ranks between Eurofever/PRINTO and American experts for CAPS.

		PRINTO				**CARRA**			
		Rank	Medium rank		%	Rank	Medium rank		%
1	Positive NLRP3 genetic test	430	7.7	810	53,1	124	8.3	210	59,0
2	Urticarial rash	377	7.3	810	46,5	102	7.3	210	48,6
3	Response to IL-1Beta blockade	377	6.7	810	46,5	92	7.1	210	43,8
4	Recurrent fever	332	8.1	810	41,0	102	7.3	210	48,6
5	Increase of acute phase reactants and serum amyloid A during fever episodes	279	6.5	810	34,4	52	5.8	210	24,8
6	Hearing loss	260	5.4	810	32,1	54	6.0	210	25,7
7	Episodes triggered by cold exposure	208	5.8	810	25,7	83	5.9	210	39,5
8	Age at onset <1 year	135	7.1	810	16,7	50	5.6	210	23,8
9	Chronic urticaria	145	6.0	810	17,9	32	6.4	210	15,2
10	Chronic meningitis	129	4.4	810	15,9	42	4.2	210	20,0
11	Increase of acute phase reactants and serum amyloid A during fever episodes and well being	134	5.6	810	16,5	32	8.0	210	15,2
12	Chronic disease course	122	5.8	810	15,1	43	5.4	210	20,5
13	Fever	143	7.5	810	17,7	14	4.7	210	6,7
14	Positive family history	80	3.5	810	9,9	71	6.5	210	33,8
15	Eye involvement	99	5.5	810	12,2	5	5.0	210	2,4
16	Neurologic involvement	81	5.4	810	10,0	21	7.0	210	10,0
17	Positive NLRP12 genetic test	62	6.9	810	7,7	32	6.4	210	15,2
18	Conjunctivitis	72	4.0	810	8,9	13	6.5	210	6,2
19	Osteo-arthropathy	69	5.3	810	8,5	5	5.0	210	2,4
20	Arthralgia	63	3.2	810	7,8	7	1.8	210	3,3
21	Cartilage overgrowth	63	4.8	810	7,8	3	3.0	210	1,4
22	Age at onset <1 month	56	5.6	810	6,9	10		210	4,8