Table 1 Criteria and definition for the classification of cryopyrin-associated periodic syndrome (CAPS)

Patient-related symptoms	
Family history of CAPS	Phenotype and or genetic
	confirmation of CAPS in
	other family members
Confirmed NLRP3 mutation	Genetic confirmation of
	NLRP3 mutation
CAPS disease course related variables	
Early disease onset	Age at onset of CAPS
	typical symptoms in infanc
	or early childhood
Episodic disease course characterized by CAPS	Disease course with
typical symptoms with or without persistent	episodes of clinically activ
inflammation	CAPS disease
Triggered inflammatory attacks	Triggers: cold or other
	stressors
Evidence of raised inflammatory marker	Evidence of CAPS clinical
associated with CAPS clinical signs	signs coupled with system
	inflammation, e.g. CRP
CAPS typical symptoms	
Amyloidosis	Evidence of organ amyloid
	deposits, e.g. kidney
Recurrent episodes of systemic symptoms	Evidence of systemic
	features like fever and/or
	chills and/or fatigue and/o
	rigors
Urticaria-like rash	Histologically characterize

	by neutrophilic dermatitis
Chronic aseptic meningitis	Evidence of clinical,
	laboratory and/or imaging
	evidence of non infectious
	inflammatation of the
	meninges
Recurrent eye inflammation,	Recurrent non-allergic, non-
	infectious conjunctivitis
	with/without other
	inflammatory ocular
	manifestations
Sensorineural hearing loss	Evidence of increased
	hearing thresholds on
	audiogram
Musculoskeletal sign and symptoms	Evidence of arthralgia,
	myalgia, arthritis and/or
	periarticular swelling
Skeletal abnormalities	Evidence of epiphyseal
	overgrowth, frontal bossing,
	clubbing and/or growth
	failure

Table 2
Systematic literature review search items

	"cryopyrin associated periodic syndromes" [MeSH Terms] OR	1056
CAPS	"Cryopyrin Associated Periodic Syndromes"[tiab] OR "Cryopyrin	
Pubmed	Associated Periodic Syndrome" [tiab] OR "Cryopyrin Associated Periodic	
	Fever Syndromes"[tiab] OR "Cryopyrin Associated Periodic Fever	
	Syndrome"[tiab OR cryopyrinopath*[tiab] OR FCAS[tiab] OR "Familial	
	Cold Autoinflammatory Syndrome"[tiab] OR "Familial Cold	
	Urticaria"[tiab] OR MWS[tiab] OR "Muckle Wells Syndrome"[tiab] OR	
	CINCA[tiab] OR (Chronic[tiab] AND Infantile[tiab] AND	
	Neurological[tiab] AND Cutaneous[tiab] AND Articular[tiab]) OR	
	NOMID[tiab] OR "Neonatal Onset Multisystem Inflammatory	
	Disease"[tiab] OR "Infantile Onset Multisystem Inflammatory	
	Disease"[tiab]	
CAPS	'cinca syndrome'/exp OR 'muckle wells syndrome'/exp OR 'familial cold	1512
Embase	autoinflammatory syndrome'/exp OR 'cryopyrin associated periodic	
	syndromes':ab,ti OR 'cryopyrin associated periodic syndrome':ab,ti OR	
	'cryopyrin associated periodic fever syndromes':ab,ti OR 'cryopyrin	
	associated periodic fever syndrome':ab,ti OR cryopyrinopath*:ab,ti OR	
	fcas:ab,ti OR 'familial cold autoinflammatory syndrome':ab,ti OR	
	'familial cold urticaria':ab,ti OR mws:ab,ti OR 'muckle wells	
	syndrome':ab,ti OR cinca:ab,ti OR (chronic:ab,ti AND infantile:ab,ti	
	AND neurological:ab,ti AND cutaneous:ab,ti AND articular:ab,ti) OR	
	nomid:ab,ti OR 'neonatal onset multisystem inflammatory	
	disease':ab,ti OR 'infantile onset multisystem inflammatory	
	disease':ab,ti	
CAPS	MeSH descriptor: [Cryopyrin-Associated Periodic Syndromes] explode	29
	all trees OR	
	"Cryopyrin Associated Periodic Syndromes" or "Cryopyrin Associated	
L		L

	Periodic Syndrome" or "Cryopyrin Associated Periodic Fever	
	Syndromes" or "Cryopyrin Associated Periodic Fever Syndrome" or	
	cryopyrinopath* or FCAS or "Familial Cold Autoinflammatory	
	Syndrome" or "Familial Cold Urticaria" or MWS or "Muckle Wells	
	Syndrome" or CINCA or (Chronic and Infantile and Neurological and	
	Cutaneous and Articular) or NOMID or "Neonatal Onset Multisystem	
	Inflammatory Disease" or "Infantile Onset Multisystem Inflammatory	
	Disease"(title/abstract)	
AID	"Hereditary Autoinflammatory Diseases"[Mesh:NoExp] OR	781 (all)
Pubmed	"autoinflammatory diseases" [tiab] OR "autoinflammatory	253[NtH1]
	syndromes"[tiab] OR "periodic fever syndromes"[tiab] OR	(new)
	"autoinflammatory disease" [tiab] OR "autoinflammatory	
	syndrome"[tiab] OR "periodic fever syndrome"[tiab]	
AID	'autoinflammatory disease'/de OR 'hereditary periodic fever'/de OR	1388 all
Embase	'autoinflammatory diseases':ab,ti OR 'autoinflammatory	508
	syndromes':ab,ti OR 'periodic fever syndromes':ab,ti OR	new[NtH2]
	'autoinflammatory disease':ab,ti OR 'autoinflammatory syndrome':ab,ti	
	OR 'periodic fever syndrome':ab,ti	
AID	MeSH descriptor: [Hereditary Autoinflammatory Diseases] this term	33
Cochrane	only	
	OR	
	"autoinflammatory diseases" or "autoinflammatory syndromes" or	
	"periodic fever syndromes" or "autoinflammatory disease" or	
	"autoinflammatory syndrome" or "periodic fever	
	syndrome"(title/abstract)	
Embase AID	syndrome"[tiab] OR "periodic fever syndrome"[tiab] 'autoinflammatory disease'/de OR 'hereditary periodic fever'/de OR 'autoinflammatory diseases':ab,ti OR 'autoinflammatory syndromes':ab,ti OR 'periodic fever syndromes':ab,ti OR 'autoinflammatory disease':ab,ti OR 'autoinflammatory syndrome':ab,ti OR 'periodic fever syndrome':ab,ti MeSH descriptor: [Hereditary Autoinflammatory Diseases] this term only OR "autoinflammatory diseases" or "autoinflammatory syndromes" or "periodic fever syndromes" or "autoinflammatory disease" or "autoinflammatory syndromes" or "periodic fever	508 new[NtH2]

Figure 1: Systematic literature review – search results

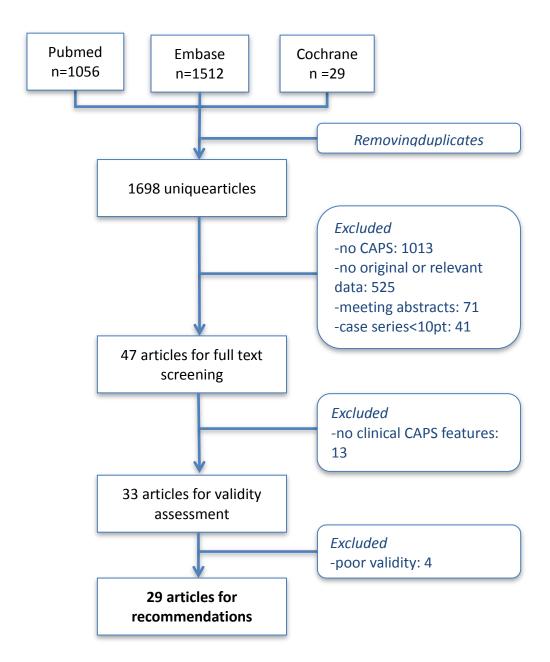
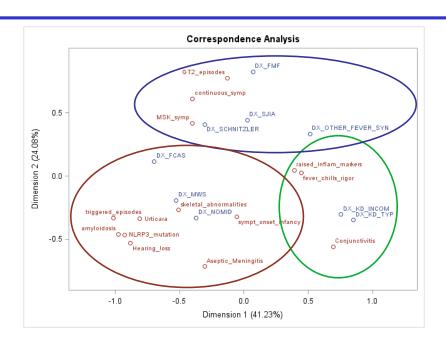


Figure 2: Correspondence analysis





Correspondence analysis revealed three distinct entities: CAPS (red circle), non-CAPS autoinflammatory diseases (blue circle) and monophasic inflammatory diseases (green circle)

Figure 3: Proposed CAPS diagnosis criteria model

Step 6: Development of a diagnostic model

Raised inflammatory markers (CRP/SAA) (Mandatory criteria)

plus

≥ 2 of 6 CAPS typical signs/symptoms:

- urticaria-like rash
- cold/stress triggered episodes
- sensorineural hearing loss
- musculoskeletal symptoms(arthralgia/arthritis/myalgia)
- chronic aseptic meningitis
- skeletal abnormalities (epiphyseal overgrowth/frontal bossing









Legend.

Specifity was 94% and sensitivity was 81% for the CAPS diagnosis criteria model. It performed equally well for all CAPS subtypes and in subgroups with or without evidence of NLRP3 mutation (p<0.001).

Figure 4 CAPS expert survey (supplemental material)

Q1 Family History 1. A positive family history of a genetically confirmed diagnosis of Cryopyrin-Associated Periodic Syndrome (CAPS) in a family member I consider this variable not often**not** undecided often very relevant relevant relevant relevant \bigcirc \bigcirc \bigcirc \otimes This ranking applies to \otimes all diseases in the CAPS spectrum OROonly some diseases in the CAPS spectrum, please check all applicable \square FCAS □ FCAS/MWS \square MWS ☐ MWS/CINCA/NOMID CINCA/NOMID 2. A positive family history of clinical signs and symptoms associated with CAPS I consider this variable oftennot undecided often very not relevant relevant relevant relevant 0 \bigcirc \bigcirc \bigcirc This ranking applies to \otimes all diseases in the CAPS spectrum OROonly some diseases in the CAPS spectrum, please check all applicable ☐ FCAS □ FCAS/MWS \square MWS ☐ MWS/CINCA/NOMID CINCA/NOMID **Q2** Characteristic of active disease 1. Age at disease onset I consider this variable often**not** undecided often very relevant relevant relevant relevant 0 \bigcirc \bigcirc 0 \otimes

This ranking applies to

all diseases in the	CAPS spectrum	n <i>OR</i>				
⊗only some disease	es in the CAPS s	pectrum, please	e check all	applicable		
□ FCAS	AS □ FCAS/MWS		VS ⊠	☑ MWS/CINCA/NOMID		X
CINCA/NOMID						
2. Epis	odic nature of s	ymptoms				
I consider this variable	not relevant	often not relevant	unc	lecided often relevant	very relevant	
This ranking applies	s to					
O all diseases in the	e CAPS spectrum	m <i>or</i>				
Oonly some disease	es in the CAPS s	spectrum, pleas	echeck all d	applicable		
□ FCAS ⊠ F	CAS/MWS over	lap 🗵 MWS	⊠ MWS/	CINCA/NOMI	D overlap	
CINCA/NOMID						
3. Dura I consider this variable	ation of episode not relevant	S often not relevant	unc	lecided often relevant	very relevant	
	0	\otimes	0	0	0	
This score applies to ⊗ all diseases in the		11 <i>01</i>				
Oonly some diseas	-		echeck all (applicable		
•	CAS/MWS over	•			D overlap	
CINCA/NOMID						
	ernal triggers re Cold-induced fla	C	ase flares			
I consider this variable	not	often not	unc	lecided often	very	

	relevant	relevant		relevant	relevant	
	0	0	0	\otimes	0	
m						
This score applies						
O all diseases in the	ne CAPS spectro	um <i>or</i>				
⊗only some diseas	ses in the CAPS	spectrum, pleas	echeck al	l applicable		
⊠ FCAS ⊠ F	FCAS/MWS over	erlap 🗵 MWS	⊠ MW	S/CINCA/NOMID	overlap	
CINCA/NOMID						
b)	Stress-induced	flares				
I consider this variable	not	often not		undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	\otimes	0	
This score applies	to					
O all diseases in the	ne CAPS spectro	um <i>or</i>				
⊗only some diseas	ses in the CAPS	spectrum, pleas	echeck al	l applicable		
□ FCAS ⊠ F	FCAS/MWS over	erlap 🗵 MWS	\square MW	S/CINCA/NOMID	overlap	
CINCA/NOMID						
c) Infe	ection-induced	flares				
I consider this variable	not	often not		undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	\otimes	0	
This score applies:	to					

This score applies to

 \bigcirc all diseases in the CAPS spectrum \emph{or}

		-	echeck all applicable		
□ FCAS 🗵	FCAS/MWS over	rlap 🗵 MWS	□ MWS/CINCA/N	OMID overlap	
CINCA/NOMID)				
Q3 Systemic	clinical symptor	ms of active o	lisease		
1. Feve	r				
I consider this variable	not	often not	undecided often	very	
	relevant	relevant	relevant	_	
	O	Ο	0 ⊗	0	
m					
This score applie					
	the CAPS spectrur				
-		•	echeck all applicable		
□ FCAS □	FCAS/MWS over	rlap \square MWS	☐ MWS/CINCA/N	OMID overlap	
CINCA/NOMID)				
2. Fatig	gue				
2. Fatig	not	often not	undecided often	very	
_	not relevant	relevant	relevan	t relevant	
_	not			•	
I consider this variable	not relevant	relevant	relevan	t relevant	
I consider this variable This score applies	relevant columns co	relevant	relevan	t relevant	
I consider this variable This score applie ⊗ all diseases in	not relevant o es to the CAPS spectrur	relevant	relevant	t relevant	
This score applie ⊗ all diseases in ○only some dis	not relevant o es to the CAPS spectrur eases in the CAPS	relevant	relevani	t relevant ⊗	
This score applie ⊗ all diseases in Oonly some dis □ FCAS	not relevant o es to the CAPS spectrur eases in the CAPS FCAS/MWS over	relevant	relevant	t relevant ⊗	
This score applie ⊗ all diseases in ○only some dis	not relevant o es to the CAPS spectrur eases in the CAPS FCAS/MWS over	relevant	relevani	t relevant ⊗	
This score applie ⊗ all diseases in Oonly some dis □ FCAS □ CINCA/NOMID	not relevant o es to the CAPS spectrur eases in the CAPS FCAS/MWS over	relevant	relevani	t relevant ⊗	
This score applie ⊗ all diseases in Oonly some dis □ FCAS □ CINCA/NOMID 3. Head	not relevant o es to the CAPS spectrur eases in the CAPS FCAS/MWS over	relevant o mor spectrum, pleas rlap MWS	relevant check all applicable MWS/CINCA/N	t relevant ⊗ OMID overlap	
This score applie ⊗ all diseases in Oonly some dis □ FCAS □ CINCA/NOMID	not relevant o es to the CAPS spectrur eases in the CAPS FCAS/MWS over	relevant	relevani	t relevant ⊗ OMID overlap very	

This score applies to						
O all diseases in the	CAPS spectrui	m <i>or</i>				
⊗only some disease	s in the CAPS s	spectrum, plea	secheck	all applicable		
□ FCAS □ FC	CAS/MWS over	·lap ⊠ MWS	⊠M	WS/CINCA/NOMID o	overlap [X
CINCA/NOMID						
4. Irritabili	ty					
I consider this variable	not	often n o	t	undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	\otimes	0	
This score applies to						
O all diseases in the	CAPS spectrum	m <i>or</i>				
⊗only some disease	s in the CAPS s	spectrum, plea	secheck	all applicable		
□ FCAS □ FC	CAS/MWS over	·lap ⊠ MWS	⊠M	WS/CINCA/NOMID o	overlap 🛭	X
CINCA/NOMID						
Q4 Organ-specif	ic clinical sy	mptoms of	active	disease		
1. Eye manifesta		•				
a) Conjunc						
I consider this variable	not	often n o	ıt	undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	0	\otimes	
This score applies to						
O all diseases in the	CAPS spectrui	m <i>or</i>				

 \otimes **only some** diseases in the CAPS spectrum, *pleasecheck* **all** applicable

□ FCAS	⊠ FCAS	S/MWS overla	ap ⊠ N	AWS	⊠ MWS	/CINCA/NOMID	overlap	
CINCA/NOMI	D							
b) Ker	atitis							
I consider this variable		not		often not	un	decided often	very	
		elevant	relevant			relevant	relevant	
)	0		O	\otimes	0	
This score appl	ies to							
O all diseases i	in the C	APS spectrum	or					
⊗only some di	seases ir	n the CAPS sp	ectrum,	please	check all	applicable		
□ FCAS	⊠ FCAS	S/MWS overla	ap ⊠ N	MWS	⊠ MWS	/CINCA/NOMID	overlap	
CINCA/NOMI	D							
c) Uve	eitis							
I consider this variable		not		often not	un	decided often	very	
		elevant	relevant			relevant	relevant	
	()	O		O	\otimes	0	
This score appl	ies to							
O all diseases i	in the C	APS spectrum	or					
⊗only some di	seases ir	n the CAPS sp	ectrum,	please	check all	applicable		
□ FCAS	⊠ FCAS	S/MWS overla	ap ⊠ N	I WS	⊠ MWS	/CINCA/NOMID	overlap	
CINCA/NOMI	D							
d) Pap	illedem	a and/or seco	ndary o	optic ne	erve dam	age (atrophy, vis	ion loss)	
I consider this variable		not		often not	un	decided often	very	
		elevant	relevant			relevant	relevant	
	()	0		0	0	\otimes	

This score applies to

 \bigcirc all diseases in the CAPS spectrumor

⊗only some d	liseases in	the CAPS spec	ctrum, please	echeck all appli	cable	
\square FCAS	□ FCAS	S/MWS overlap	\square MWS	⊠ MWS/CIN	CA/NOMID o	verlap 🗵
CINCA/NOM	ID					
2. Oral /ear	-nose-th	roat (ENT)	manifestat	tions		
a) Or	al ulcers	or aphtous sto	matitis			
I consider this variab		not	often not	undecided	l often	very
			levant		relevant	relevant
) ()	O	\otimes	0
This score app	olies to					
		APS spectrum <i>or</i>				
⊗only some d	liseases in	the CAPS spec	etrum, <i>please</i>	echeck all appli	cable	
□ FCAS	ĭ FCAS	S/MWS overlap	⊠ MWS	⊠ MWS/CIN	CA/NOMID ov	verlap
CINCA/NOM	ID					
b) Ex	udative j	pharyngitis				
I consider this variab	le	not	often not	undecided	d often	very
			levant		relevant	relevant
) C)	O	\otimes	0
This score app	olies to					
		APS spectrum <i>or</i>	•			
		the CAPS spec		echeck all appli	icable	
□ FCAS		S/MWS overlap	•			verlap \square
CINCA/NOM	ID	•				•
c) Ce	rvical lyr	nphadenopath	y			
I consider this variab	•	not	often not	undecided	l often	very
			levant		relevant	relevant
) C)	0	\otimes	0

This score appl	lies to						
O all diseases	in the CAPS spec	ctrum <i>or</i>					
⊗only some di	seases in the CA	PS specti	rum, <i>please</i>	check a	all applicable		
□ FCAS	ĭ FCAS/MWS	overlap	⊠ MWS	⊠ MV	VS/CINCA/NOMID	overlap	
CINCA/NOMI	ID						
d) Sen	sorineural hear	ing loss					
I consider this variable	e no	t	often not		undecided often	very	
	relevant	relev	ant		relevant	relevant	
	0	0		0	0	\otimes	
This score appl	lies to						
O all diseases	in the CAPS spec	etrum <i>or</i>					
⊗only some di	seases in the CA	PS specti	um, <i>please</i>	check a	ll applicable		
\square FCAS	☑ FCAS/MWS	overlap	⊠ MWS	⊠ MV	VS/CINCA/NOMID	overlap	X
CINCA/NOMI	ID						
3. Chest ma	nifestations						
a) Ple	uritis / pericard	itis / ches	st pain				
I consider this variable	_		oftennot		undecided often	very	
	relevant	relev	ant		relevant	relevant	
	0	0		0	\otimes	0	
This score appl	lies to						
O all diseases	in the CAPS spec	ctrum <i>or</i>					
⊗only some di	iseases in the CA	PS specti	um, <i>please</i>	check a	ll applicable		
□ FCAS	ĭ FCAS/MWS	overlap	⊠ MWS	⊠ MV	VS/CINCA/NOMID	overlap	
CINCA/NOMI	ID	_				-	

4. Abdominal manifestations

a) Abdominal pain

I consider this variable	not	often not		undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	\otimes	0	
This score applies to)					
O all diseases in the	CAPS spectru	m <i>or</i>				
⊗only some disease	es in the CAPS	spectrum, pleas	echeck al	ll applicable		
□ FCAS ⊠ FC	CAS/MWS ove	rlap 🗵 MWS	⊠ MW	S/CINCA/NOMID	overlap	
CINCA/NOMID						
b) Splenom	negaly					
I consider this variable	not	often not		undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	\otimes	0	
This score applies to)					
O all diseases in the	CAPS spectru	m <i>or</i>				
⊗only some disease	es in the CAPS	spectrum, pleas	echeck al	ll applicable		
□ FCAS ⊠ FC	CAS/MWS ove	rlap 🗵 MWS	⊠ MW	S/CINCA/NOMID	overlap	
CINCA/NOMID						
c) Renal m	anifestations:	proteinuria				
I consider this variable	not	often not		undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	0	\otimes	
This score applies to)					
O all diseases in the	CAPS spectru	m <i>or</i>				
⊗only some disease	es in the CAPS	spectrum, pleas	echeck al	l l applicable		
-		•		S/CINCA/NOMID	overlap	X
CINCA/NOMID		-			•	

d) Re	nal manifestatio	ons: amyloidosi	S			
I consider this variable	e i	not of	ten not	undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	0	\otimes	
	in the CAPS spoiseases in the CA FCAS/MWS	APS spectrum, p		all applicable WS/CINCA/NOMII) overlap	×
5. Skin man	nifestations ticaria-like rasl					
a) Ur I consider this variable		_	ten not	underided often	***	
i consider uns variabi	e relevant	of relevant	tennot	undecided often relevant	very relevant	
	0	0	0	0	\otimes	
This score applies to ○ all diseases in the CAPS spectrum or ⊗only some diseases in the CAPS spectrum, pleasecheck all applicable □ FCAS □ FCAS/MWS overlap □ MWS □ MWS/CINCA/NOMID overlap CINCA/NOMID						
b) Ma	culo-papular r	ash				
I consider this variable	e r	of of	ten not	undecided often	very	
	relevant	relevant	_	relevant	relevant	
	0	0	0	\otimes	0	

This score applies to

	n the CAPS spectr	u111 01				
⊗only some dis	eases in the CAPS	spectrum, pleas	echeck all d	applicable		
□ FCAS □	▼ FCAS/MWS ov	erlap 🗵 MWS	⊠ MWS/	CINCA/NOMII	O overlap	
CINCA/NOMII)					
6. Musculosk	xeletal manifest	ations				
I consider this variable	not	often not	und	decided often	very	
	relevant	relevant		relevant	relevant	
	0	O	O	\otimes	Ο	
This sacra appli	as to					
This score appli						
	n the CAPS spectr		1 1 11	1. 11		
•	eases in the CAPS	•		• •		_
	▼ FCAS/MWS ov	erlap 🗵 MWS	⊠ MWS/	CINCA/NOMII) overlap	
CINCA/NOMII)					
	e deformities (pat	_				
I consider this variable	not relevant	often not relevant	und	decided often relevant	very relevant	
	0					
	\circ	0	\circ	0	\otimes	
	O	0	0	0	\otimes	
This score appli	J	O	0	0	\otimes	
11	J	-	0	0	\otimes	
O all diseases in	es to n the CAPS spectr	um <i>or</i>			\otimes	
○ all diseases in ⊗only some dis	es to n the CAPS spectr seases in the CAPS	um <i>or</i> s spectrum, <i>pleas</i>	echeck all d	applicable		\boxtimes
○ all diseases in ⊗only some dis	es to In the CAPS spectroseases in the CAPS FCAS/MWS over	um <i>or</i> s spectrum, <i>pleas</i>	echeck all d	applicable		\boxtimes
○ all diseases in ⊗only some dis	es to In the CAPS spectroseases in the CAPS FCAS/MWS over	um <i>or</i> s spectrum, <i>pleas</i>	echeck all d	applicable		X
○ all diseases in ⊗only some dis	es to In the CAPS spectroseases in the CAPS FCAS/MWS over	um <i>or</i> s spectrum, <i>pleas</i>	echeck all d	applicable		☒
○ all diseases in ⊗only some dis	es to In the CAPS spectroseases in the CAPS FCAS/MWS over	um <i>or</i> s spectrum, <i>pleas</i>	echeck all a ⊠ MWS⁄	applicable		\boxtimes
○ all diseases in ⊗only some dis □ FCAS □ CINCA/NOMII b) Gro	es to n the CAPS spectr seases in the CAPS FCAS/MWS over with retardation	um <i>or</i> 5 spectrum, <i>pleas</i> erlap □ MWS	echeck all a ⊠ MWS⁄	applicable CINCA/NOMII	O overlap	\boxtimes

This score app	olies to					
O all diseases	in the CAPS spectro	ım <i>or</i>				
Oonly some o	liseases in the CAPS	spectrum, plea	secheck	all applicable		
□ FCAS	☐ FCAS/MWS over	erlap 🗆 MWS	⊠MV	WS/CINCA/NOMII	O overlap	X
CINCA/NOM	ID					
7. CNS ma	nifestations					
a) Mo	eningitis headaches	/ aseptic menir	ngitis			
I consider this variab	le not	often no	t	undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	\otimes	0	
This score app	olies to					
O all diseases	in the CAPS spectro	ım <i>or</i>				
Oonly some of	diseases in the CAPS	spectrum, plea	secheck	all applicable		
\square FCAS	☐ FCAS/MWS over	erlap \square MWS	⊠MV	WS/CINCA/NOMII	O overlap	X
CINCA/NOM	ID					
8. other ma	anifestations, ple	ase specify				
a)						
I consider this variab	le not	often no	t	undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	0	0	
This score app	olies to					
O all diseases	in the CAPS spectro	ım <i>or</i>				
Oonly some of	liseases in the CAPS	spectrum, plea	secheck	all applicable		
\square FCAS	☐ FCAS/MWS over	erlap \square MWS	\square MV	WS/CINCA/NOMII	O overlap	
CINCA/NOM	ID					

b)					
I consider this variable	not	often not	undecided	often	very
	relevant	relevant	_	relevant	relevant
	0	0	0	0	0
This score applies to					
O all diseases in the C	CAPS spectrum	n <i>or</i>			
Oonly some diseases	in the CAPS s	pectrum, please	echeck all appli	cable	
□ FCAS □ FCA	AS/MWS overl	ap □ MWS	□ MWS/CIN	CA/NOMID ov	verlap \square
CINCA/NOMID					
Q5 Inflammatory	markers				
a) Elevated (CRP or SAA <u>v</u>	<u>vith</u> clinical dis	sease activity		
I consider this variable	not	often not	undecided	often	very
	relevant	relevant		relevant	relevant
	O	0	O	O	O
This score applies to					
O all diseases in the C	CAPS spectrum	n or			
Oonly some diseases	in the CAPS s	pectrum, please	echeck all appli	cable	
□ FCAS □ FCA	AS/MWS overl	ap □ MWS	□ MWS/CIN	CA/NOMID ov	verlap □
CINCA/NOMID					
b) Elevated (CRP or SAA v	<u>vithout</u> clinical	disease activi	ty in symptom	free interval
I consider this variable	not	often not	undecided		very
	relevant	relevant		relevant	relevant
	0	\circ	0	0	0
This score applies to					
O all diseases in the C	CAPS spectrum	n <i>or</i>			
Oonly some diseases	•		echeck all appli	cable	

☐ FCAS [☐ FCAS/MWS o	verlap 🗆 MV	VS □ M	WS/CINCA/NOMI	D overlap	
CINCA/NOMI	D					
c) othe	er:					
I consider this variable	not	oft	en not	undecided often	very	
	relevant	relevant		relevant	relevant	
	0	0	0	0	0	
This score apple	ies to					
O all diseases i	n the CAPS spect	trum <i>or</i>				
Oonly some di	seases in the CAI	PS spectrum, p	leasecheck	all applicable		
□ FCAS I	□ FCAS/MWS o	verlap \square MV	WS □ M	WS/CINCA/NOMI	D overlap	
CINCA/NOMI	D					