

**Table 1**

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

<b>Patient-related symptoms</b>	
Family history of CAPS	Phenotype and or genetic confirmation of CAPS in other family members
Confirmed NLRP3 mutation	Genetic confirmation of NLRP3 mutation
<b>CAPS disease course related variables</b>	
Early disease onset	Age at onset of CAPS typical symptoms in infancy or early childhood
Episodic disease course characterized by CAPS typical symptoms with or without persistent inflammation	Disease course with episodes of clinically active CAPS disease
Triggered inflammatory attacks	Triggers: cold or other stressors
Evidence of raised inflammatory marker associated with CAPS clinical signs	Evidence of CAPS clinical signs coupled with systemic inflammation, e.g. CRP
<b>CAPS typical symptoms</b>	
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/or rigors
Urticaria-like rash	Histologically characterized

	by neutrophilic dermatitis
Chronic aseptic meningitis	Evidence of clinical, laboratory and/or imaging evidence of non infectious inflammation 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**

<b>CAPS</b>  <b>Pubmed</b>	“cryopyrin associated periodic syndromes”[MeSH Terms] OR “Cryopyrin Associated Periodic Syndromes”[tiab] OR “Cryopyrin 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]	1056
<b>CAPS</b>  <b>Embase</b>	'cinca syndrome'/exp OR 'muckle wells syndrome'/exp OR 'familial cold 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	1512
<b>CAPS</b>	MeSH descriptor: [Cryopyrin-Associated Periodic Syndromes] explode all trees OR "Cryopyrin Associated Periodic Syndromes" or "Cryopyrin Associated	29

	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)	
<b>AID Pubmed</b>	"Hereditary Autoinflammatory Diseases"[Mesh:NoExp] OR "autoinflammatory diseases"[tiab] OR "autoinflammatory syndromes"[tiab] OR "periodic fever syndromes"[tiab] OR "autoinflammatory disease"[tiab] OR "autoinflammatory syndrome"[tiab] OR "periodic fever syndrome"[tiab]	781 (all) 253 <sup>[NtH1]</sup> (new)
<b>AID Embase</b>	'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	1388 all 508 new <sup>[NtH2]</sup>
<b>AID Cochrane</b>	MeSH descriptor: [Hereditary Autoinflammatory Diseases] this term only OR "autoinflammatory diseases" or "autoinflammatory syndromes" or "periodic fever syndromes" or "autoinflammatory disease" or "autoinflammatory syndrome" or "periodic fever syndrome"(title/abstract)	33

**Figure 1: Systematic literature review – search results**

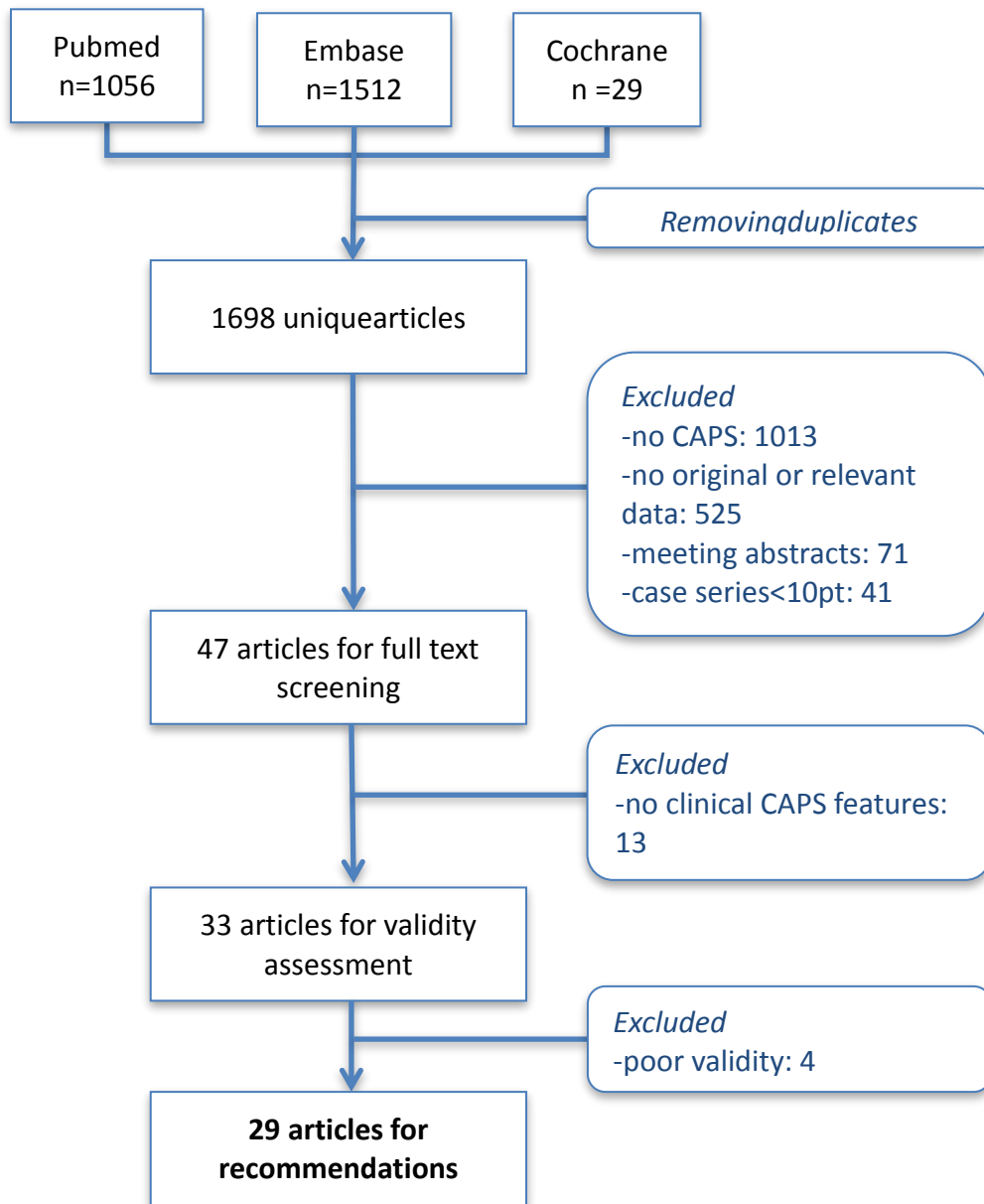
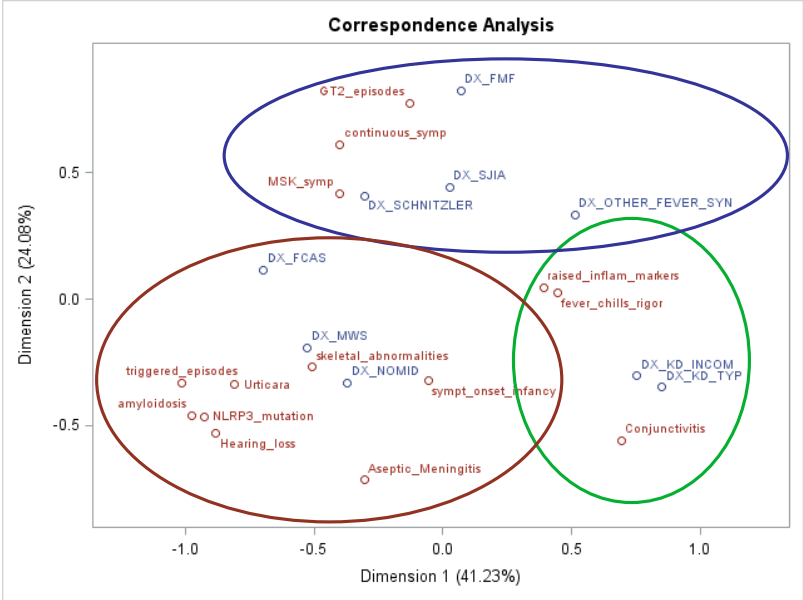


Figure 2: Correspondence analysis

## Results 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

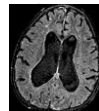
## Step 6: Development of a diagnostic model

**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 .

Specificity 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 relevant	oftennot relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This ranking applies to

**all** diseases in the CAPS spectrum **OR**

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS       FCAS/MWS       MWS       MWS/CINCA/NOMID       CINCA/NOMID

### 2. A positive family history of clinical signs and symptoms associated with CAPS

I consider this variable

	not relevant	oftennot relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This ranking applies to

**all** diseases in the CAPS spectrum **OR**

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS       FCAS/MWS       MWS       MWS/CINCA/NOMID       CINCA/NOMID

## Q2 Characteristic of active disease

### 1. Age at disease onset

I consider this variable

	not relevant	oftennot relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This ranking applies to

**all** diseases in the CAPS spectrum **OR**

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS       FCAS/MWS       MWS       MWS/CINCA/NOMID        
CINCA/NOMID

## 2. Episodic nature of symptoms

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>	<input type="radio"/>

This ranking applies to

all diseases in the CAPS spectrum **OR**

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS       FCAS/MWS overlap       MWS       MWS/CINCA/NOMID overlap        
CINCA/NOMID

## 3. Duration of episodes

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum **OR**

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS       FCAS/MWS overlap       MWS       MWS/CINCA/NOMID overlap        
CINCA/NOMID

## 4. External triggers resulting in disease flares

### a) Cold-induced flares

I consider this variable

	not	often not	undecided	often	very
--	-----	-----------	-----------	-------	------

	relevant	relevant		relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

**b) Stress-induced flares**

I consider this variable	<b>not</b>		<b>often not</b>		<b>undecided</b>	<b>often</b>		<b>very</b>
	relevant	relevant			relevant	relevant	relevant	
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

**c) Infection-induced flares**

I consider this variable	<b>not</b>		<b>often not</b>		<b>undecided</b>	<b>often</b>		<b>very</b>
	relevant	relevant			relevant	relevant	relevant	
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

### Q3 Systemic clinical symptoms of active disease

#### 1. Fever

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant		relevant	relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

#### 2. Fatigue

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant		relevant	relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

#### 3. Headaches

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant		relevant	relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

#### 4. Irritability

I consider this variable

	<b>not</b>	<b>often</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant		relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

## Q4 Organ-specific clinical symptoms of active disease

### 1. Eye manifestations

#### a) Conjunctivitis

I consider this variable

	<b>not</b>	<b>often</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant		relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

**b) Keratitis**

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant			relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

**c) Uveitis**

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant			relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

**d) Papilledema and/or secondary optic nerve damage (atrophy, vision loss)**

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant			relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

## 2. Oral /ear-nose-throat (ENT) manifestations

### a) Oral ulcers or aphtous stomatitis

I consider this variable	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

### b) Exudative pharyngitis

I consider this variable	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

### c) Cervical lymphadenopathy

I consider this variable	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

#### d) Sensorineural hearing loss

I consider this variable	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

### 3. Chest manifestations

#### a) Pleuritis / pericarditis / chest pain

I consider this variable	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID



## 4. Abdominal manifestations

### a) Abdominal pain

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

### b) Splenomegaly

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

### c) Renal manifestations: proteinuria

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

#### d) Renal manifestations: amyloidosis

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant			relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

### 5. Skin manifestations

#### a) Urticaria-like rash

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant			relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

#### b) Maculo-papular rash

I consider this variable	<b>not</b>	<b>often</b>	<b>not</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant			relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

## 6. Musculoskeletal manifestations

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

### b) Bone deformities (patella overgrowth, clubbing, frontal bossing)

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

### b) Growth retardation

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

## 7. CNS manifestations

### a) Meningitis headaches / aseptic meningitis

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

## 8. other manifestations, please specify

### a) \_\_\_\_\_

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

**b)**

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

- all diseases in the CAPS spectrum *or*
- only some** diseases in the CAPS spectrum, *please check all applicable*
- FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

## Q5 Inflammatory markers

### a) Elevated CRP or SAA with clinical disease activity

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

- all diseases in the CAPS spectrum *or*
- only some** diseases in the CAPS spectrum, *please check all applicable*
- FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap     CINCA/NOMID

### b) Elevated CRP or SAA without clinical disease activity in symptom free interval

I consider this variable

	not relevant	often not relevant	undecided	often relevant	very relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

- all diseases in the CAPS spectrum *or*
- only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID

**c) other:** \_\_\_\_\_

I consider this variable	<b>not</b>	<b>often</b>	<b>undecided</b>	<b>often</b>	<b>very</b>
	relevant	relevant		relevant	relevant
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

This score applies to

all diseases in the CAPS spectrum *or*

**only some** diseases in the CAPS spectrum, *please check all applicable*

FCAS     FCAS/MWS overlap     MWS     MWS/CINCA/NOMID overlap      
CINCA/NOMID