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 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
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/or rigors
Urticaria-like rash	Histologically characterized

	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

CAPS Pubmed	"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
CAPS Embase	'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
CAPS	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)	
AID Pubmed	"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[NtH1] (new)
AID Embase	'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[NtH2]
AID Cochrane	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

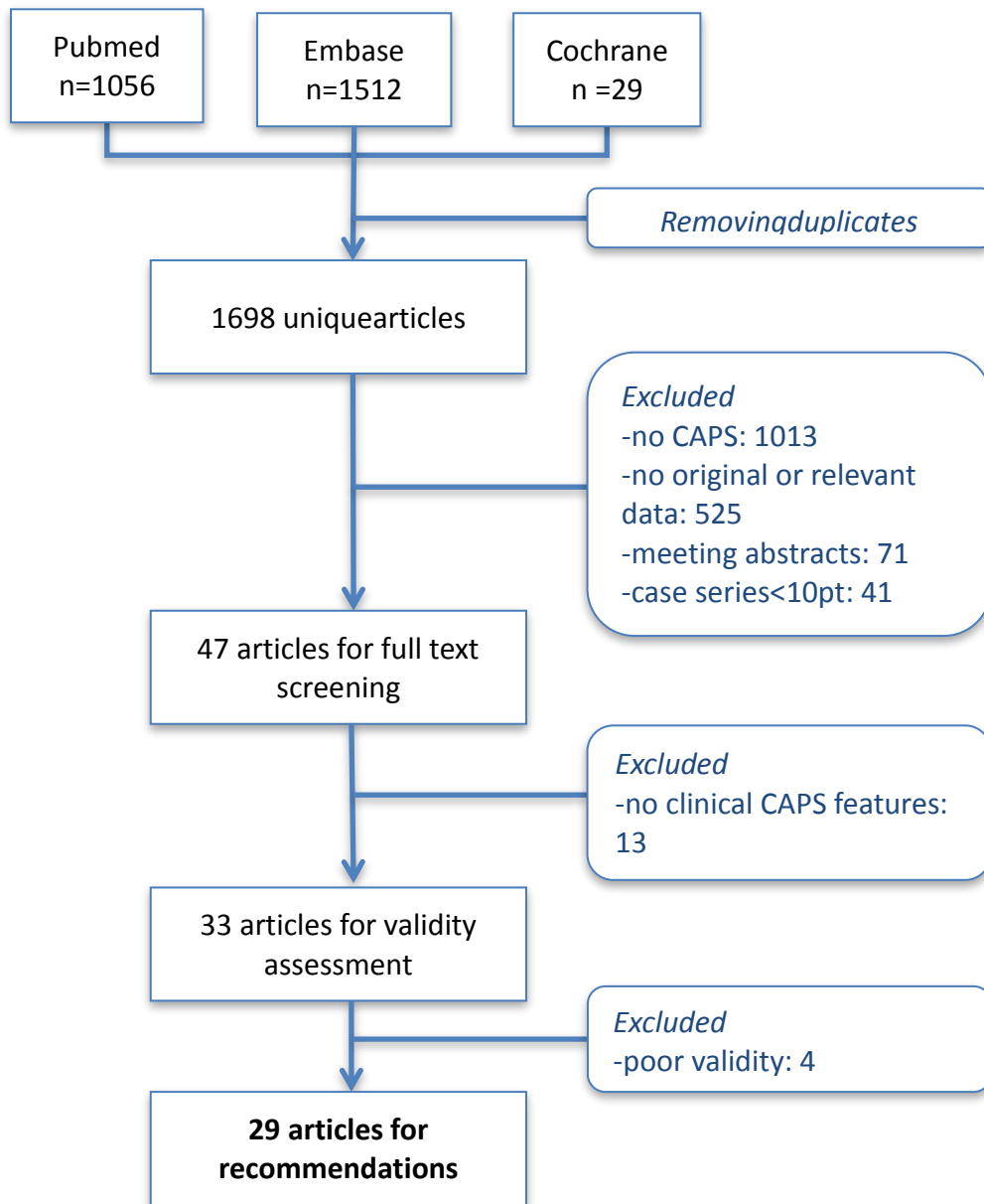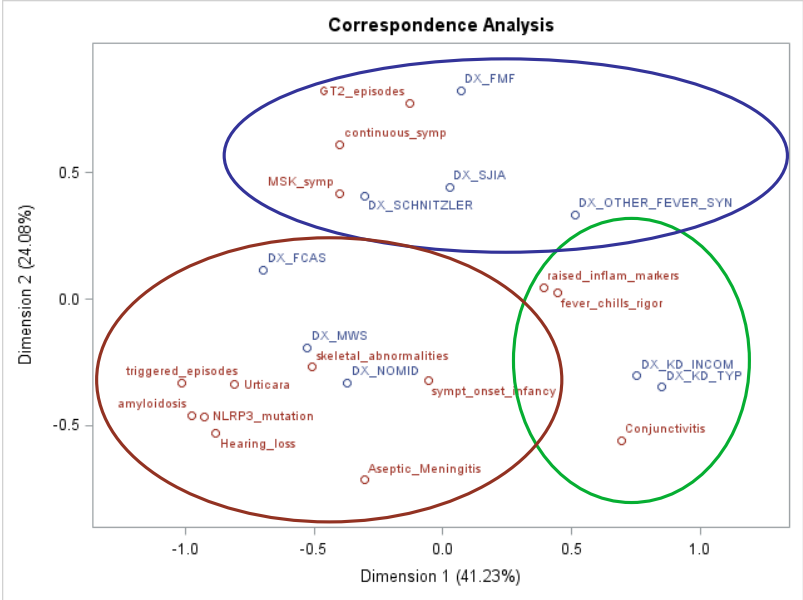**Figure 1: Systematic literature review – search results**

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┌──────────────┐  ┌──────────────┐  ┌──────────────┐
│   Pubmed     │  │   Embase     │  │  Cochrane    │
│   n=1056     │  │   n=1512     │  │   n =29      │
└──────────────┘  └──────────────┘  └──────────────┘
         │                │                │
         └────────────────┼────────────────┘
                          │         ┌─────────────────────────┐
                          │─────────│  Removingduplicates      │
                          ▼         └─────────────────────────┘
              ┌──────────────────────┐
              │  1698 uniquearticles │
              └──────────────────────┘
                          │         ┌─────────────────────────┐
                          │         │ Excluded                │
                          │         │ -no CAPS: 1013          │
                          │─────────│ -no original or relevant│
                          │         │ data: 525               │
                          │         │ -meeting abstracts: 71  │
                          │         │ -case series<10pt: 41   │
                          ▼         └─────────────────────────┘
              ┌──────────────────────┐
              │ 47 articles for full │
              │ text screening       │
              └──────────────────────┘
                          │         ┌─────────────────────────┐
                          │         │ Excluded                │
                          │─────────│ -no clinical CAPS       │
                          │         │ features: 13            │
                          ▼         └─────────────────────────┘
              ┌──────────────────────┐
              │ 33 articles for      │
              │ validity assessment  │
              └──────────────────────┘
                          │         ┌─────────────────────────┐
                          │─────────│ Excluded                │
                          │         │ -poor validity: 4       │
                          ▼         └─────────────────────────┘
              ┌──────────────────────┐
              │ 29 articles for      │
              │ recommendations      │
              └──────────────────────┘
```

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)
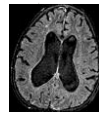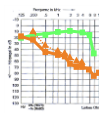
Figure 3: Proposed CAPS diagnosis criteria model

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** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	⊗

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	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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
	○	○	⊗	○	○

This ranking 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

3. Duration of episodes

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	⊗	○	○	○

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

4. External triggers resulting in disease flares

a)   Cold-induced flares

I consider this variable	**not**	often**not**	undecided	**often**	**very**

	not relevant	oftennot relevant		often relevant	very relevant
relevant	relevant		relevant	relevant	
○	○	○	⊗	○	

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)   Stress-induced flares

I consider this variable	**not**	oftennot	undecided **often**	**very**
	relevant	relevant		relevant	relevant
	○	○	○	⊗	○

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

c) Infection-induced flares

I consider this variable	**not**	oftennot	undecided **often**	**very**
	relevant	relevant		relevant	relevant
	○	○	○	⊗	○

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

Q3 Systemic clinical symptoms of active disease

1. Fever

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

2. Fatigue

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	⊗

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

3. Headaches

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

4. Irritability

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

Q4 Organ-specific clinical symptoms of active disease

1. Eye manifestations

a) Conjunctivitis

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	⊗

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) Keratitis

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

c) Uveitis

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

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

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	⊗

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

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
	○	○	○	⊗	○

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) Exudative pharyngitis

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

c) Cervical lymphadenopathy

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

d) Sensorineural hearing loss

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	⊗

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

3. Chest manifestations

a)   Pleuritis / pericarditis / chest pain

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

4. Abdominal manifestations

a) Abdominal pain

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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) Splenomegaly

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

c) Renal manifestations: proteinuria

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	⊗

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

d) Renal manifestations: amyloidosis

I consider this variable	not relevant	often not relevant	undecided	often relevant	very relevant
	○	○	○	○	⊗

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

5. Skin manifestations

a) Urticaria-like rash

I consider this variable	not relevant	often not relevant	undecided	often relevant	very relevant
	○	○	○	○	⊗

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) Maculo-papular rash

I consider this variable	not relevant	often not relevant	undecided	often relevant	very relevant
	○	○	○	⊗	○

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

6. Musculoskeletal manifestations

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	⊗

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)   Growth retardation

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

7. CNS manifestations

a) Meningitis headaches / aseptic meningitis

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	⊗	○

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

8. other manifestations, please specify

a) _____

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	○

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) _____

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	○

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 <u>with</u> clinical disease activity

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	○

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 <u>without</u> clinical disease activity in symptom free interval

I consider this variable	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	○

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	**not** relevant	often**not** relevant	undecided	**often** relevant	**very** relevant
	○	○	○	○	○

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