<table>
<thead>
<tr>
<th><strong>Patient-related symptoms</strong></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Family history of CAPS</td>
<td>Phenotype and or genetic confirmation of CAPS in other family members</td>
</tr>
<tr>
<td>Confirmed NLRP3 mutation</td>
<td>Genetic confirmation of NLRP3 mutation</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>CAPS disease course related variables</strong></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Early disease onset</td>
<td>Age at onset of CAPS typical symptoms in infancy or early childhood</td>
</tr>
<tr>
<td>Episodic disease course characterized by CAPS</td>
<td>Disease course with episodes of clinically active CAPS disease</td>
</tr>
<tr>
<td>typical symptoms with or without persistent inflammation</td>
<td></td>
</tr>
<tr>
<td>Triggered inflammatory attacks</td>
<td>Triggers: cold or other stressors</td>
</tr>
<tr>
<td>Evidence of raised inflammatory marker associated with CAPS</td>
<td>Evidence of CAPS clinical signs coupled with systemic inflammation, e.g. CRP</td>
</tr>
<tr>
<td>clinical signs</td>
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</tbody>
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<table>
<thead>
<tr>
<th><strong>CAPS typical symptoms</strong></th>
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<tbody>
<tr>
<td>Amyloidosis</td>
<td>Evidence of organ amyloid deposits, e.g. kidney</td>
</tr>
<tr>
<td>Recurrent episodes of systemic symptoms</td>
<td>Evidence of systemic features like fever and/or chills and/or fatigue and/or rigors</td>
</tr>
<tr>
<td>Urticaria-like rash</td>
<td>Histologically characterized</td>
</tr>
<tr>
<td>Condition</td>
<td>Description</td>
</tr>
<tr>
<td>------------------------------------------------</td>
<td>---------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Chronic aseptic meningitis</td>
<td>Evidence of clinical, laboratory and/or imaging evidence of non infectious inflammation of the meninges</td>
</tr>
<tr>
<td>Recurrent eye inflammation,</td>
<td>Recurrent non-allergic, non-infectious conjunctivitis with/without other inflammatory ocular manifestations</td>
</tr>
<tr>
<td>Sensorineural hearing loss</td>
<td>Evidence of increased hearing thresholds on audiogram</td>
</tr>
<tr>
<td>Musculoskeletal sign and symptoms</td>
<td>Evidence of arthralgia, myalgia, arthritis and/or periarticular swelling</td>
</tr>
<tr>
<td>Skeletal abnormalities</td>
<td>Evidence of epiphyseal overgrowth, frontal bossing, clubbing and/or growth failure</td>
</tr>
<tr>
<td>CAPS</td>
<td>Search Items</td>
</tr>
<tr>
<td>------</td>
<td>------------------------------------------------------------------------------</td>
</tr>
<tr>
<td><strong>Embase</strong></td>
<td>'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</td>
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<tr>
<td><strong>MeSH</strong></td>
<td>MeSH descriptor: [Cryopyrin-Associated Periodic Syndromes] explode all trees OR &quot;Cryopyrin Associated Periodic Syndromes&quot; or &quot;Cryopyrin Associated Periodic Syndrome&quot;</td>
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<tr>
<td>AID</td>
<td>Search</td>
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<tr>
<td>Embase</td>
<td>'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</td>
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<tr>
<td>Cochrane</td>
<td>MeSH descriptor: [Hereditary Autoinflammatory Diseases] this term only OR &quot;autoinflammatory diseases&quot; or &quot;autoinflammatory syndromes&quot; or &quot;periodic fever syndromes&quot; or &quot;autoinflammatory disease&quot; or &quot;autoinflammatory syndrome&quot; or &quot;periodic fever syndrome&quot;(title/abstract)</td>
</tr>
</tbody>
</table>
Figure 1: Systematic literature review – search results

- **Pubmed n=1056**
- **Embase n=1512**
- **Cochrane n =29**

1698 unique articles

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
Correspondence analysis revealed three distinct entities: CAPS (red circle), non-CAPS autoimmune inflammatory diseases (blue circle) and monophasic inflammatory diseases (green circle)
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).
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 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    often      not    undecided    often    very
                                      relevant relevant relevant relevant 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    often      not    undecided    often    very
                                      relevant relevant relevant relevant relevant

   This ranking applies to
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

<table>
<thead>
<tr>
<th>Variable</th>
<th>Not relevant</th>
<th>Often relevant</th>
<th>Undecided</th>
<th>Often relevant</th>
<th>Very relevant</th>
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<td>O</td>
<td>O</td>
<td>X</td>
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This ranking applies to

all diseases in the CAPS spectrum

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

<table>
<thead>
<tr>
<th>Variable</th>
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<th>Often relevant</th>
<th>Undecided</th>
<th>Often relevant</th>
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<td>O</td>
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<td>O</td>
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</table>

This score applies to

all diseases in the CAPS spectrum

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

<table>
<thead>
<tr>
<th>Variable</th>
<th>Not relevant</th>
<th>Often relevant</th>
<th>Undecided</th>
<th>Often relevant</th>
<th>Very relevant</th>
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</table>
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

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

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 not relevant not often relevant undecided often relevant very relevant
   - O
   - O
   - O
   - X
   - O

   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 not relevant not often relevant undecided often relevant very relevant
   - O
   - O
   - O
   - O
   - X

   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 not relevant not often relevant undecided often relevant very relevant
   - O
   - O
   - O
   - X
   - O
This score applies to
○ all diseases in the CAPS spectrum or
.prod 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  not  oftennot  undecided  often  very
          relevant        relevant        relevant        relevant
         ☐       ☐       ☐       ☒            ☒

This score applies to
○ all diseases in the CAPS spectrum or
.prod 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  not  oftennot  undecided  often  very
          relevant        relevant        relevant        relevant
         ☐       ☐       ☐       ☒            ☒

This score applies to
○ all diseases in the CAPS spectrum or
.prod only some diseases in the CAPS spectrum, please check all applicable
b) Keratitis

I consider this variable

<table>
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This score applies to

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

CINCA/NOMID

c) Uveitis

I consider this variable

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This score applies to

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

CINCA/NOMID

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

I consider this variable

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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 aphthous stomatitis

I consider this variable

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<th>often relevant</th>
<th>undecided relevant</th>
<th>often relevant</th>
<th>very relevant</th>
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This score applies to

○ all diseases in the CAPS spectrum or

只有一些疾病在CAPS谱系中，检查所有适用

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

b) Exudative pharyngitis

I consider this variable

<table>
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<th>often relevant</th>
<th>very relevant</th>
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This score applies to

○ all diseases in the CAPS spectrum or

只有一些疾病在CAPS谱系中，检查所有适用

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

c) Cervical lymphadenopathy

I consider this variable

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CINCA/NOMID
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 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, 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 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, please check all applicable
☐ FCAS ☒ FCAS/MWS overlap ☒ MWS ☒ MWS/CINCA/NOMID overlap ☐ CINCA/NOMID
4. Abdominal manifestations

a) Abdominal pain

<table>
<thead>
<tr>
<th>I consider this variable</th>
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<th>often not relevant</th>
<th>undecided relevant</th>
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☑ only some diseases in the CAPS spectrum, please check all applicable

☐ FCAS  ☒ FCAS/MWS overlap  ☒ MWS  ☒ MWS/CINCA/NOMID overlap  ☐ CINCA/NOMID

b) Splenomegaly

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<tr>
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<th>undecided relevant</th>
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☑ 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

<table>
<thead>
<tr>
<th>I consider this variable</th>
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<th>often not relevant</th>
<th>undecided relevant</th>
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☑ 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

<table>
<thead>
<tr>
<th>I consider this variable</th>
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This score applies to

- [ ] all diseases in the CAPS spectrum
- 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

<table>
<thead>
<tr>
<th>I consider this variable</th>
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<th>undecided relevant</th>
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This score applies to

- [ ] all diseases in the CAPS spectrum
- 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

<table>
<thead>
<tr>
<th>I consider this variable</th>
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<th>oftennot relevant</th>
<th>undecided relevant</th>
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This score applies to
1. 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

<table>
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☐ FCAS  ✗ FCAS/MWS overlap  ✗ MWS  ✗ MWS/CINCA/NOMID overlap  ☑
CINCA/NOMID

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

<table>
<thead>
<tr>
<th>I consider this variable</th>
<th>not</th>
<th>often</th>
<th>not</th>
<th>undecided</th>
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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

<table>
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<tr>
<th>I consider this variable</th>
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<th>often</th>
<th>not</th>
<th>undecided</th>
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☐ FCAS  ☑ FCAS/MWS overlap  ☑ MWS  ✗ MWS/CINCA/NOMID overlap  ✗
CINCA/NOMID
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 often not undecided often very
       relevant relevant relevant relevant 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

8. other manifestations, please specify
   a) _________________________________________
   I consider this variable
       not often not undecided often very
       relevant relevant relevant relevant 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 with clinical disease activity

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

This score applies to
- all diseases in the CAPS spectrum
- 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 often not undecided often very relevant relevant relevant relevant relevant relevant

This score applies to
- all diseases in the CAPS spectrum
- only some diseases in the CAPS spectrum, please check all applicable
☐ FCAS  ☐ FCAS/MWS overlap  ☐ MWS  ☐ MWS/CINCA/NOMID overlap  ☐ CINCA/NOMID

c) other: _______________________________________________________

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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