



<https://printo.it/pediatric-rheumatology/TW/intro>

## Cryopyrin syndrome (CAPS)

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### 1. Cryopyrin syndrome

#### 1.1 Cryopyrin syndrome

Cryopyrin syndrome (CAPS) is a group of autoinflammatory syndromes characterized by recurrent febrile episodes (FCAS), Muckle-Wells syndrome (MWS), CINCA syndrome, and NOMID. These syndromes are caused by mutations in the NLRP3 gene. The clinical features of CAPS include recurrent febrile episodes, skin rashes, conjunctivitis, and hearing loss. The severity of the disease varies among the different syndromes, with NOMID being the most severe and FCAS being the least severe.

#### 1.2

CAPS is a group of autoinflammatory syndromes characterized by recurrent febrile episodes, skin rashes, conjunctivitis, and hearing loss.

#### 1.3

CAPS is a group of autoinflammatory syndromes characterized by recurrent febrile episodes (FCAS, MWS, CINCA/NOMID) caused by mutations in CIAS1 (or NLRP3). Treatment with cryopyrinase (cryopyrin inhibitor) is effective in reducing the frequency and severity of the febrile episodes. In a study, 30% of CINCA/NOMID patients treated with cryopyrinase showed a significant improvement in their clinical symptoms.

#### 1.4

CAPS is a group of autoinflammatory syndromes characterized by recurrent febrile episodes caused by mutations in CIAS1. Treatment with cryopyrinase is effective in reducing the frequency and severity of the febrile episodes. In a study, 50% of patients treated with cryopyrinase showed a significant improvement in their clinical symptoms.

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**1.5** CAPS

**1.6**

FCAS 1-2 24 6 FCAS

MWS 70% 25% CAPS

CINCA NOMID 3 25% CAPS MWS FCAS CINCA NOMID CAPS FCAS MWS CINCA NOMID

**1.7** CAPS FCAS MWS CI

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NCA/NOMID

## 2.

### 2.1

CAPS FCAS MWS MWS CINCA/NOMID

### 2.2

CAPS CAPS

### 2.3

CAPS IL-1 $\beta$  IL-1 $\beta$  IL-1 $\beta$  CAPS Anakinra etanercept IL-1 FDA 11 FCAS MWS Canakinumab IL-1 FDA EMA 4 CAPS MWS 4-8 IL-1

### 2.4

CAPS

### 2.5

FCAS MWS CINCA CINCA/NOMID IL-1 CAPS

## 3

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### 3.1

### 3.2

### 3.3

### 3.4

### 3.5

### 3.6

### 3.7