



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

Cryopyrin-associated periodic fever syndrome (CAPS)

0000

1. Cryopyrin-associated periodic fever syndrome

1.1 Cryopyrin-associated periodic fever syndrome

Cryopyrin-associated periodic fever syndrome (CAPS) is a group of autoinflammatory syndromes characterized by recurrent febrile episodes (FCAS), Muckle-Wells syndrome (MWS), CINCA syndrome, and NOMID. CAPS is caused by mutations in the NLRP3 gene, which encodes the cryopyrin protein. FCAS, CINCA, and NOMID are characterized by recurrent febrile episodes, while MWS is characterized by skin rash and hearing loss. CAPS is a rare genetic disorder that affects the immune system.

1.2

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

1.3

CAPS is a group of autoinflammatory syndromes characterized by recurrent febrile episodes (FCAS, MWS, CINCA/NOMID) and skin rash. CAPS is caused by mutations in the CIAS1 (or NLRP3) gene, which encodes the cryopyrin protein. cryopyrin is a protein that is involved in the immune system. CAPS is a rare genetic disorder that affects the immune system. 30% of CINCA/NOMID patients have mutations in the CIAS1 gene. CAPS is a group of autoinflammatory syndromes characterized by recurrent febrile episodes, skin rash, and hearing loss.

1.4

CAPS is a group of autoinflammatory syndromes characterized by recurrent febrile episodes, skin rash, and hearing loss. 20% of patients have mutations in the CIAS1 gene. 50% of patients have mutations in the CIAS1 gene. CAPS is a group of autoinflammatory syndromes characterized by recurrent febrile episodes, skin rash, and hearing loss.

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

NCA/NOMID

2.

2.1

CAPS FCAS MWS MWS CINCA/NOMID

2.2

CAPS CAPS

2.3

CAPS IL-1 β IL-1 β IL-1 β 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

3.1

3.2

3.3

3.4

3.5

3.6

3.7