

# Sensorineural hearing loss in anti-Interleukin-1 treated Cryopyrin-Associated Periodic Syndrome (CAPS) patients: Modifiable risk factors and real-life barriers

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**Background:** Evidence on long-term outcomes of targeted therapies for hearing preservation in CAPS is limited. Identifying risk factors for hearing loss progression may guide monitoring, while real-life barriers to personalized treatment need to be addressed.

**Objective:** To identify modifiable risk factors associated with progression of hearing impairment from a longitudinal cohort of anti-IL-1-treated children and adults with Cryopyrin-Associated Periodic Syndromes (CAPS) and explore real-life barriers to optimal long-term management.

## Methods:

- A single-center longitudinal study included pediatric and adult anti-IL-1-treated CAPS patients with sensorineural hearing loss (2006–2024).
- Data included demographics, disease characteristics, genotype, treatment, and hearing assessments (4PTA, HF-PTA).
- Primary outcome: WHO grade of hearing impairment at last follow-up.
- Factors influencing hearing impairment and therapy escalation barriers were identified.

## Results

- 36 CAPS patients (20M/16F) with SNHL

### Age (Median, Range):

- Onset: 11.8y (0.5–50), Diagnosis: 35y (0.5–75), HL Diagnosis: 40y (6–75), Treatment Delay: 24.8y (0.5–71), Follow-up: 8.2y (1–18)

### Phenotypes:

- Moderate 83%, Severe 17%

### NLRP3 Variants:

- Pathogenic 39%, Likely Pathogenic 39%, VUS 14%, No Variant 8%

### Therapy:

- Initial: Anakinra 44%, Canakinumab 56%;
- Last Follow-up: Anakinra 21%, Canakinumab 71%, Combo 3%

### Hearing Loss:

- Baseline 83%, Follow-up 88%
- WHO Grades: Stable 86%, Improved 8%, Worsened 6%

### Therapy Escalation (SNHL Progression):

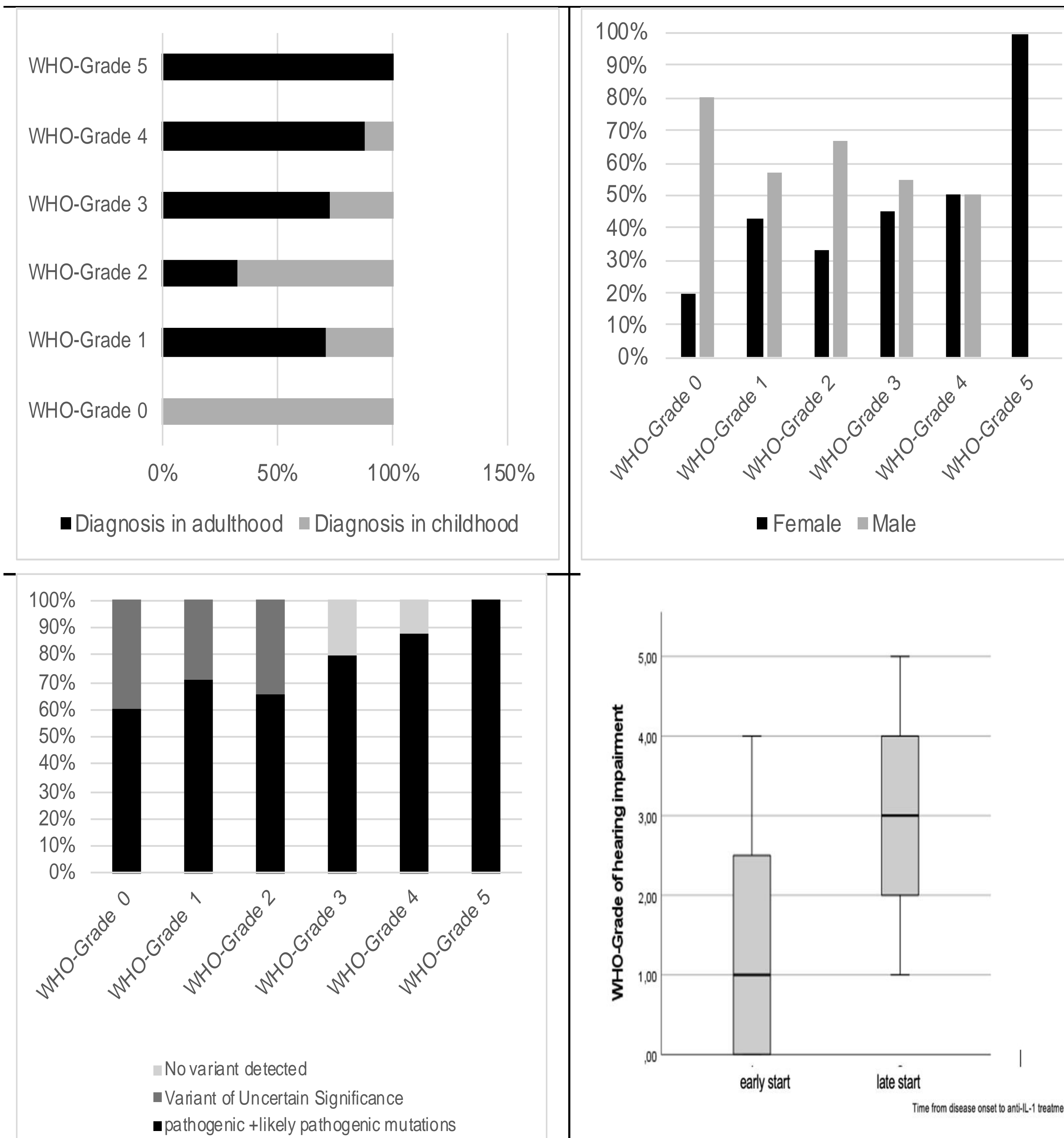
- 28% (n=10), Mean 5.4y

### Progression:

- Early: Partially controlled (n=3);
- Late: Noncompliance (n=4), Puberty (n=2), Pregnancy (n=1)

**Post-Escalation:** Improved 6, Stable 4

Figure : Association of patient- and treatment related factors with WHO grade of hearing impairment in CAPS patients



## Conclusions:

Early diagnosis, timely intervention, and a refined T2T approach are vital for hearing in lifelong CAPS management. Precision care and continuous monitoring are key to improving long-term outcomes.

## References:

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