THE ROLE OF AGE AND OTITIS MEDIA IN HEARING LOSS PATTERNS IN MUCOPOLYSACCHARIDOSIS

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Introduction

- Mucopolysaccharidosis (MPS)is a group of inherited metabolic disorders leading to glycosaminoglycan (GAG) accumulation
- 7 distinct clinical types; many subtypes
- MPS patients initially develop typically but later experience a decline in physical and/or cognitive function
- Hearing loss (HL) is a known complication
- What is the prevalence and relationship of otitis media (OM) with HL in MPS patients?

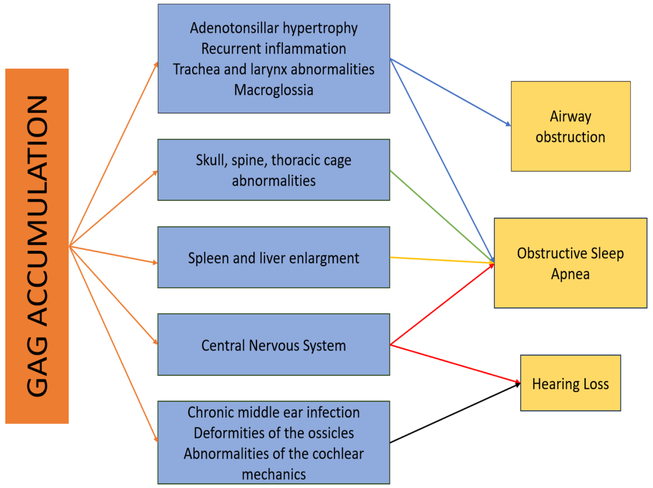

Figure 1. Otolaryngological problems due to GAG accumulation.

Methods

- Retrospective cohort study using TriNetX US Network
- Data from Jan 01, 2006 to Dec 31, 2024
- Data collected on Jan 26, 2025
- MPS patients of all age groups were identified via ICD-10 E76
- Split in 2 groups: OM and Non-OM
- 1:1 propensity score matching (PSM) by demographics
- Comparison of HL prevalence between matched cohorts
- OM patients of all age groups were identified via ICD-10 H65-67
- Split in two groups MPS and Non-MPS
- 1:1 PSM by demographics
- Comparison of HL prevalence between matched cohorts.

Results

- All types of HL were more frequent in MPS patients with OM compared to Non-OM:
  - CHL (21.4% vs. 3.2%; **OR=8.4**, p<.001)
  - SNHL (29.7% vs. 9.3%; **OR=4.1**, p<.001)
  - MHL (16.9% vs. 2.5%; **OR=8.0**, p<.001)
- All types of HL were more frequent in OM patients with MPS compared to Non-MPS OM patients:
  - CHL (22.2% vs. 6.7%; **OR=4.0**, p<.001)
  - SNHL (30.7% vs. 3.2%; **OR=13.2**, p<.001)
  - MHL (17.7% vs. 1.7%; **OR=12.4**, p<.001)

Discussion

- Our data indicate a significant occurrence of sensorineural hearing loss (SNHL) in MPS patients, particularly among older children and adolescents
- This trend suggests a progressive nature of SNHL in the MPS population, necessitating regular auditory assessments

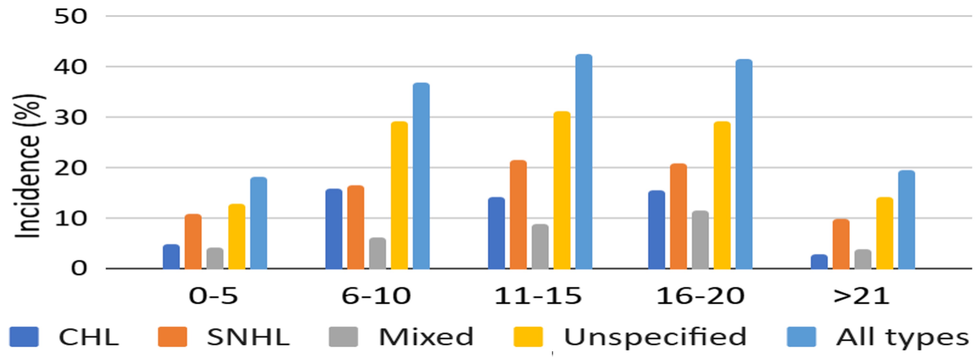

Figure 2. Incidence of Hearing Loss in MPS Patients: Age at Diagnosis

Hearing Loss Type, N(%)	OM	Non-OM
Sensorineural	169 (29.7)	53 (9.3)
Conductive	122 (21.4)	18 (3.2)
Mixed	96 (16.9)	14 (2.5)
Unspecified	233 (40.9)	78 (13.7)
All Hearing Loss	325 (57.1)	113 (19.9)

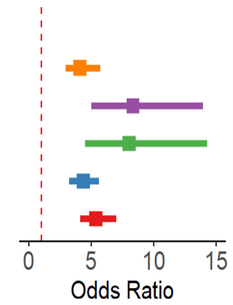

Figure 3. The effect of OM on HL outcomes in MPS patients

Hearing Loss Type, N(%)	MPS	Non-MPS
Sensorineural	189 (7.2)	44 (1.7)
Conductive	53 (2.0)	15 (0.6)
Mixed	53 (2.0)	10 (0.4)
Unspecified	248 (9.4)	61 (2.3)
All Hearing Loss	376 (14.3)	92 (3.5)

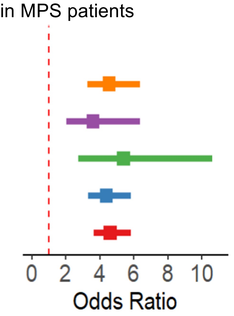

Figure 4. The effect of MPS on HL outcomes in Non-OM patients

- Strong association between OM and increased risk of all types of hearing loss in MPS patients
- Highlights MPS as an independent risk factor for more severe auditory complications following OM episodes

Hearing Loss Type, N(%)	MPS	Non-MPS
Sensorineural	180 (30.7)	19 (3.2)
Conductive	130 (22.2)	39 (6.7)
Mixed	104 (17.7)	10 (1.7)
Unspecified	244 (41.6)	49 (8.4)
All Hearing Loss	338 (57.7)	81 (13.8)

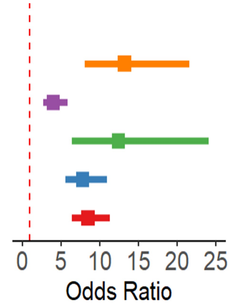

Figure 5. The effect of MPS on HL outcomes in OM patients

Conclusion

- Significant otolaryngological complications in patients with MPS, notably an **increased risk of hearing loss** associated with **OM**
- Interplay between MPS, OM, and hearing loss necessitates a proactive and comprehensive approach to auditory health in MPS patients
- Further research is warranted to explore targeted interventions that can improve hearing outcomes and overall quality of life for MPS patients

References