

Exploring the Benefits of Personal Remote Microphones in the Rehabilitation of Central Auditory Processing Disorder in Mitochondrial Disease Patients

Nehzat Koohi; Sarah Holmes; Amada Male; Doris-Eva Bamiou; Yusra El-Makhzoumi; Komul Vaghela; Shivani Dattani; Robert Pitceathly; Diego Kaski
University College London

BACKGROUND

- Auditory processing disorders impact speech perception, sound localisation, and temporal processing.
- These issues are common in Mitochondrial Disease (MD) and Friedreich's Ataxia.
- Conventional hearing aids often fail to address these challenges, particularly in noisy environments.
- Personal remote microphones (PRMs) show promise by improving speech perception through improved signal-to-noise ratios.
- Usability, ergonomics, and cost challenges remain barriers to widespread PRMs adoption.

Aim

To evaluate the potential benefits of hearing aids coupled with PRMs on speech perception in background noise, and assess usability and patient-reported outcomes over six weeks

METHODS

Sample: 5 MD patients (m.3243A>G) with mild to moderate SNHL and APD. Group included 2 males and 3 females (mean age 52; SD = 10.10).

Measures: PRM Efficacy: Bamford-Kowal-Bench Speech-in-Noise Test (BKB-SIN) test with/without hearing aids, Roger Pen,

Subjective Change: Speech, Spatial, and Qualities questionnaire (SSQ), Listening Effort Assessment Scale (LEAS).

Focus Group: Feedback on PRM usability and challenges.

Analysis: SPSS 29 ($p < 0.05$), One-way ANOVA (week 0 vs week 6), Repeated measures ANOVA (pre/post-test).

RESULTS

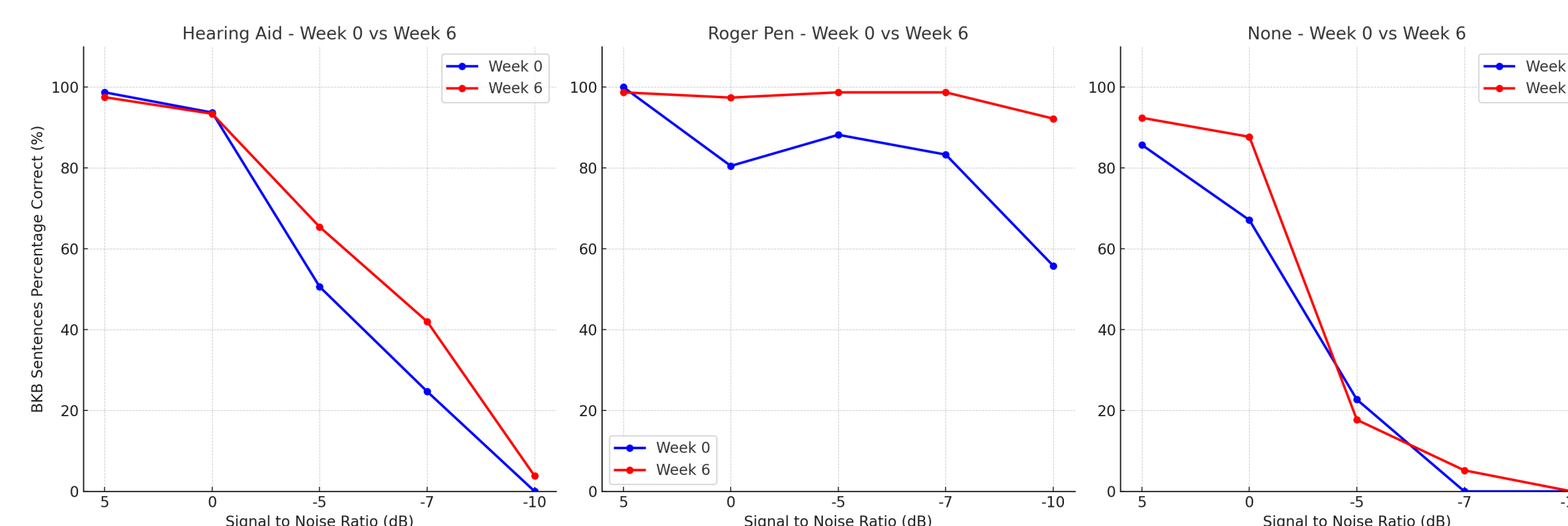


Figure 1: The graph compares the BKB sentence recognition percentages across different signal-to-noise ratios (dB) at Week 0 and Week 6, using Roger Pen, hearing aids, and no devices. Significant improvements were observed with the Roger Pen, especially in challenging listening conditions.

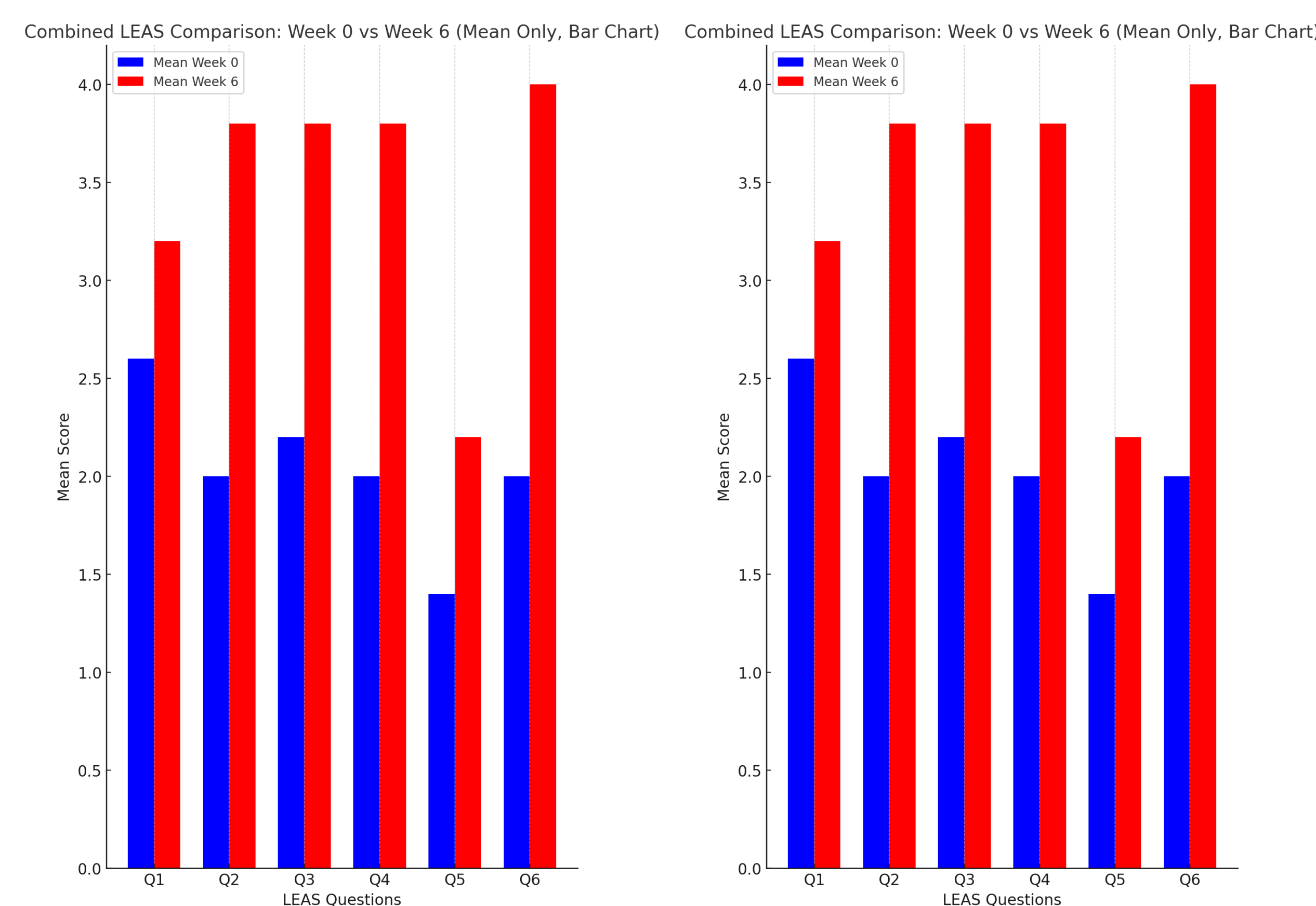


Figure 2: Mean SSQ scores for Speech, Spatial, and Qualities components at Week 0 and Week 6. No significant changes were observed across these components between the two time points ($p > 0.05$).

Figure 3: Mean LEAS scores for Week 0 and Week 6, covering effort in quiet (Q1), moderate noise (Q2), loud environments (Q3), fatigue (Q4), group conversations (Q5), and concentration in daily listening (Q6). A significant difference was observed between Week 0 and Week 6 ($p < 0.05$).

DISCUSSION

- **Hearing rehabilitation challenges:** Patients with mitochondrial disease often face sensory overload and social isolation due to inadequate auditory care. Objective speech-in-noise testing showed that hearing aids were ineffective in signal-to-noise ratios below 0 dB, complicating rehabilitation.
- **PRM benefits:** Significant improvements in listening effort (LEAS) and BKB sentence scores were observed with the PRM in speech-in-noise perception.
- **Patient feedback:** Most participants reported overall improvements in speech clarity and listening effort. However, difficulties with setup and use in noisy environments were common.
- **Device limitations:** Complex settings and limited functionality in certain environments. A simplified, technology-integrated solution and adaptable PRM designs are needed.
- **Broader impact:** Early, individualised rehabilitation is essential to mitigate the link between hearing loss and cognitive decline, supporting better quality of life.

REFERENCES

1. Rance G. Auditory neuropathy/dys-synchrony and its perceptual consequences. *Trends Amplif.* 2005;9(1):1-43. doi: 10.1177/108471380500900102. PMID: 15920648; PMCID: PMC4111505.
2. Koohi et al., 2024. Beyond the Cochlea: Exploring the Multifaceted Nature of Hearing Loss in Primary Mitochondrial Diseases. *Brain Communications* (in press). Crum R, Chowsilpa S, Kaski D, Giunti P.
3. Bamiou DE, Koohi N. Hearing rehabilitation of adults with auditory processing disorder: a systematic review and meta-analysis of current evidence-based interventions. *Front Hum Neurosci.* 2024 Jun 21;18:1406916. doi: 10.3389/fnhum.2024.1406916. Erratum in: *Front Hum Neurosci.* 2024 Aug 13;18:1468962. doi: 10.3389/fnhum.2024.1468962. PMID: 38974481; PMCID: PMC11224551.

ACKNOWLEDGEMENTS

We are grateful to the Lily foundation, and participants and families involved in this study.