Cochlear Implantation in Individuals with Usher Syndrome: Speech Perception Outcomes and Implications for Quality of Life

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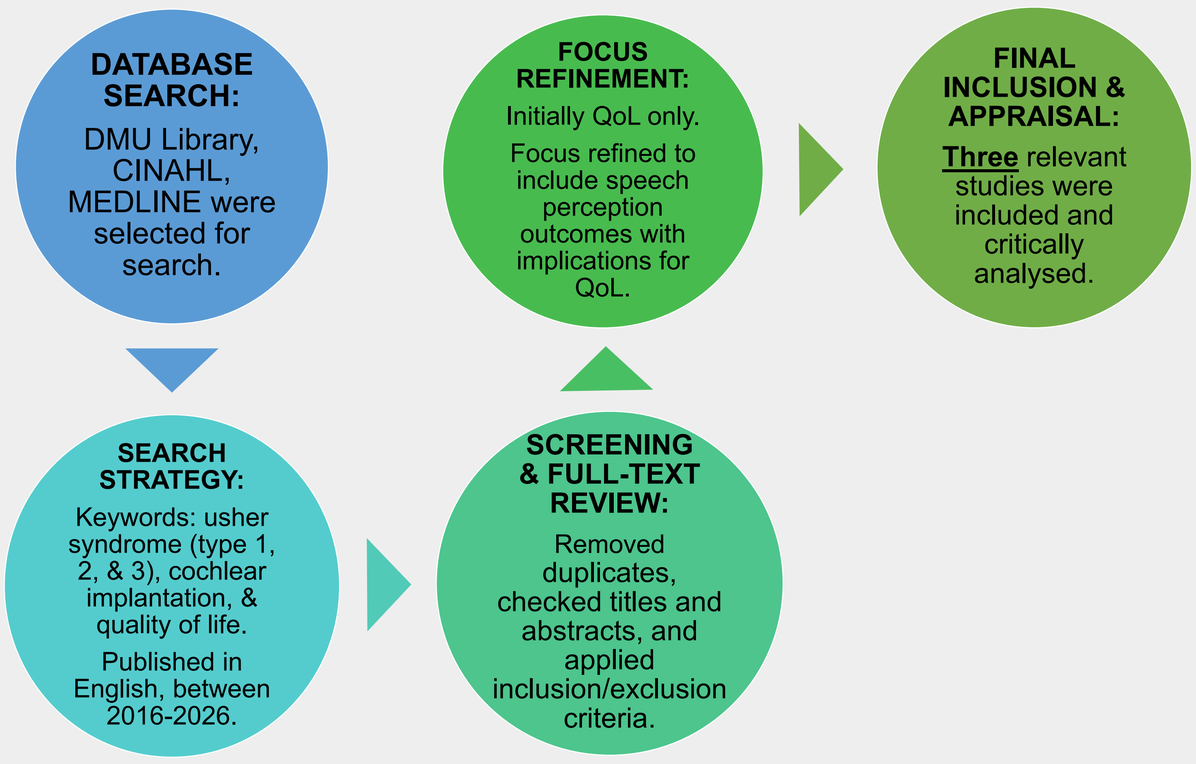
INTRODUCTION

Usher syndrome (USH) is a rare, inherited condition and a cause of sensorineural hearing loss (HL) and retinitis pigmentosa (RP).
Cochlear implantation (CIs) is a key intervention used to manage severe to profound HL, providing access to sounds and supporting speech perception (NIDCD, 2024; NICE, 2019). Dual sensory impairment significantly impacts quality of life (QoL), with increased risks of social isolation, depression, and reduced independence (Dean et al., 2017; Cosh et al., 2017; Chou, 2008).
CI is associated with improvements in speech perception, although outcomes are highly variable and have multiple influencing factors (Cornwall et al., 2023).
However, evidence is often limited due to small sample sizes and retrospective designs, which impact generalisability (Davies et al., 2021).
Accordingly, the current study aims to critically evaluate current evidence on speech perception outcomes following cochlear implantation in individuals with USH and to explore the implications of these outcomes for QoL.

METHODOLOGY



RESULTS

Author (Year)	Sample & Hierarchy	Speech Perception	Factors Affecting Speech	Timing of CI	QoL Outcomes
Fehrmann et al. (2024)	n=35 Level 3	Significant improvement with stable long-term scores	Age and auditory experience influence outcomes	Early and simultaneous CI resulted in better outcomes	Not directly measured, but included in discussion of results
Hartel et al. (2017)	n=8 Level 3	Improved phoneme scores	Prior hearing aid use may support outcomes	Not explored	Improved QoL and highlighted that ongoing support is needed
Hoshino et al. (2017)	n=10 Level 3	Variable improvement with some limited benefit	Limited pre-implantation stimulation resulted in poorer outcomes	Later implantation linked to poorer results	Emphasised patient-centred care and benefits from sound awareness

THEMES

Speech Perception Following Cochlear Implantation

- CI consistently improved phoneme scores across the USH types.
- Significant gains in USH2 (41% to 86%) and USH3 (15% to 71%).
- Improvements in 3/4 individuals with USH1.
- Overall, findings support CI as effective for improving functional speech perception in USH.

Factors Affecting Speech Perception

- Better scores occur when the hearing loss is post-lingual and if auditory stimulation is consistent pre-CI
- Rehabilitation engagement and cognitive factors may further influence outcomes, especially in late implantation
- Overall, evidence supports early CI and auditory stimulation to optimise outcomes, and is consistent with NICE (2019) guidance.

Outcomes Associated with the Timing of Implantation

- Early CI is associated with substantially higher long-term phoneme scores compared to late CI.
- Simultaneous bilateral CI showed better speech perception outcomes than sequential, supporting current NICE (2019) guidance.
- Overall, early and simultaneous CI optimise speech perception and functional hearing.

Implications for Quality of Life

- CI improves speech perception, with positive impacts on communication and daily functioning.
- Significant improvements in general wellbeing, reflecting increased confidence and social engagement.
- However, ULS highlighted persistent functional limits in independence, dual sensory functional challenges, and reliance on assistive devices and support in complex tasks.

CONCLUSIONS AND FUTURE DIRECTIONS

The present findings support CI as a successful alternative for individuals with USH. Additionally, the importance of early identification and intervention, consistent auditory stimulation, structured rehabilitation, and patient-centred counselling is also highlighted.

Unfortunately, evidence is limited by small sample sizes, retrospective designs, and controlled clinical settings, which reflects the available literature on USH. This highlights the need for further, large-scale and longitudinal research, particularly focusing on the functional and QoL outcomes within this population.

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