



Auditory Performance Outcomes for Cochlear Implant Users with Mondini Dysplasia: the Need for Consistency and Collaboration

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Why study Mondini Dysplasia and Cochlear Implant Outcomes?

- Cochleae with Mondini Dysplasia are often amenable to cochlear implantation but knowledge about performance outcomes is limited.
- The degree of benefit and long-term prognosis is poorly understood since patient populations are small.
- There are no recent studies within the UK that have considered performance outcomes for this particular cohort of implant users.
- An understanding of current research coupled with the identification of gaps in the knowledge base can inform clinical practice and guide future research.

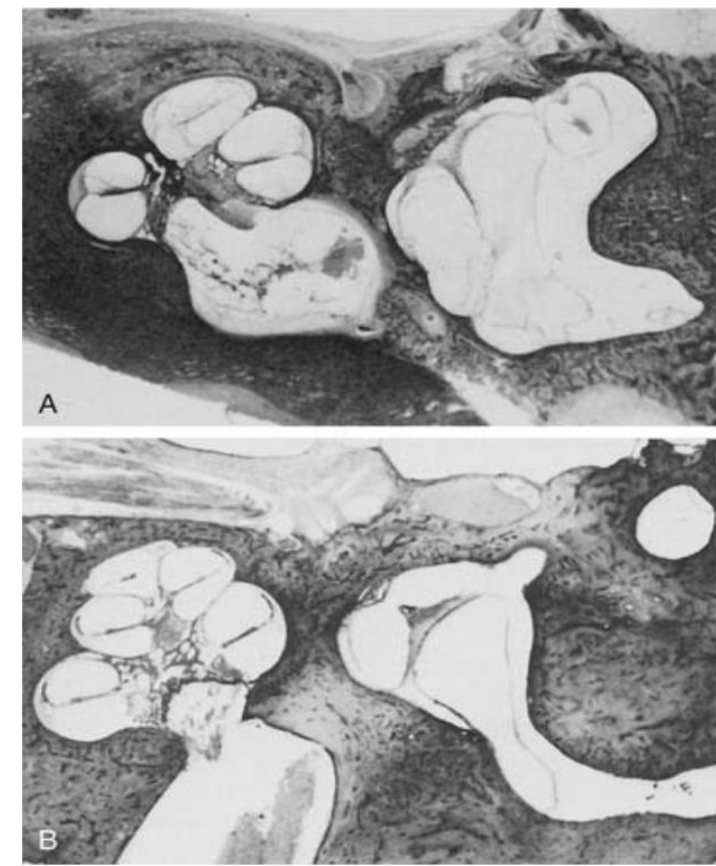


Figure 1 (A) Mondini cochlea with the characteristic **Triad of Malformation**: cystic cochlear apex, enlarged vestibular aqueduct, dilated vestibule (B) Normal cochlea. [https://entokey.com/congenital-malformations-of-the-inner-ear/]

Scoping Review: Aims

- To identify performance outcomes for implant users with Mondini Dysplasia reported in the published literature
- To appraise the available evidence (Quality Assessment and Risk of Bias Assessment)
- To assess whether device-related data is utilised in the determination of auditory performance

Method

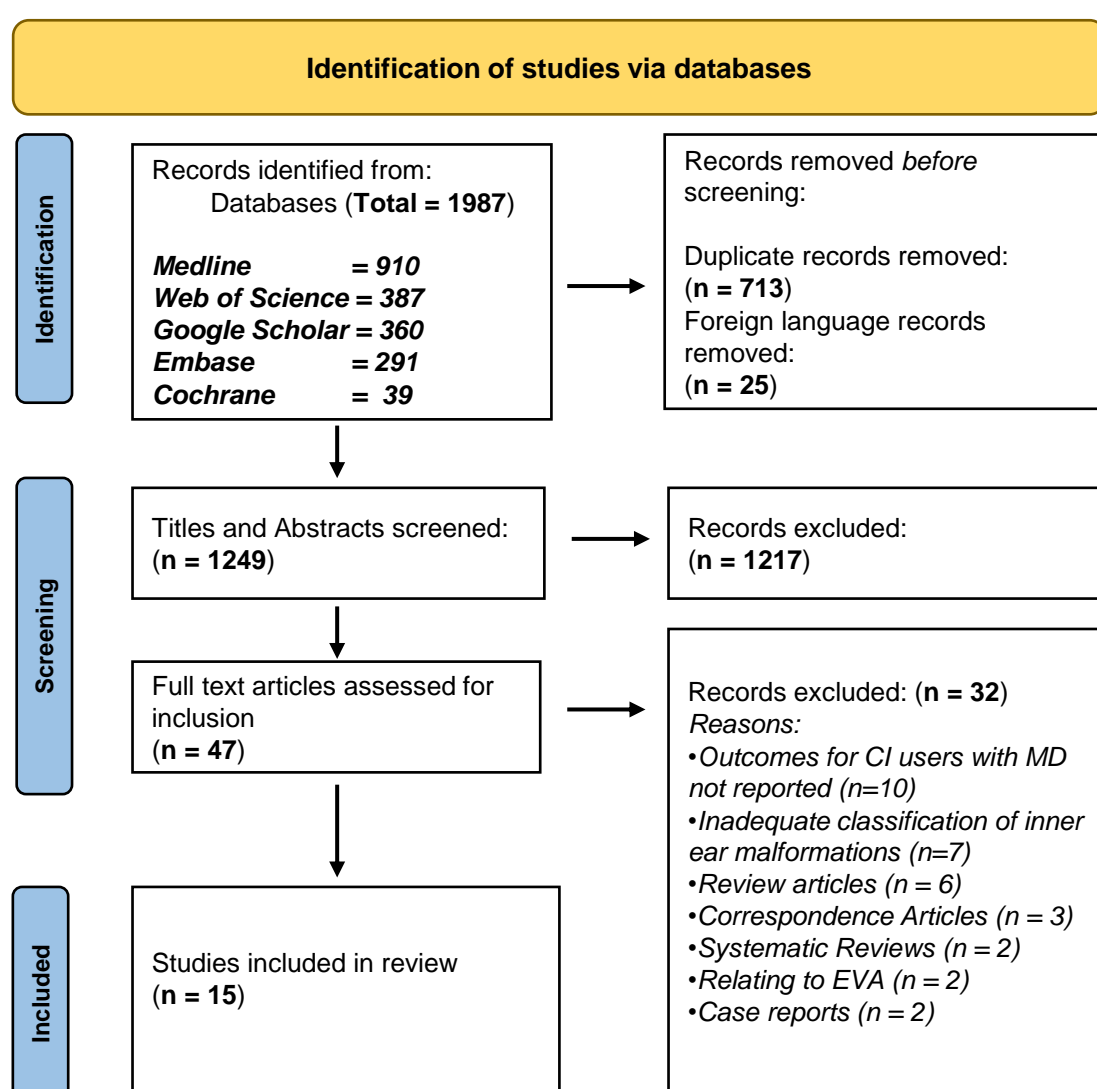


Figure 2. PRISMA* Flow Diagram. The search and selection process involved in identifying relevant articles for inclusion in the scoping review. *PRISMA: Preferred Reporting Items for Systematic Reviews and Meta-Analyses

Results: 15 Studies with Limited Follow-Up

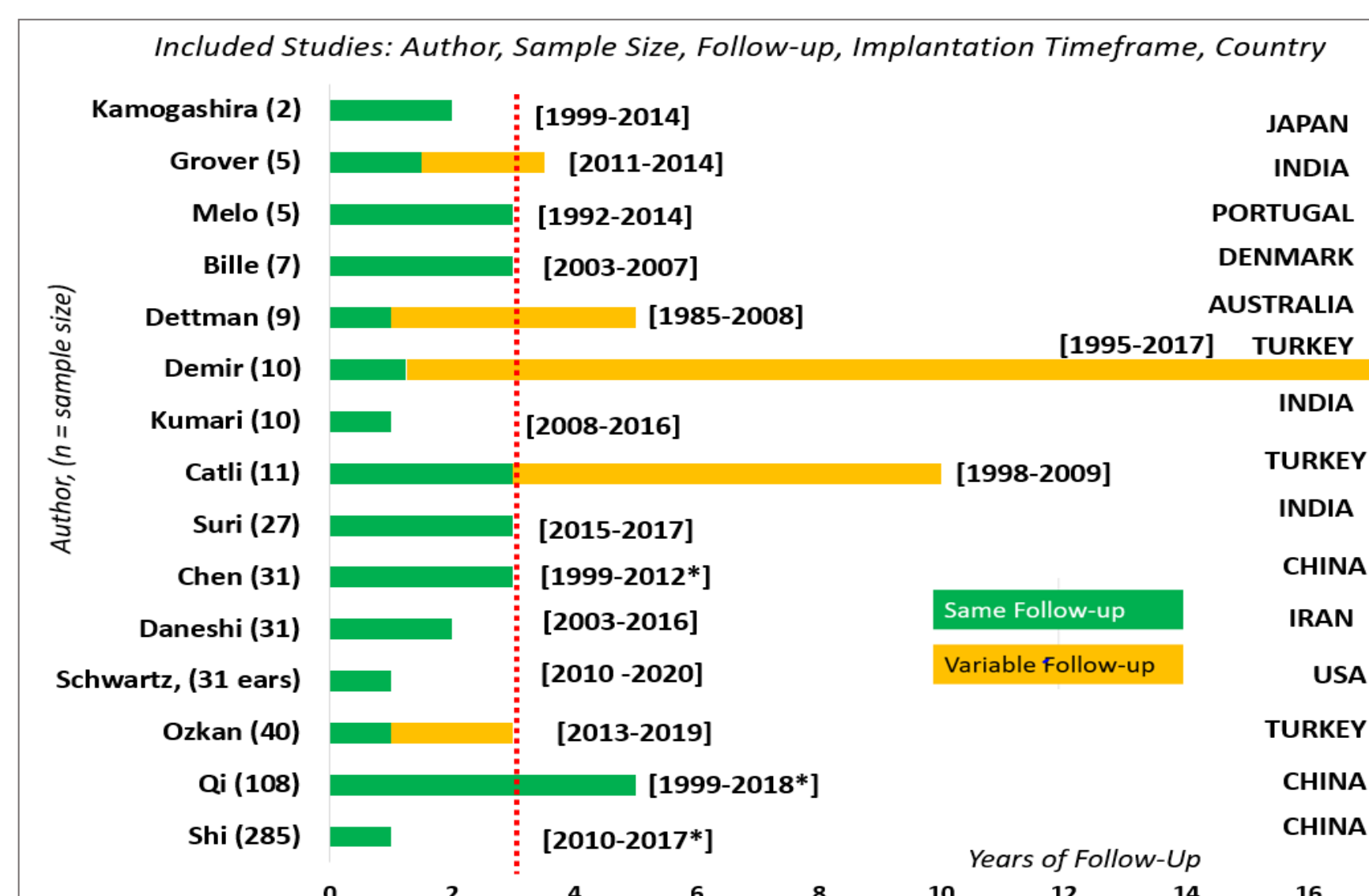


Figure 3. Follow-up Periods in Included Studies. A summary of included studies with sample size in brackets beside author name. Follow up was generally 3 years or less denoted by the vertical dotted line. The implantation timeframe for each study is included in square brackets. The country in which the study took place is indicated in the right margin. (*) denotes studies that took place within the same institution

Risk of Bias Assessment

Author, Year of publication	Selection	Assessment of exposure	Outcome absent at start	Matching of variables	Prognostic factors	Assessment of outcome	Adequate follow up	Similar co-interventions	Overall Risk of Bias
Bille, 2015	+	Y	-	Y	N	+	+	Y	Medium
Catli, 2015	+	+	-	Y	N	+	+	N	Medium
Chen, 2015	+	N	-	Y	N	N	+	N	Medium-High
Kamogashira, 2015	+	+	-	Y	N	N	N	N	Medium-High
Kumari, 2021	+	+	-	Y	N	+	-	Y	Medium-High
Melo, 2017	+	+	-	Y	N	+	N	N	Medium-High
Ozkan, 2021	+	+	-	Y	N	Y	N	Y	Medium
Qi, 2019	+	+	-	Y	N	N	-	N	Medium-High
Suri, 2021	+	+	-	Y	N	+	+	Y	Medium

Figure 4. Appraisal of Evidence. Included studies with a good quality rating on the Newcastle-Ottawa Rating Scale were assessed using the Cochrane Risk of Bias Tool for Cohort Studies

+	Y	N	-
Definitely yes (low risk of bias)	Probably yes	Probably no	Definitely no (high risk of bias)

Conclusions:

Auditory Performance Outcomes (1-3 years post implantation)

- **Categories of Auditory Performance score (CAP) = 5-7**
Able to follow conversation without lipreading
Speech Intelligibility Rating (SIR) = 3-5
Able to produce connected speech that is mostly intelligible
(Evidence from 4 good quality studies; medium risk of bias)
- The development of auditory perception and speech occurred at a **slower rate** compared to those with normal cochlear architecture but there was no difference between groups 1-2 years post implantation
(Evidence from 3 good quality studies; medium-high risk of bias)
- None of the studies utilised device-related data in the assessment of auditory performance

Future Research: The need for consistency & collaboration

- Consistency in the classification of Mondini Dysplasia (ensuring all three features of the triad of malformation are present), and consistency in the type of outcome measure used are necessary to enable collaborative studies that are sufficiently powered.
- Those who become non-users are typically excluded from studies which may skew understanding of the true benefit of cochlear implants for these users.
- Future studies must have longer follow-up periods and utilise objective measurements from the implant itself (data-logs, impedance measures, patterns in electrode deactivation) to better understand performance over time.