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How Complete is the Reporting of the Mondini Triad in a Cohort of Patients with Cochlear Implants? A Service Evaluation.

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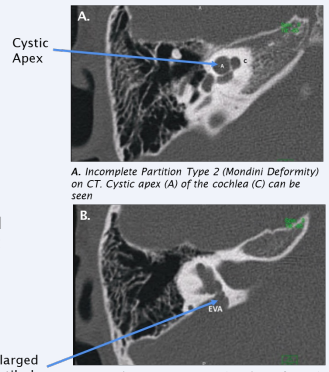
Introduction

Identification of the Mondini triad is needed for planning surgical interventions as the malformation is associated with a greater risk of complications (1). Advising patients about possible complications is important to allow for informed decision making. Prompt, accurate diagnosis of malformations would allow children with hearing loss to undergo surgery early, enabling the benefits of cochlear implantation within the first year of life to be realised (2,3).

A Mondini deformity is a congenital inner ear malformation. A shortened cochlea with a cystic apex; dilated vestibule and an enlarged vestibular aqueduct comprise the triad (4). The triad should be present on radiological examination for a Mondini's diagnosis to be made. People with Mondini malformations can achieve similar speech acquisition to people with anatomically normal ears, however this is after a longer rehabilitation period, and not seen in all people (4).

There is a need for more research into the the association between Mondini malformations and hearing outcomes. This could enhance patient counselling and inform rehabilitation after implantation. Studies need sizeable cohorts from multiple centers', and accurate reporting of anatomy and hearing performance. This study paves the way for this by evaluating the consistency of reporting of the triad in patients (figure 2) from a single center.



A. Incomplete Partition Type 2 (Mondini Deformity) on CT. Cystic apex (A) of the cochlea (C) can be seen

B. Incomplete Partition Type 2 (Mondini Deformity) on CT. Shows an enlarged vestibular aqueduct (EVA).

Figure 1: CT Images of Mondini's Dysplasia. Sourced from Sennaroğlu et al. 2017 (4).

Methods

Patient data for this evaluation [ERGO 76600] was collected from medical records stored at USAIS.

The records of all patients with a structural inner ear malformation and a syndromal cause of hearing loss associated with Mondini's were checked.

All participants were assigned anonymised study numbers.

Data from participants was considered against the inclusion and exclusion criteria.

Data was analysed using descriptive statistics in Excel and in some cases binomial regression.

Findings

64% (n=36) of the ears identified had a complete Mondini triad (Figure 3). A dilated vestibule was the feature most commonly not reported. A cystic apex or an enlarged vestibular aqueduct, features that were the surgically relevant or affected candidacy for implantation, were recorded more frequently.

44% of the Mondini cohort were implanted aged 5 or under (Figure 2), comparatively older than the wider USAIS cohort, where 9% of patients were implanted aged 5 or under. This is likely due to the increased identification of congenital hearing loss through new-born hearing screening.

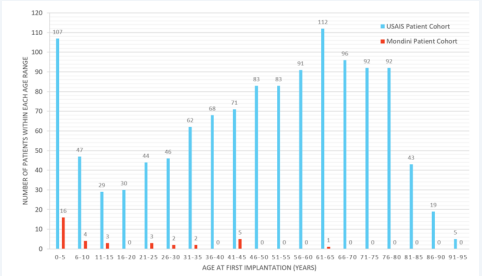


Figure 2: Age range of the patients within the Mondini cohort (red bar) at the time they were implanted compared with those from the entire USAIS patient cohort (blue bar).

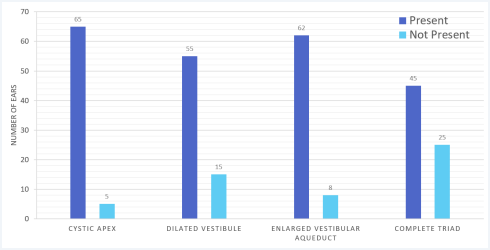


Figure 3: Presence of the Mondini triad features within the Cohort
Presence of the three features of the Mondini triad in the cohort, and the presence of the complete and incomplete triad within the cohort. 65 ears (92%) had CA reported as present, and 5 ears (8%) did not have the feature reported. An EV was reported as present in 55 ears (78%) within the Mondini cohort. The other 15 ears (21%) did not have the feature reported. An EVA is reported as present in 62 ears (89%). 8 ears (11%) within the cohort did not have an EVA reported.
CA = Cystic Apex, DV = Dilated Vestibule, EVA = Enlarged Vestibular Aqueduct.

To determine whether patients were diagnosed consistently regardless of their age when they were scanned, we looked to see if a specific feature was commonly reported as not present in any particular age range across the cohort. When looking at dilated vestibules this identified that a large proportion of the patients without the feature recorded, were aged between three – ten yrs. Similarly, when looking at the recording of an enlarged vestibular aqueduct we observed that a large proportion of the patients without the feature recorded, were aged under 3 yrs. Binomial regression analysis of these findings demonstrated a statistically significant difference between the test group and control groups.

Clinical Impact

Our evaluation showed that USAIS was effective in identifying features of the Mondini triad which effected patient management. Development of precise criteria for a Mondini malformation in instances where the triad is complete, along with alternatives for patients without the full triad would improve the completeness of malformation reporting at the service.

Changes to the classification of malformations would allow for improved surgical planning to prevent complications. People with Mondini's are at increased risk for a variety of surgical complications, but most commonly CSF leaks of varying severity. Within the USAIS cohort 55% of patients had a CSF leak. Complications have an adverse effect on post-operative audiological outcomes, changes to surgical planning to reduce them could help mitigate this effect.

Future Work

Following on from this work we would like to carry out similar evaluative studies in collaboration with other UK centres, using this service evaluation as a comparison for the data required. This could pave the way for statistically well powered research that enables the features of Mondini's to be prognostic of audiological outcomes and which may inform implant development.

Multi-centre data could better inform pre-operative patient counselling, enabling patients, parents and guardians to more fully informed about the balance of risks and benefits of implantation. This in turn could be used to improve the standard of patient care.

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All Ears PPIE Group Website

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AUDITORY IMPLANT SERVICE USAIS MAKING CONNECTIONS