Preliminary Trends Observed Following Cochlear Implantation of Children with Auditory Neuropathy Spectrum Disorder or Hypoplastic VIII Nerves or Both

- Marsha Jenkins, Jannet Horton, Linda Baxter, Lauren Hegarty - St Thomas' Hearing Implant Centre, Guy's and St Thomas' NHS Foundation Trust, London, UK
- Mark Chung, Lisa Nash, Azhar Shaida, Hala Kanona - Auditory Implant Programme, Royal National ENT and Eastman Dental Hospitals, University College London Hospitals NHS Foundation Trust, London, UK.
- Peter Keating – Ear Institute, University College London (UCL), London, UK.

NHS University College London Hospitals NHS Foundation Trust

NHS Guy's and St Thomas' NHS Foundation Trust

Background

- The current UK NHSP protocol for children diagnosed with Auditory Neuropathy Spectrum Disorder (ANSD) recommends longer monitoring of behavioural results before consideration for cochlear implant (CI) referral. It follows then that this cohort of children are often referred to CI centres after the age of one (BSA, 2019).
- It has long been established that optimal outcomes with CI are achieved the younger the age of implantation (Cowan et al, 2018).
- The challenge in predicting outcomes for children with ANSD and hypoplastic VIII nerves is well recognised and families are often counselled conservatively into what outcomes may be achieved.
- This is a multi-centre retrospective review, including children implanted either unilaterally or bilaterally who have a diagnosis of ANSD and/or hypoplastic VIII nerves or both.

Methods

A total of 52 children implanted between 2008-2023 at the Hearing Implant Centre (St. Thomas') and the Auditory Implant Programme (UCLH) were included.
Both audiological and functional outcomes were analysed in relation to the 3 categories of presentation below:

1. **TN = Hypoplastic nerve/s (T) with SNHL (N)**
2. **NA = Normal nerves (N) and ANSD (A)**
3. **TA = Hypoplastic nerve/s (T) and ANSD (A)**

Children with complex needs, within each group, have been highlighted to take these factors into consideration.

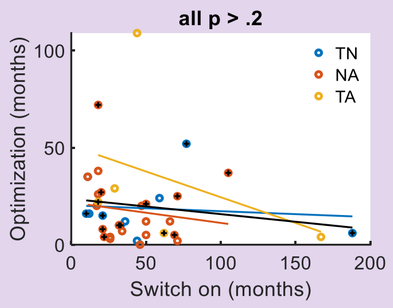
Results

- Audiological outcomes were analysed, namely: age of implantation; device use and mapping parameters required. Particular interest was focussed on time to optimisation for the 3 groups as it has been questioned whether this cohort takes longer to reach this point. Optimisation is the timeline from implantation to good aided access achieved in at least 3 frequencies.

- Functional outcomes were also analysed, including: auditory perception; mode of communication and speech intelligibility. These outcomes were highly correlated with one another, and showed similar trends (data not shown). Consequently, different outcome measures were normalized (z-score conversion) and averaged to create combined functional outcomes.

Results: Audiological

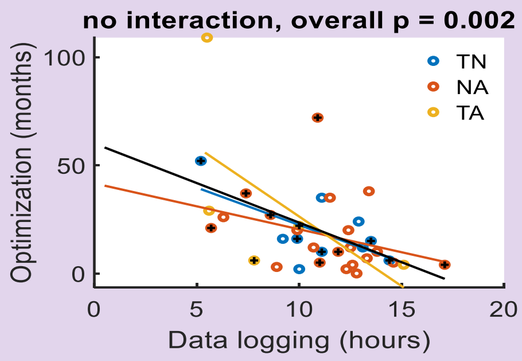
1. Does time to optimisation depend on age at initial activation?



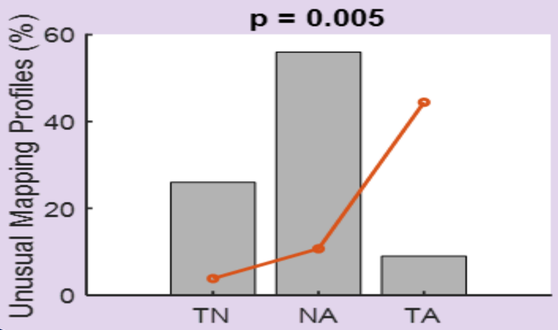
Age of activation does not make a difference on optimisation rate for any of the groups (ANCOVA; p>0.05).

2. Does time to optimisation depend on datalogging?

Higher data logging values are associated with faster optimisation times overall (ANCOVA; p<0.05) but the relationship does not differ significantly across groups (ANCOVA; p > 0.05) .



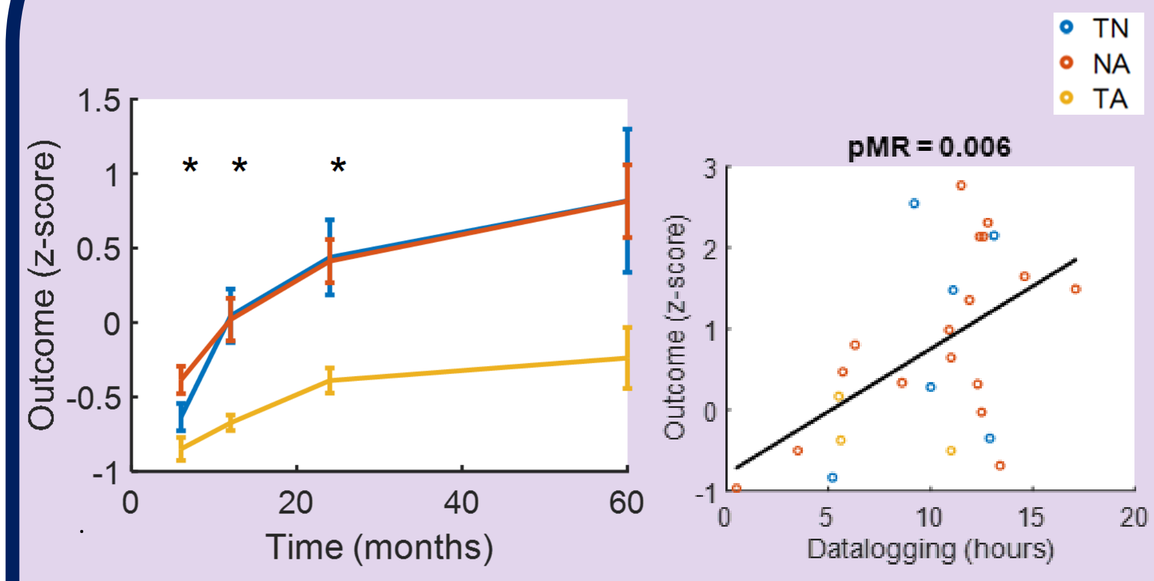
3. Is there a difference between groups for mapping parameters used?



The TA group is significantly more likely to show unusual mapping parameters compared to the other groups (post-hoc test; p<0.05; Chi-Square test). The prevalence of complex needs was similar across groups (Chi-Square test; p>0.05; data not shown).

Results: Functional

1. How does overall outcome improve over time?



- Combined functional outcomes were worse for TA (yellow) than the other two groups (post-hoc tests; p<0.05; mixed effects ANOVA)

- Combined functional outcomes also improved at each time-point within the first 2 years (post-hoc tests; p<0.05; mixed effects ANOVA)

- However, combined functional outcomes improved at similar rates in different groups (no significant interaction; p>0.05; mixed effects ANOVA).

- Higher datalogging is associated with greater benefits over time (ANCOVA, p<0.05) however there is no difference between groups (ANCOVA, p>0.05).

Preliminary Conclusions and Trends

- The time taken to optimisation did not depend on the age at switch-on across groups. This may be because it's easier to optimize older children.
- The time taken to optimisation does however depend on the datalogging and the relationship does not differ significantly across the 3 groups.
- The TA group are more likely to have unusual mapping parameters compared to the TN & NA groups.
- The TA group tend to perform worse overall than either TN or NA groups. The trends are similar across different functional outcome measures. This is at least partly because different outcome measures are highly correlated with one another.
- Specific counselling for TA families could be recommended as their outcomes are significantly different from the TN and NA groups.
- ***Future directions – continued monitoring of these groups to observe if these trends continue. Compare this cohort with age matched controls for further information on how the 3 groups compare to CI children without ANSD and/or hypoplastic VIII nerves***.

Title: Trends observed following Cochlear implantation of children with ANSD or ANSD with hypoplastic nerves or hypoplastic nerves only.

Authors: Marsha Jenkins, Jannet Horton, Linda Baxter, Lauren Hegarty - St Thomas' Hearing Implant Centre, Guy's and St Thomas' NHS Foundation Trust, London, UK.
Mark Chung, Lisa Nash - Auditory Implant Programme, Royal National ENT and Eastman Dental Hospitals, University College London Hospitals NHS Foundation Trust, London, UK.
Peter Keating - University College London (UCL), London, UK.

Abstract

Introduction:
The GSTT and the UCLH Paediatric Implant teams have come together to perform a multi-centre retrospective review. The review includes children implanted either unilaterally or bilaterally who have been diagnosed with ANSD only, or ANSD with hypoplastic nerves, or only hypoplastic nerves. These categories have been defined as they can be confused in the literature. The main objective of this review is to see if these different categories yield different outcomes. The current NHSP protocol used in the UK for children diagnosed with ANSD however require longer monitoring for behavioural results before being considered for an implant referral and thus these children are often referred to the implant centres after the age of one (BSA, 2019). It has long been established that the best outcomes with Cochlear implants are achieved the younger the child is implanted (Cowan et al, 2018). The difficulty in predicting outcomes for children with hypoplastic nerves has long been known and often families are counselled conservatively into what outcomes could be achieved. This review will look in detail at the outcomes of the various Anatomical and Audiological presentations.

Method:
A total of 51 patients implanted between 2008-2023 were included. We will be analysing Audiological and Functional outcomes in relation to the 3 categories of presentation. The presentations are as follows:
TN = Hypoplastic (T) nerve with SNHL (N)
NA = Normal nerve (N) and ANSD (A)
TA = Hypoplastic (T) and ANSD (A)
The patient groups have also been classified to take into consideration children presenting with different complex needs.

Results:
The following Audiological outcomes will be analysed, Age of implantation, Device use and Mapping parameters required. Particular interest will be focussed on time to optimisation for the above groups as it has been questioned whether this cohort takes longer to reach this point. Optimisation is the time line from implantation to good aided access achieved in at least 3 frequencies. Functional outcomes were also be assessed including Auditory perception, Mode of Communication and Speech intelligibility.

Discussion:
The review of joint work between GSTT and UCLH of the presentations and outcomes of this cohort will be presented in detail. Trends appearing from the above analyses will be discussed and possible recommendations made.