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## Abstract

It is estimated that 7 to 10% of children with permanent hearing loss have a diagnosis of auditory neuropathy spectrum disorder (ANSD). Due to the unique nature of this disorder, audiologic management of children with ANSD is particularly challenging. This study analyzed the timeline from diagnosis to intervention for a group of children with ANSD followed at UNC Hospitals. Implications for audiologic management are also considered.

## Background

### What is ANSD?

ANSD is a disorder characterized by a disruption in the transmission of an auditory signal from the inner ear to the brain. Clinical findings typically include absent or markedly abnormal auditory brainstem responses (ABR) combined with evidence of outer hair cell activity, as demonstrated by present otoacoustic emissions (OAEs) or a present cochlear microphonic (CM). Functional hearing status in children with ANSD is highly variable, with pure tone thresholds ranging from within normal limits to profound hearing loss. Some individuals with ANSD may also present with abnormally poor speech understanding abilities relative to their degree of hearing loss.

### What are the implications of this diagnosis on early intervention?

In children with cochlear hearing loss, ABR testing can be used to estimate hearing thresholds and guide audiologic intervention. Due to the lack of neural synchrony along the auditory pathway, ABR testing is not useful in estimating thresholds for children with ANSD. Consequently, audiologic intervention cannot be initiated until hearing sensitivity is assessed with behavioral audiometry. Moreover, co-existing conditions and developmental delays may set back the age at which reliable behavioral testing can be performed in this medically complex population. For the aforementioned reasons, the timeline for diagnosis and intervention in children with ANSD may diverge from the 1-3-6 guidelines recommended by the Joint Committee on Infant Hearing.

## Methods

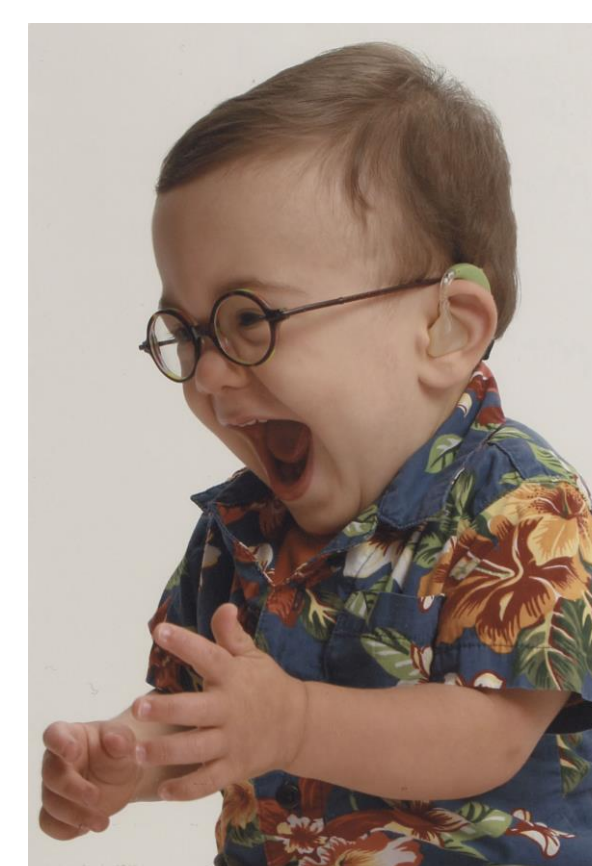
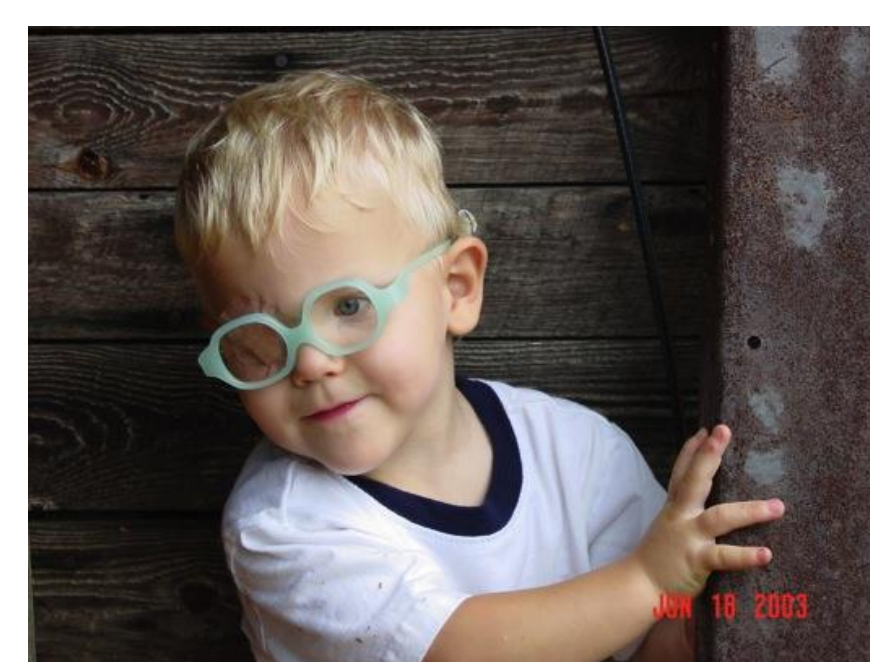
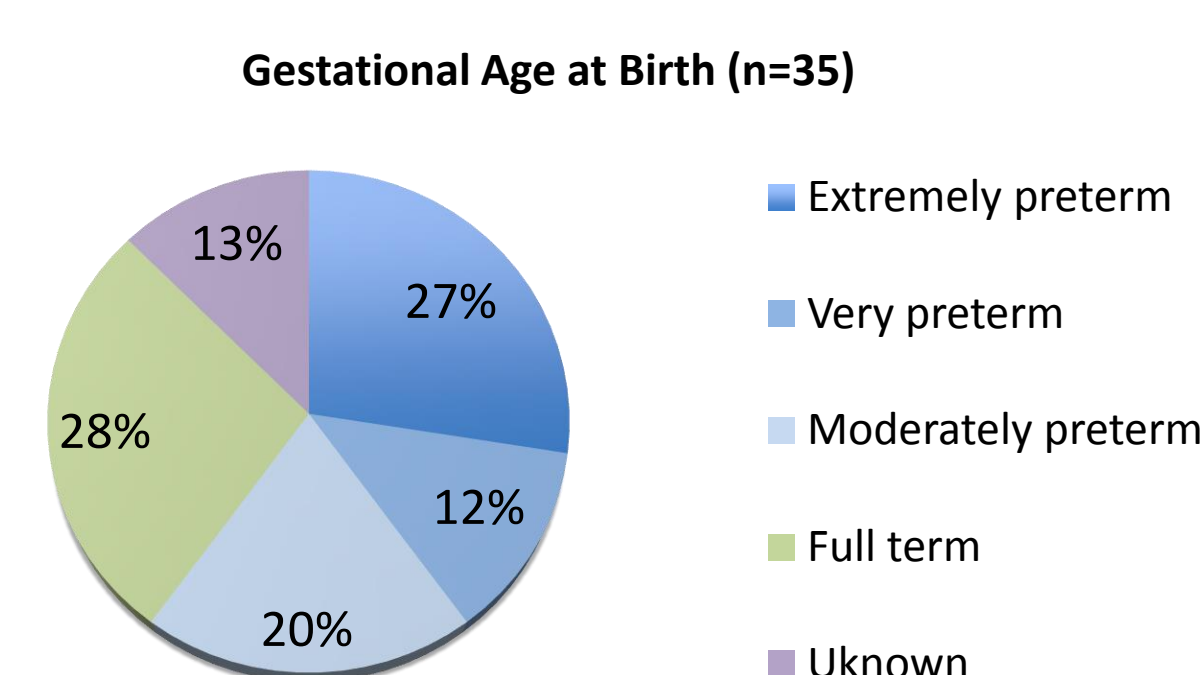
A random sample of 40 patients with a diagnosis of bilateral ANSD followed by the Audiology Department at UNC Hospitals was identified. Medical records were reviewed and the following data collected:

1. Gestational age at birth
2. Age at time of diagnosis of ANSD
3. Age at time of intervention (e.g. fitting of hearing technology)
4. Number of attempts at behavioral testing between time of diagnosis and intervention

## Results

### I. Gestational age at birth

Gestational age at birth was available for 35 of 40 subjects. The 35 with known gestational age were then placed into categories of preterm status, including extremely preterm (<28 weeks), very preterm (28 to <32 weeks), moderately preterm (32 to <37 weeks), and full-term ( $\geq 37$  weeks). Results indicate that 59% of the sample for whom gestational age at birth was available were born prematurely.

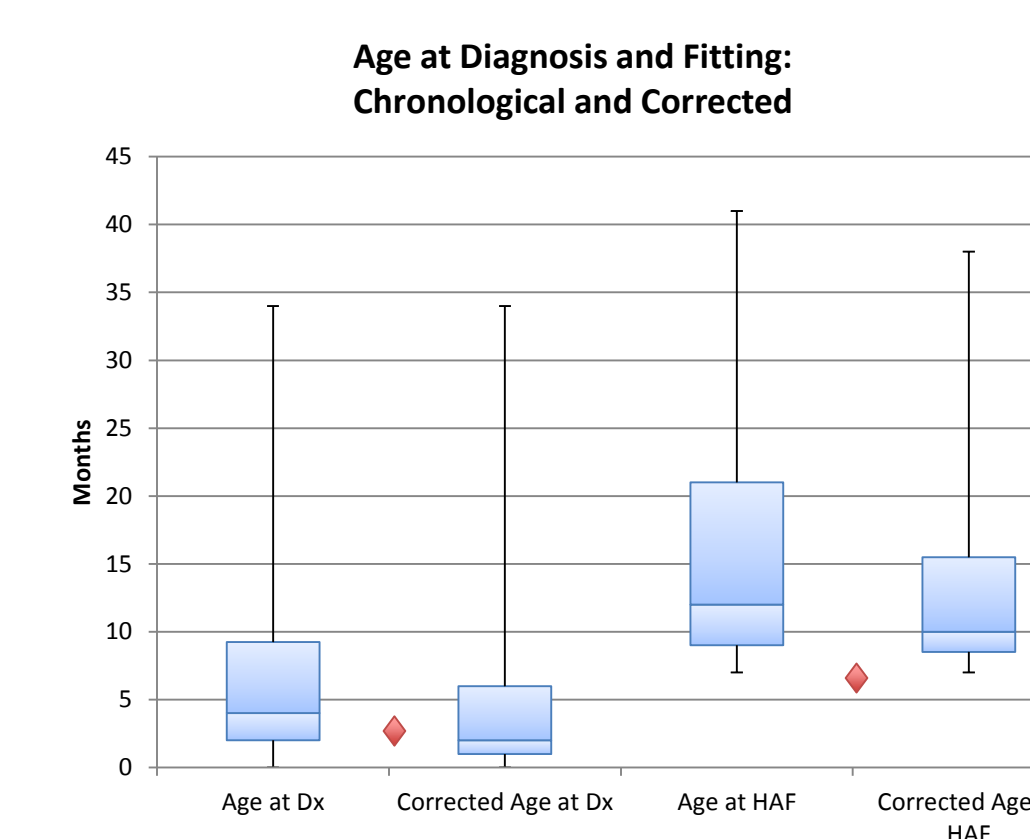


### II. Age at diagnosis and fitting (chronological and corrected)

Patient medical records were analyzed to determine chronological age at the time of diagnosis of ANSD for each subject (n=40). Corrected age at diagnosis was then calculated when information on gestational age at birth was available (n=35). The same process was used to identify the age at which each child was fit with hearing aids (n=36) and the corrected age at the time of the fitting (n=36).

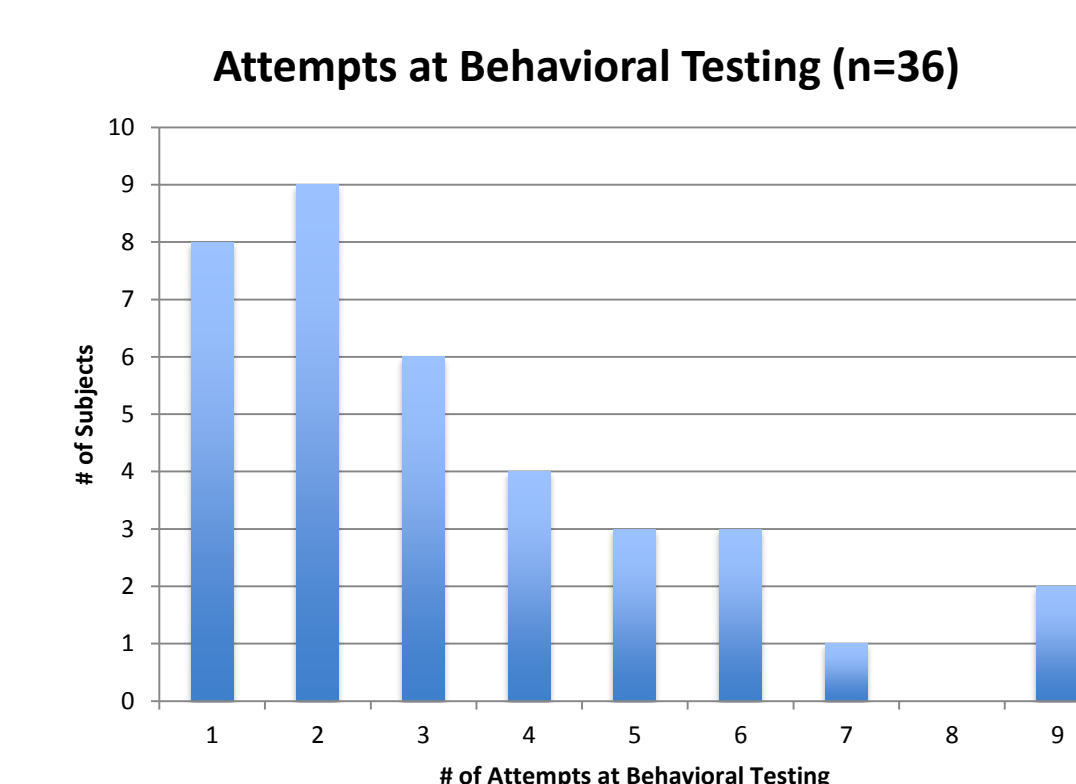
## Results, continued

Below is a representation of these findings, with red diamonds used to represent the recommended age of diagnosis (no later than 3 months) and intervention (no later than 6 months) for children with typical sensorineural hearing loss.



### III. Number of Attempts at Behavioral Testing

The number of times a child was seen for behavioral testing prior to the fitting of hearing aids was also determined. Results indicate that the mean number of attempts at behavioral testing was 3.2, with some children seen as many as 9 times before reliable thresholds could be established and intervention could be initiated.



Mean	3.3
Median	3
Range	3-9

## Impressions

- An encouraging finding for this cohort of children with ANSD is that the majority of children were able to reliably perform behavioral audiometry after an average of 3 visits.
- Within the current sample, children with severe-to-profound hearing loss required fewer attempts at behavioral testing before thresholds could be established and intervention initiated.
- As expected, children with the most complicated health histories generally required the greatest number of attempts at behavioral testing, with some children needing as many as 9 visits before reliable behavioral thresholds could be established.

## Conclusions

- Health histories, developmental trajectories, and audiologic profiles are highly variable among this clinical population, making it difficult to establish a "one-size-fits-all" timeline of audiologic care.
- Appropriate counseling is particularly critical for parents of children with ANSD. Families may receive conflicting advice from professionals or confusing information from the internet and other resources. Careful counseling can help families understand the complexity of this disorder, reduce frustration, and provide encouragement to return to the clinic for behavioral testing as needed.
- Continued research is needed to explore the role of cortical evoked potentials and other electrophysiologic tests in the evaluation and management of children with ANSD.

## References

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