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Does parental experience of the diagnosis and intervention process differ for children with auditory neuropathy?

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ABSTRACT

Objectives: This study compared parental experience of the audiological diagnosis and intervention process in children with auditory neuropathy spectrum disorder and sensory neural hearing loss.
Methods: A matched group survey was used with parents of children with auditory neuropathy spectrum disorder (ANSD) matched with a control group of parents and children with sensorineural hearing loss (SNHL). The two groups were matched in terms of the child's gender, age, amplifications used, social background and utilisation of private or public health care sectors. An interview questionnaire, consisting of 45 questions in six categories (1. biographic information, 2. experiences of audiological diagnosis, 3. hearing aid benefit, 4. parental experience of the rehabilitation decision making process, 5. parental needs for emotional support and 6. parental needs for information) using a 5-point Likert scale for categories 2–7, was administered by the same audiologist.
Results: Children with ANSD experienced a significantly longer waiting period from diagnosis to hearing aid fitting ($p = 0.025$) and/or cochlear implantation ($p = 0.036$). Parents of children with ANSD reported significantly different experiences of the diagnostic process ($p = 0.001$) with poorer understanding of the diagnosis and reporting insufficient time allowed for asking questions. During the rehabilitation decision-making process 47% of parents with ANSD children (vs. 0% of parents with SNHL children) reported receiving conflicting information. Parents of children with ANSD were also less likely to recommend hearing aids to other parents. Information needs were similar between groups.
Conclusions: Parents of children with ANSD have different experiences and greater uncertainty during the diagnostic and rehabilitation process. Providing regular consultation and structured timelines through the diagnostic process and decision-making process may facilitate this process with less uncertainty.

1. Introduction

The term auditory neuropathy or auditory dys-synchrony is used to describe a form of hearing impairment in which cochlear outer hair cell function is evident, but afferent neural transmission is disordered [1–3]. The term 'auditory neuropathy spectrum disorder' (ANSD) was adopted more recently as a way of describing its heterogeneous and multifaceted nature [4].

Evidence supports multiple etiologies and multiple locations as the site of lesion in ANSD, ranging from the cochlear receptors or inner hair cells (IHC), to kernicteric deposits anywhere from the spiral ganglion fibres to the brainstem, to a paucity of myelinated fibres in the VIIIth nerve [2,3].

By clinical definition, patients with this disorder have normal otoacoustic emissions (OAE's) and/or cochlear microphonic (CM) potentials, but exhibit an absent or severely abnormal auditory brainstem response (ABR) beyond the expected for the degree of hearing loss [1–3,5,6]. Although the diagnosis is clear with an absent ABR, no clear guideline exists on what exactly defines an abnormal ABR in cases of ANSD but it includes abnormal wave morphology with ABR thresholds significantly elevated above the expected degree of hearing loss [6].

The degree of hearing loss found in ANSD patients range from mild to profound. Clinical presentation in ANSD typically involves inordinate difficulty listening in noise, possible fluctuations in

Abbreviations: ANSD, auditory neuropathy spectrum disorder; SNHL, sensorineural hearing loss; NICU, neonatal intensive care unit; CAEP, cortical auditory evoked potentials; ABR, auditory brainstem response; ASSR, auditory steady state response; MVOS, my view on services.

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hearing over time, delayed speech and language development, and speech perception performance that are not easily predicted based on the level of residual hearing [3,5,7].

Variation is typical of ANSD patients and may relate to time of onset, underlying mechanisms, genetics, possible risk factors, ability to understand speech and changes over time. Some patients display no overt delays or auditory complaints until adulthood or in some cases until MEMR or ABR testing is completed. On the other end of the spectrum patients may display an apparent total lack of sound awareness, reflected in their severely affected communication and speech production abilities [8]. Most patients fall between these two extremes, showing inconsistent auditory responses with best responses in quiet and poorest in noise [5–7].

In light of the variability inherent to ANSD, families may experience contradicting information regarding the diagnosis, choices in communication, amplification and intervention for their child [9,10]. In the case of children with ANSD and their families, this process is intensified because of the complexity and heterogeneity inherent to the condition [6].

Most of the documented research on the experiences and support of families of children with hearing loss is focused on children's early stages of development [11]. The impact of deafness on the family, including the various challenges associated with raising a child with hearing loss and families' support needs have been well documented, but little has been documented about the specific experiences, perceptions and needs of parents with ANSD children. In a qualitative narrative study by Uus [12], experiences of ANSD parents were described. The diagnosis of ANSD in this study was made following newborn hearing screening. It was found that these parents did not prioritise hearing very highly at the time of diagnosis because of other medical problems. ANSD was not seen as a standalone diagnosis but as part of a bigger picture as all of the ANSD babies were graduates from the NICU [12]. The fact that multiple risks and developmental delays are associated with ANSD mean there are additional factors that influence the parental experiences [13]. Investigating parental experiences for children with ANSD compared to those with SNHL may inform early health provider and parent interactions to be responsive to particular needs. The aim of this study therefore, was to compare parental experiences of the audiological diagnosis and intervention process in children with SNHL and ANSD.

2. Methods

This study was conducted with parents of children who attended a centre for hearing impaired children. Children at this centre are exposed to an auditory/oral method of communication that aims to empower parents and children to manage in the mainstream environment. Institutional ethical committee approval was obtained before data collection was initiated.

A matched-group survey was used with parents of children with ANSD matched with a control group of parents of children with SNHL.

2.1. Study population

Two groups of 15 parents each were sampled, one with children with ANSD and the control group with children with SNHL. The two groups were matched in terms of the child's gender, age, amplification used, social background and utilisation of private or public health care sectors (Table 1).

Table 1 describes the demographic characteristics of the families who participated in this survey.

Table 1
Characteristics of participants (parents and children).

Characteristics	ANSD ($n=15$)	SNHL ($n=15$)
Relationship with child with hearing loss		
Father	1	2
Mother	13	13
Grandparent	1	–
Highest qualification		
Primary school (grade 1–7)	1	2
High school (grade 8–12)	4	4
Matric completed	2	3
Tertiary education	8	6
Enrolled in parent guidance programme		
At time of study	10	10
Discharged from programme	5	5
Medical service provider		
State	6	6
Private	9	9
Ages of children at time of survey		
Average	5.9 years (SD = 2.8)	6.1 years (SD = 2.9)
Range	1.10–12.3 years	2–12.3 years
Risk factors		
Unknown	2	8
Heredity		5
Prematurity (≤34 weeks)	12	
Blood transfusions	8	2
Loss of oxygen	6	
Other	–	
Newborn hearing screening		
Yes	10	5
No	4	9
Unsure	1	1
Average age at time of identification	1.4 years (SD = 1.1)	1.4 years (SD = 1.1)
Average age at time of hearing aid fitting	1.9 years (SD = 1.3)	1.7 years (SD = 1.1)
Average age of cochlear implant		
First	3.9 years (SD = 1.6)	2.8 years (SD = 1.9)
Second	4.10 years (SD = 2.0)	3.0 years (SD = 1.5)
Amplification		
Bilateral hearing aids	4	6
Bilateral cochlear implants	6	6
Bimodal	3	3
Unilateral hearing aid	1	-
None	1	

2.2. Procedures

An interview questionnaire, consisting of 45 questions divided into six subcategories (biographic information, experiences of audiological diagnosis, hearing aid benefit, parental experiences of the rehabilitation decision making process, parental needs for emotional support and parental needs for information) was administered. The same independent audiologist administered the questionnaire to all parents/caregivers. These interviews were conducted in a personal or telephonic interview. On average an interview was conducted in 20 min. Instructions about the completion of the questionnaire were conveyed to each parent/caregiver and informed consent was required by each participant before commencing.

Questions were derived and adopted from an existing questionnaire, namely the MVOS (My View On Services) [14]. Additional questions were referred from qualitative report on parents whose infants have been identified with ANSD [12]. Fourteen questions were asked to gain demographic and case history information. A 5-point Likert scale was implemented for the remaining 31 questions in order to obtain information