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Vestibular function in children with auditory neuropathy spectrum disorder

Robert Nash [a,*], John Veness [b], Michelle Wyatt [a], Ewa Raglan [b], Kaukab Rajput [c]

[a] Department of ENT, Great Ormond Street Hospital, WC1N 3JH London, UK
[b] Department of Audiology, Great Ormond Street Hospital, WC1N 3JH London, UK
[c] Cochlear Implant Department, Great Ormond Street Hospital, WC1N 3JH London, UK

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ABSTRACT

Objective: Children with auditory neuropathy spectrum disorder (ANSD) account for about 10% of paediatric patients referred for cochlear implantation. Vestibulopathy may be associated with ANSD, and may have implications when formulating management plans in this patient group. We wanted to determine the incidence and predictive factors for vestibulopathy in this patient group to guide vestibular testing in this patient population, and give insight to the aetiology of ANSD.
Methods: We reviewed the outcomes of vestibular function testing in a cohort of paediatric patients with ANSD.
Results: Probable or definite vestibulopathy was seen in 42% of patients who were tested. Vestibulopathy was associated with medical co-morbidities, but was not associated with imaging findings.
Conclusions: Vestibulopathy is relatively prevalent in this patient group, and should be considered when planning the investigation and management of children with ANSD.

Introduction

Auditory neuropathy spectrum disorder (ANSD) is a syndrome that is increasingly recognised as a major cause of hearing loss in a paediatric population. The term ANSD refers to hearing impairment due to pathology at any point between the inner hair cells, and the distal auditory pathway. It is characterised by a disparity between central measures of hearing such as auditory brainstem responses, and measures of outer hair cell function such as otoacoustic emissions (OAEs), or cochlear microphonics. However, it is important to note that in established ANSD, OAEs may be negative as they may disappear over time. It is thought that ANSD may account for approximately 5–10% [1–3] of paediatric patients with profound hearing loss.

In addition to its prevalence, ANSD is important for other reasons. Firstly, neonates may 'pass' newborn screening programmes with normal OAE responses, but in the presence of auditory neuropathy, they may still have a significant hearing loss [4]. Secondly, ANSD is associated with conditions such as prematurity which may already put the patient at risk of developmental delay. Additional hearing loss may exacerbate this disability. Thirdly, some patients with ANSD may be less amenable to treatment with cochlear implantation [5,6] and early development of alternative communication strategies may have to be considered.

ANSD is a spectrum of conditions that produce hearing loss distal to the outer hair cells. These include disorders of the inner hair cells, neuropathy in the cochlear or vestibulocochlear nerve, and central pathologies. As the vestibular and auditory systems are intrinsically linked, these conditions can affect the vestibular system. The overlap between auditory and vestibular neuropathies in adult and paediatric patients has previously been demonstrated.

A number of studies have tested the superior vestibular nerve using calorics and clinical examination in patients with ANSD. Starr [7] showed that five of a series of ten had vestibulopathy – three clinical and an additional two subclinical. Sheykholeslami [8] and Kaga [9] demonstrated vestibulopathy in all ANSD patients they assessed (three and five respectively).

The vestibulopathy is frequently subclinical. When testing 14 asymptomatic patients, Fujikawa [10] found nine with evidence of objective vestibulopathy. Indeed, in the largest series, only 18% reported balance difficulties, despite vestibular dysfunction being detected in 44% [11]. Auditory and vestibular neuropathy was frequently associated with peripheral neuropathy, and indeed in this large series, the overlap was noted with 62% of ANSD patients having abnormal somatosensory evoked potentials – a marker for peripheral neuropathy. Masuda [12] showed that vestibular

* Corresponding author. Tel.: +44 20 7405 9200.
E-mail address: mr.robert.nash@gmail.com (R. Nash).

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function had declined over time with hearing in three 60 year old patients, and therefore when considering a paediatric population, it is important to consider these patients separately.

Two studies have looked at this association in children using a combination of examination, calorics, pursuit testing, and optokinetic nystagmus. Both Konradson [13] and Akdogan [14] demonstrated normal vestibular function in four and two patients respectively.

The saccule and inferior vestibular nerve have also been investigated. In a trial of eight adults with adult onset auditory neuropathy, the vast majority (81%) of tested ears had concomitant abnormal vestibular evoked myogenic potentials (VEMPs) [15]. In three 19–21 year old patients with recent diagnoses of auditory neuropathy, all had some element of vestibulopathy; one patient had symptomatic vestibular hypofunction, one asymptomatic but clinically detectable, and one was only detectable on objective testing [16]. Sheykholeslami and Kaga's series mentioned above also included VEMP testing, which was also abnormal in all eight patients. Whereas, in Akdogan's paediatric series, whilst superior vestibular nerve function was normal, two out of the three patients had abnormal VEMPs.

In summary, whilst there have been investigations of vestibulopathy in adult patients with ANSD, we have very limited information on vestibulopathy in paediatric patients. Given the frequency of subclinical vestibulopathy, all paediatric patients at our institution with ANSD who were referred for consideration of cochlear implantation underwent objective vestibular testing. We summarise the results of the vestibular assessment. We also investigated the past medical history and imaging findings to see if these were associated with vestibulopathy.

Methods

Setting

Tertiary and quaternary care paediatric hospital in London, UK.

Patients

Patients were retrospectively identified using a local database of children diagnosed with ANSD at our institution. This patient list was correlated with those who had undergone vestibular function testing. Patients with ANSD who were considered for cochlear implantation were routinely referred for vestibular function testing.

Diagnosis of ANSD

The diagnosis of ANSD was clinical, but heavily based on results of hearing performance, OAEs, cochlear microphonics and ABRs.

Vestibular testing

Vestibular function testing can be highly challenging in a paediatric population. At our institution we use rotational chair testing [17] and clinical examination to make an assessment of vestibulopathy. Our institution solely treats paediatric patients, and we therefore have a significant experience of vestibular function testing in this group. Obtaining valid rotational chair testing can require encouragement, support and patience. Tests can be performed sitting on a parent's lap. It is not uncommon for tests to be abandoned and scheduled for a different day. The use of confectionery can also calm children and make them more compliant to testing. Examples of normal vestibular function and vestibulopathy on sinusoidal rotational testing are shown in Figs. 1 and 2. Measurement of spectral purity ensured that the child's performance of the test was adequate. Gain records the magnitude of vestibular response to a stimulus, phase provides an estimate of the vestibular system time constant, and asymmetry determines the percentage difference between the strength of the response of the right and left side peripheral horizontal response.

Imaging

Three dimensional imaging of the temporal bone was reviewed for our patient group. This was correlated with vestibular function testing.

Results

Patient demographics

35 patients were identified. 20 were male and 15 were female. The mean age was 5.8 years at the time of assessment at our institution (range 0.5–17.4 years). 20 patients underwent cochlear implantation,

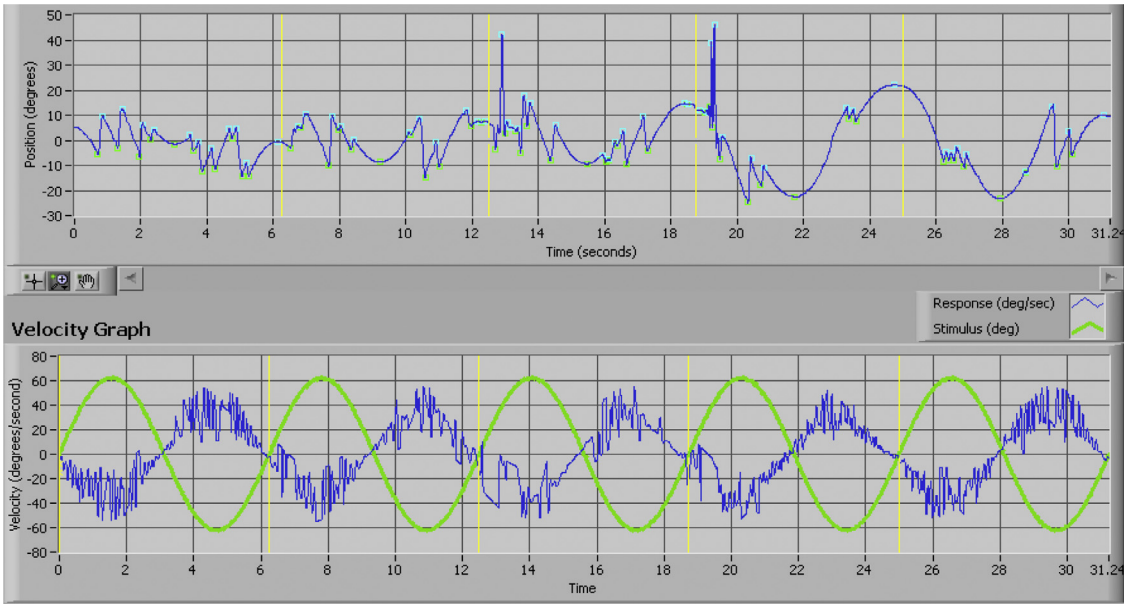

Fig. 1. Normal vestibular function. The rotation of the chair is matched by an inverse rotation of the eye in order to stabilise gaze using the vestibulo-ocular reflex.