Chapter 1
Late Onset Auditory Neuropathy Spectrum Disorder: A Psychosocial Perspective

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ABSTRACT

The chapter attempts to understand the enigma of late-onset auditory neuropathy spectrum disorder (ANSD) from a psychosocial perspective. The focus of research has always been on accurate diagnosis and management of individuals with ANSD. However, there are limited studies on the psychological problems faced by individuals with late-onset ANSD. In the present chapter, the studies on late-onset ANSD and the psycho-social problems faced by them are reviewed. They experience serious psychosocial issues which need to be addressed urgently for appropriate management of individuals with ANSD. It is highlighted that a psychologist/psychiatrist should be a part of the rehabilitation team for management of individuals with late-onset ANSD. In addition, the focus of rehabilitation should not be restricted to physiological problems, but the psychological problems should also be addressed. Thus, a patient-centric approach in management would definitely improve the quality of life of some individuals with ANSD.

INTRODUCTION

Auditory neuropathy spectrum disorder (ANSD) is a retro-cochlear pathology in which the outer hair cell functioning is normal but there is an abnormality in the auditory nerve (Berlin et al., 2010; Berlin, Hood, Morlet, Rose, and Brashears, 2003; Deltenre et al., 1999; Roush, Frymark, Venediktov, and Wang, 2011; Starr, Picton, Sininger, Hood, and Berlin, 1996). ANSD was first explained by Starr et al. (1991) as a single case study in detail. The condition was later described in detail on ten subjects and named as auditory neuropathy (Starr et al., 1996). The condition is also described using the terminology ‘auditory dys-synchrony’ as this would better indicate the pathophysiology of the disorder (Berlin, Hood, and Rose, 2001; Berlin, Jeanfreau, Hood, Morlet, and Keats, 2001).

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The term ‘auditory neuropathy’ was not preferred as the eighth cranial nerve (Vestibulocochlear nerve/auditory nerve) itself may not be affected as the pathology can be at inner hair cells (IHC) level or at the level of the synapse of IHC and auditory nerve. In addition, the term auditory neuropathy may lead clinicians not to consider cochlear implant as a management option. It has been shown later that the individuals with auditory dys-synchrony benefit from cochlear implants (Berlin et al., 2003; Peterson et al., 2003). A group of audiologists, neonatologists, medical geneticists, hearing scientists and neurologists had a conference for the development of guidelines on identification and appropriate management of auditory neuropathy. One of the decisions taken in the conference was to replace the current terminology used for describing the clinical condition as auditory neuropathy spectrum disorder (ANSD).

ANSD can affect the functioning of inner hair cells, synapse between the inner hair cells and auditory nerve, or the auditory nerve itself (Starr, Sininger, and Pratt, 2000). Individuals with ANSD may manifest asynchronous firing of the auditory nerve fibers due to demyelinization (Starr, Picton, and Kim, 2001). The demyelinization leads to temporal asynchrony in the firing of the auditory nerve fibers and thus reducing the compound action potential of the auditory nerve. Asynchronization not only affects auditory brainstem responses, but also influences auditory perception dependent on temporal cues (Kraus et al., 2000; Rance, McKay, and Grayden, 2004; Starr, Zeng, Michalewski, and Moser, 2010; Zeng, Kong, Michalewski, and Starr, 2005; Zeng, Oba, Garde, Sininger, and Starr, 1999).

The prevalence of ANSD which is reported in Western countries varies from 11% to 0.5% (Berlin et al., 2000; Cone-Wesson and Rance, 2000; Kraus, Ozdamar, Stein, and Reed, 1984; Rance et al., 1999; Tang, McPherson, Yuen, Wong, and Lee, 2004). There are studies on the prevalence of ANSD on ‘at risk’ children (Rance et al., 1999) and school-aged hearing impaired children (Tang et al., 2004). Tang et al. (2004) reported that the prevalence was 2.44% in school-going children diagnosed as having hearing impairment. Rance et al. (1999) studied 5199 at-risk children and reported that one in 433 (0.23%) was the prevalence of ANSD and the prevalence was one in nine (11.01%) in children with sensorineural hearing loss of degree greater than the moderate degree. Cone-Wesson and Rance, (2000) concluded from their study that everyone in every 200 children with hearing-impaired will have audiological findings which are consistent with the diagnosis of ANSD. Berlin et al. (2000) assessed the prevalence and reported that one percent out of 1000 children studied with severe to profound hearing loss showed robust otoacoustic emissions in one or both ears and 10% of the children showed robust or OAE of lesser amplitude in one ear at least. In the Indian context, Kumar and Jayaram (2006) reported that one in 183 in individuals who are diagnosed as having sensorineural hearing loss had ANSD. The study was conducted retrospectively and reported that among 11,205 clients (5854 males and 5351 females) who had sensorineural hearing loss, sixty-one of them were identified as having ANSD. Bhat, Kumar, and Sinha (2007) suggested that in children with hearing impairment who going to school, the prevalence of ANSD was 2.47%. They studied 220 children who were of four to 16 years old and reported that ANSD is not an extremely rare disorder.

The onset of symptoms related to ANSD can fall into two different age groups. The symptoms of ANSD can be reported from infancy and childhood or these symptoms may begin to appear in adolescence or early adulthood (Berlin et al., 2010; Kumar and Jayaram, 2006; Sininger and Oba, 2001). Majority of the studies carried out in the Western population suggest that approximately only one out of four individuals with ANSD are over the age of 10 years (Berlin et al., 2010; Sininger and Oba, 2001; Starr et al., 2000). However, as the stark contrast, studies carried out on Indian population have reported that the onset of the symptoms of ANSD was majorly in adolescence (16 to 25 years) referred to as late-onset
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