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A Note on Neonatal Auditory Neuropathy and Its Epidemiology

Ruby Jose*

Department of Obstetrics and Gynecology, KJ Somaiya Medical College & Research Centre, Mumbai, India

***Corresponding author:** Ruby Jose, Department of Obstetrics and Gynecology, KJ Somaiya Medical College & Research Centre, Mumbai, India, E-mail: roserb@gmail.com

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Editorial Note

Congenital hearing loss occurs when the ear's capability to transform the vibratory mechanical energy of sound into the electrical energy of nerve impulses is impeded. Hearing loss is classified based on the location of the lesion; conductive hearing loss affects the outer or middle ear, whereas sensorineural hearing loss affects the inner ear, auditory nerve, or central auditory pathway. The term "Mixed hearing loss" refers to both conductive and sensorineural hearing loss. Sound waves cannot propagate through the ear in conductive hearing loss, which occurs as a result of mal-development of the middle ear, external ear, or both, or as a result of acute blockage of the middle ear caused by effusion.

Sensorineural hearing loss is classified as sensory hearing loss (when hair cells are impaired), central hearing loss (when the cause is found in the central auditory pathway), and Auditory Neuropathy Spectrum Disorder. Auditory Neuropathy Spectrum Disorder refers to a group of clinical conditions characterized by the presence of otoacoustic emissions (sounds of cochlear origin, which can be recorded by a microphone fitted into the ear canal) and cochlear microphony, as well as abnormal or lacking auditory brainstem responses, which leads to impaired speech discrimination. Auditory Neuropathy Spectrum Disorder can be caused by primary lesion in the inner hair cells, the auditory nerve, or an intermediate synapse.

For this common issue, new-born hearing screening programs are available in the major developed countries. The goal of these programs is to test all infants within one month after birth. Early detection, followed by early intervention and therapy, promotes better developmental outcomes later in life. Since hearing loss can worsen over time, new-born hearing screening programs may overlook children with progressive hearing loss. As a result, periodic screening at regular intervals is recommended for newborns to reduce the risk. The medical and supportive treatment of congenital hearing loss is determined by the origin and the type of hearing impairment. Genetic factors (including both non-syndromic forms in which hearing loss is the sole clinical symptom and syndromes such as Usher or Jervell and Lange–Nielsen syndromes), craniofacial abnormalities, and prenatal infections are the most common factors of hearing loss.

Neonatal hearing screening programs have been accessible in North America, Europe, and many developed countries since the beginning of the century. These programs are for universal neonatal hearing screening (checking all infants rather than only those with risk factors for hearing loss). According to these screening programs, the prevalence of permanent bilateral hearing loss is expected to be 1.33 per 1,000 live births. The incidence rises to 2.83 per 1,000 children in primary school and 3.5 per 1,000 in adolescents. This rise over time is most likely due to the inclusion of individuals with hearing loss caused by progressive, acquired, or late-onset hereditary factors. Diagnostic results for several kinds of hearing loss, such as Auditory Neuropathy Spectrum Disorder, are frequently inconclusive in infants since language skills are still developing and not abnormal at that time; as a result, prevalence estimates vary greatly. In countries lacking universal neonatal hearing screening programs, prevalence estimates range from 19 per 1,000 newborns in Sub-Saharan Africa to 24 per 1,000 in South Asia.

The existence of risk variables is the most important predictor. The study of hereditary hearing loss has substantially enhanced the understanding of the normal auditory function and the pathophysiological mechanisms that might impair it. Genetic mutations can disrupt any component of the hearing pathway.