

감각 신경성 난청의 원인과 재활

이승환

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Etiology and Rehabilitation of Sensorineural Hearing Loss

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Hearing is a special sense by which we perceive sound by detecting vibrations. Hearing ability is important for us to be able to interact, communicate and socialize. Sound is captured by the human ear which is a peripheral sensory organ composed of the external ear, middle ear and inner ear. The external ear serves to focus sound energy through the external auditory canal toward the tympanic membrane. The middle ear amplifies the sound energy via the ossicular chain which transfers sound vibrations to the inner ear. The inner ear includes the cochlea where mechano-electrical transduction of sound energy takes place. Humans perceive the vibrations of sound as transient auditory signals of vibratory mechanical forces transmitted through the external ear and middle ear, where they are transduced to neural signals in the inner ear passing via the auditory nerve to the central auditory system of the auditory cortex where the signal is processed and interpreted and passed on for perceptive processing. Understanding the central auditory processing in the central nervous system is essential to understanding how the sound information is discriminated and analysed to allow good communication.

Any single problem in the peripheral or central auditory pathway may cause hearing impairment. Conductive hearing loss is defined as the hearing loss due to inappropriate mechanical transmission. Conditions that can cause conductive hearing loss are

foreign body obstruction of the external auditory canal, perforation of the tympanic membrane, middle ear effusion, the destruction of ossicles, etc. Depending on the specific cause of conductive hearing loss, medical or surgical treatment can restore hearing.

Sensorineural hearing loss refers the hearing loss caused by inner ear problems such as hair cell damage or neural degeneration. Unlike the conductive hearing loss, sensorineural hearing loss is usually permanent. Sensorineural hearing loss is divided into congenital and acquired. Congenital hearing loss may be caused by a congenital anomaly, a chromosomal syndrome or a congenital infection such as rubella or cytomegalovirus. Acquired sensorineural hearing loss might have numerous etiologic causes, including noise-induced hearing loss, presbycusis, trauma, inflammatory causes and sudden sensorineural hearing loss, etc [1]. Since restoration of hearing is a difficult and challenging issue in sensorineural hearing loss, treatment focuses on the personalized care for hearing rehabilitation according to the hearing level.

Hearing aids are the most common method of hearing rehabilitation. Because hearing aids are very specific to a patient's needs, it is important to determine the proper amplification based on lifestyle and listening demands as well as audiologic tests. In addition, verification and fitting procedures are essential to achieve successful hearing aid use.

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With the development of cochlear implants came a turning point in the treatment of hearing loss. A cochlear implant is a surgically implanted electronic device that provides sufficient recovery of hearing for speech recognition in patients with profound hearing loss who cannot benefit fully from hearing aids. Especially, it is considered as an important treatment method to allow language development in congenital sensorineural hearing loss patients identified in newborn hearing screening.

Recently, a middle ear implant has been developed and is considered to be an alternative to conventional hearing aids. A middle ear implant converts sound energy into enhanced mechanical vibration which directly stimulate the middle ear structures including ossicular chain or round window.

The hair cells in the cochlea are well differentiated cells that do not regenerate after damage. However, active research is in progress to develop a biological method using stem cell technique to repair the damaged cochlea thereby restoring hearing without prosthesis. We expect to see favorable outcomes utilizing stem cell therapy in the future.

Uncorrected hearing loss is associated with lower quality of life, reduced social activity and social isolation, leading to an increased prevalence of depression. Early identification of hearing loss and tailored rehabilitation strategies are the focus to reduce the social burden of hearing loss.

Herein, we review the etiology and characteristics of genetic hearing loss [2], the importance of early diagnosis and screening

strategy for neonatal hearing loss [3], presbycusis which is the most common type of sensorineural hearing loss [4], recently increasing noise-induced hearing loss due to the increase of environmental noise exposure, and especially, various exposures of noise to youth [5], and sudden sensorineural hearing loss [6]. Rehabilitation by hearing aids [7], middle ear implant [8], cochlear implant [9], and recent research regarding stem cell therapy [10] are also discussed. We expect that this review will be an important guideline for physicians to understand and help hearing loss patients.

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