

The link between Brain and Hearing system

Abstract

There are some studies which confirmed that dysfunction in Central Nervous System(CNS) may cause a malfunction in the Peripheral Auditory system (Cochlea_ Auditory Nerve, Auditory Neuropathy), but the question is could Brain Disorder without any lesion in the Cochlea and/or Auditory nerve cause Sensorineural Hearing Loss? It means that the Audiogram shows that the patient suffers from sensorineural hearing loss but the site of the lesion is neither Sensory nor Neural while Brain may be involved in charge of this. And if the answer is yes then could we hear with our Brain and without Cochlea and /or Auditory nerve?

- A. We deal with this subject in this paper by:
- B. Otosclerosis and Meniere's disease and The Brain Involvement.
- C. Hearing Loss following dysfunction in the Central Auditory and/or central non auditory system.
- D. Auditory Brainstem Implant in Patients who suffer from Neurofibromatosis Type two compare to Non Tumor cases, Mondini Syndrome, Michel aplasia.
- E. Possible role of Utricle and Saccule in Auditory (Hearing) System
- F. We propose a new Hypothesis that the External Ear Canal is not the only input of Auditory Signals, Sounds could transfer by our eyes and skin to the Cerebral Cortex and approach to the Cochlea (Backward Auditory input pathway of Sounds).

Keywords: otosclerosis, meniere's syndrome, central auditory system

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Discussion

Otosclerosis and Meniere's disease and the brain involvement

Meniere's disease (MD) is a disease which is specified by episodes of feeling Vertigo(rotating, spinning) Tinnitus, Hearing loss, Fullness in the ear.¹ Typically, only one ear is initially affected then, over time both ears may become involved. Episodes generally last from 20 minutes to 24 hours. The time between episodes varies. The hearing loss and Tinnitus in the ears can become permanent over time.¹ Otosclerosis is a genetic disorder that causes hearing loss due to middle ear 's inability to conduct the sounds to the inner ear because of stiffness on middle ear bones (ossicles), in the early stage of the disease the patient suffer from conductive hearing loss but the disease is progressive and eventually it causes Mixed hearing loss. Symptoms may include Hearing loss or Tinnitus. In some cases, vertigo may occur. Treatment options are Hearing aids or surgery (Stapedectomy), some medications such as bisphosphonates, Sodium Fluoride may slow the progression of the disease.² There are some studies which showed that the Central Nervous System is involved in both pathologies.^{1,2}

In Meniere's disease presumably Increasing Cortisol Level following Dopamine deficiency in Hypothalamus and infection of the CNS by Measles Virus in Otosclerosis.^{1,2} According to these studies a combination of medications like Ribavirin and Intraventricular Interferon -Alpha and Inosiplex may be a cure or treatment for the Otosclerosis and Medications which increase the Dopamine level (Dopamine Agonist) may be helpful for the treatment of Meniere's

Disease, further studies need to be done to find out the usefulness of these medications for these disorders and side effect, interaction and so on.^{3,4}

Hearing loss following dysfunction in central auditory pathways and/or central non auditory pathways

There are some papers which indicated Auditory Neuropathy occurred secondary to Inferior Colliculus Disorder and Sudden Deafness following infarction of the Primary Auditory Cortex. It seems that dysfunction in Central Auditory pathways may cause damage in Peripheral Auditory system, however, there are some patients with normal Audiogram but they suffer from Central Auditory Processing Disorder(CAPD) , therefore, malfunction in Central Nervous System (CNS) may cause damage in peripheral Auditory system but it depends on the site and degree of the Lesion in the CNS, probably mild lesion in some part of the Brain does not affect the peripheral Auditory system.⁵⁻¹⁴

We had a patient who referred to us because of the unilateral Sudden Deafness. The imaging result showed a Tumor on Occipital Lobe which is a Visual Central System, not an Auditory section , so not only disorder in Central Auditory pathways may cause peripheral hearing loss malfunction in other parts of Brain may involve in causing hearing loss.³ We have seen a lot of patients with Sensorineural hearing loss and normal Otoacoustic Emissions (OAE) this could be because of Auditory Neuropathy, lack of cooperation of patient during the Audiometry test, neural lesion, the collapse of the ear canal, Cochlear Synaptopathy, but it is not always the case. There are some

studies which verified normal or near normal OAE and Compound Action Potential and the Audiometry results indicate Sensorineural hearing loss but these tests show Cochlea and the Auditory nerve are intact therefore the reason of the hearing loss could be because of the lesions in the brain with normal function of the Cochlea and/or Auditory nerve.^{3,15-18}

Advance functional images of the Brain show that Prefrontal and Temporal Lobes are more responsible for recognition of High-Frequencies and Cerebellum and brainstem typically subcortical areas are more in charge of the comprehension of the Low-Frequency Signals.^{5,8,13,19} Reticular Formation and Prefrontal Lobe has a tremendous role in the Auditory system typically in Speech recognition and selective hearing and the correlation between these part and other parts of the brain such as whole Cortex and Hippocampus are involved in speech recognition and Listening skills.^{19,20}

Auditory brainstem implant (ABI) outcome in tumor vs non tumor cases

Non tumor pathologies

Mondini malformation is a type of inner ear disease which is congenital. Individuals with Mondini dysplasia have one and a half turns of the cochlea instead of the normal two turns. It may occur unilaterally or bilaterally and causes profound sensorineural hearing loss.²¹ Michel aplasia or complete labyrinthine aplasia (CLA), is a congenital disorder as well. It is specified by the bilateral absence of differentiated inner ear structures and results in Anacusis. Michel aplasia should not be confused with michel dysplasia (common cavity inner ear deformity) It may affect one or both ears.²² Aplasia is the medical term for body parts that are completely absent or undeveloped. In Michel aplasia, the bony labyrinth of the inner ear is Anaplastic. Other nearby structures such as vestibule and semicircular canals may be underdeveloped or completely absent as well.²²

Neurofibromatosis type II (NF2, Tumor)

Neurofibromatosis type II is a genetic disease which may arise spontaneously. The main feature of the disease is the development of symmetric, benign brain tumors in the region of the cranial nerve VIII. Auditory Brainstem Implant (ABI) is a Prosthesis for individuals with hearing loss due to Neurofibromatosis Type 2 which Bypass both the inner ear and the auditory nerve and stimulates the cochlear nucleus (CN) and provides users with a variety of hearing sensations to assist patients with sound awareness and communication.²² ABI is also an option for Non- tumor cases which have been mentioned earlier.

ABI outcome in tumor vs non tumor cases

There are some case studies which verified that non- tumor cases such as cochlear hypoplasia, Mondini dysplasia who underwent Auditory Brainstem Implant could even understand the speech over the phone typically patients with non-Neurodevelopmental disorders, so it seems there is a possibility to hear with brain and without cochlea or Auditory Nerve and if hearing without Cochlea and/or Auditory Nerve is possible then hearing loss because of lesions in some part of the brain without any or minor lesions in Cochlea and/or Auditory Nerve is likely.^{3,23}

Speech recognition in some patients with NF2 after ABI surgery would be worsen and ABI could not provide a well speech recognition for these patients unlike non Tumor patients, This could be because of the damage of the Cochlear Nucleus or Central Auditory system during

surgery or because of the Tumor. Cochlear Implantation has been done for some Tumor cases and the result was better than patients with NF2 who underwent ABI.²¹ A device which is a combination of Cochlear Implant and Auditory Brainstem Implant may be more helpful for the patients who suffer from NF2 as the cochlea is involved in this disease as well as Auditory/Vestibular Nerve.²³

Possible role of the utricle and saccule in auditory (hearing) system

The utricle and Saccule are the two otolith organs and they are part of the balancing system in the vestibule section of the labyrinth . Both Utricle and Saccule contain a sensory epithelium (Macula) which consists of Hair cells and associate supporting cells. The utricle detects linear accelerations and head-tilts in the horizontal plane.²⁴ The saccule is a small membranous sac, paired with the utricle. It converts head movements into neural impulses for the brain to interpret. The saccule detects linear accelerations and head tilts in the vertical plane. When the head moves vertically, the sensory cells of the saccule are evoked and the neurons connected to them begin transmitting impulses to the brain. These impulses travel along the vestibular portion of the eighth cranial nerve to the vestibular nuclei in the brainstem. Although Utricle and Saccule are responsible for Vestibular System there are some findings which show there is a possibility of their role in Auditory (Hearing) System as well.²⁵ The vestibular evoked myogenic potential (VEMP or VsEP) is a neurophysiological evaluation technique used to determine the function of the utricle and saccule . It complements the information provided by caloric testing and other forms of vestibular apparatus testing such as Videonystagmography, Video Head Impulse test (VHIT) and so on. There are two different types of VEMPs, oVEMP and cVEMP. The oVEMP measures integrity of the utricle and superior vestibular nerve and the cVemp evaluates the saccule and the inferior vestibular nerve.²⁶

Many studies have been done in recent years and investigate VEMPs in Idiopathic Sudden Hearing Loss, Meniere's disease, Noise Induced Hearing Loss, High frequencies hearing loss, Profound Sensorineural hearing loss and so on. These studies indicated that the Utricle and Saccule is not only responsible for Vestibular system, they are in charge of the auditory system too. It seems that Utricle and Saccule deliver the auditory information to the Cochlea and integrate the Audiovestibular information for transferring to the Brain and that could be one of the reasons for the link between hearing loss and falling. It seems that Saccule is more responsible in High frequencies and Utricle is more in charge of low frequencies.²⁷⁻³⁵

The vestibular implant consists of motion sensors strictly fixed to the patient's head and of electronic components which are processor and stimulator that interpret the received motion information into electrical signals transmitted to the brain via electrodes implanted in the surrounding of vestibular nerve endings.³⁶ Vestibular Implant is under observation and research right now and in the near future the clinical trial of this device will be initiated. A combination of Vestibular Implant and Cochlear Implant may be more helpful for the patients who suffer from profound sensorineural hearing loss compared to Cochlear Implant only typically for those patients with Cochlear Implant Failure and abnormal VEMPs.

Backward auditory input pathway hypothesis

We not only hear with our ears, but also through our skin and our eyes, according to a study. The finding, based on experiments

in which participants listened to certain syllables while puffs of air hit their skin, suggests our brains engage and integrate information from various senses to build an image of our surroundings. The new cochlear Implant is combined with Haptic Neuroprosthetic for pitch discrimination and this enhanced pitch discrimination has the potential to improve music perception, speech recognition, and speech prosody perception in CI users. We also not only see by our eyes, we may hear with our eyes as well. There is a significant relationship between Eye disorders and hearing impairments.^{37,38} When we are in a dark place our Pupils dilated to absorb more lights and it seems that is the same reaction to sounds. Auditory stimuli changes the pupil size and some parts of the brain such as Locus coeruleus and some Central Auditory pathways such as Inferior Colliculus and Superior Olivary Complex are involved in pupil dilation following Auditory stimuli. Visual information from a speaker's face can reinforce or interfere with accurate auditory perception. This integration of information across auditory and visual streams has been observed in functional imaging studies, and has typically been attributed to the frequency and *stoutness* with which perceivers jointly encounter event-specific information from these two modalities.³⁹⁻⁴² It seems that Sounds transmit from two paths, Forward Auditory input pathway which are the information from the external ear canal which approach to the Cochlea and the other one Backward Auditory input pathway which are the information of signals that transfer from Skin and eyes to the Cerebral Cortex and come down and reach to the Cochlea.^{3,38,39,43,44}

It seems that forward Auditory input pathway is more in charge of low frequencies and Backward Auditory input pathway is more responsible for high frequencies.^{3,40} The cochlea is the rendezvous between signals which come from Forward Auditory input pathway and signals which come from the Backward Auditory input pathway then all information will be combined and organized and coding by Cochlea and will be sent to the Brain again for the final processing.^{3,39,40} There is a high possibility that the Brain process the initial processing of the sounds when it receive the signals from Skin and eyes (backward Auditory input pathway) and before sending the information to the cochlea.³⁹

Conclusion

There are many patients with sensorineural hearing loss Audiogram but the site of the lesion is neither Sensory nor Neural, The site of the lesion is Central Nervous System. Hearing loss because of peripheral auditory dysfunction secondary to CNS disorders is possible also Hearing loss because of CNS disorders with mild lesion in Peripheral Auditory sections or even intact Peripheral Auditory sections is possible. A combination of Cochlear Implant and Auditory Brainstem Implant may be more helpful for the patients who suffer from NF2 compared to Auditory Brainstem Implant only. A combination of Vestibular Implant and Cochlear Implant may be more helpful for the patients who suffer from Profound Hearing Loss compare to Cochlear Implant only, since most of the patients who suffer from profound hearing loss have Vestibular End -organ dysfunction (hidden loss of otolithic function). A combination of Cochlear Implant and Haptic Neuroprosthetic and a specific eyeglass or the Goggles with the ability to receive the sounds and transfer it through the eyes may boost speech recognition significantly. Most Peripheral pathologies such as Meniere's and Otosclerosis initiated from CNS and medications which used for the treatment of Neurological disorders and mentioned in this paper before may be useful for the treatment of these pathologies as well. There are a lot of patients with similar Audiograms and similar type and degree of hearing loss, but inside the site of lesions may be completely different from one patient to another.

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Conflicts of interest

The author declares that there is no conflict of interest to disclose.

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