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Cochlear implant (CI) + auditory brainstem implant (ABI) for neurofibromatosis type 2(NF2) patients

Abstract

A cochlear implant (CI) is a surgically implanted electronic device that provides a sense of sound to a person who is profoundly deaf or severely hard of hearing.

Cochlear implants may help provide hearing in patients who are deaf because of damage to sensory hair cells in their cochlea. In those patients, the implants often can enable sufficient hearing for better understanding of speech. The quality of sound is different from natural hearing, with less sound information being received and processed by the brain. However, many patients are able to hear and understand speech and environmental sounds. Newer devices and processing-strategies allow recipients to hear better in noise, enjoy music, and even use their implant processors while swimming.

An auditory brainstem implant (ABI)[1] is a surgically implanted electronic device that provides a sense of sound to a person who is profoundly deaf, due to retro cochlear hearing impairment (due to illness or injury damaging the cochlea or auditory nerve, and so precluding the use of a cochlear implant).[2]

The auditory brainstem implant uses technology similar to that of the cochlear implant, but instead of electrical stimulation being used to stimulate the cochlea, it is used to stimulate the brainstem of the recipient.

Neurofibromatosis type 2(NF2)[3] is an inherited disease. The main manifestation of this disease is the developmental of symmetric and nonmalignant brain tumor in the region of cranial nerve VIII which is the Auditory- Vestibular Nerve that transmit sensory information from the Inner ear to the brain. NF2 is caused by mutations of the Merlin gene which seems to impact the form and movement of the cell. The principal treatment consists of Neurosurgical removal of tumors. The underlying disorder does not have cure due to the cell function caused by genetic mutation.

ABI is used for patients who suffer from NF2 or other retro cochlear pathologies such as Inner ear Aplasia, but this device is not as successful as CI. Speech recognition in some patients with NF2 after surgery would be worsen and ABI could not provide speech recognition as well as CI for these patients, however in some patients speech recognition or even speaking with telephone has been provided by ABI usually non-tumor disorders.[4]

We propose a theory in this paper which indicates that a device which is a combination of CI and ABI might be more useful for patients who suffer from NF2.

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Abbrevations: CI, cochlear implant; ABI, auditory brainstem implant; NF2, neurofibromatosis type 2; OAE, oto acoustic emission

Discussion

NF2 is an inherited disease. The main manifestation of this disease is the developmental of symmetric and non malignant brain tumor in the region of cranial nerve VIII which is the Auditory- Vestibular Nerve that transmit sensory information from the Inner ear to the brain. NF2 is caused by mutations of the Merlin gene which seems to impact the form and movement of the cell. The principal treatment consists of Neurosurgical removal of tumors. The underlying disorder does not have cure due to the cell function caused by genetic mutation. ABI is used for patients who suffer from NF2 or other Retro cochlear pathologies such as Inner ear Aplasia. But this device is not as successful as CI. Speech recognition in some patients with NF2 after surgery would be worsen and ABI could not provide speech recognition as well as CI for these patients, however in some patients speech recognition or even speaking with telephone has been provided by ABI usually non-tumor disorders. Hearing Loss which is occurred following NF2 is not just because of Auditory Nerve. There are some evidences which confirm cochlear lesion even in the early stage of the disease including absence of Oto Acoustic Emission (OAE) and Cochlear Micro phonic. Probably Cochlear lesion is because of ischemia of cochlea by tumor. Cochlear Implant solely is done for some NF2 patients and results were relatively successful, so Hearing loss which is occurred by Tumors is a combination of Cochlea and Auditory Nerve lesions and CNS disorder following Peripheral lesions.

Conclusion

Prosthesis shall be invented by scientists which may be a combination of CI and ABI for patients who suffer from NF2. Perhaps double electrical stimulation on both Cochlea and Brainstem with the same Speech Processor.

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Conflicts of interest

Author declares there are no conflicts of interest.

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