

Review Article

Cochlear Implantation in Patients with Neurofibromatosis Type 2 and Patients with Vestibular Schwannoma in the Only Hearing Ear

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Cochlear implants are a new surgical option in the hearing rehabilitation of patients with neurofibromatosis type 2 (NF2) and patients with vestibular schwannoma (VS) in the only hearing ear. Auditory brainstem implant (ABI) has been the standard surgical treatment for these patients. We performed a literature review of patients with NF2 and patients with VS in the only hearing ear. Cochlear implantation (CI) provided some auditory benefit in all patients. Preservation of cochlear nerve integrity is crucial after VS resection. Results ranged from environmental sound awareness to excellent benefit with telephone use. Promontory stimulation is recommended although not crucial. MRI can be performed safely in cochlear implanted patients.

1. Introduction

1.1. Treatment Approach in Bilateral Hearing Loss. Bilateral hearing loss represents a great disability for patients with neurofibromatosis type 2 (NF2) and patients with vestibular schwannoma (VS) in the only hearing ear. The treatment for VS patients (unilateral and bilateral) is diverse, including observation, surgery, and radiotherapy [1].

Treatment of the worst hearing ear in NF2 patients could leave an only hearing ear with tumor and the risk of further hearing loss. Moreover, treatment of VS in an only hearing ear can also lead to deafness. Both situations represent a therapeutic dilemma [2]. Developments of new hearing rehabilitation strategies have changed the management of these patients. Figure 1 shows the MRI of a patient with bilateral vestibular schwannomas.

1.2. Surgical Options in Hearing Rehabilitation. Since 1979, auditory brainstem implant (ABI) has been the only auditory rehabilitation option in patients with no serviceable hearing and previous schwannoma resection. ABI was designed to restore hearing in patients with nonfunctional cochlear

nerves who were not candidates for cochlear implantation [3]. Although ABI provides environmental sound and significant lip reading assistance, they have not reached consistent results in speech discrimination [3–7].

Recently cochlear implants have emerged as a reasonable therapeutic option in selected cases. Initially patients should undergo schwannoma resection with preservation of the cochlear nerve as the main goal. Subsequently CI is done in a standard fashion through a cochleostomy or directly through the round window. Results of CI in NF2 patients and VS in an only hearing ear are quite promising [8–10] and may provide outcomes comparable to those of postlingually implanted nontumor patients.

In this study we reviewed cochlear implantation as hearing rehabilitation in patients with NF2 and in patients with VS in the only hearing ear.

1.3. Cochlear Nerve Preservation and Function. Bilateral vestibular schwannomas in a NF2 patient can invade and grow within the cochlear nerve, while unilateral sporadic vestibular schwannoma (VS) only compresses it [11]. Also, identification of the surgical plane between the tumor and