

Disease Characteristics of Neurofibromatosis Type II and Cochlear Implantation

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Abstract : This study analyzes the clinical manifestations, hearing rehabilitation methods and outcomes of a complex case of neurofibromatosis type II (NF2). Methods: The clinical manifestations, medical history, clinical data, surgical methods and postoperative hearing rehabilitation outcomes of an NF2 patient were analyzed to determine the hearing reconstruction method and postoperative effect for a special type of NF2 acoustic neuroma. Results: The patient had bilateral acoustic neuromas with profound sensorineural hearing loss in both ears. Peripheral blood genetic testing did not reveal pathogenic gene mutations, suggesting mosaicism. The patient had an intracochlear schwannoma in the right ear and severely impaired vision in both eyes. Cochlear implantation with tumor retention was performed in the right ear. After 2 months of family-based auditory and speech rehabilitation, the Categories of Auditory Performance (CAP) score improved from 0 to 5. Conclusion: NF2 has complex clinical manifestations and poor prognosis. For NF2 patients with intracochlear tumors, cochlear implantation with tumor retention can be used to reconstruct hearing.

Keywords : NF2, intracochlear schwannoma, hearing reconstruction, cochlear implantation

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