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Hearing optimisation in neurofibromatosis type 2: A systematic review.

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BACKGROUND: It is common for patients with neurofibromatosis type 2 to develop bilateral profound hearing loss hearing loss, and this is one of the main determinants of quality of life in this patient group.

OBJECTIVES: The aim of this systematic review was to review the current literature regarding hearing outcomes of treatments for vestibular schwannomas in neurofibromatosis type 2 including conservative and medical management, radiotherapy, hearing preservation surgery and auditory implantation in order to determine the most effective way of preserving or rehabilitating hearing.

SEARCH STRATEGY: A MESH search in PubMed using search terms (('Neurofibromatosis 2' [Mesh]) AND 'Neuroma, Acoustic'[Mesh]) AND 'Hearing Loss' [Mesh] was performed. A search using keywords was also performed. Studies with adequate hearing outcome data were included. With the exception of the cochlear implant studies (cohort size was very small), case studies were excluded.

EVALUATION METHOD: The GRADE system was used to assess quality of publication. Formal statistical analysis of data was not performed because of very heterogenous data reporting.

RESULTS: Conservative management offers the best chance of hearing preservation in stable tumours. The use of bevacizumab probably improves the likelihood of hearing preservation in growing tumours in the short term and is probably more effective than hearing preservation surgery and radiotherapy in preserving hearing. Of the hearing preservation interventions, hearing preservation surgery probably offers better hearing preservation rates than radiotherapy for small tumours but recurrence rates for hearing preservation surgery were high. For patients with profound hearing loss, cochlear implantation provides significantly better auditory outcomes than auditory brainstem implantation. Patients with untreated stable tumours are likely to achieve the best outcomes from cochlear implantation. Those who have

had their tumours treated with surgery or radiotherapy do not gain as much benefit from cochlear implantation than those with untreated tumours.

CONCLUSIONS: This review summarises the current literature related to hearing preservation/rehabilitation in patients with NF2. Whilst it provides indicative data, the quality of the data was low and should be interpreted with care. It is also important to consider that the management of vestibular schwannomas in NF2 is complex and decision-making is determined by many factors, not just the need to preserve hearing.

KEYWORDS: acoustic neuroma; cochlear implants; outcomes; sensorineural hearing loss; systematic reviews

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