HEARING AND REHABILITATION IN NEUROFIBROMATOSIS TYPE 2 (NF2)

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ETIOLOGY OF VS IN NF2

• Likely local genetic event-
• NF2 gene
  • Low-dose radiation
  • (Cohort studies, Hiroshima & Nagasaki)
MANAGEMENT CONSIDERATIONS

- Preservation of life
- Preservation of function:
  - Facial motion
  - Hearing
  - Balance
MANAGEMENT OPTIONS

- Observation
- Surgery
- Radiation
DECISION MAKING FACTORS

- Age (life expectancy vs average growth)
- General Health
- Tumor size
- Symptoms/Signs
- Patient Desire
HEARING – NATURAL HISTORY

(n = 491)

(Stangerup et al, O & N; 2010)
EFFECT OF TUMOR GROWTH

Sughrue 2010, UCSF

- Systematic review
- 34 studies; 982 pts
- VS < 25mm, Class A/B
- f/u 26 – 52 mo
- Better hearing
  - Growth < 2.5 mm/yr
  - 75% vs 32% (p < 0.0001)
  - Not related to initial size

(Sughrue et al, J Neurosurg; 2010)
Impact of SSD

The diagram illustrates various sub-scales of speech quality (SSQ) and their scores in different contexts. The axes represent SSQ sub-scale scores, and the data points show the impact of acoustic neuroma patients compared to population controls. The error bars indicate 95% confidence intervals.
RADIOSURGICAL TECHNIQUES

- Gamma Knife
- LINAC
- Cyber Knife
- Proton Beam
- All rarely considered in NF2
Long-term hearing outcomes following stereotactic radiosurgery for vestibular schwannoma: patterns of hearing loss and variables influencing audiometric decline

Clinical article

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Object. The goals of this retrospective cohort study were as follows: 1) to describe the long-term prevalence and timing of hearing deterioration following low-dose (12- to 13-Gy marginal dose) stereotactic radiosurgery (SRS) for vestibular schwannoma (VS); and 2) to identify clinical variables associated with long-term preservation of useful hearing following treatment.

Methods. Patients with serviceable hearing who underwent SRS for VS between 1997 and 2002 were studied. Data including radiosurgery treatment plans, tumor characteristics, pre- and posttreatment pure tone average, speech discrimination scores, and American Academy of Otolaryngology-Head and Neck Surgery hearing class were collected. Time to non-serviceable hearing was estimated using the Kaplan-Meier method. Univariate and multivariate associations with time to non-serviceable hearing were evaluated using Cox proportional hazards regression models.

Results. Forty-four patients met the study criteria and were included. The median duration of audiometric follow-up was 9.3 years. Thirty-six patients developed non-serviceable hearing at a mean of 4.2 years following SRS. The Kaplan-Meier estimated rates of serviceable hearing at 1, 3, 5, 7, and 10 years following SRS were 80%, 55%, 48%, 38%, and 23%, respectively. Multivariate analysis revealed that pretreatment ipsilateral pure tone average (p < 0.001) and tumor size (p = 0.009) were statistically significantly associated with time to non-serviceable hearing.

Conclusions. Durable hearing preservation a decade after low-dose SRS for VS occurs in less than one-fourth of patients. Variables including preoperative hearing capacity and tumor size may be used to predict hearing outcomes following treatment. These findings may assist in pretreatment risk disclosure. Furthermore, these data demonstrate the importance of long-term follow-up when reporting audiometric outcomes following SRS for VS.

(http://thejns.org/doi/abs/10.3171/2012.9.JNS12919)

Key Words • hearing preservation • stereotactic radiosurgery • Gamma Knife surgery • vestibular schwannoma • cerebelloponine angle
SURGICAL OPTIONS

• Translabyrinthine
• Middle Fossa
• Retrosigmoid
EARLY AND PROACTIVE MANAGEMENT
MIDDLE FOSSA CRANIOTOMY
William E. Hitselberger, MD

1931-2014
MIDDLE FOSSA CRANIOTOMY

• Indications
  • Up to 1 cm in the CPA
  • Laterally placed
  • Good hearing
CURRENT SMALL SERIES (6 MONTHS)

- 10/13 patients with preserved serviceable hearing (76.9%)
- 11/13 patients with SDS within 20% of preoperative score (84.6%)
Retrosigmoid Approach
POOR HEARING PREOP OR LARGE TUMORS

- Indications
  - Up to 1 cm in the CPA
  - Laterally placed
  - Good hearing
(Adapted from "Neuroatology"; Jackler and Brackmann)
Auditory Performance: Group Means

SERT
MTS-Word
CID

Sentences

Chance performance level
Lipreading Enhancement: Group Means

Iowa Vowels

CUNY Sentences

Sound
Vision
CUNY Sentence Scores Over Time: Group Means
PEDiatric ABI

- Implantation of congenitally deaf children who cannot benefit from cochlear implantation
  - Cochlear aplasia or malformation
  - Absence of cochlear nerve
  - Surgery for purposes of ABI only: No tumor resection!
Eisenberg et al. 2012
J Am Acad Audiol 23:412–421
SUMMARY AND CONCLUSION

• NF2
  • Proactive treatment for hearing preservation
  • ABI is the best we have for larger tumors
  • CI is possible in older patients
• Pediatric ABI
  • Promising early results
THANK YOU!