Hearing rehabilitation in patients with neurofibromatosis type II: the Quebec’s experience with auditory implants

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Auditory Brainstem Implants
Use of a closed set questionnaire to measure primary and secondary effects of neurofibromatosis type 2

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Affiliations + expand

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Quality of Life (QoL) Assessment in Patients with Neurofibromatosis Type 2 (NF2)

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Comparative Study

The relationship between patients' perception of the effects of neurofibromatosis type 2 and the domains of the Short Form–36

W J Neary, V F Hillier, T Flute, S D G Stephens, R T Ramsden, D G R Evans
Hearing loss in NF2

- Bilateral vestibular schwannomas
- Progressive bilateral retrocochlear SNHL → anacusis
- Unpredictable evolution
- Infiltrative tumor
- Destructive therapies

Conservative management + regular MRI
Cochlear Implants (CI)

- Considered in
  - untreated stable VS
  - Treated VS with intact cochlear nerve

- Open set speech : 40%

- deteriorates?

Objectives

1) To review the **hearing rehabilitation** methods in NF2 patients

2) To describe the **auditory outcomes** on short and long term
METHODS

• Retrospective case series (2000-2021)
• All NF2 patients > 16-year-old
• Single tertiary care referral center (CHU de Québec-Université Laval, Quebec City, Canada)
DATA COLLECTION

- Patients’ demographic information
- Surgery, radiotherapy and/or chemotherapy (e.g. bevacizumab)
- Hearing evolution
- Hearing rehabilitation methods implant details (CI and/or ABI)
- Auditory outcomes following implantation (short-term and long-term).
• Multimedia Audiological Test
  – *battery of tests developed at our center for French Canadian users*

Pure tone detection
Word identification in a closed-set paradigm
Word and sentence recognition in an open-set paradigm
Speech tracking
Results

12 patients

9 ♀ 3 ♂

43 yo (17-74)

10 Cochlear implants

5 Auditory Brainstem Implant
CI with stable VS in situ

Patient #3 (64 ♂)
Patient #6 (24 ♂)
Patient #9 (26 ♀)
Patient #11 (24 ♀)

CI

Regular users (1 year post op)

Retrosigmoid surgery + ABI

Translab + Retrosigmoid surgery + ABI

Hearing benefits

loss /2 years

Hearing benefits

loss /3 years

ABIs

ABI nonuser

ABI regular user

CI

Hearing benefits

loss /2 years
CI after VS surgical resection

Patient #7 (71♀)

Retrosigmoid surgery + cochlear nerve preservation

34 years later CI

CI regular user
Radiation therapy prior to CI

Patient #1 (74 ♀)
- Stereotactic RT (50 Gy)
- Hearing benefits loss /2 years
- Stable VS in situ
- Satisfying and stable results (4 years post op)

Patient #2 (48 ♀)
- Radiosurgery (gamma knife)
- CI Satisfying and stable results

Patient #4 (68 ♂)
- Radiosurgery (gamma knife)
- Deceiving CI results
- Tumor regrowth Stereotactic RT (18 Gy)
- CI nonuser

Patient #5 (43 ♀)
- Stereotactic RT (50 Gy)
- Hearing benefits loss /2 years
- Stable VS in situ
- Satisfying and stable results (4 years post op)
Multimodality therapy prior to ABI insertion

Patient # 8 (25 ♀)
- Translab surgery x2 + ABI radiosurgery
- ABI regular user (8 years po)

Patient # 10 (34 ♀)
- Radiosurgery Translab+retrosigmoid +ABI
- ABI regular user (2 years po)

Patient # 12 (17 ♀)
- Bevacizumab Retrosigmoid + ABI
- Retrosigmoid x2 Proton beam therapy
- ABI regular user (10 years po)
Hearing performances: Cochlear implant

- 6/10 regular users – follow up period: 2.3 years (1-6 years)

- PTA:
  - Pre op: 105 dB
  - Post op: 21.5 dB

- Open set unaided sentence recognition:
  - Preop: 2% (0 – 7%)
  - Post op: 55.8% (0 – 83%)
  - Most recent audiological evaluation: 56.2% (0-98%)
Hearing performances : ABI

• 3/5 regular ABI users 1/5 occasional user 1/5 nonuser

• Follow up mean 5.2 years (1 – 11 years)

• Mean PTA post op 25.5 dB

• Mean unaided open set sentence recognition :
  – Preop : 7% (0 – 18%)
  – Post op : 23% (0 – 44%)
  – Most recent evaluation : 14% (0 – 42%)
Discussion: ABI

• Invasive surgery
• Inferior audiological performances
  <12 % of sentence recognition scores (1-3)
• 23 % of sentence recognition in our study
• 80% regular use,
  – added benefit to lip-reading.
• Stable results

Discussion: CI

- 55.8% of sentence recognition scores
- 2.3 years of follow up (1, 6 years)
- Superior performance to ABI
Hearing performance: CI

4 CI failure

Patient #1
contralateral functional CI

Patient #5
CI nonuser

Patient #9
ABI nonuser

Patient #11
ABI regular user

Predictor of CI failure - Tumor size?

Discussion

• Cochlear nerve monitoring
  - Promontory stimulation test
  - eABR
  - CNAP
  - Stapedial reflexes

CN integrity before CI (in unoperated VS)

CN preservation during VS surgery
MRI Related challenges post CI
Conclusion

- ABI:
  - aid to lip reading
  - stable results
  - High user rate (80%)

- Role of CI in:
  - Non growing VS in situ
  - Post radiation
  - Post CN preserving – surgery

- Cochlear nerve monitoring