

Cochlear Implantation with Hearing Preservation in Neurofibromatosis Type II

Neil S. Patel, M.D.¹, Michael J. Link, M.D.^{1,2}, Amy P. Olund, Au.D.³, Matthew L. Carlson, M.D.¹
¹ = Department of Otorhinolaryngology, ² = Department of Neurologic Surgery, ³ = Department of Audiology
 Mayo Clinic, Rochester, MN

Case Presentation

A 52-year-old male was evaluated for hearing impairment in the setting of neurofibromatosis type II (NF2). At approximately age 24 he underwent resection of a large right sided vestibular schwannoma (VS) with resultant facial paresis and profound deafness. At age 37 he presented for management of a growing 1.1 cm left-sided VS in his only-hearing ear (Figure 1). He was treated with Gamma Knife radiosurgery (marginal dose 12 Gy, maximum dose 24 Gy) with durable tumor control. His hearing in the left ear declined progressively, with initial word recognition score (WRS) of 100% to 20% 16 years after radiosurgery (Figure 2). His standard four-frequency PTA was 89 dB HL and low-frequency PTA (averaged over 250 and 500 Hz) was 20 dB HL. His residual right-sided VS was treated with Gamma Knife radiosurgery due to recent growth.

The patient underwent cochlear implantation (CI) on the left side with a conventional length lateral wall electrode. Systemic corticosteroids and topical corticosteroids on the round window membrane were utilized. A slow, steady, full insertion was performed through the round window membrane.

MRI at Presentation

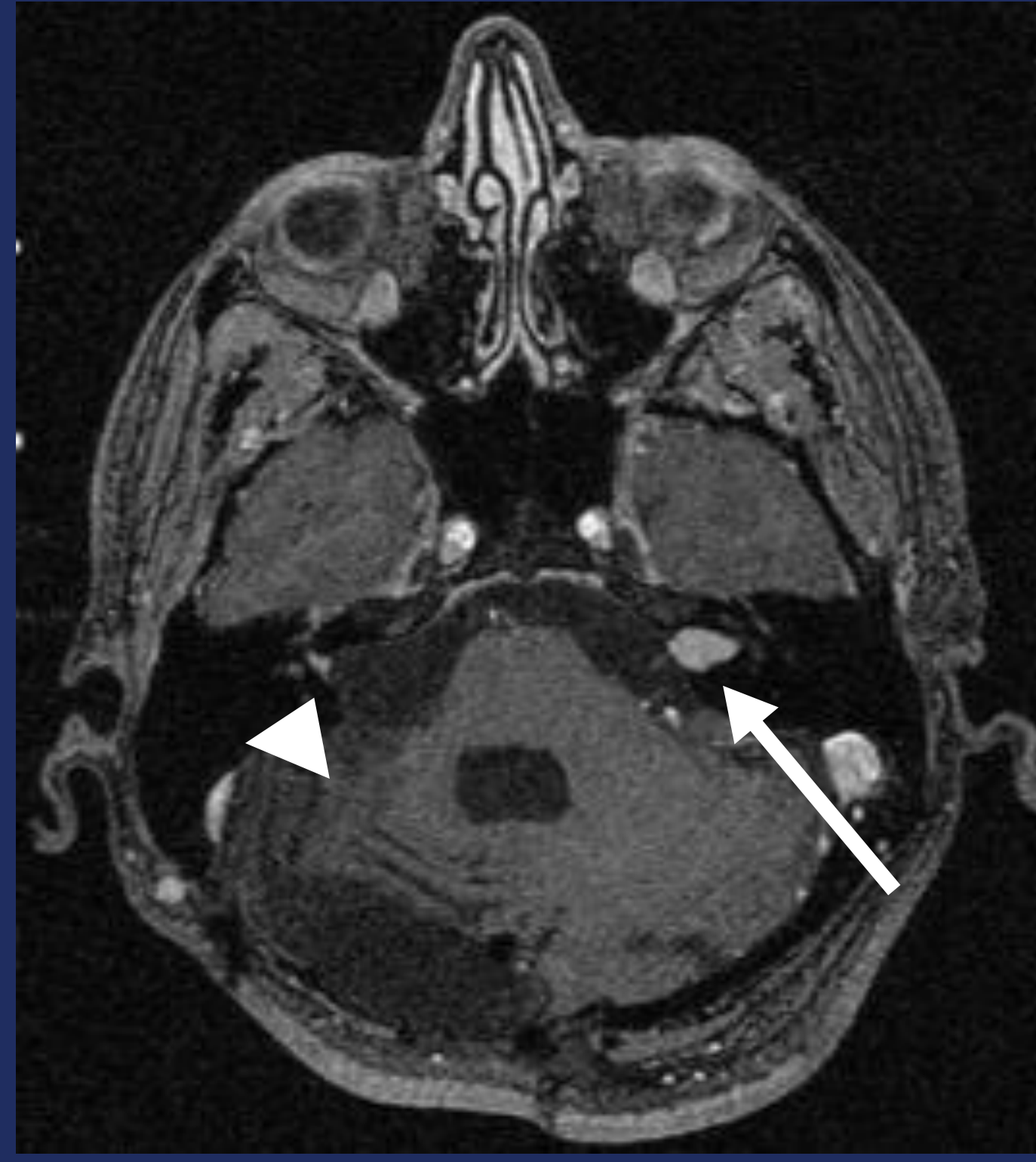


Figure 1 (above): Contrast-enhanced MRI demonstrating residual right-sided VS (white arrowhead) and 1.1 cm left-sided VS (white arrow)

Initial Audiometry

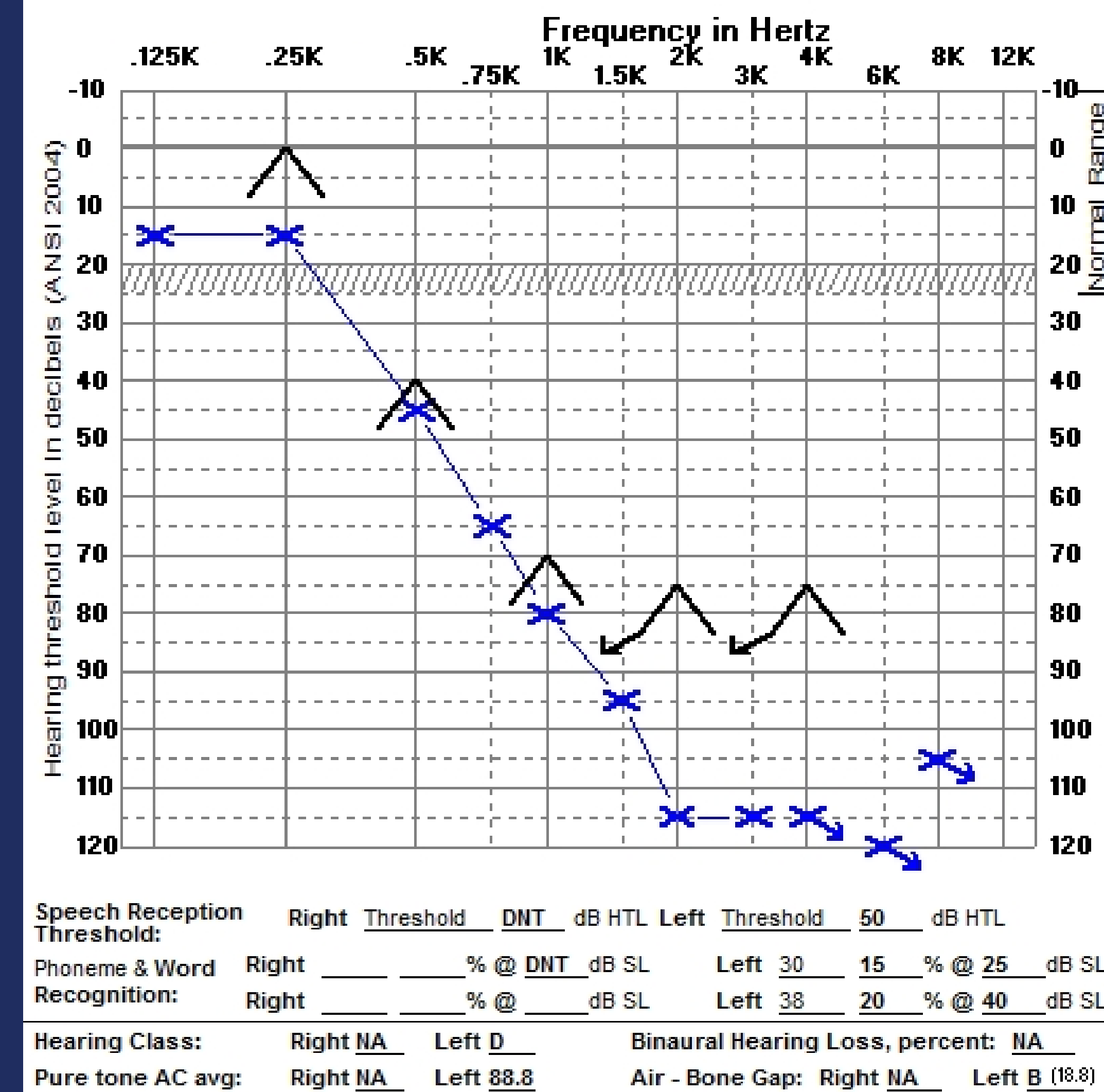


Figure 2 (left): Pure tone audiometry and speech testing at the time of cochlear implant evaluation. The right ear had a known profound hearing loss from prior VS resection. His PTA in his only-hearing ear was 89 dB HL. Cochlear implant candidacy speech testing was performed. The patient scored 8% of words on the Consonant-Nucleus-Consonant (CNC) word test list and 7% on AzBio sentences in quiet.

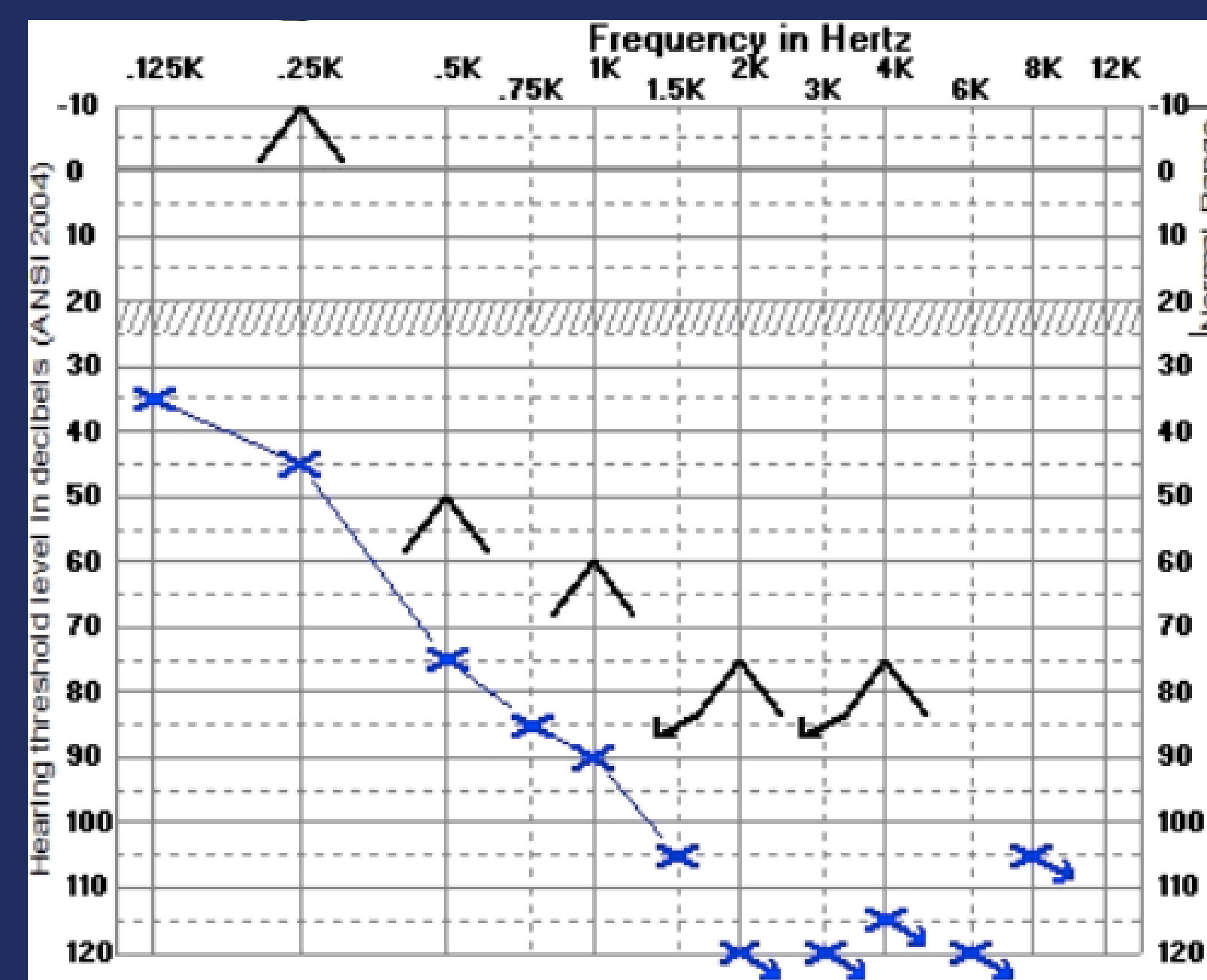
Results

A 3 week post-implantation pure-tone audiogram was obtained (Figure 3). This demonstrated a low-frequency PTA of 20 dB HL (unchanged from preoperative) consistent with complete hearing preservation, according to the 2013 HEARING Group guidelines¹.

One year after implantation, the patient exhibited aided audiometric thresholds of 25-40 dB HL across frequencies 125-8000 Hz. Speech testing results are listed in Table 1.

Surveillance of both VSs has been feasible with his cochlear implant receiver-stimulator in place (Figure 4). He is now 1.5 years from implantation and continues to be a daily CI user.

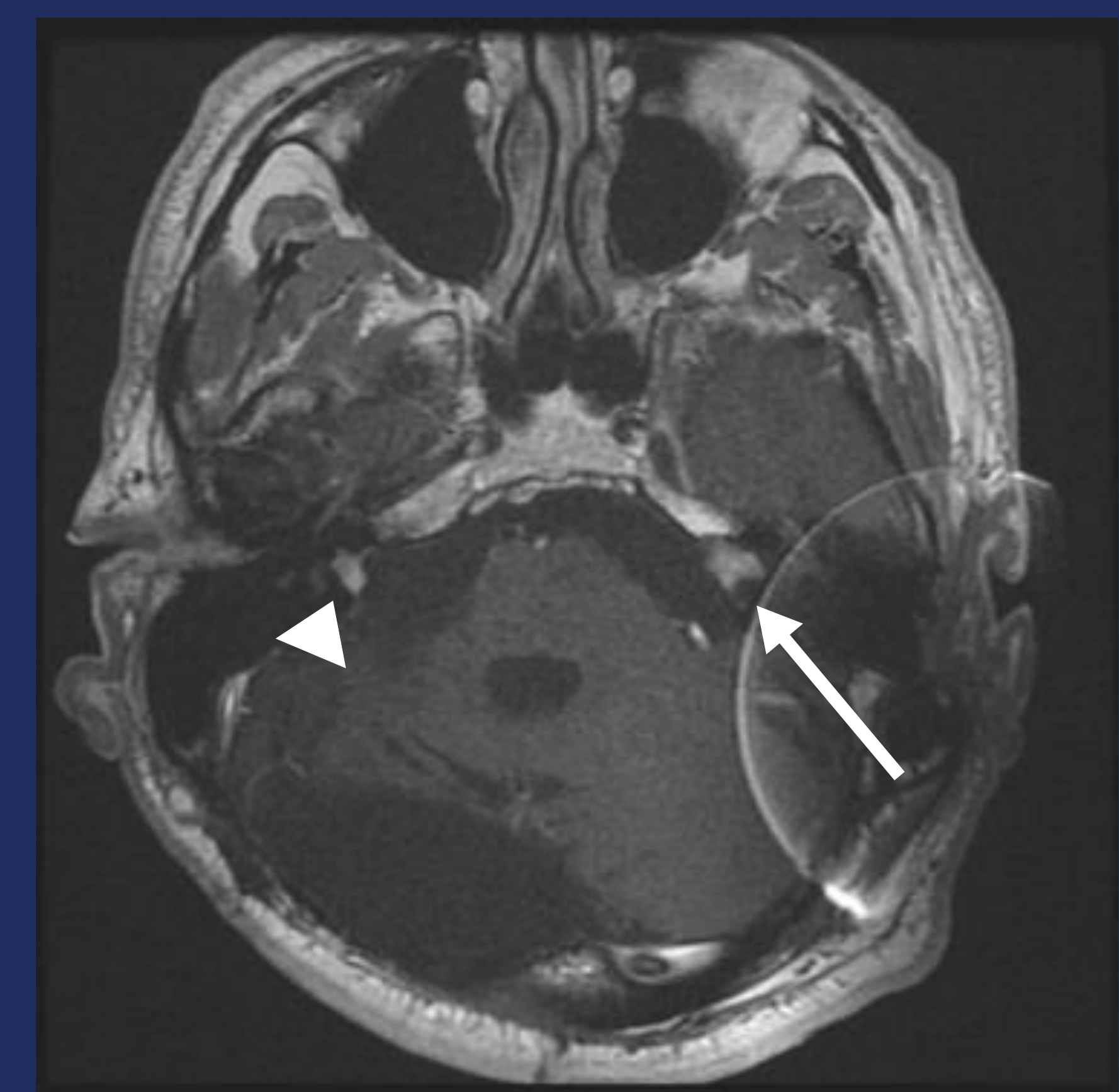
Post-Implantation Hearing Results



	Pre-CI	Post-CI
CNC word list	8%	52%
AzBio sentences in quiet	7%	44%

Table 1 (above): Pre- and 1 year post-implantation word and sentence scores. Figure 3 (left): Post-cochlear implantation audiometry demonstrating preserved low-frequency thresholds. One year after implantation, his CI-aided thresholds were in the 25-40 dB HL range. Figure 4 (right): Surveillance MRI with CI in place. The L sided VS is 15 years status post GKRS and has decreased in size. The R sided VS is 1 year status post GKRS and is stable.

Follow Up MRI



Discussion

- Hearing rehabilitation strategies in NF2 traditionally consist of hearing aids for serviceable hearing (AAOHNS class A or B) and auditory brainstem implantation (ABI) for more severe hearing loss. However, ABI users rarely achieve open-set word recognition.
- Multiple series have demonstrated rates of open-set speech recognition of up to 70%^{2,3} with a cochlear implant in the setting of NF2.
- As radiosurgery is employed frequently for the treatment of vestibular schwannoma in NF2, early cochlear implantation while residual hearing is available may allow patients to take advantage of electroacoustic stimulation and its well described benefits⁴.
- Consistent with a recent meta-analysis⁵, our experience suggests that hearing preservation rates are similar between hybrid and full length electrodes, provided that atraumatic surgical technique is employed.
- A full length electrode is preferred over a hybrid electrode in patients expected to lose residual hearing due to disease progression or treatment effect.
- In summary, cochlear implantation with preservation of residual hearing is possible in NF2 and should be offered to patients progressing to profound deafness.

References

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