UC San Diego
School of Medicine

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

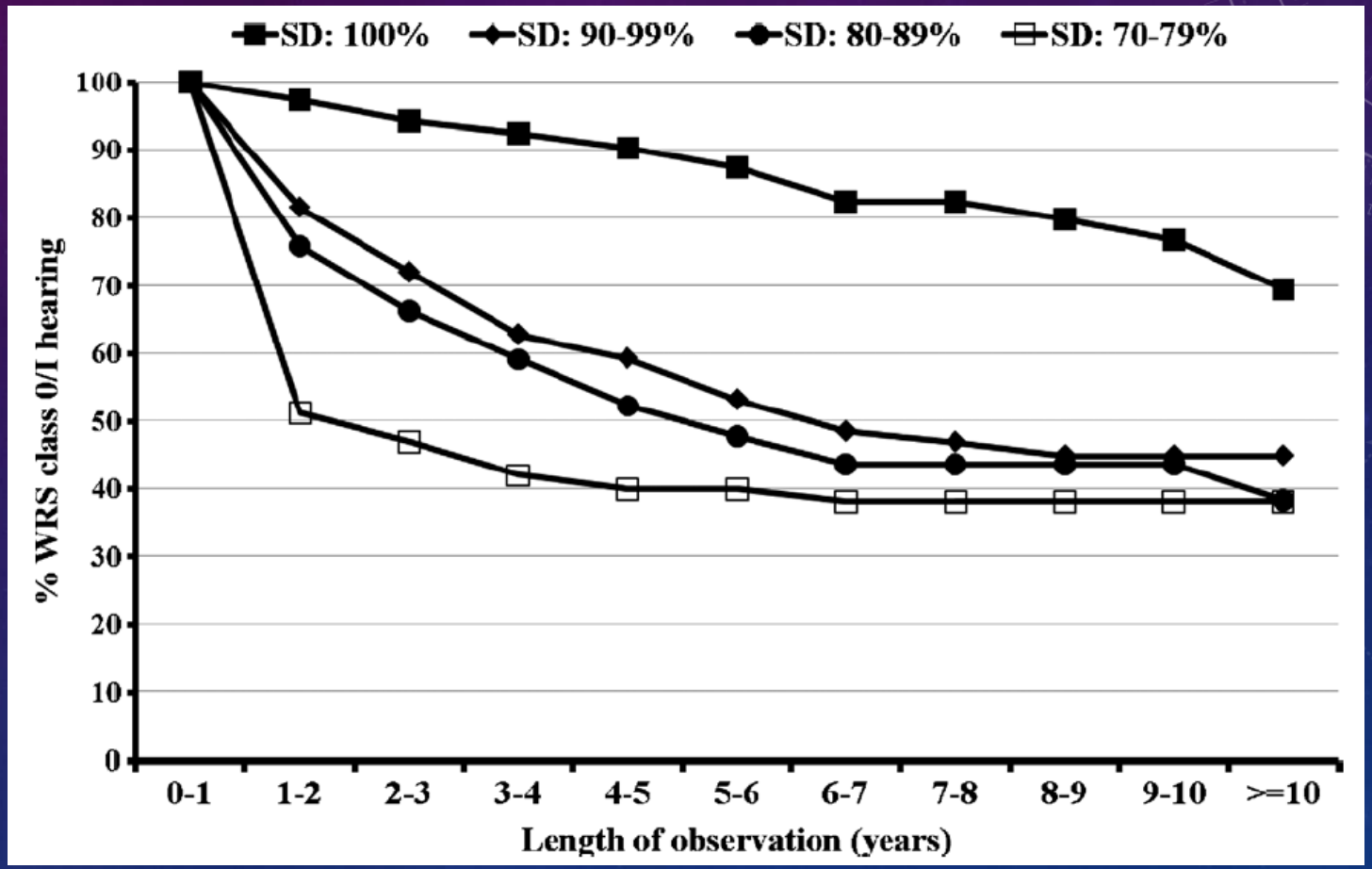
- Observation
- Surgery
- Radiation

DECISION MAKING FACTORS

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

HEARING – NATURAL HISTORY

(n = 491)

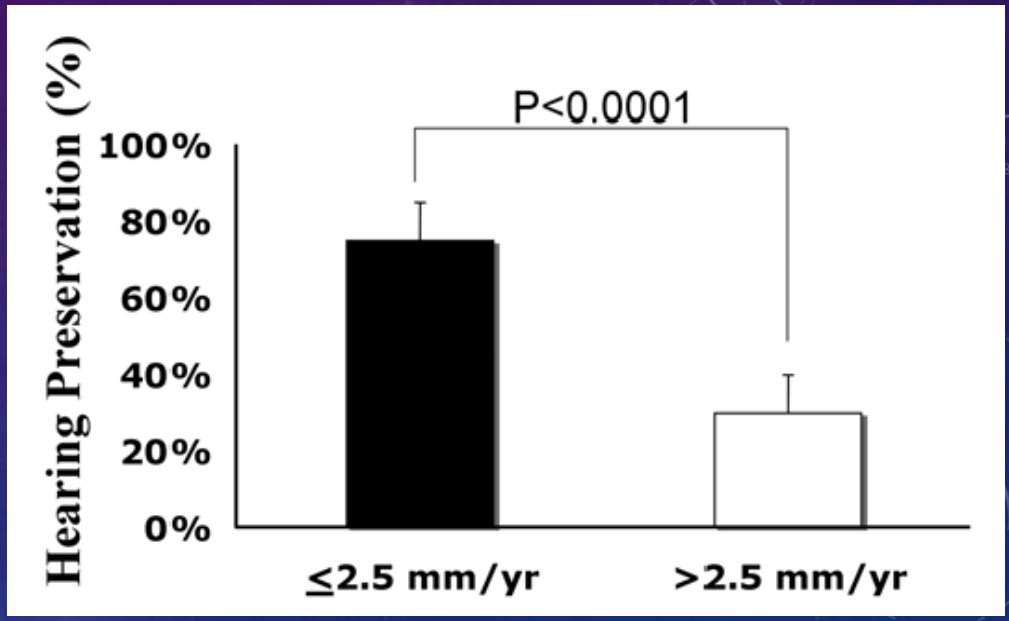


(Stangerup et al, O & N; 2010)

EFFECT OF TUMOR GROWTH

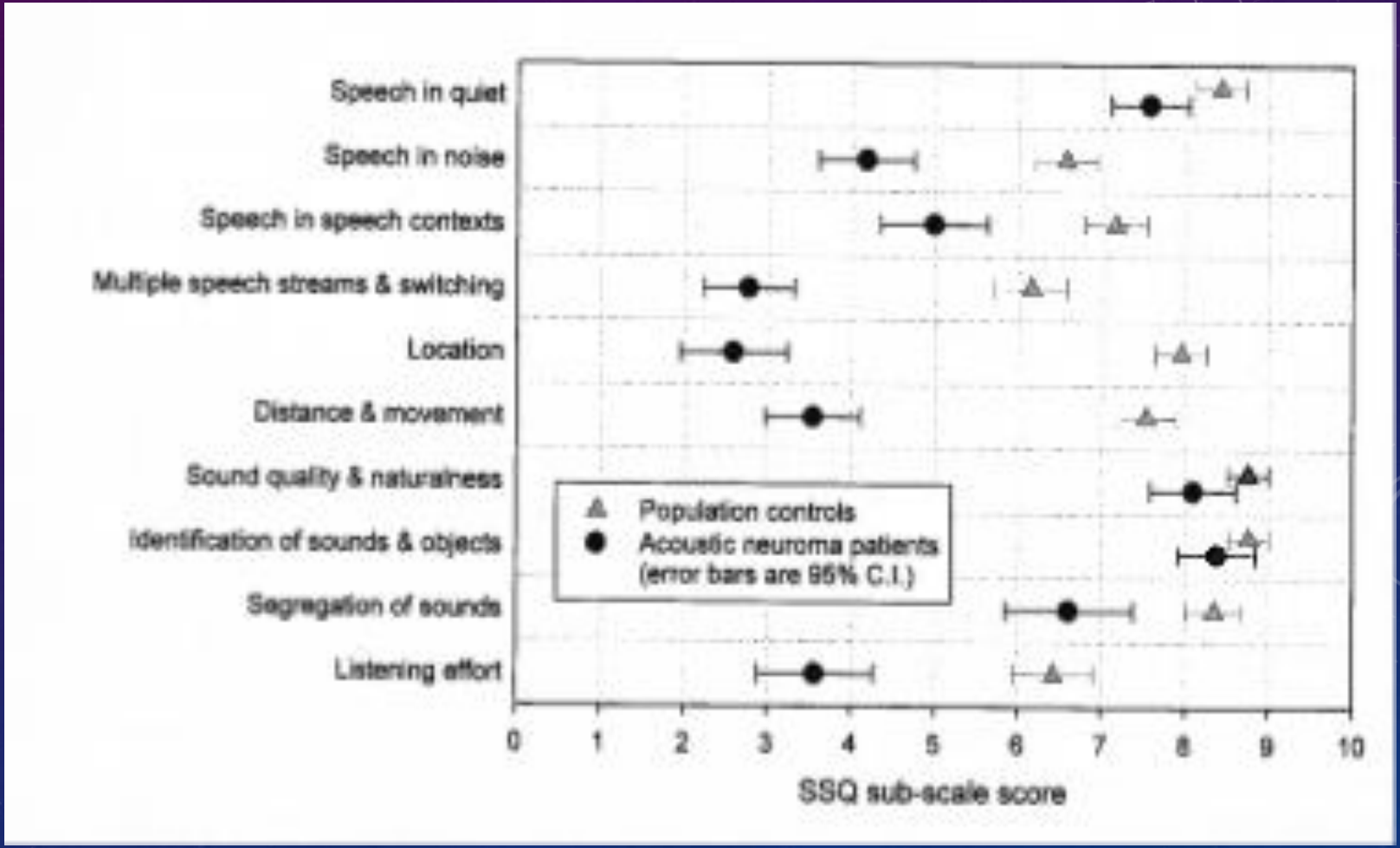
Sughrue 2010, UCSF

- 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



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 nonserviceable hearing was estimated using the Kaplan-Meier method. Univariate and multivariate associations with time to nonserviceable hearing were evaluated using Cox proportional hazards regression models.

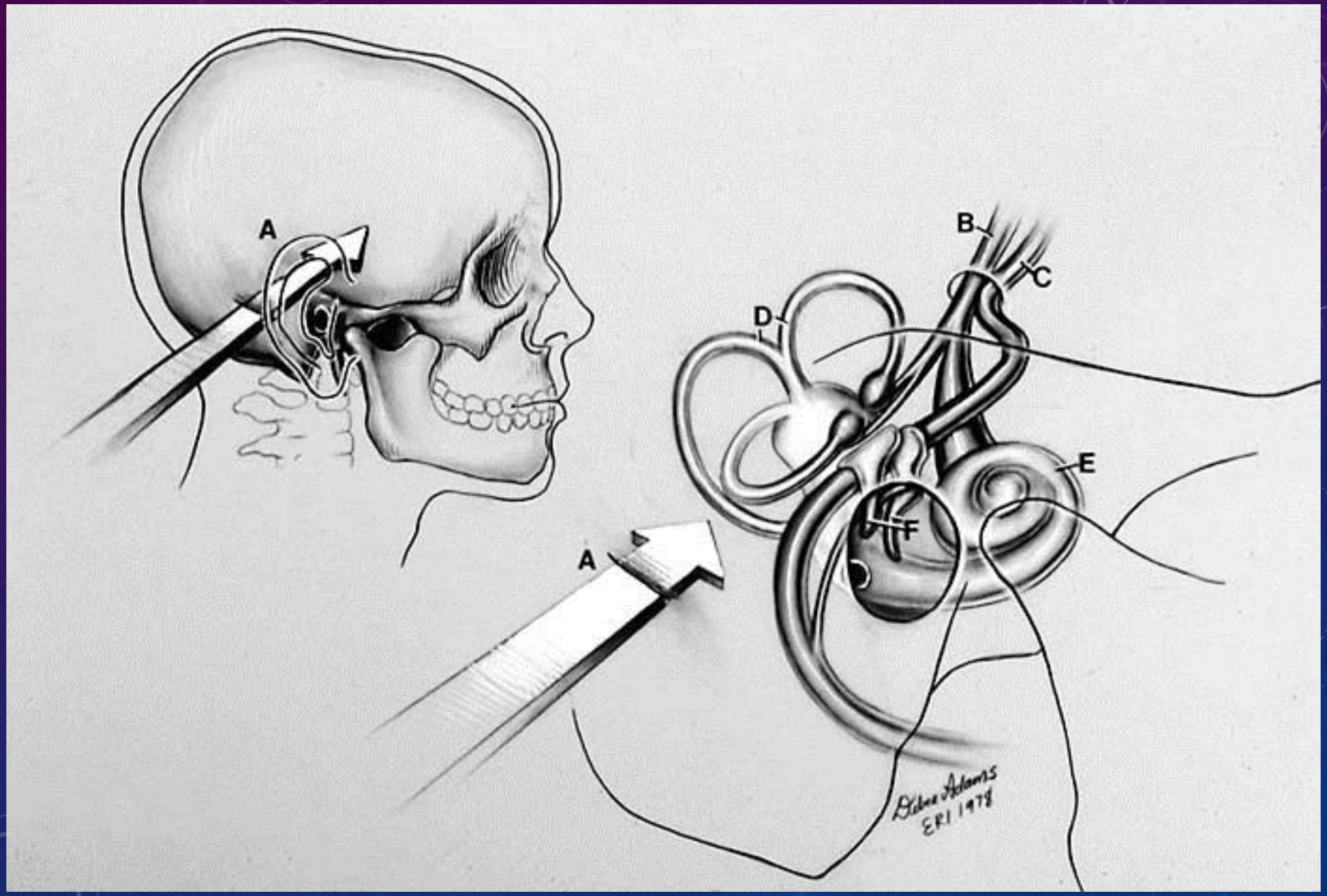
Results. Forty-four patients met the study criteria and were included. The median duration of audiometric follow-up was 9.3 years. Thirty-six patients developed nonserviceable hearing at a mean of 4.2 years following SRS. The Kaplan-Meier estimated rates of serviceable hearing at 1, 3, 5, 7, and 10 years 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 nonserviceable hearing.

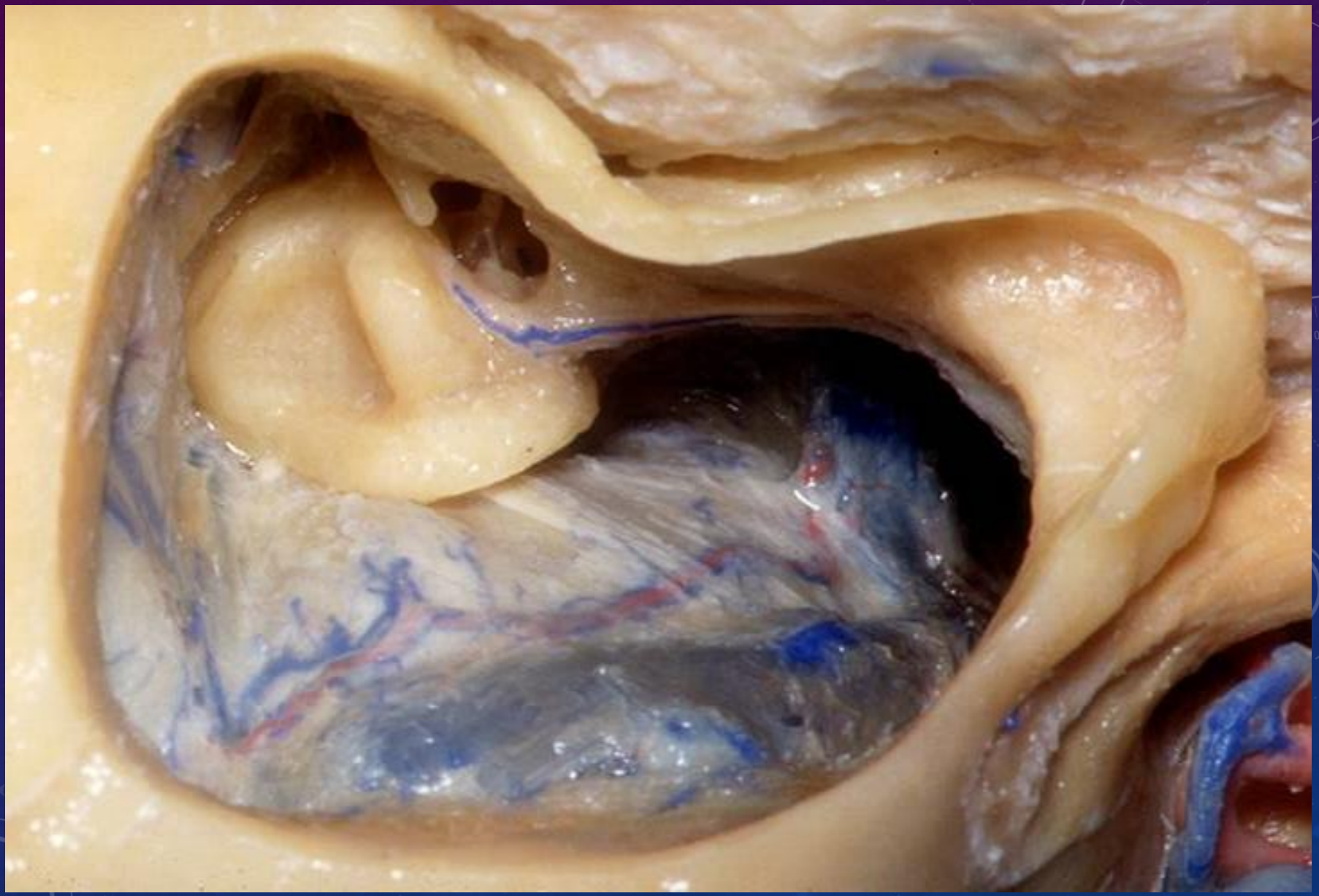
Conclusions. Durable hearing preservation a decade after low-dose SRS for VS occurs in less than one-fourth of patients. Variables including preoperative hearing capacity and tumor size may be used to predict hearing outcomes following treatment. These findings may assist in pretreatment risk disclosure. 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** • **cerebellopontine angle**

SURGICAL OPTIONS

- Translabyrinthine
- Middle Fossa
- Retrosigmoid


A
B
C
D
E
F
A
Debra Adams
ERI 1978

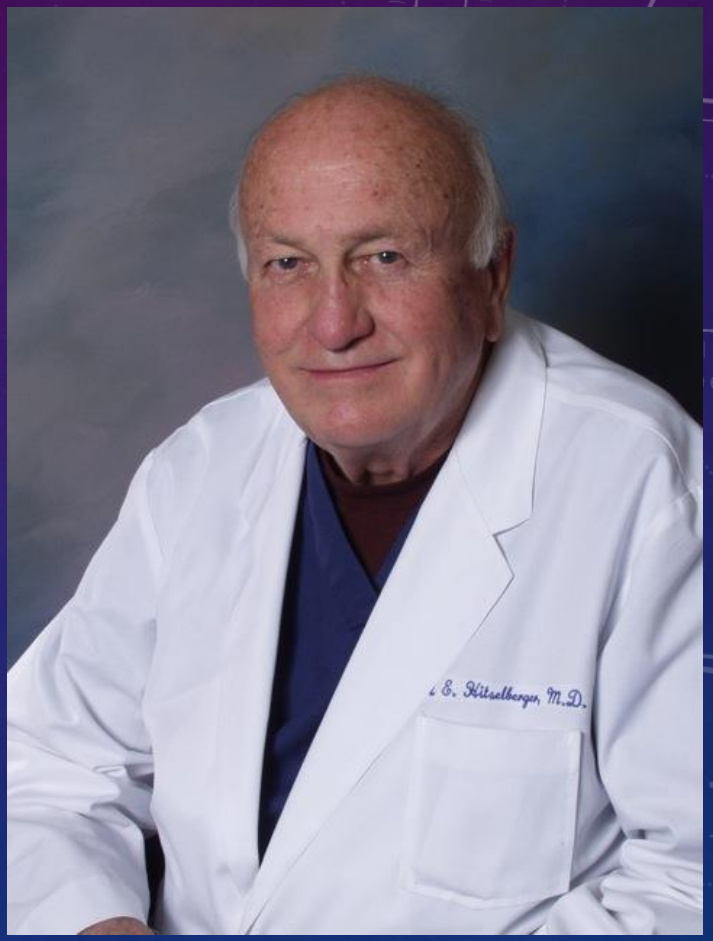


EARLY AND PROACTIVE MANAGMENT

MIDDLE FOSSA CRANIOTOMY

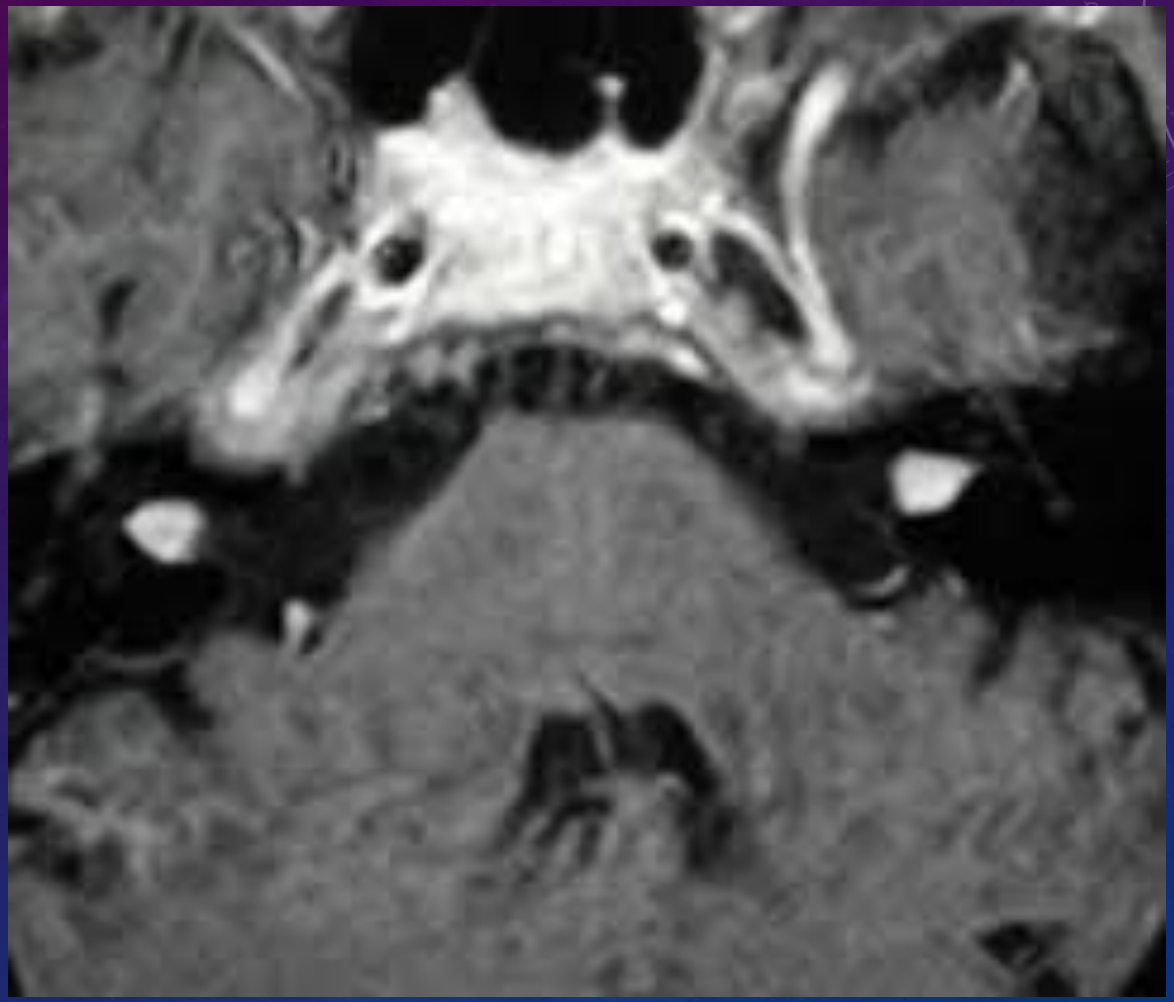
William E. Hitselberger, MD

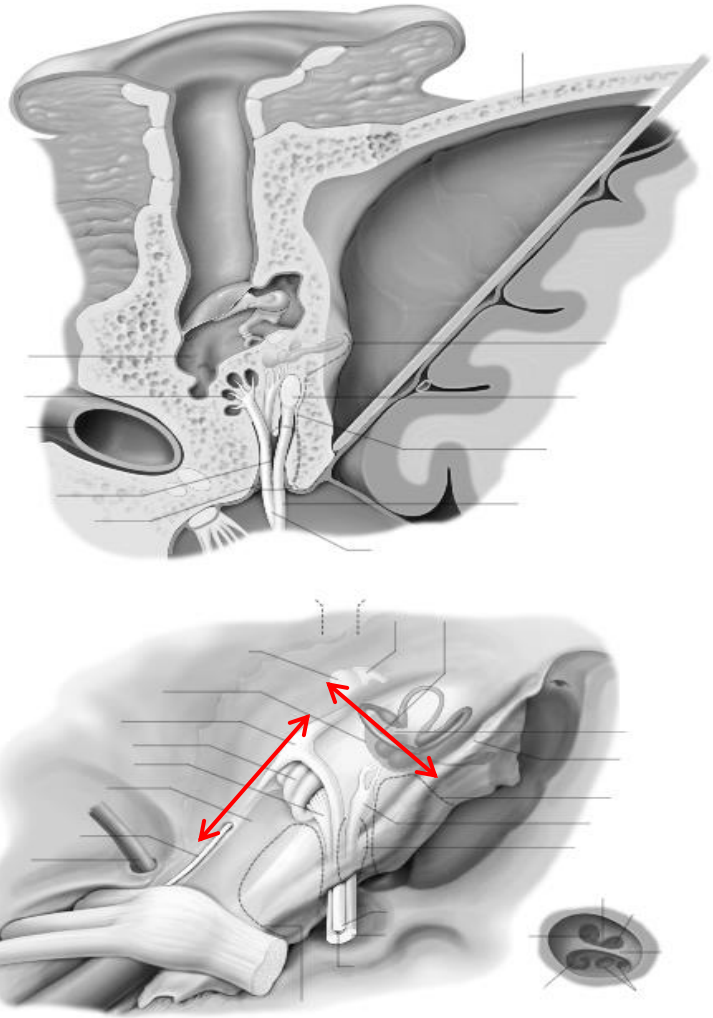
1931-2014

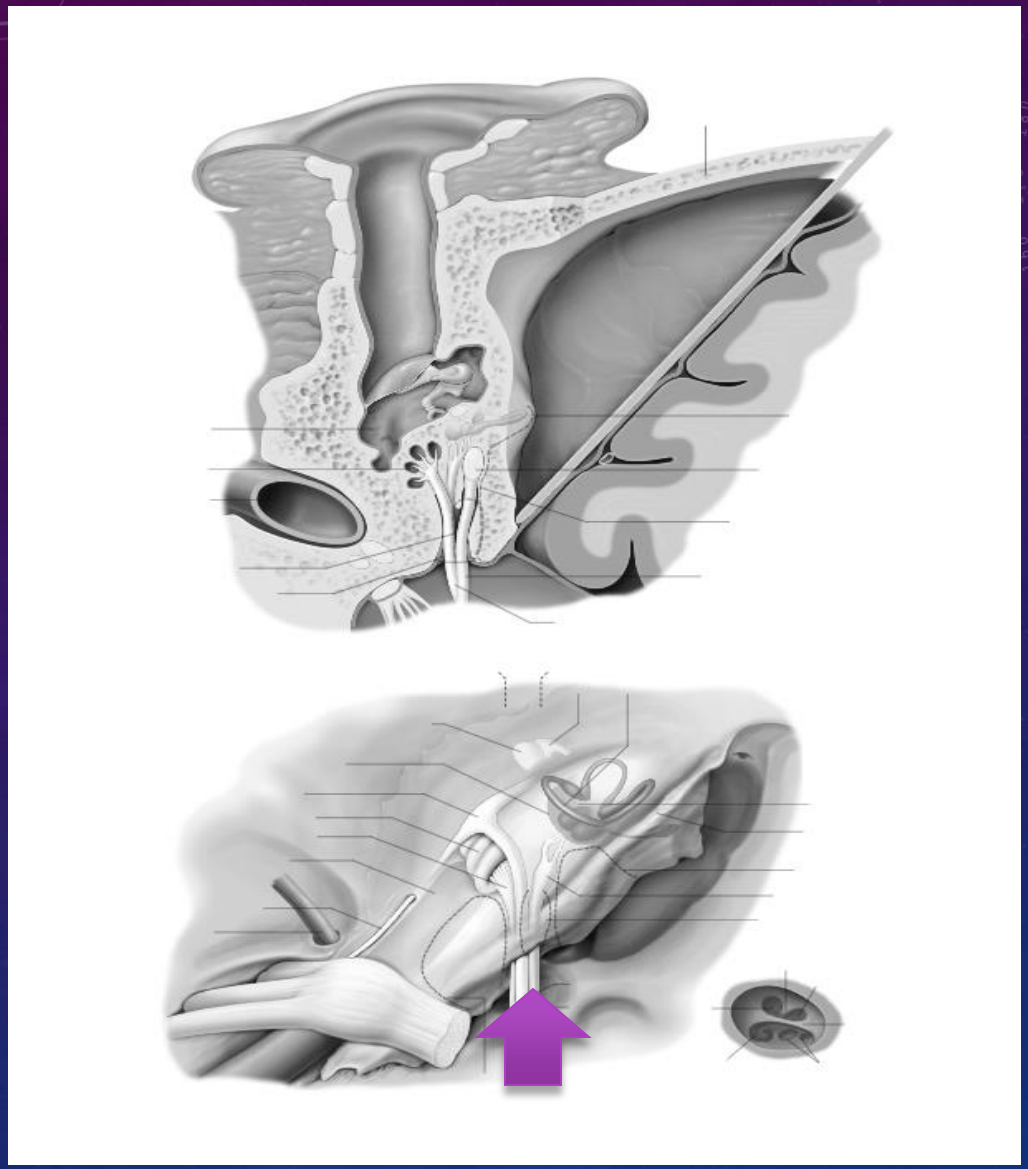


MIDDLE FOSSA CRANIOTOMY

- Indications
  - Up to 1 cm in the CPA
  - Laterally placed
  - Good hearing



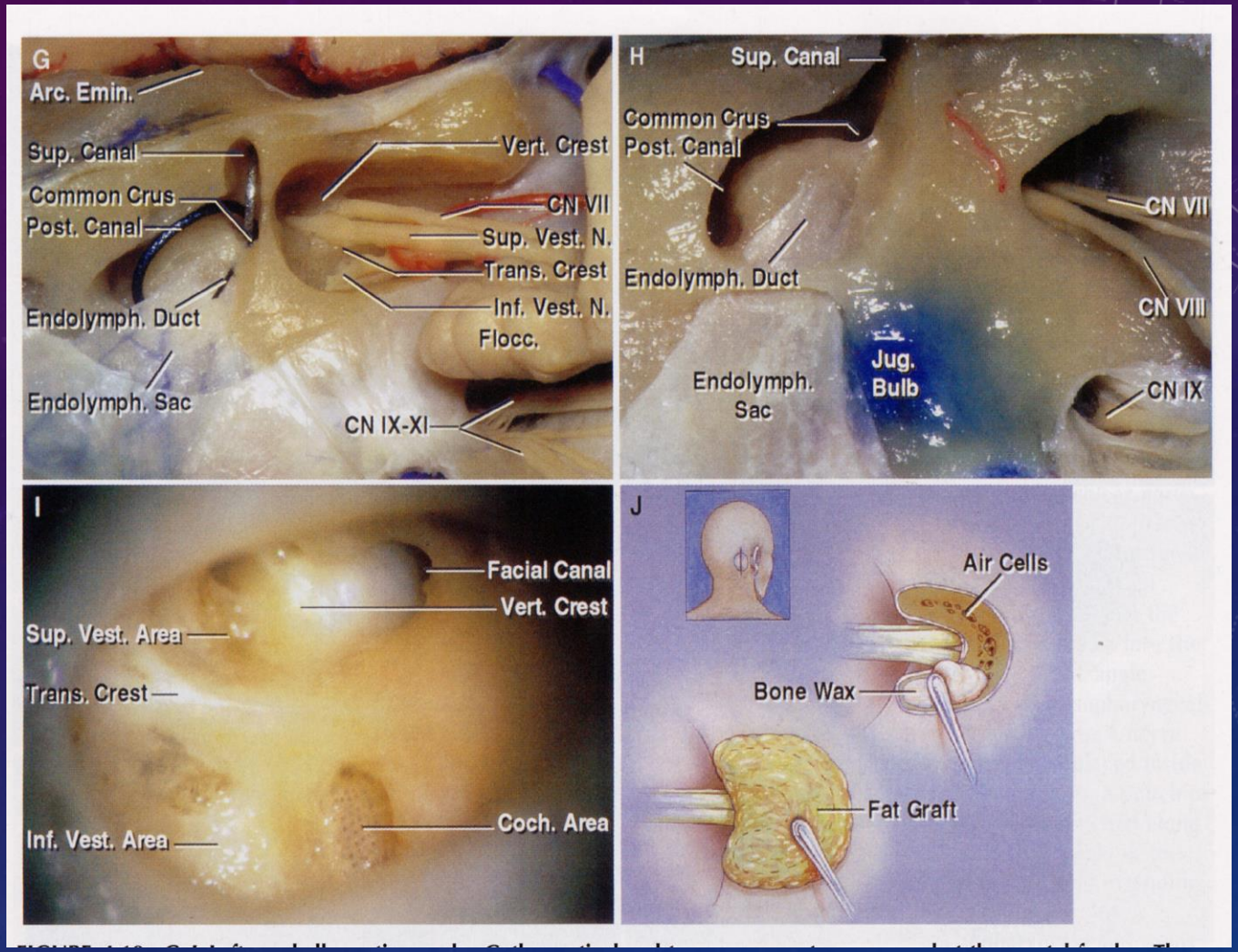




CURRENT SMALL SERIES (6 MONTHS)

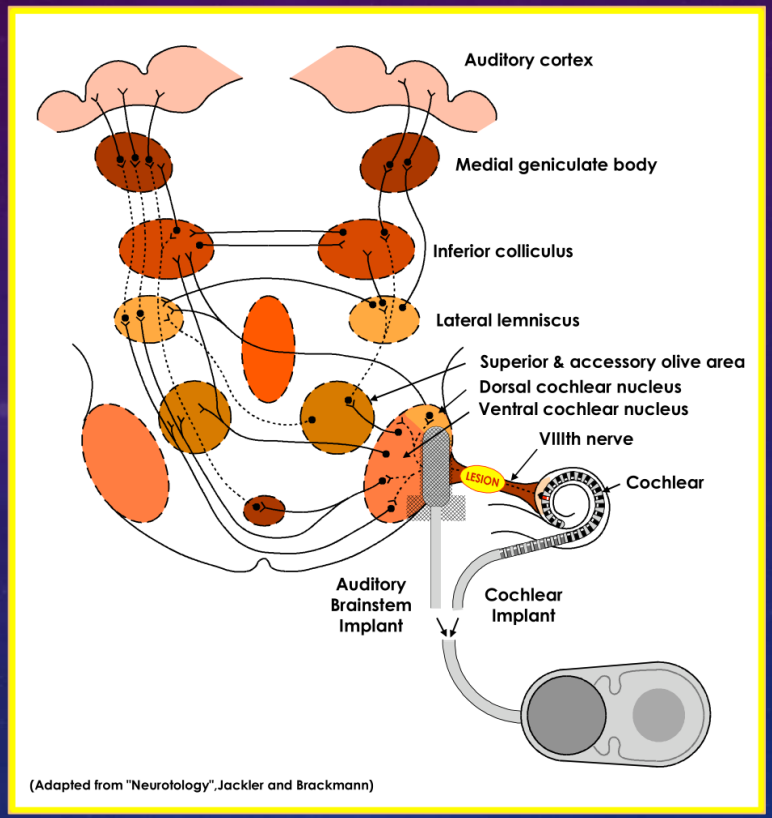
- 10/13 patients with preserved serviceable hearing (76.9%)
- 11/13 patients with SDS within 20% of preoperative score (84.6%)

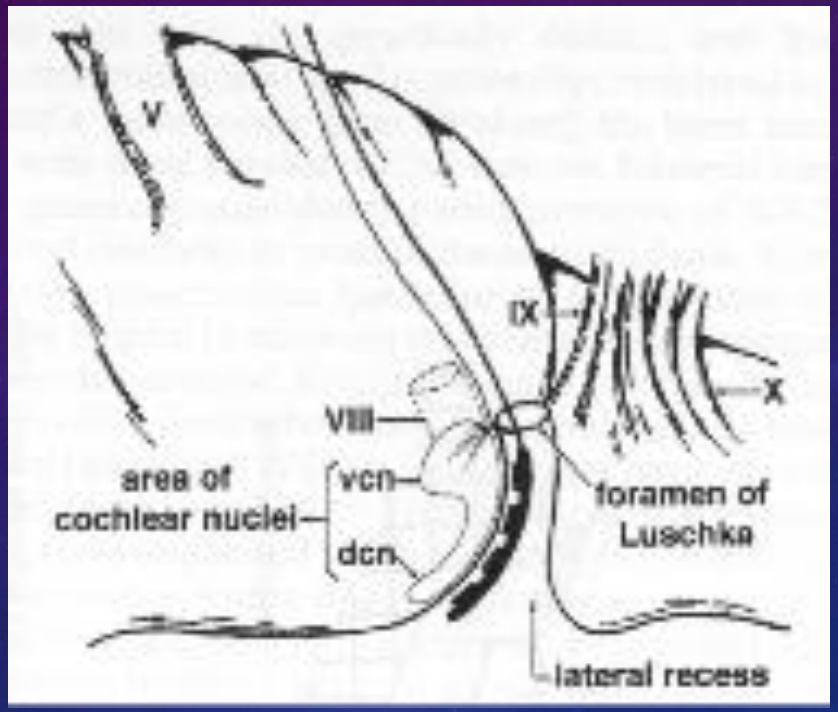
Retrosigmoid Approach

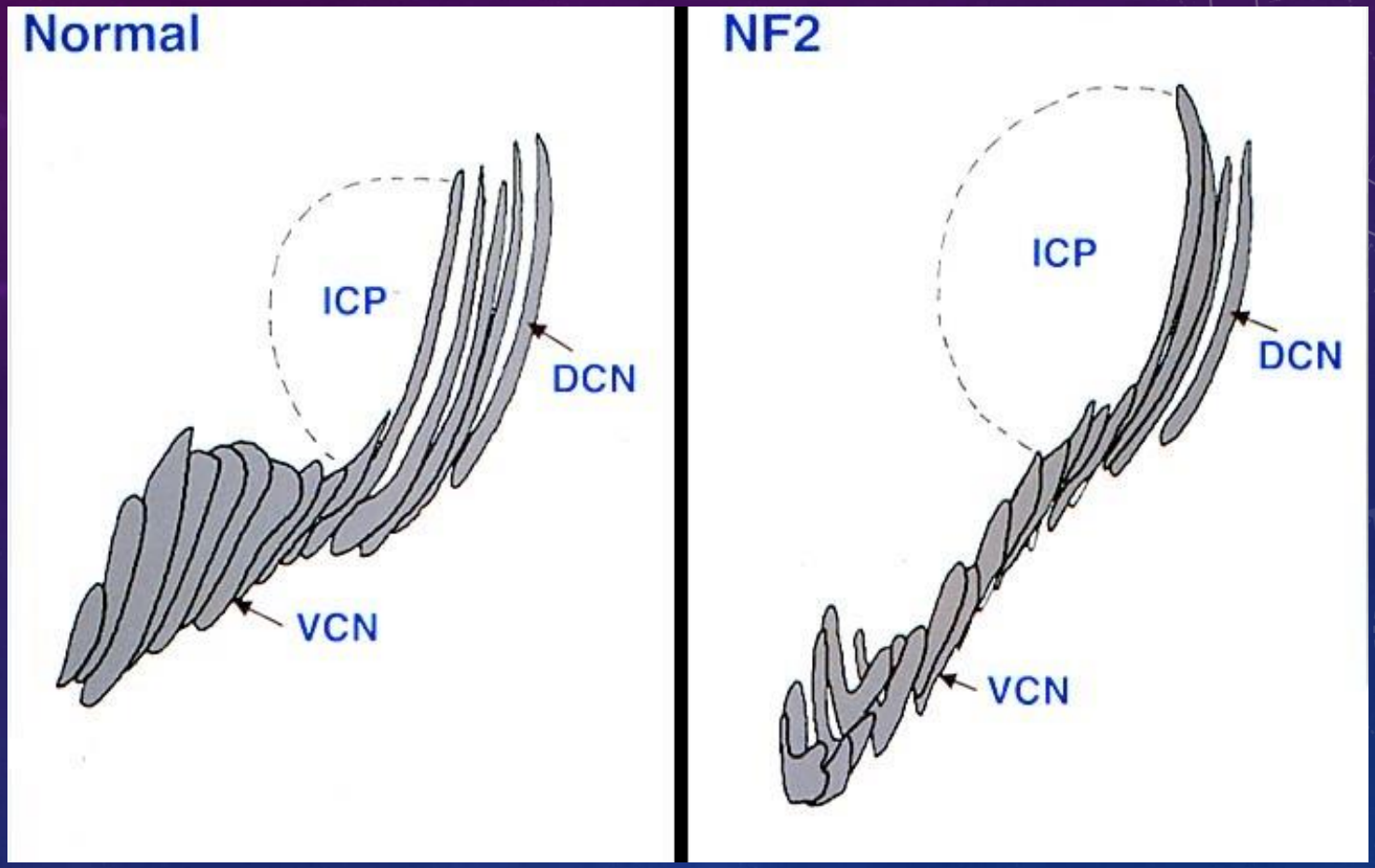


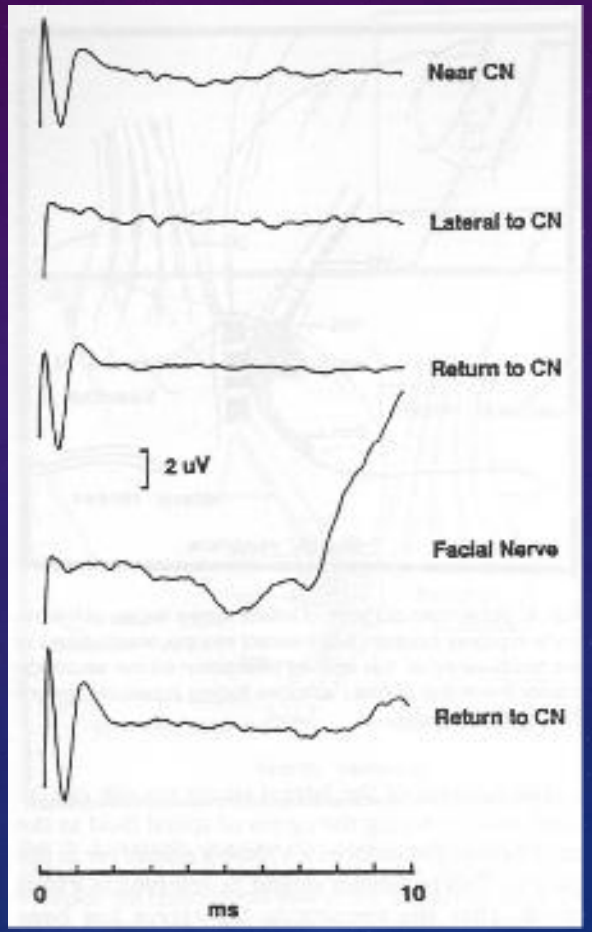
POOR HEARING PREOP OR LARGE TUMORS

- Indications
  - Up to 1 cm in the CPA
  - Laterally placed
  - Good hearing

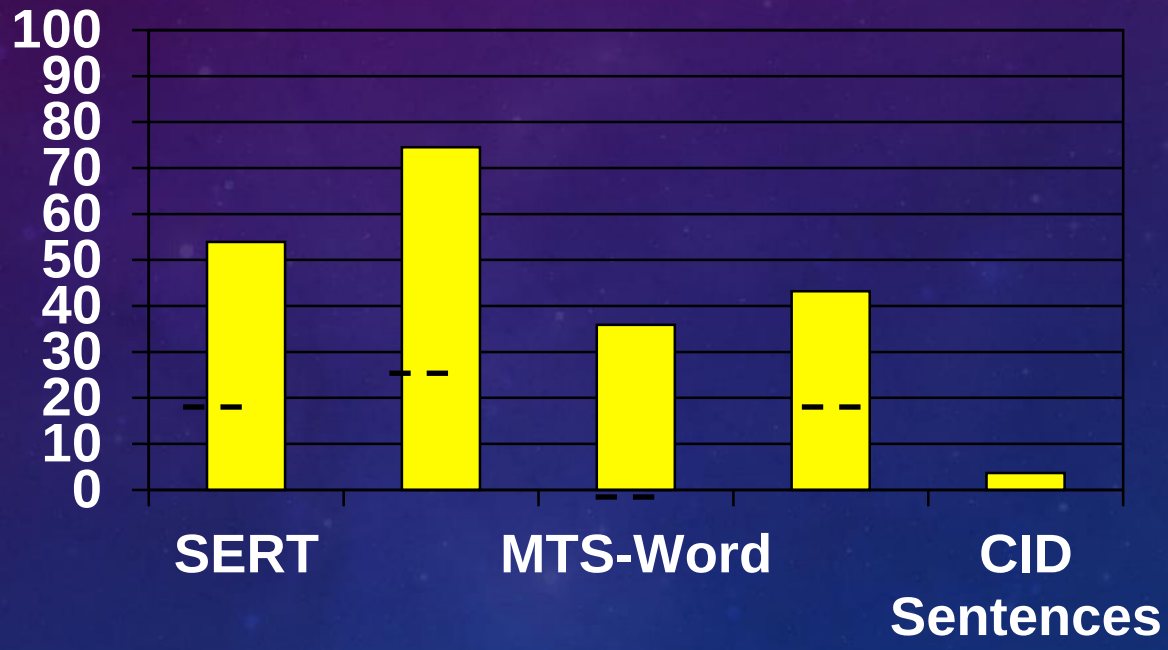

Auditory cortex
Medial geniculate body
Inferior colliculus
Lateral lemniscus
Superior & accessory olive area
Dorsal cochlear nucleus
Ventral cochlear nucleus
VIIIth nerve
LESION
Cochlear
Auditory Brainstem Implant
Cochlear Implant
(Adapted from "Neurotology",Jackler and Brackmann)


V
IX
X
VIII
area of
cochlear nuclei
vcn
dcn
foramen of
Luschka
lateral recess


Normal
ICP
DCN
VCN
NF2
ICP
DCN
VCN

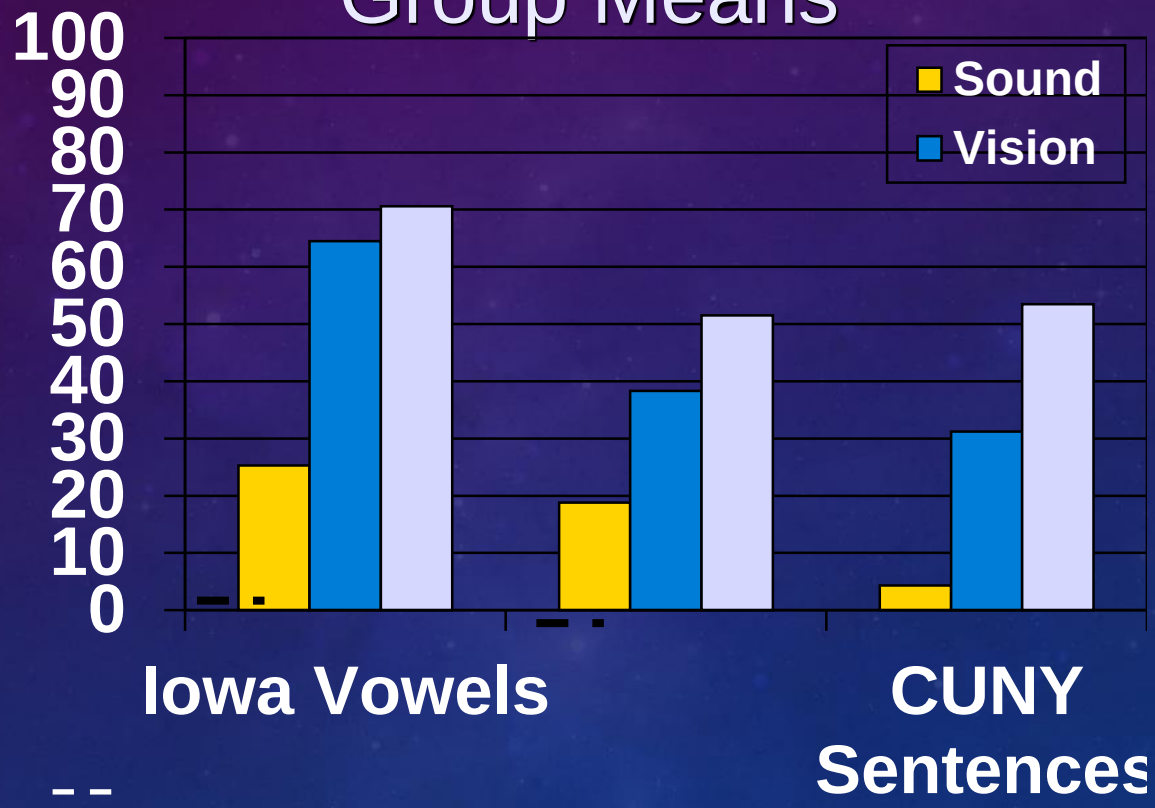

Near CN
Lateral to CN
Return to CN
2 uV
Facial Nerve
Return to CN
0
10
ms

Auditory Performance: Group Means

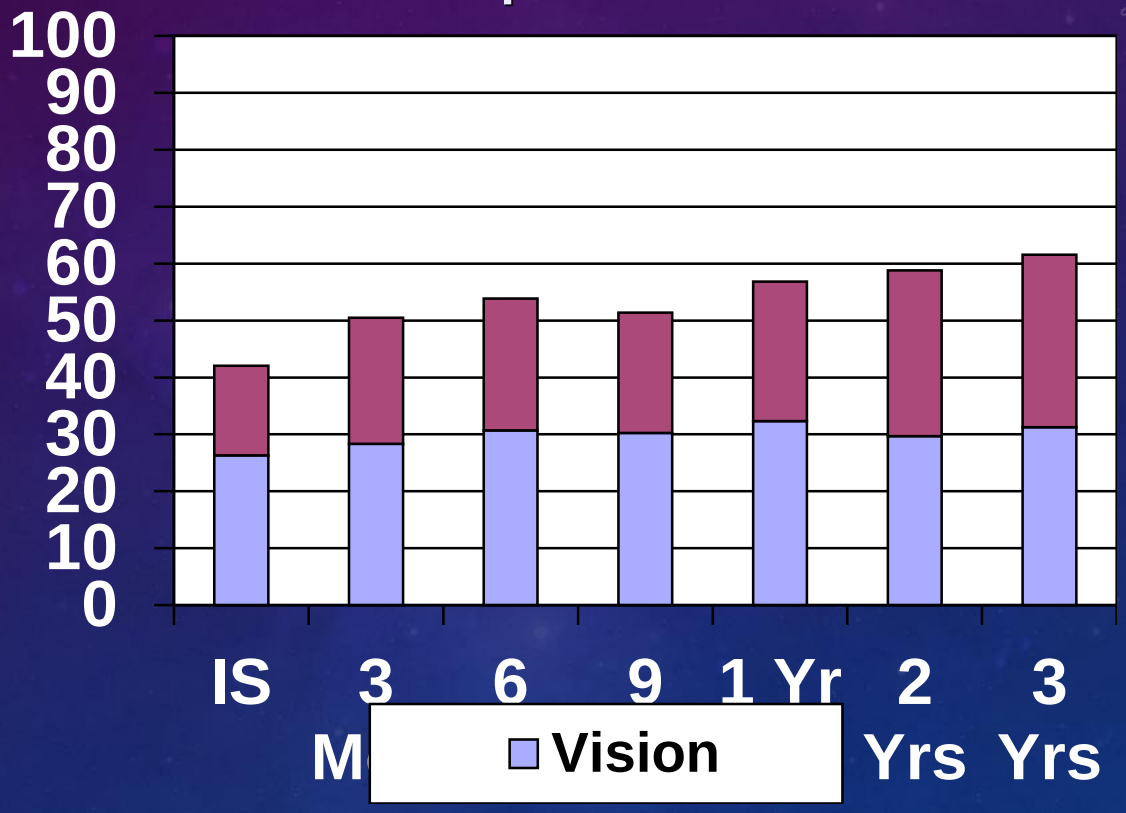


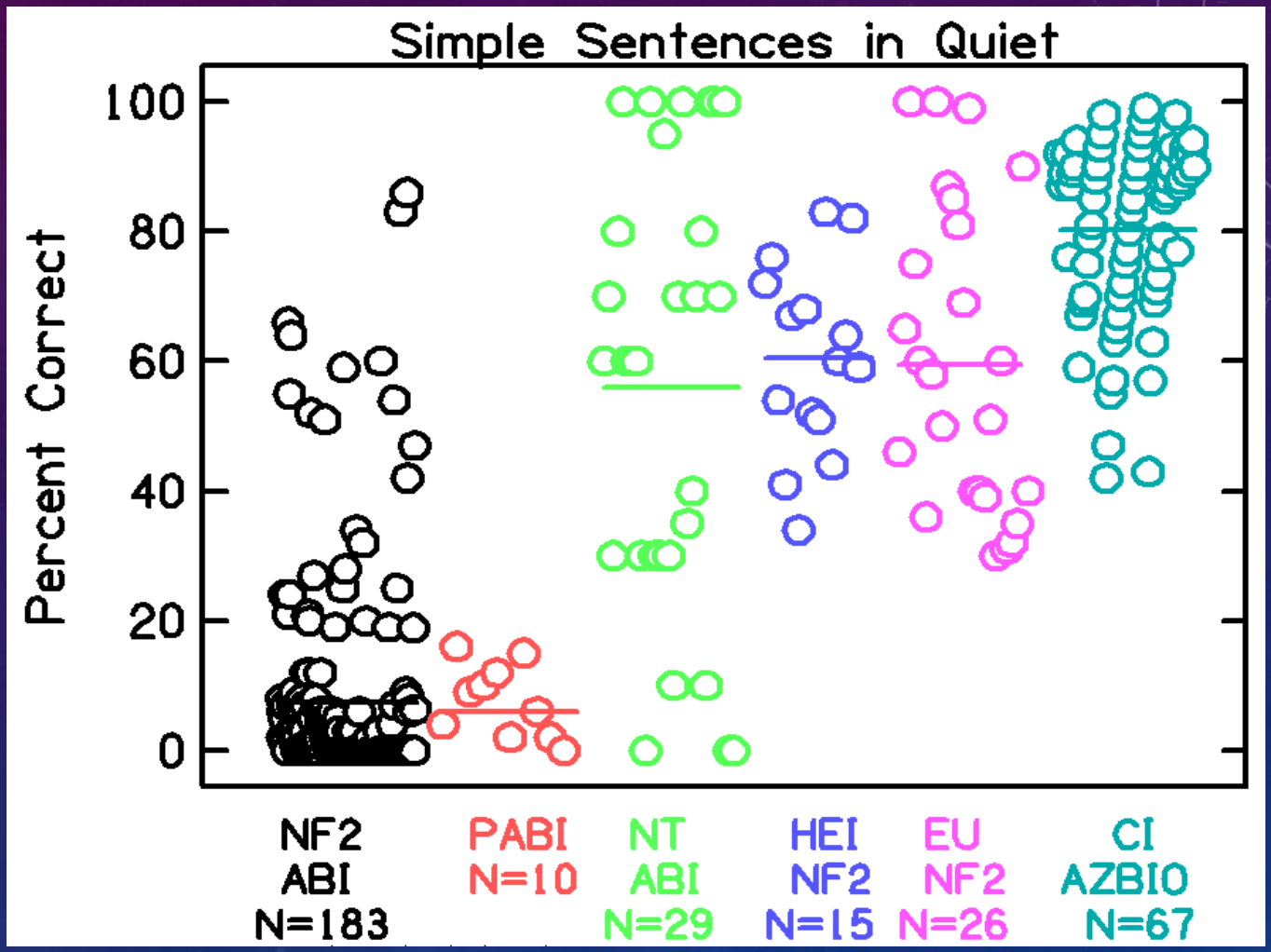
– – Chance performance level

Lipreading Enhancement: Group Means



CUNY Sentence Scores Over Time: Group Means

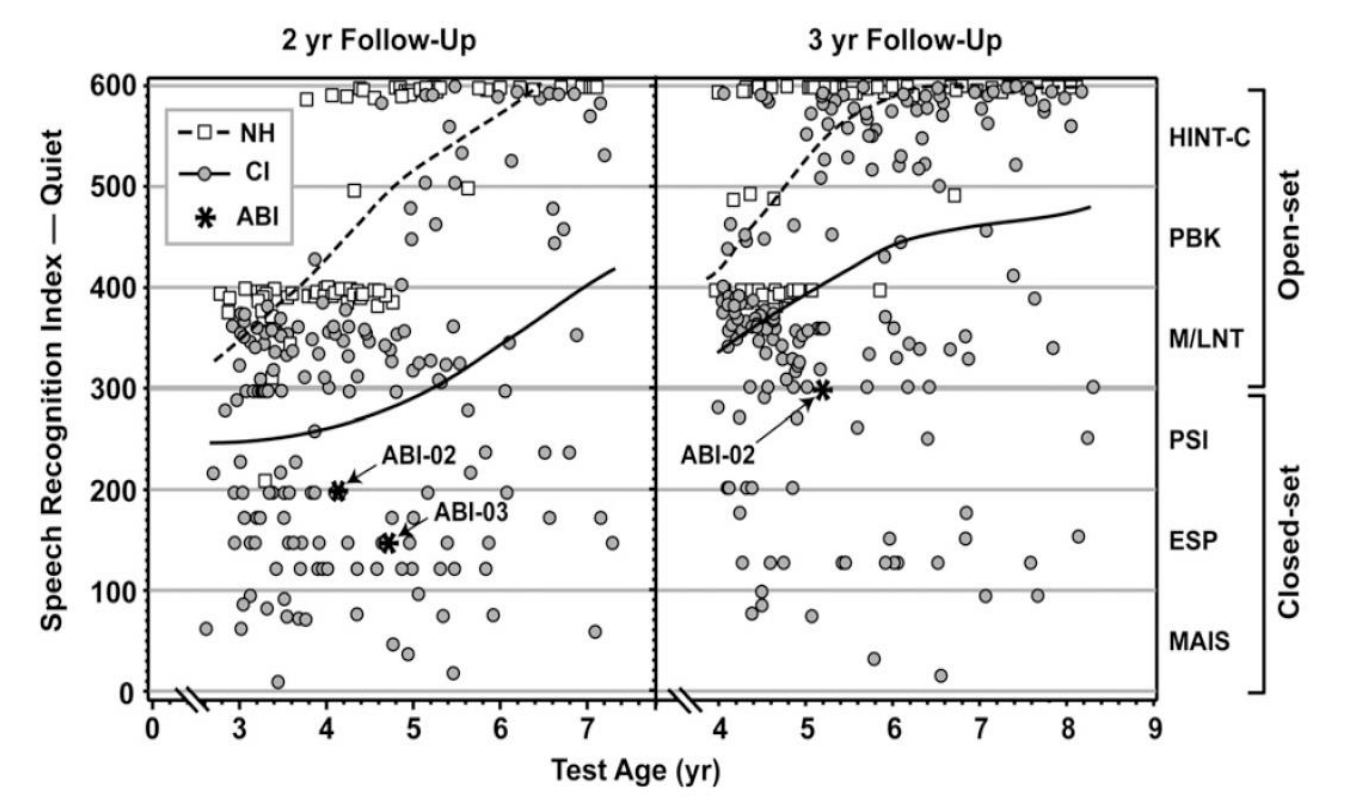


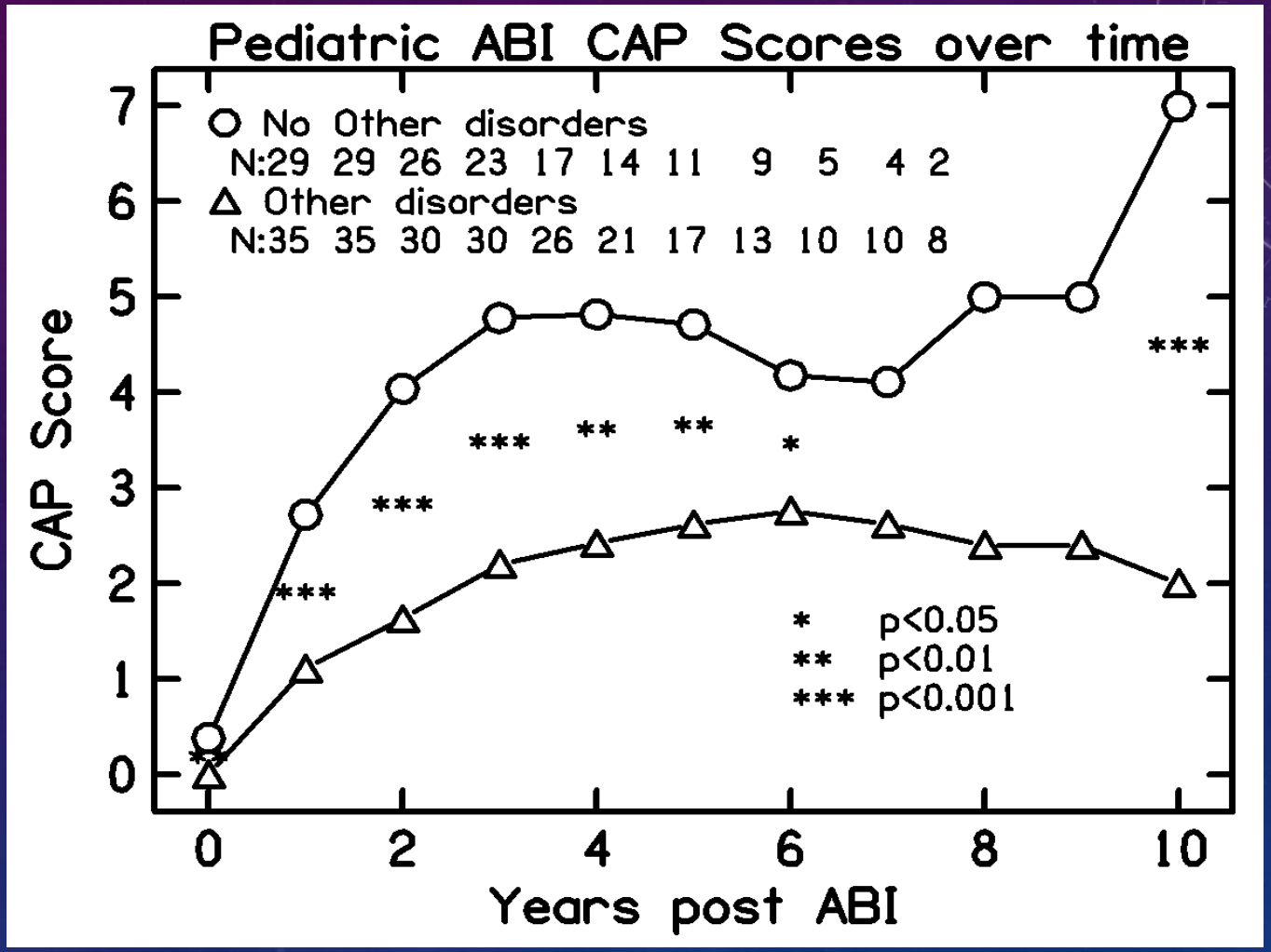

Simple Sentences in Quiet
Percent Correct
100
80
60
40
20
0
NF2
ABI
N=183
PABI
N=10
NT
ABI
N=29
HEI
NF2
N=15
EU
NF2
N=26
CI
AZBIO
N=67

PEDIATRIC ABI

- Implantation of congenitally deaf children who cannot benefit from cochlear implantation
  - Cochlear aplasia or malformation
  - Absence of cochlear nerve
  - Surgery for puposes of ABI only: No tumor resection!

Eisenberg et al. 2012
J Am Acad Audiol 23:412–421




Pediatric ABI CAP Scores over time
No Other disorders
N:29 29 26 23 17 14 11 9 5 4 2
Other disorders
N:35 35 30 30 26 21 17 13 10 10 8
CAP Score
Years post ABI
0
1
2
3
4
5
6
7
0
2
4
6
8
10
* p<0.05
** p<0.01
*** p<0.001

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!