Auditory Brainstem Implantation in Children

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SCHOOL OF MEDICINE

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  - Amy Edge
  - UNC Hospitals
  - NIH/NIDCD
  - Cochlear Corporation

ABI is investigational under an IDE from the US FDA

Pediatric Hearing Loss-The Problem

- Estimates
  - 3-4/1000 have HL (450-500 children/yr)
  - 1:1000 have severe to profound HL (~120 children in NC in 2011)
- Impact
  - Sound awareness
  - Speech understanding and language development
  - Educational Impact
  - Employment opportunities and Earning potential
  - ~$1 million per child lifetime costs ***when untreated***
    - 35% direct and 65 % indirect (lost earning potential, …)

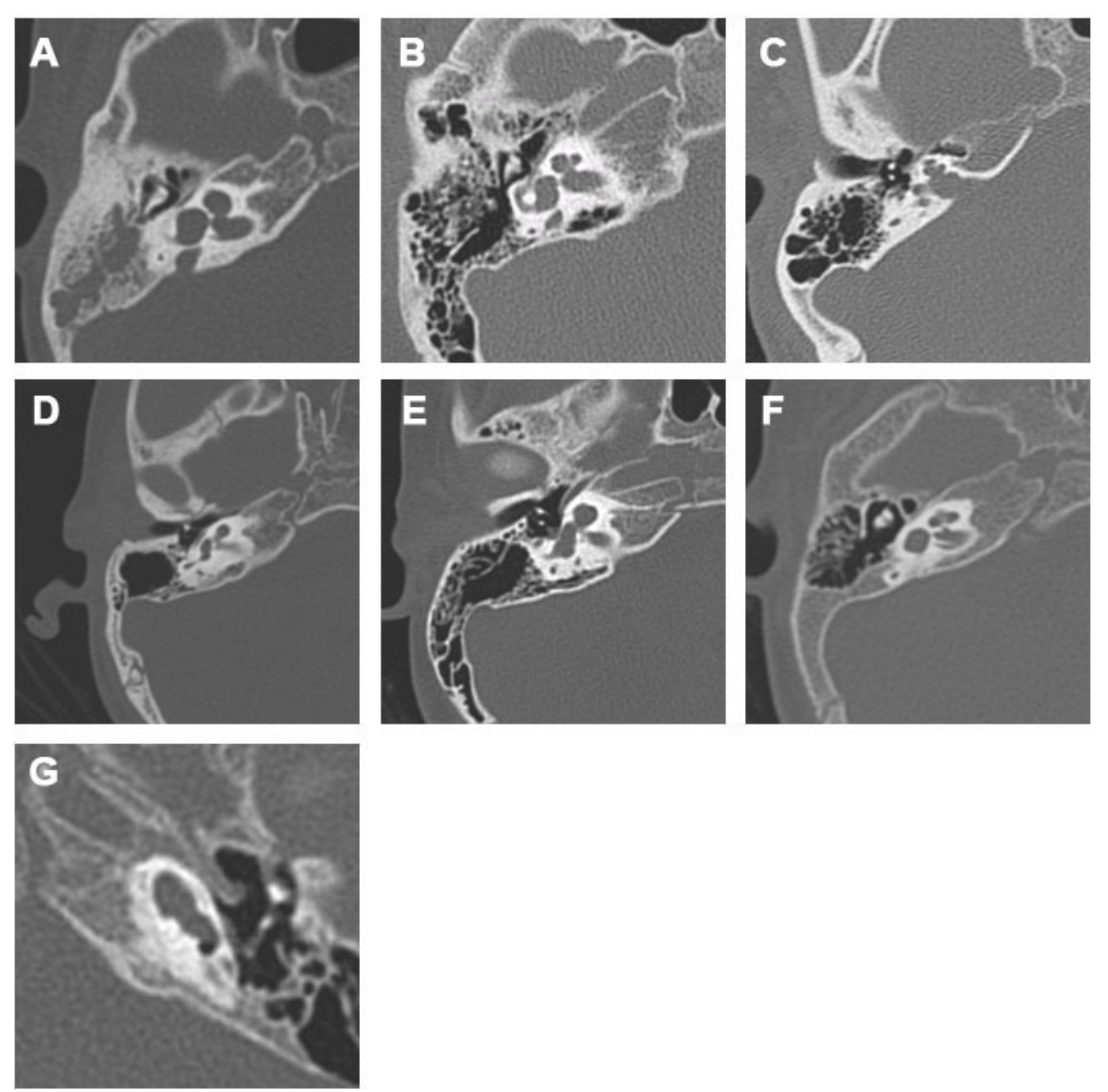
Criteria for Cochlear Implantation in Children

- Severe to profound SNHL
- Limited benefit from hearing aids
- No active middle ear pathology
- Normal eighth nerve and present cochlea

Factors that Delay Implantation and Outcome

- Auditory
  - Delay in diagnosis
  - Significant residual hearing
  - Fluctuating hearing
  - Unreliable or conflicting test results
  - ANSD
  - Underfit amplification
- Speech development
  - Good progress despite profound HL
- Parental issues
  - Missed appointments
  - Don't wear devices
  - No educational buy-in
  - Socioeconomic
- Medical
  - Anatomic uncertainty
    - CN deficiency
    - Severe inner ear malformation
  - Multiple Challenges
    - Cerebral palsy
    - Autism
    - Other

Inner Ear Malformations

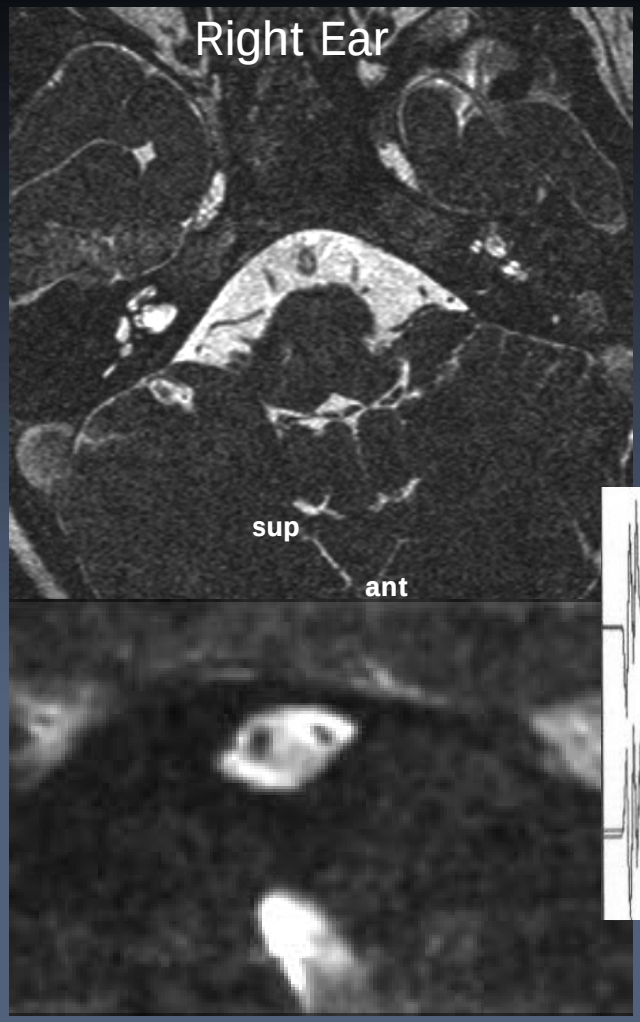


Cochlear Nerve “Aplasia”

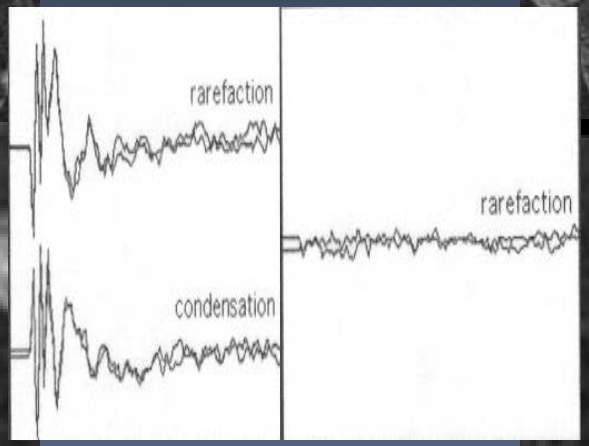
Jackler & Luxford, *Laryngoscope* (1987)

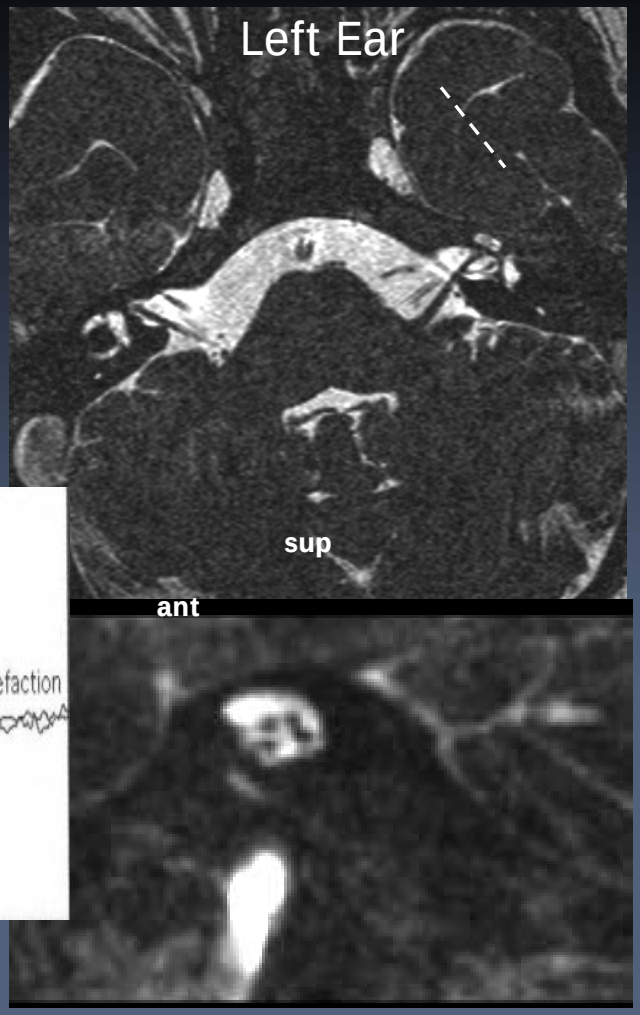
Shelton et al, *Otolaryngol Head Neck Surg* (1989)

MRI Evidence of Cochlear Nerve Deficiency



ABR



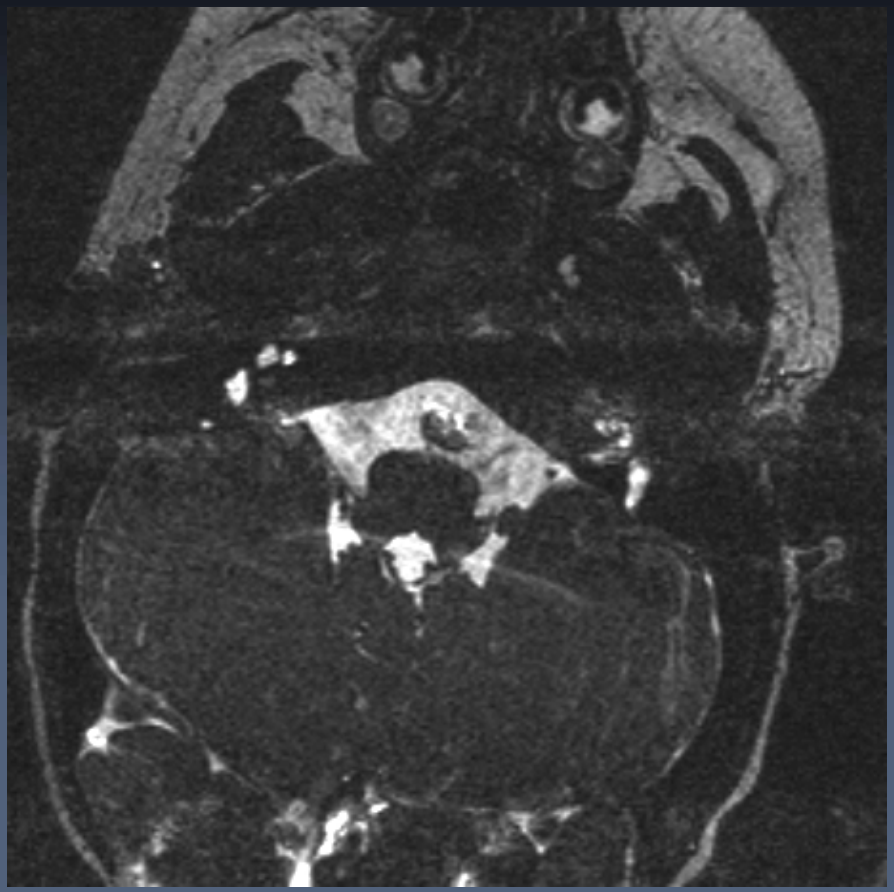


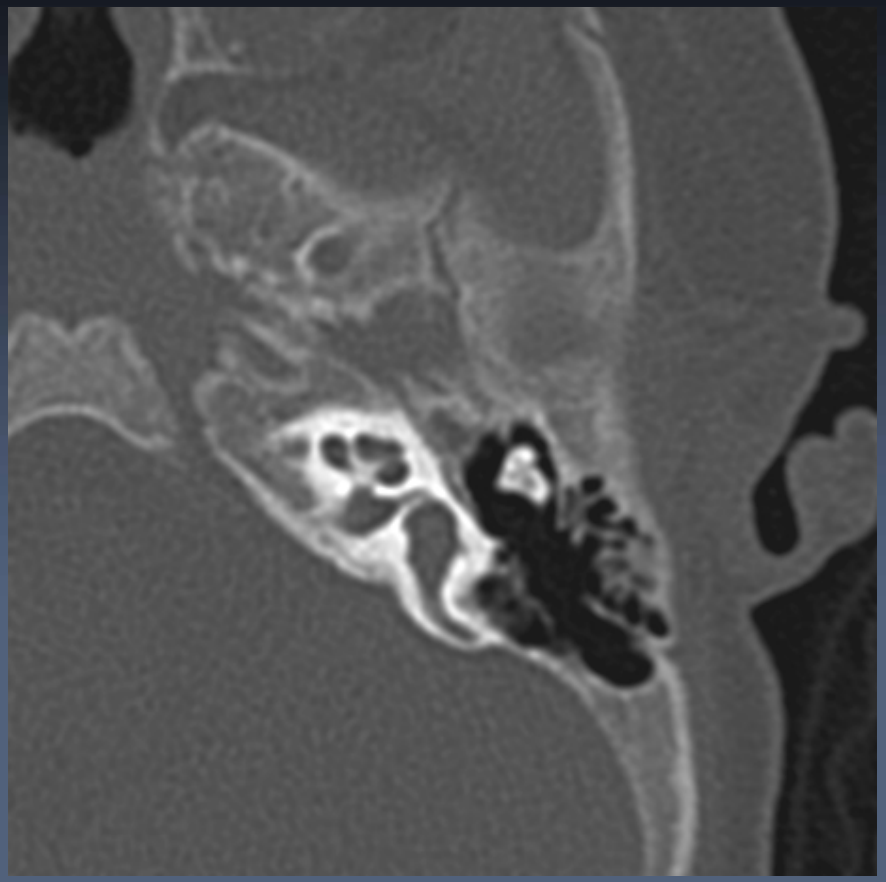
Adunka et al 2006, 2007, Buchman et al 2006

Cochlear Nerve Deficiency

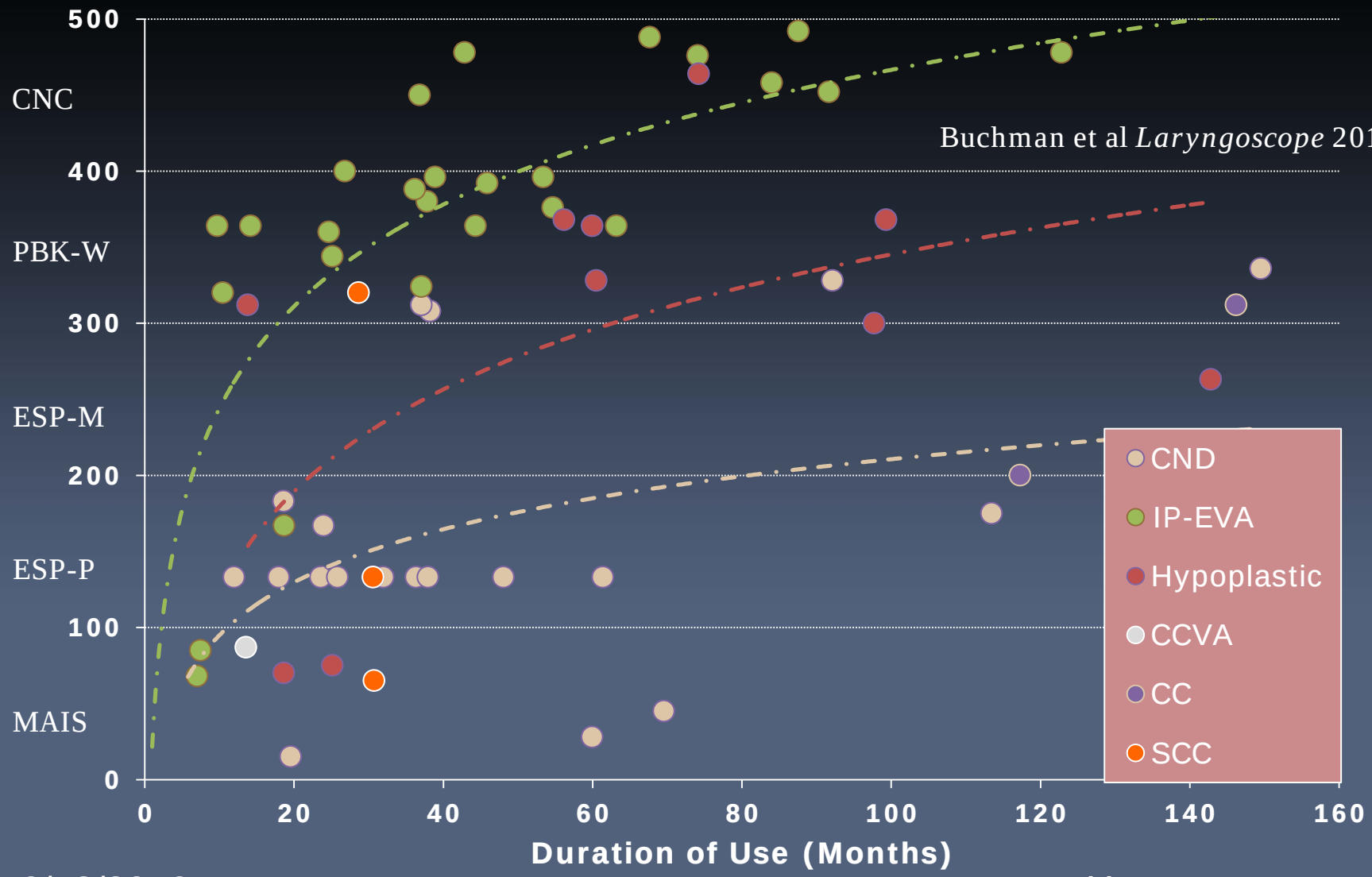
Nelson & Hinojosa, *Otology & Neurotology* (2001)

Cochlear Implant or Not?





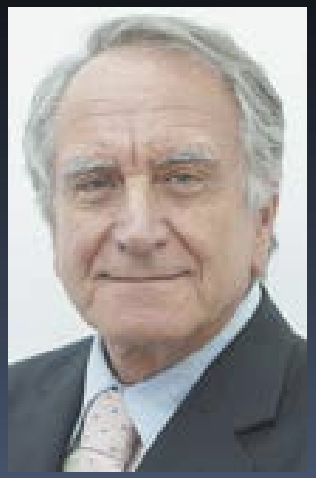
Speech Perception (SRI-Q) by Malformation



Auditory Brainstem Implant (ABI)

Possible Indications

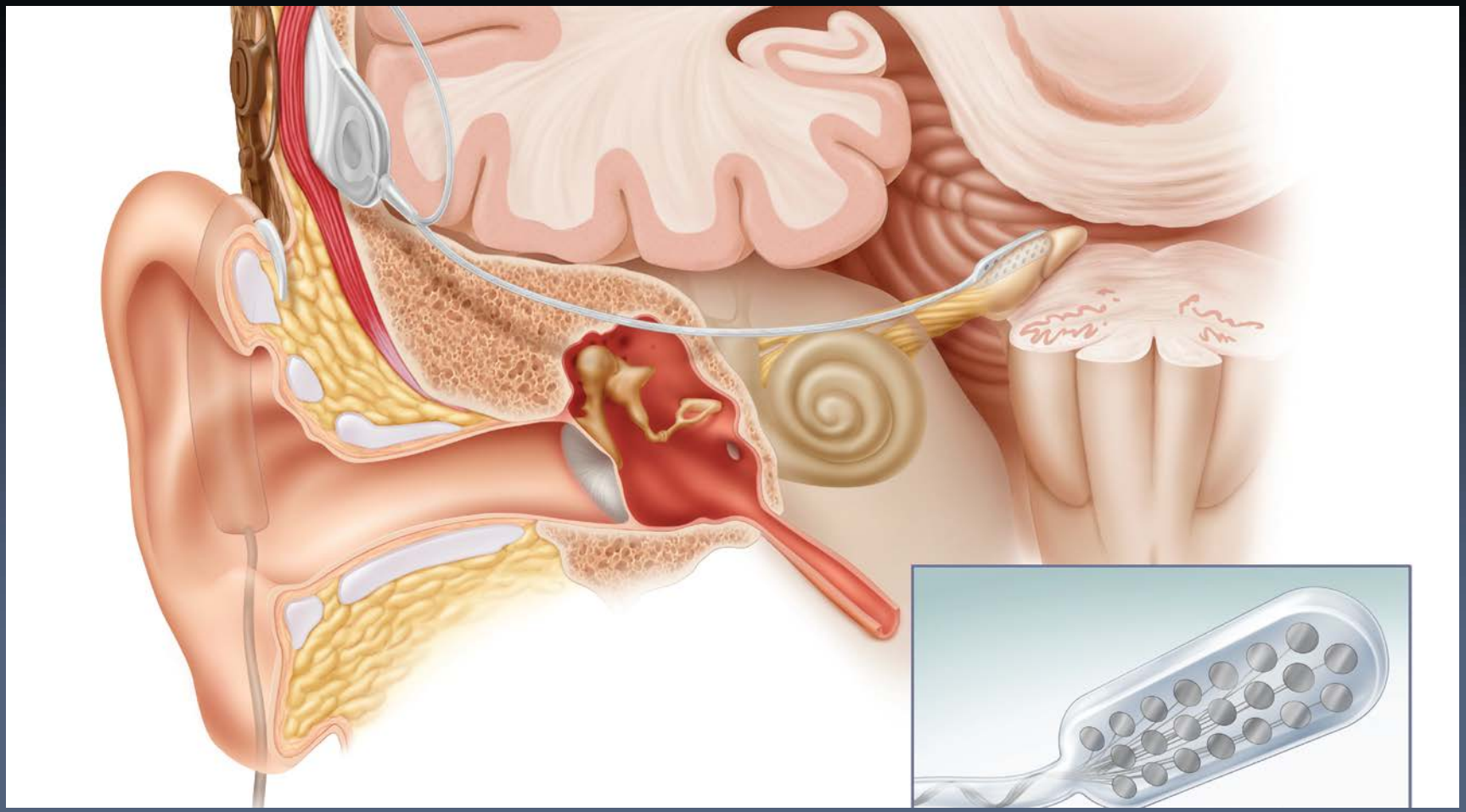
- Absent Cochlea or Cochlear Nerves
  - NF2
  - Congenital absence
  - Total ossification
  - Traumatic transection
- Unable to or failed benefit from CI
  - Severe malformations, progressive ossification, other?
- Committed Parents or Patients
- Cognitively normal or near normal



Vittorio Colletti

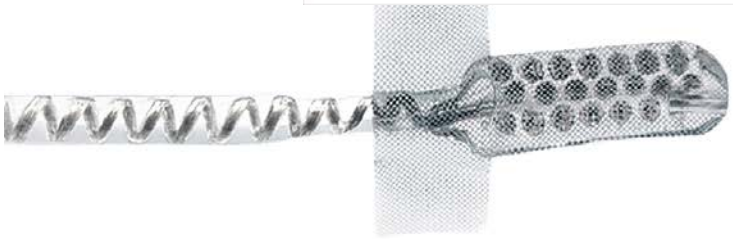
Levent Sennaroglu

Auditory Brainstem Implant



ABI Device

- Developed by William House
- Similar to Cochlear Implant
- Foramen of Luschka
- Removable Magnet



UNC Pediatric ABI Feasibility Study

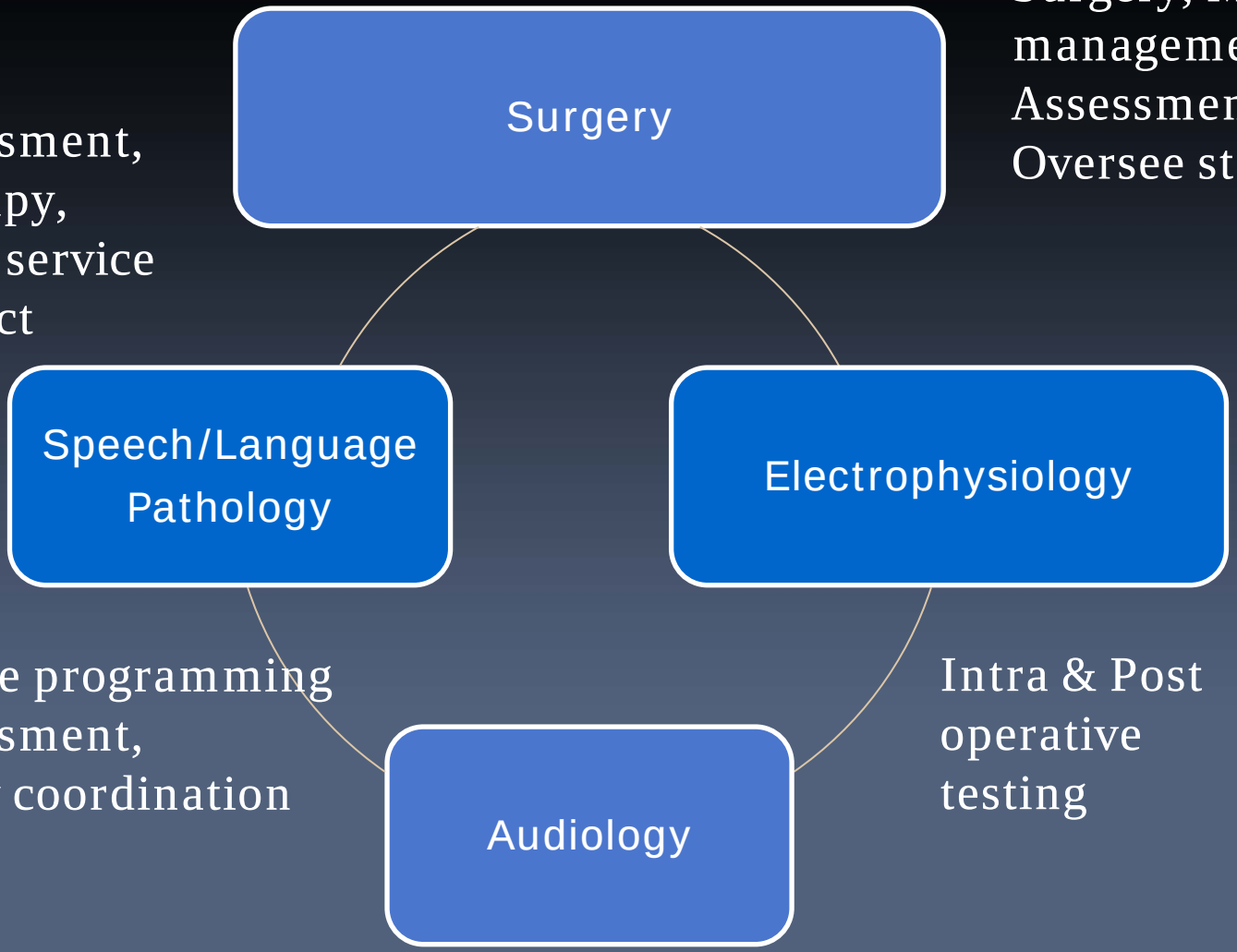
- Safety and efficacy of the Nucleus 24 Multichannel ABI:
  - to demonstrate safety of the surgical procedure
  - tolerance of device stimulation, and
  - the potential for auditory benefit beyond that experienced with their CI
- May provide the preliminary experience for a larger scale clinical trial
- Requires a team approach among surgeons, audiologists, speech/language pathologists & electrophysiologists and families
- Investigational Device Exemption from the FDA
- Institutional Review Board Approval at UNC-CH

UNC Pediatric ABI Feasibility Study

Candidates

- Subjects
  - 10 pre-linguistic young children (18 mos to 5 yrs. of age)
  - 5 post-linguistic children (<18 yrs of age)
- Failed CI OR unable to receive a CI
- No developmental/cognitive delays that would impede progress
- Appropriately motivated family

ABI Team



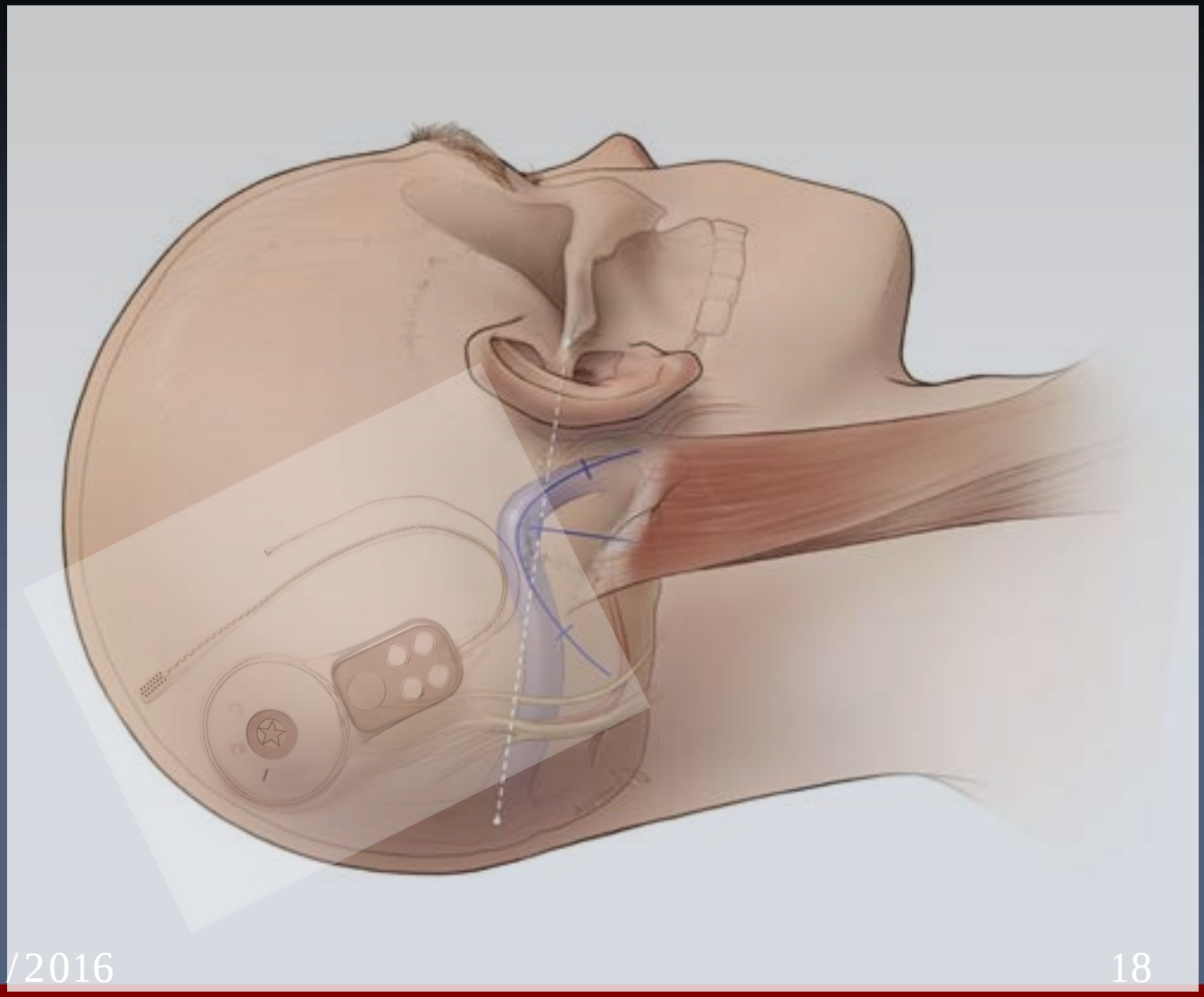
Surgery, Medical management, Assessment, Oversee study

Assessment, Therapy, Local service contact

Device programming Assessment, Study coordination

Intra & Post operative testing

Retrosigmoid Craniotomy



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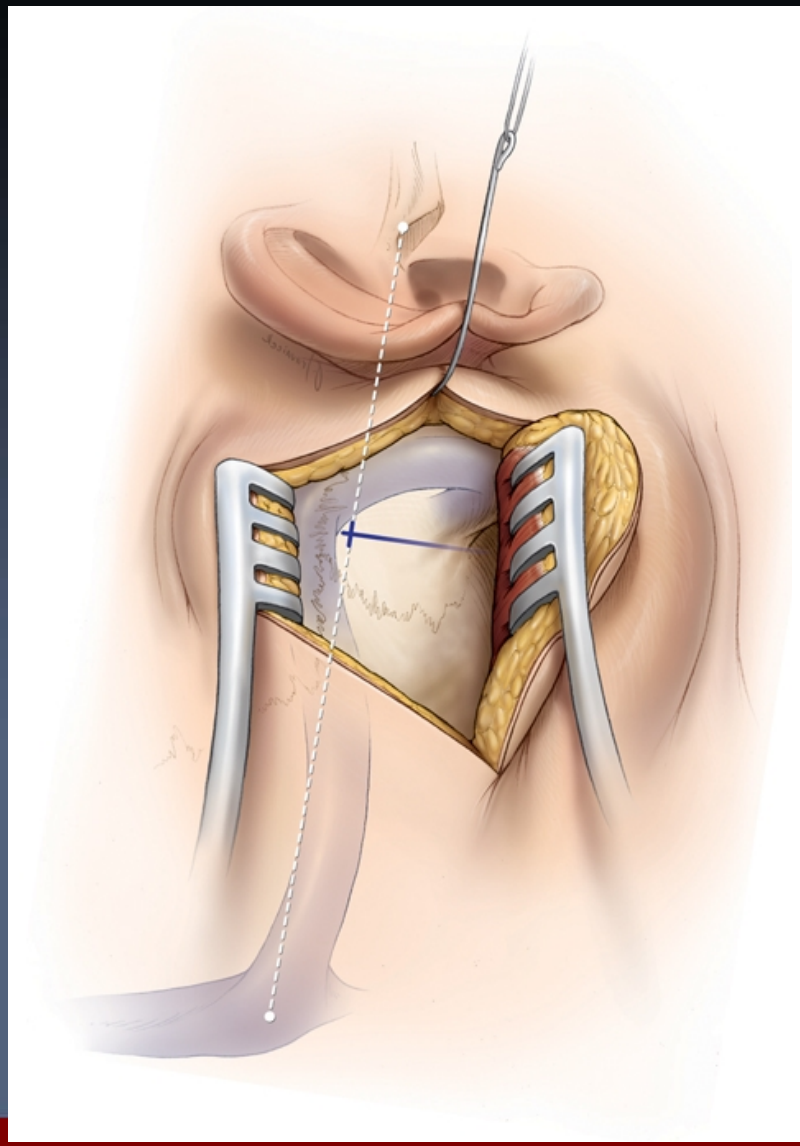

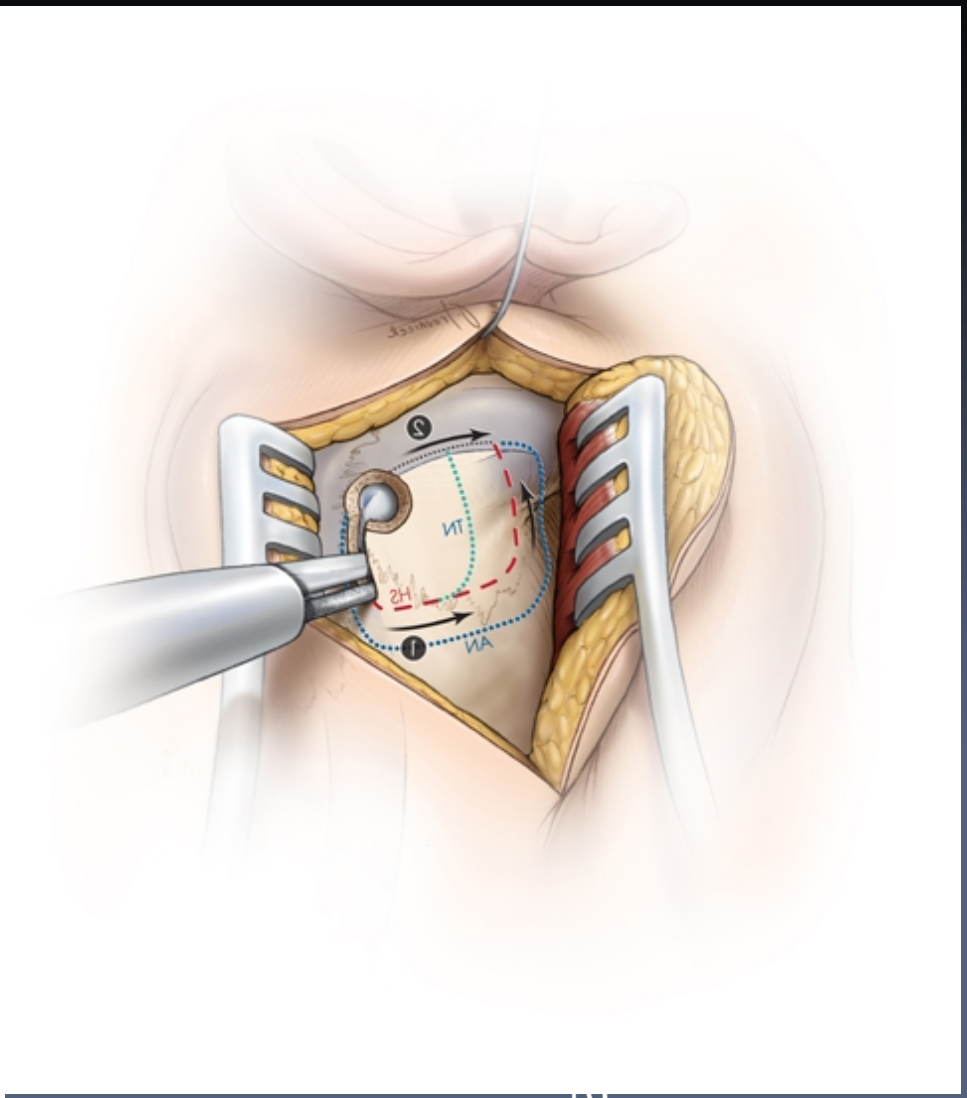

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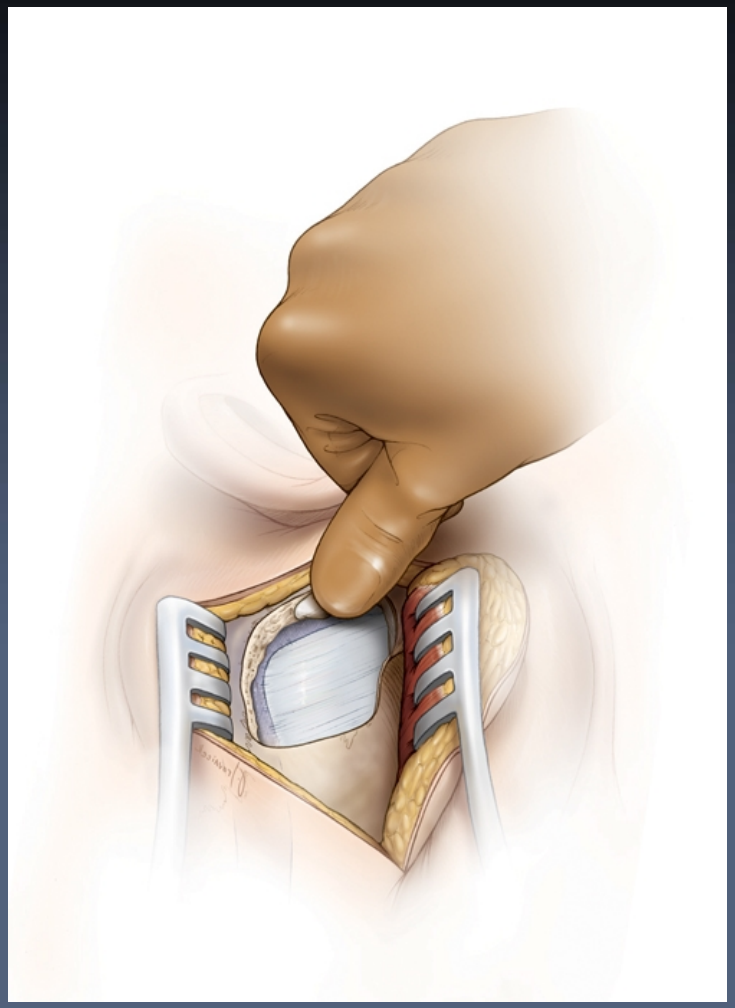
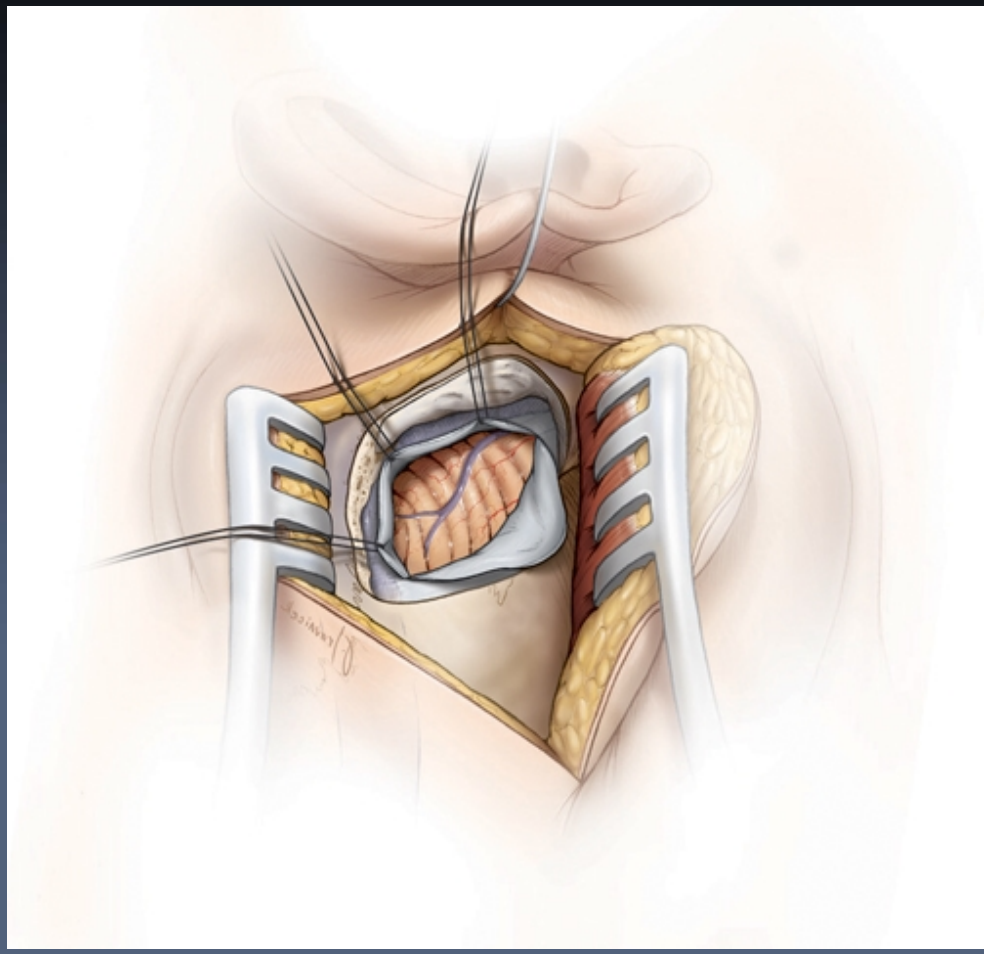
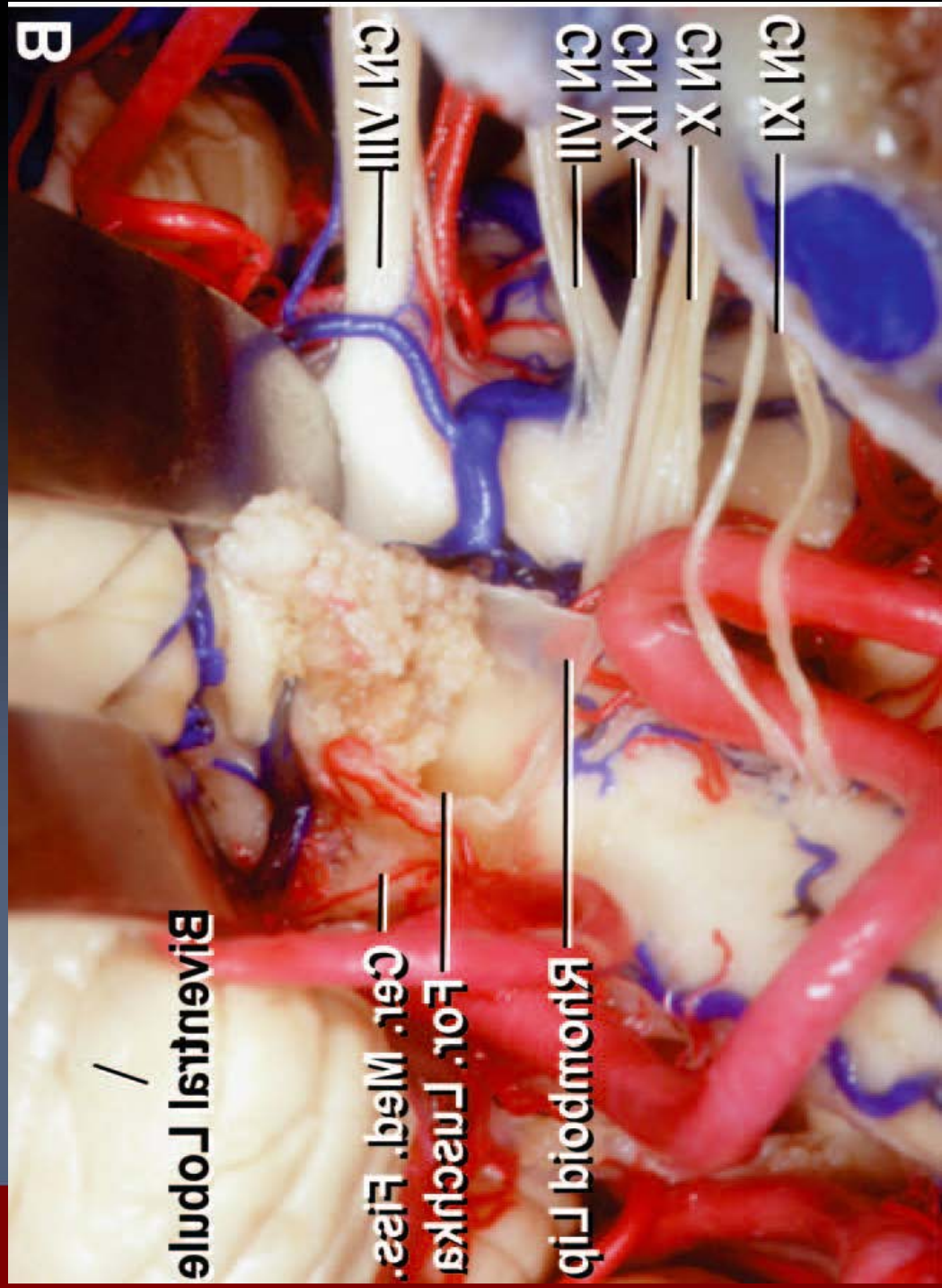




Retrosigmoid View

- Intraop maneuvers:
  - Cranial opening to sinus
  - Drain cisterna magna
  - Retract cerebellum
  - Widely open arachnoid
  - Identify lower CN (9-11)
  - Identify (7, 8)
  - Move vessels
  - Retract choroid plexus
  - Retract flocculus
  - Enter Foramen of Luschka



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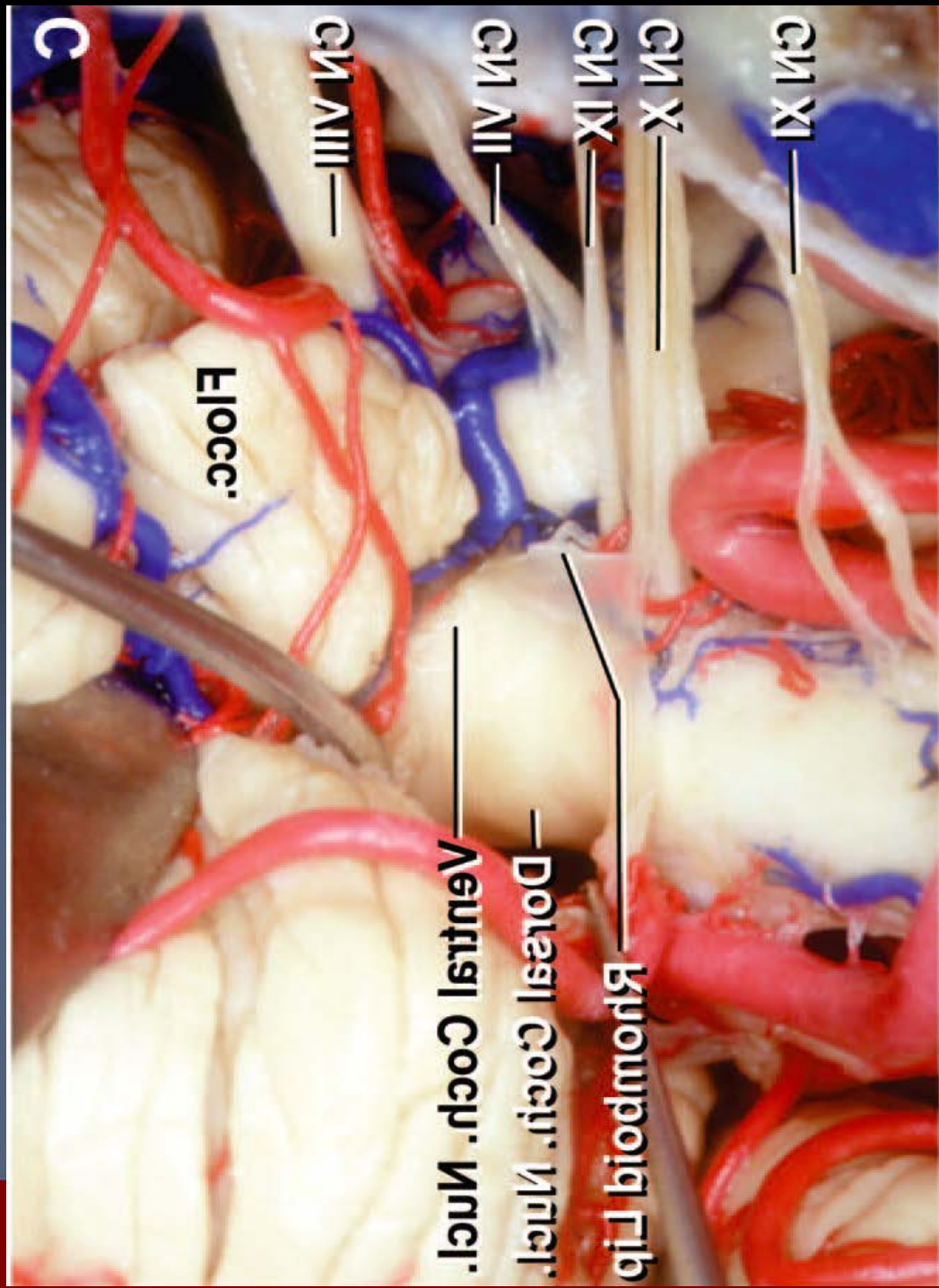
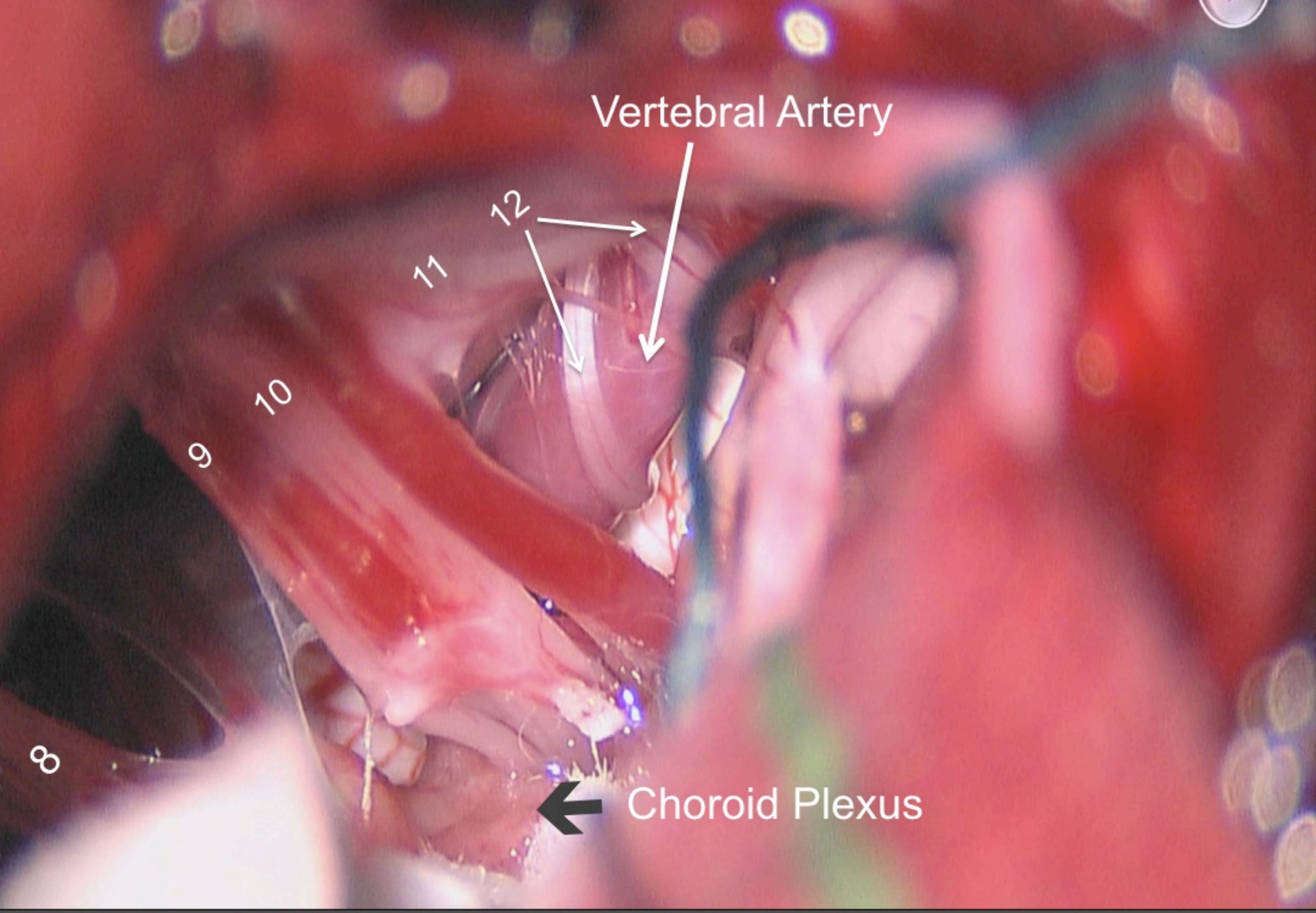
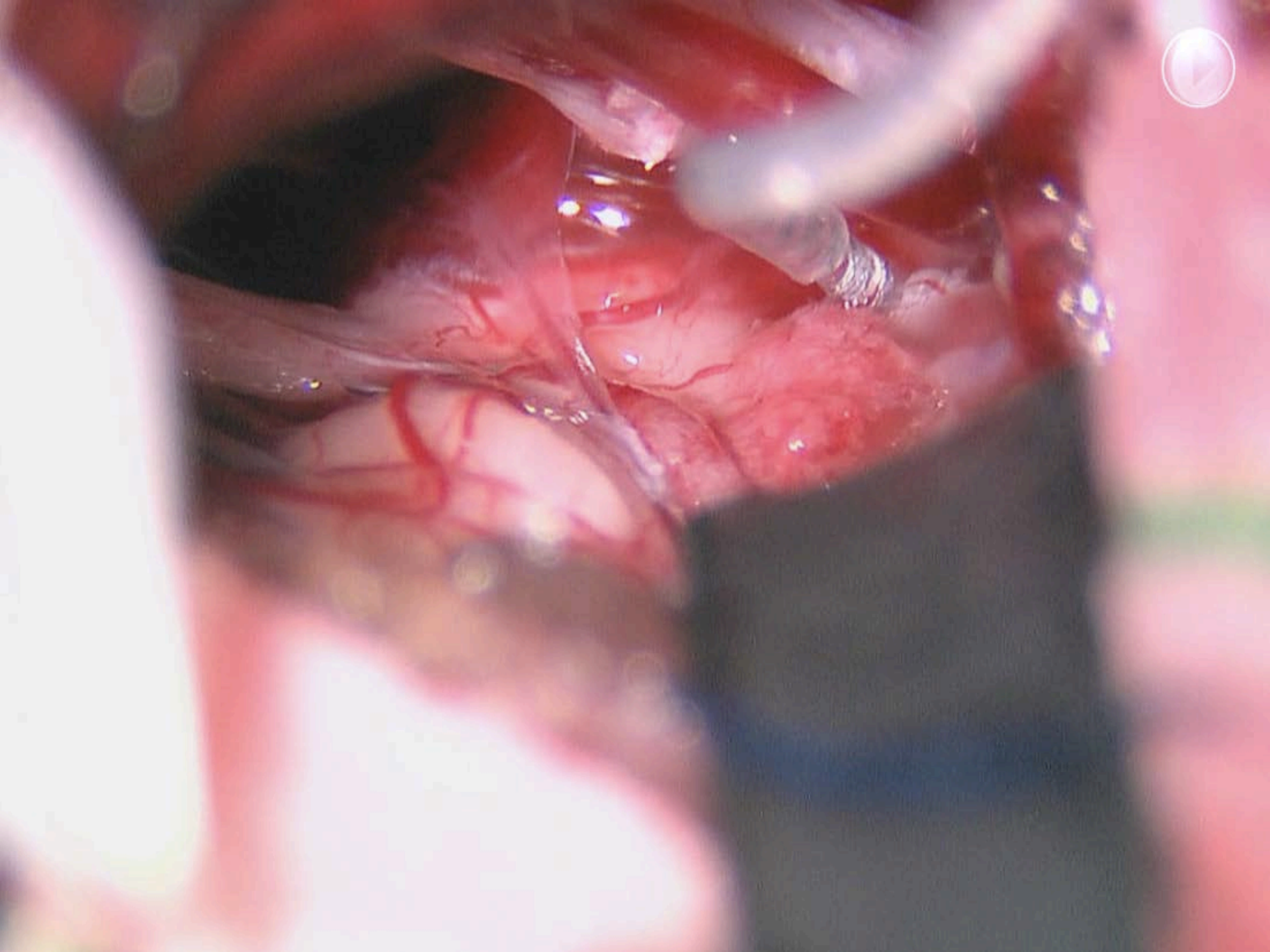


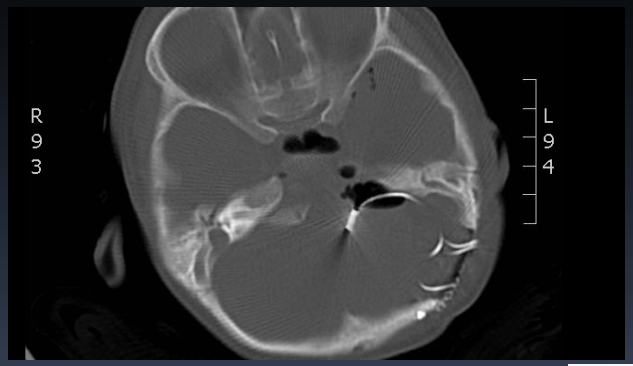
Protocol

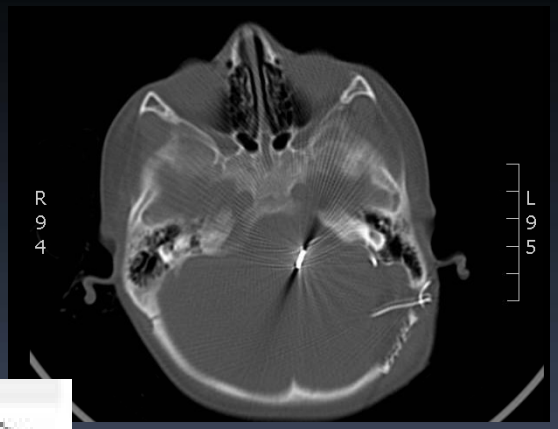
- Retrosigmoid Craniotomy
  - Nucleus 24 ABI (Cochlear Corp)
  - Monitor CN 7, 9, 10, 11
  - Implant evoked ABR
- Postop CT
- Pediatric ICU
- OR Stim prior to activate
- Activation under monitoring
- Ongoing device programming
- Speech perception/Speech & Language Assessments similar to cochlear implantation

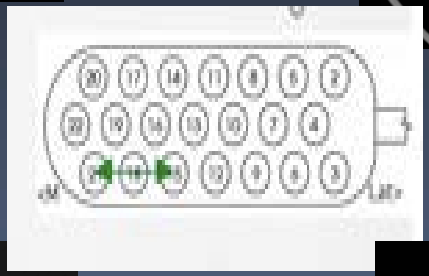

Vertebral Artery
12
11
10
9
8
Choroid Plexus

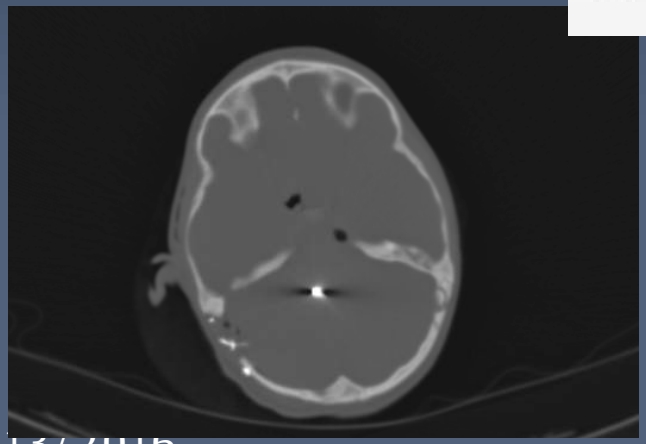


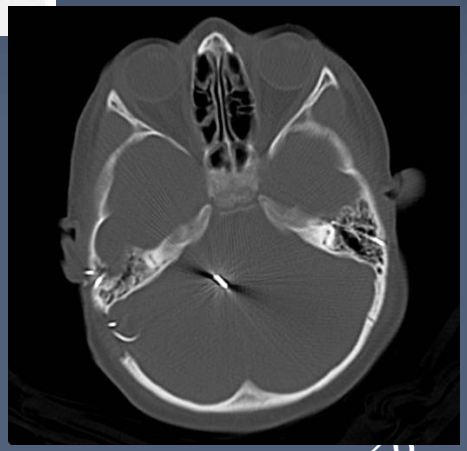
Postoperative CT Scans



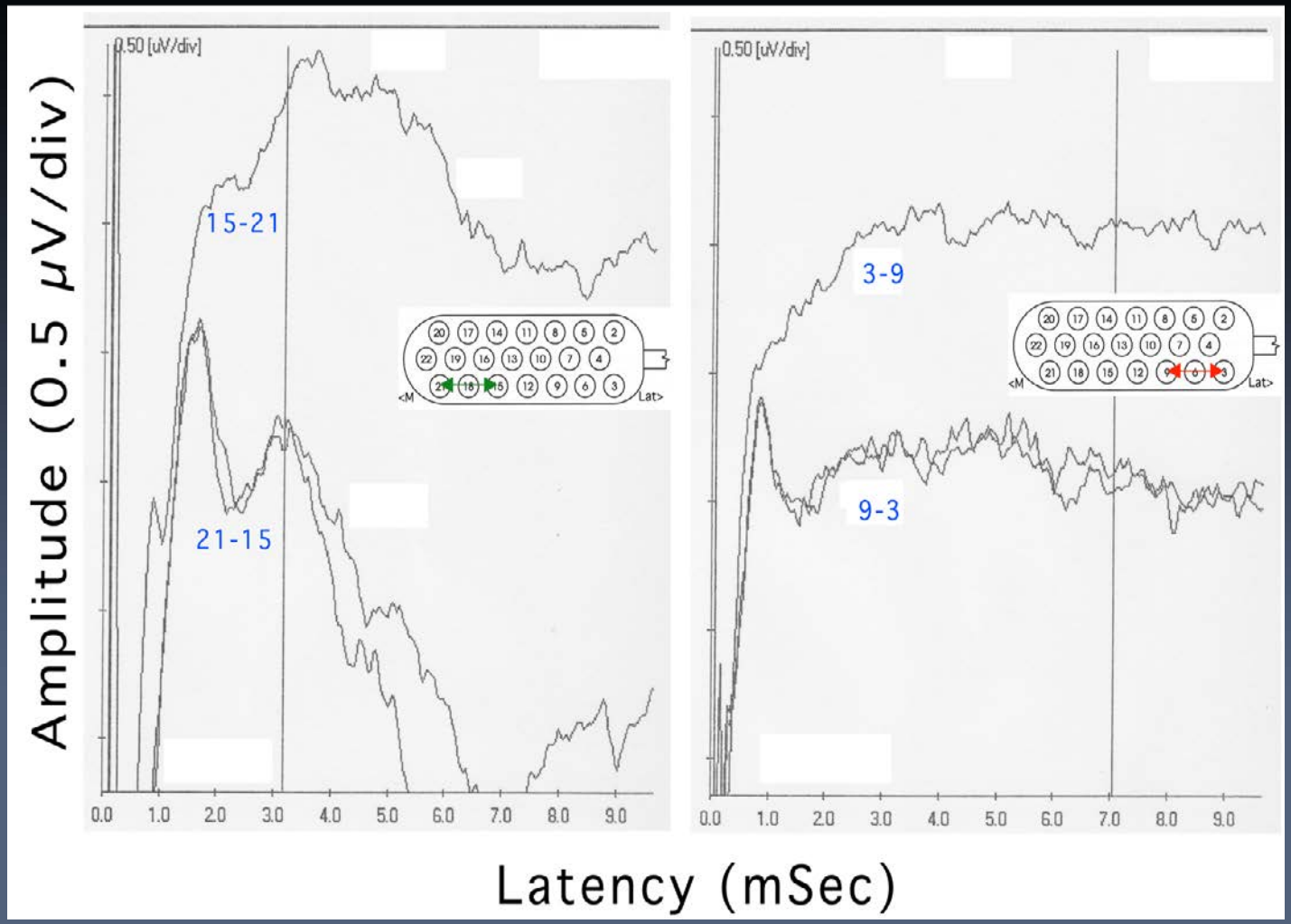








Intraop Electrically-Evoked ABR



Audiology: Device Programming

Goal is similar to CI patient programming

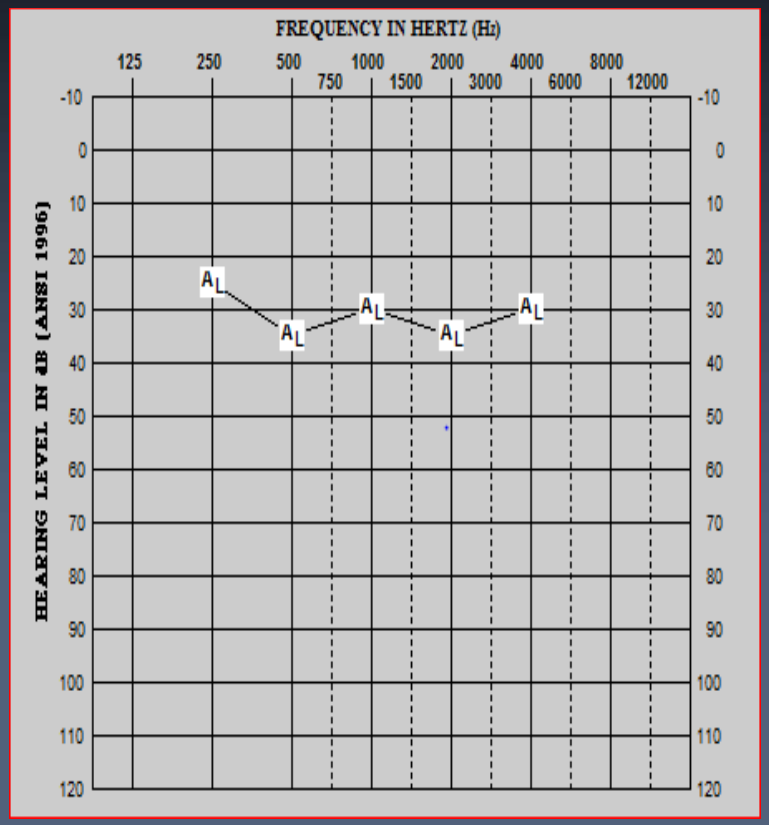
- Establish electrical threshold (T) and comfort (C) levels
- Allocation based on array design
  - Creating the auditory template that will develop with time
- Avoid stimulation of other cranial nerves (initial stim in OR)
  - Facial twitching,
  - Balance disturbance (head & trunk tilt),
  - Coughing, choking, sensation in mouth, throat, tongue, palate,
  - Heart rate changes

Demographics

	UNC1	UNC2	UNC3	UNC4	UNC5
Previous/Current CI	Yes/No	No/No	No/No	Yes/Yes	Yes/Yes
Age at ABI	3.3	2.5	3.5	5.5	2.1
Gender	M	F	M	F	F
Side	L	L	R	R	R
Etiology/Syndrome	CND/ CHARGE	CND-Michel	CND/ CHARGE	CND	CND

Case 1

- Dx at 11 mos with HL
- CHARGE Syndrome
- Previous CI use
- Cued Speech



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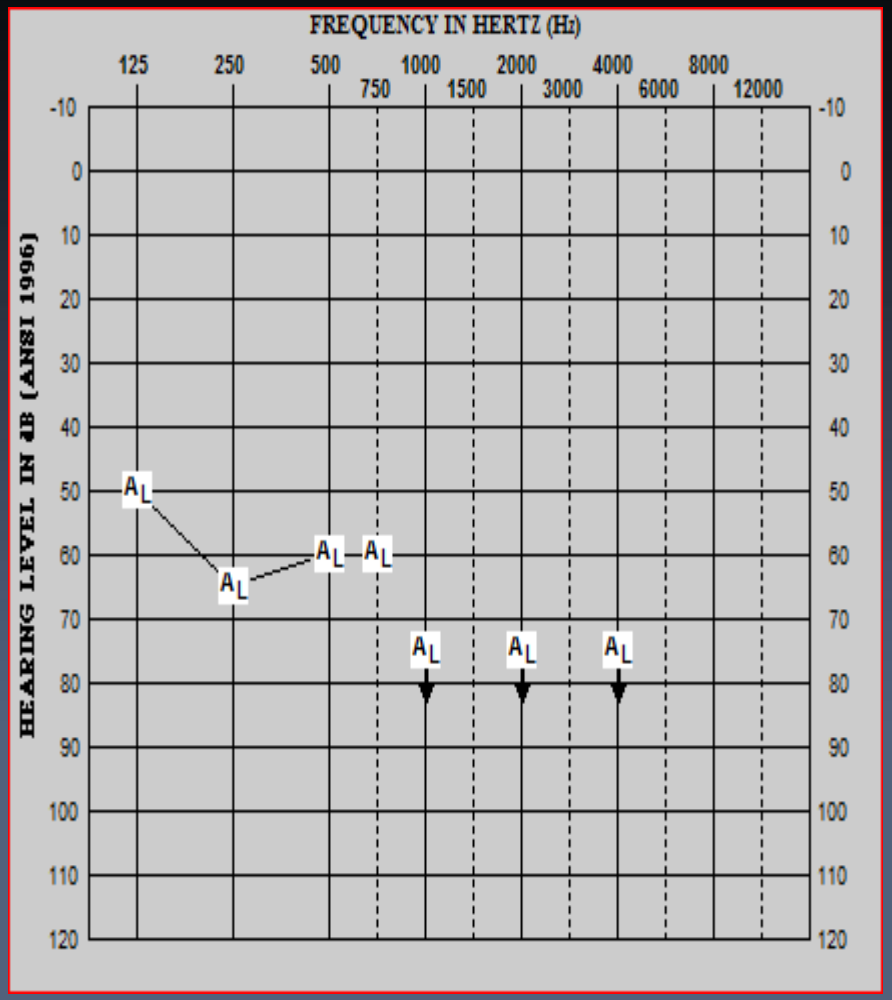
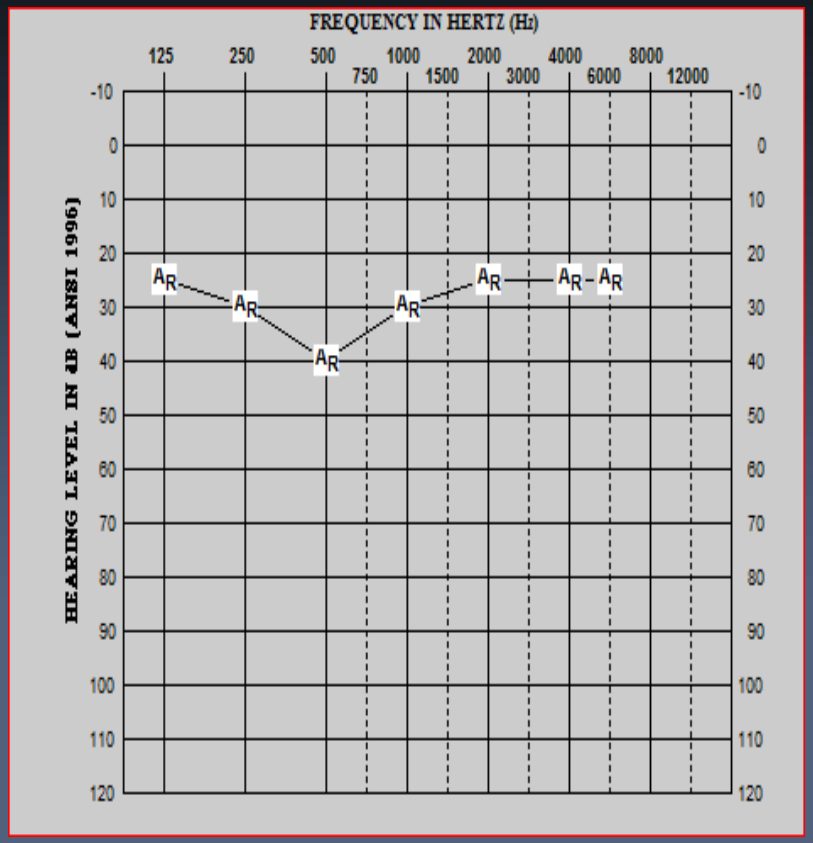
Case 2

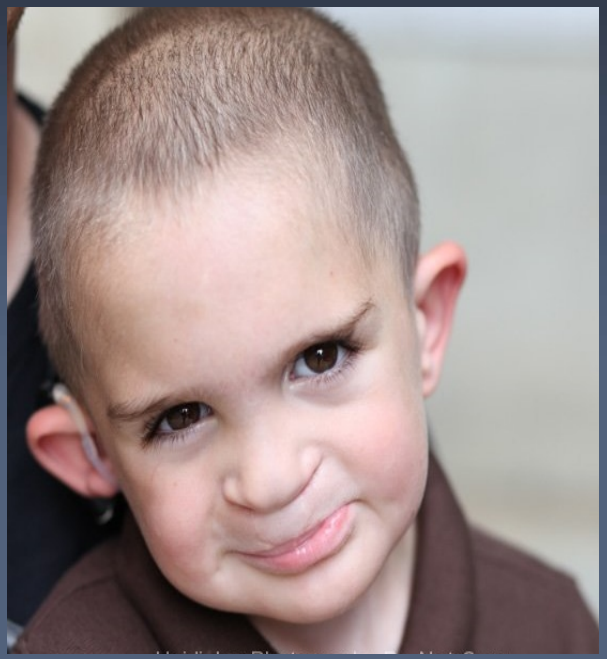


- Identified at birth
- Profound HL
- Absent cochlea and cochlear nerve
- No previous CI
- SEE

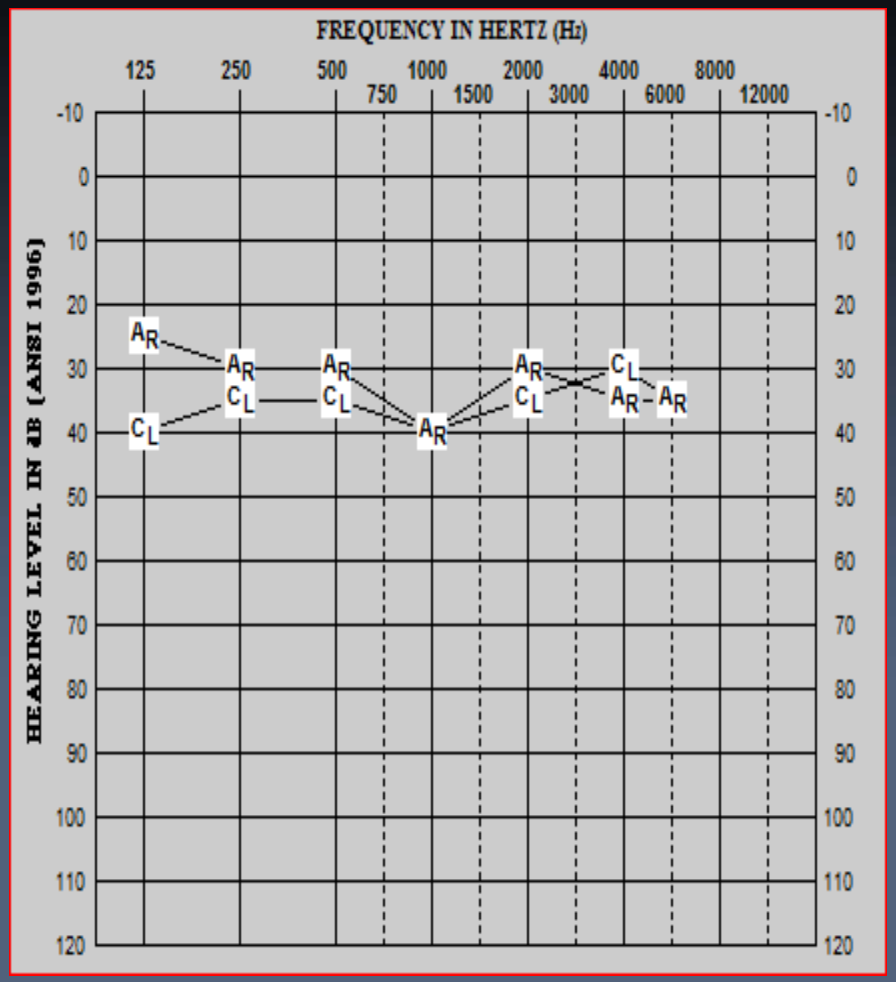
Case 3



- CHARGE syndrome
- Cleft lip & palate (repaired)
- No previous CI
- Total Communication / SEE

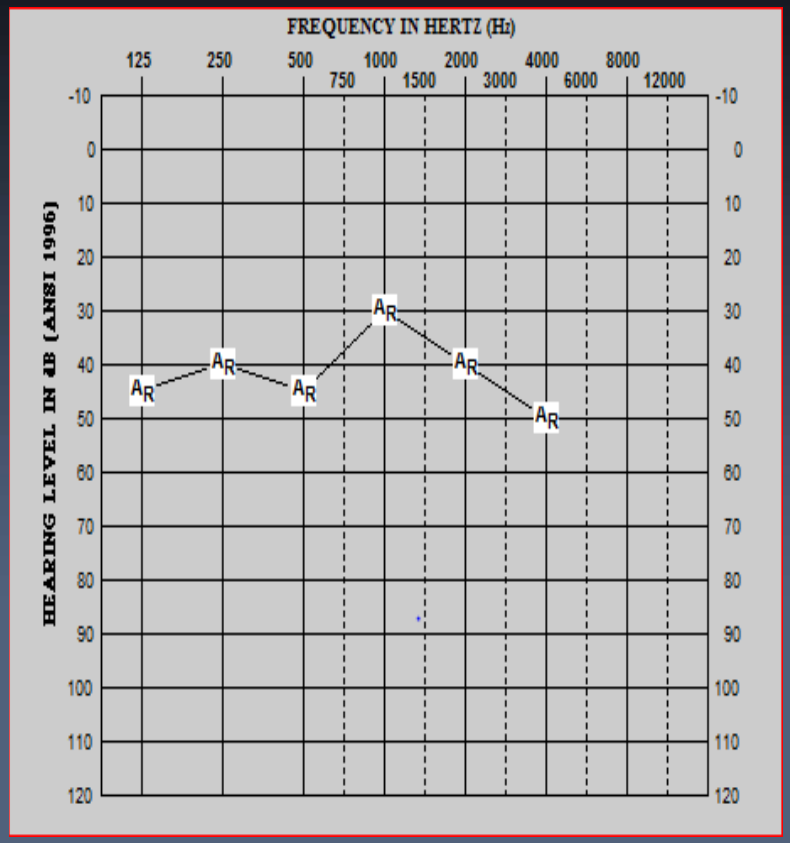


Case 4



- CIs at 10 & 13 months
- Received sound detection with Cis
- Total Communication / SEE

Case 5



- **CI at 15 mos – no sound awareness**
- **ASL**

Surgery Results

Auditory

Listening Skills and Speech Production

	UNC1	UNC2	UNC3	UNC4	UNC5
Duration of Use	3	2.5	2	18	18
Communication mode/Speech Production	Cued Speech 92% Word Patterns 53% Vowels	SEE 55%Word Patterns 29% Vowels	TC CNT	TC 48% Word Patterns 17% Vowels	ASL CNT
OWLS **Composite (85-115)**	71	71	77	44	70
Learning to Listen	100%	0% chance?	100%	47%	0%
Song Identification	100%	50%	80%	75%	0%
Phrase Identification	100%	5%	75%		0%

Speech Skills-1 year post ABI

100% vocalize

80% have increased their quality voice for pitch and duration

80% use initial sounds to imitate words /b, m/

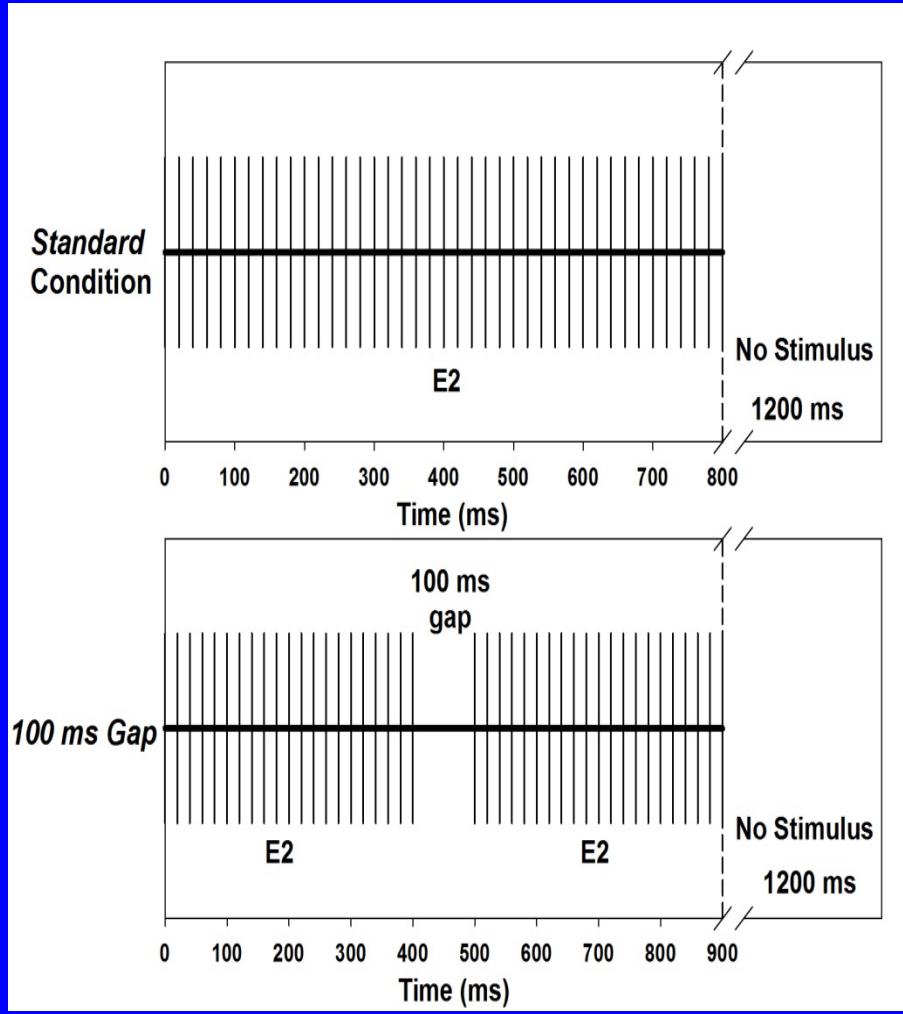
60% use vowel variety

Electrophysiology (Shuman He)

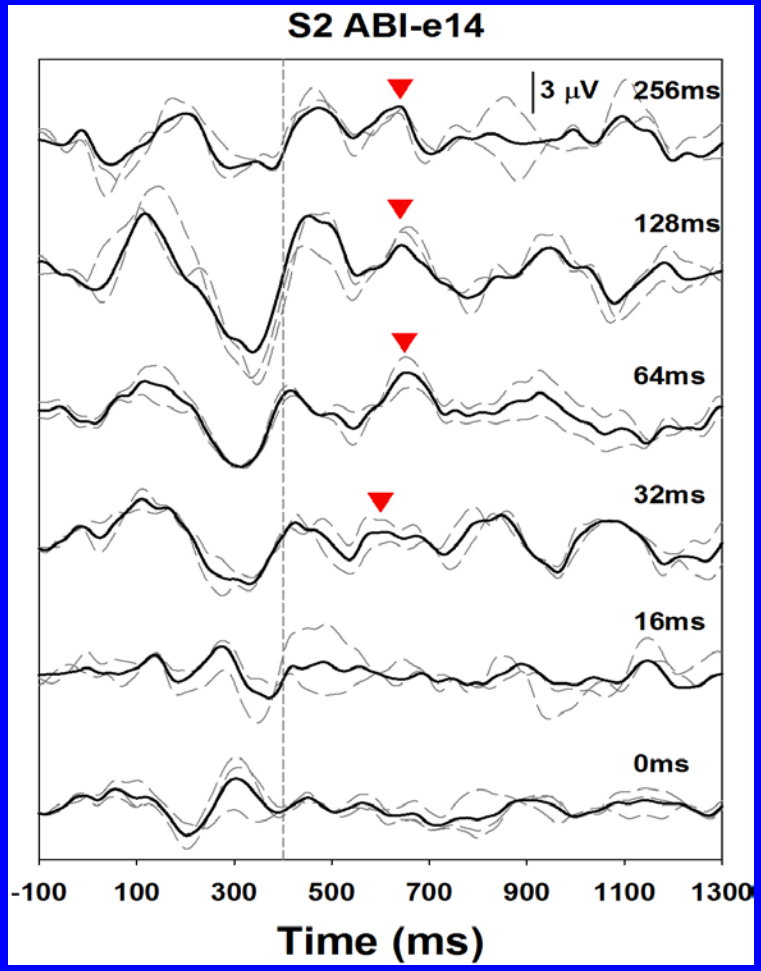
- Cortical Potentials
  - Threshold Detection
  - Gap and Electrode Change

Electrophysiology

- *Objective Measures:*
  - *Standard condition*: 800-ms biphasic pulse train.
  - *Gap condition*: Two 400-ms stimulus bursts separated by a silent interval (i.e. gap).



Area of Investigation



Conclusions

- ABI in Young Children is in very early stages in US
- Safe so far
  - CSF Leaks
  - Aseptic meningitis
- Early results
  - Sound detection in all
  - Limited speech perception –No open set in US
  - Speech Production—Some emerging—very delayed
- Objective Measures--