

Auditory Brainstem Implantation in Children

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SCHOOL OF MEDICINE

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 - 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, ...)

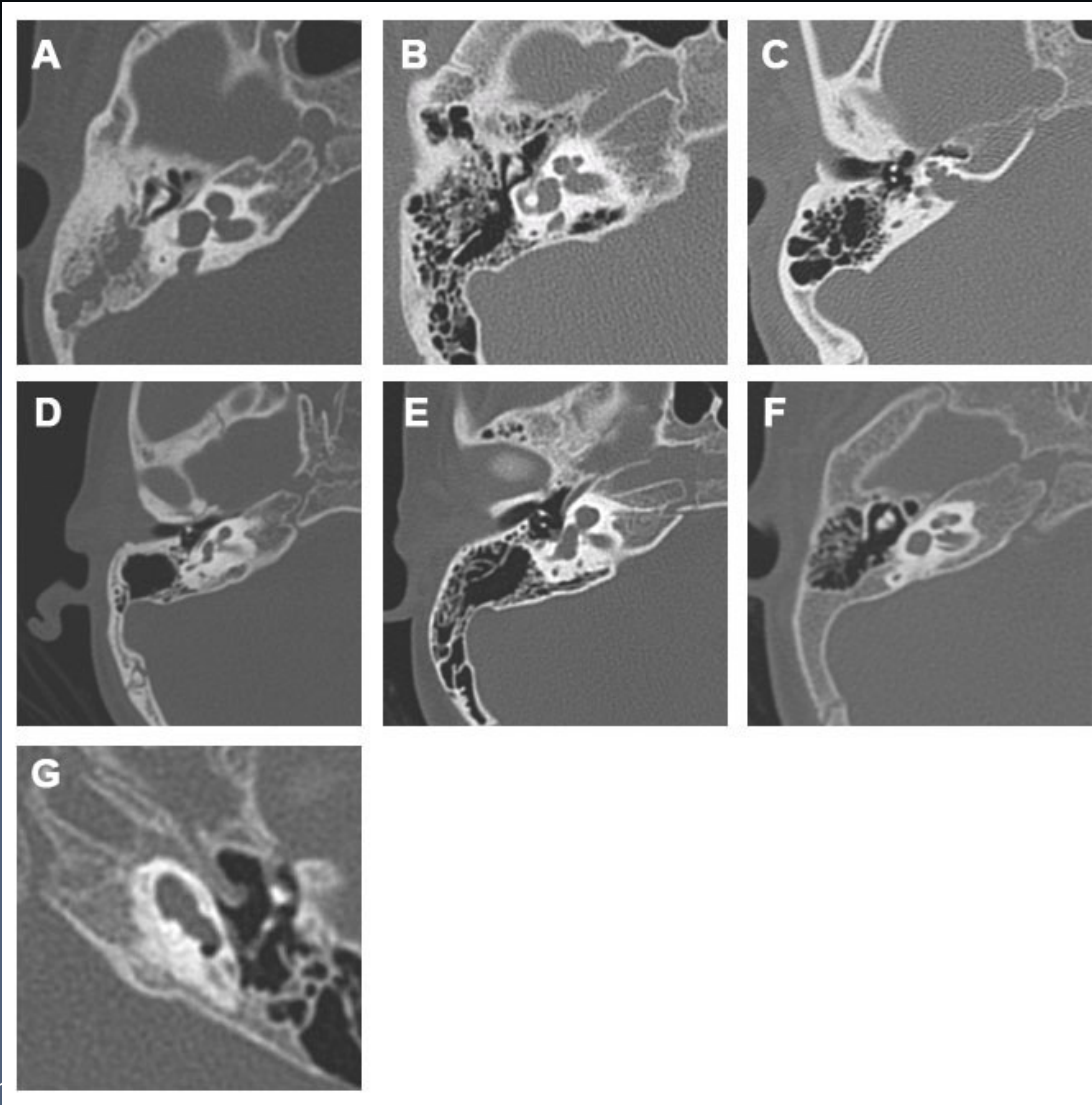
Criteria for Cochlear Implantation in Children

- 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

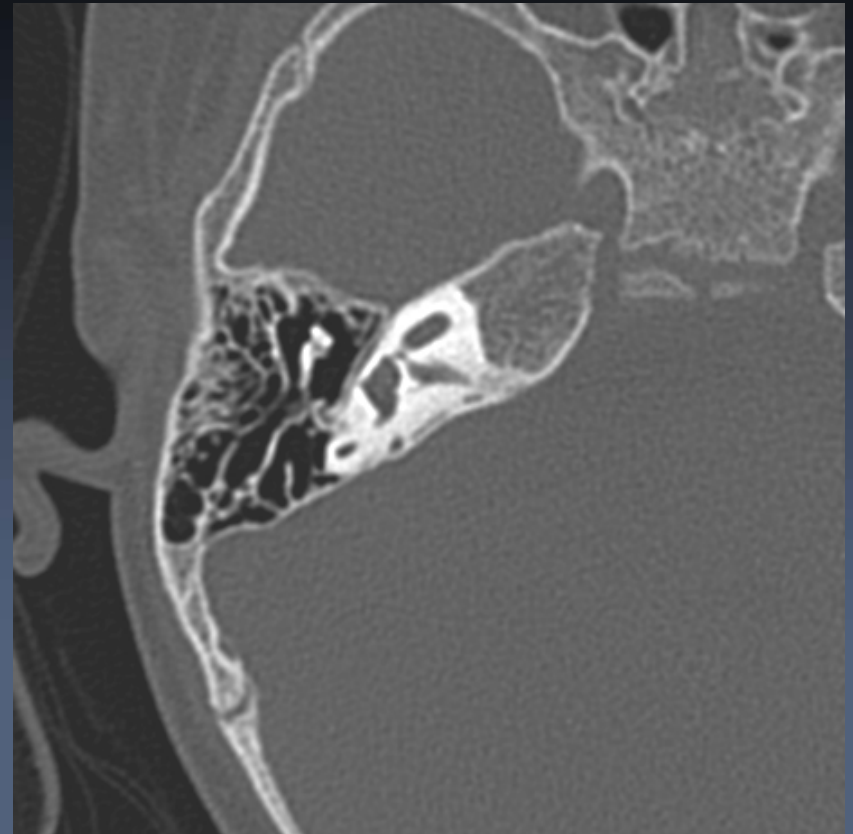
Inner Ear Malformations



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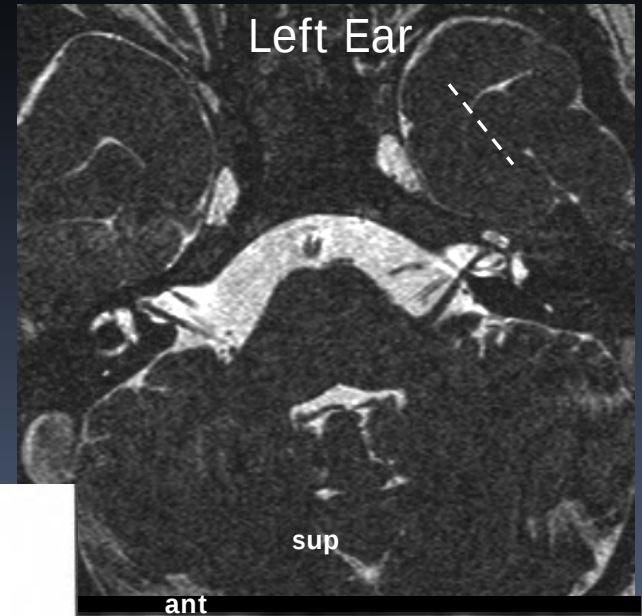
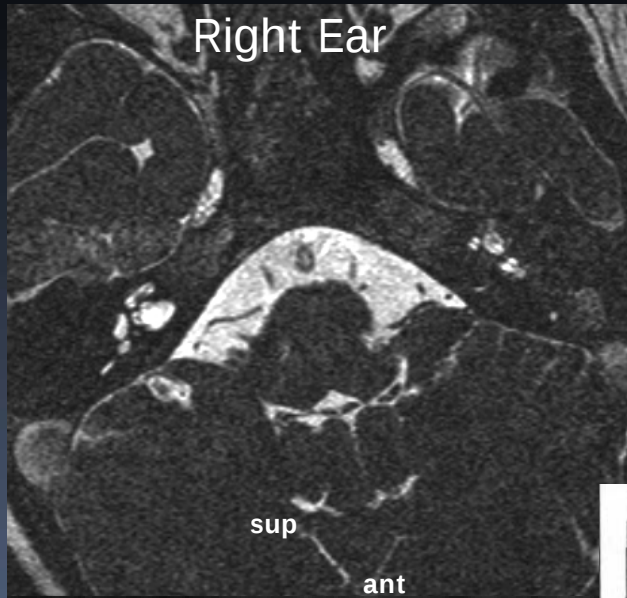
Cochlear Nerve “Aplasia”



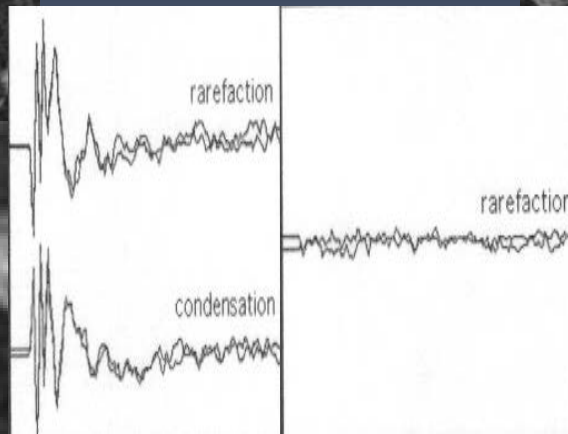
Jackler & Luxford, *Laryngoscope* (1987)

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

MRI Evidence of Cochlear Nerve Deficiency

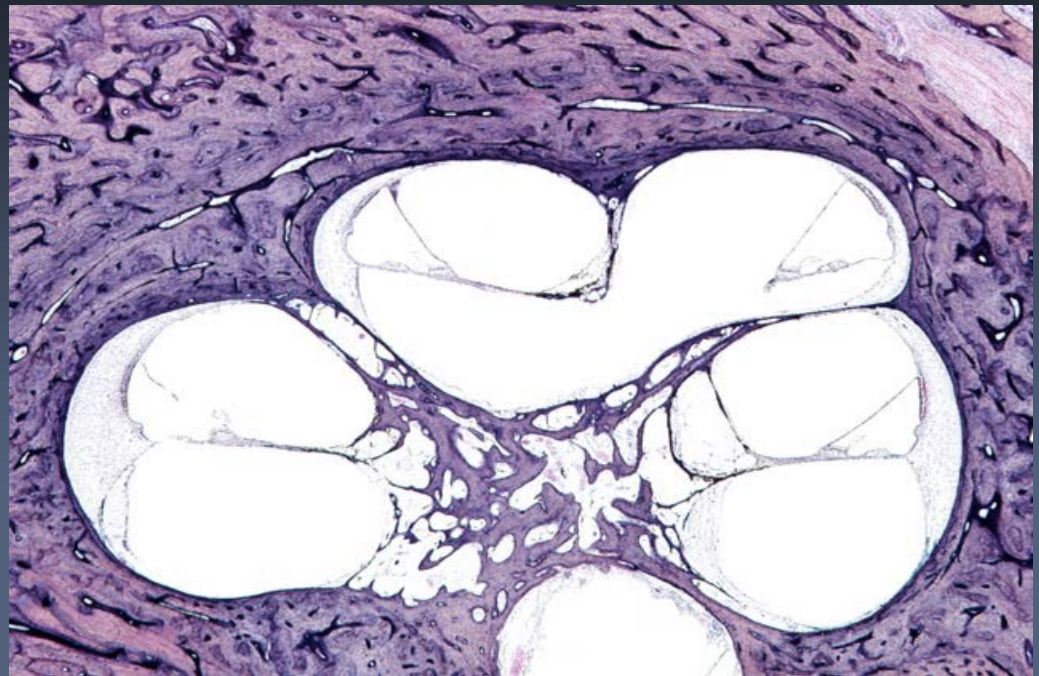


ABR



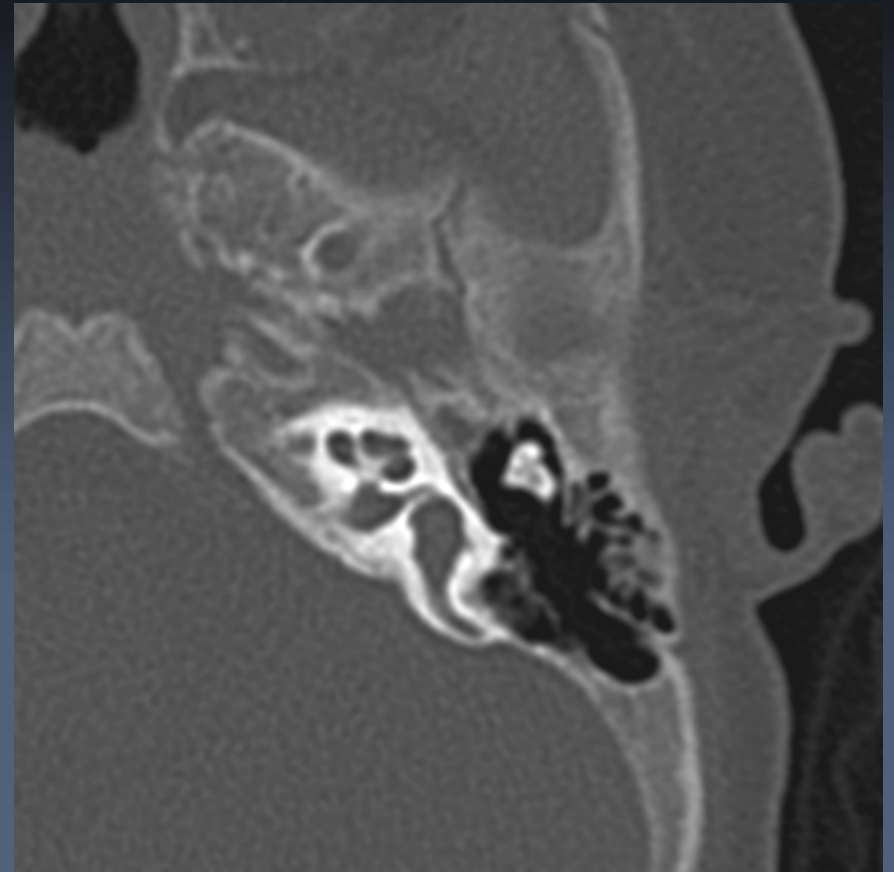
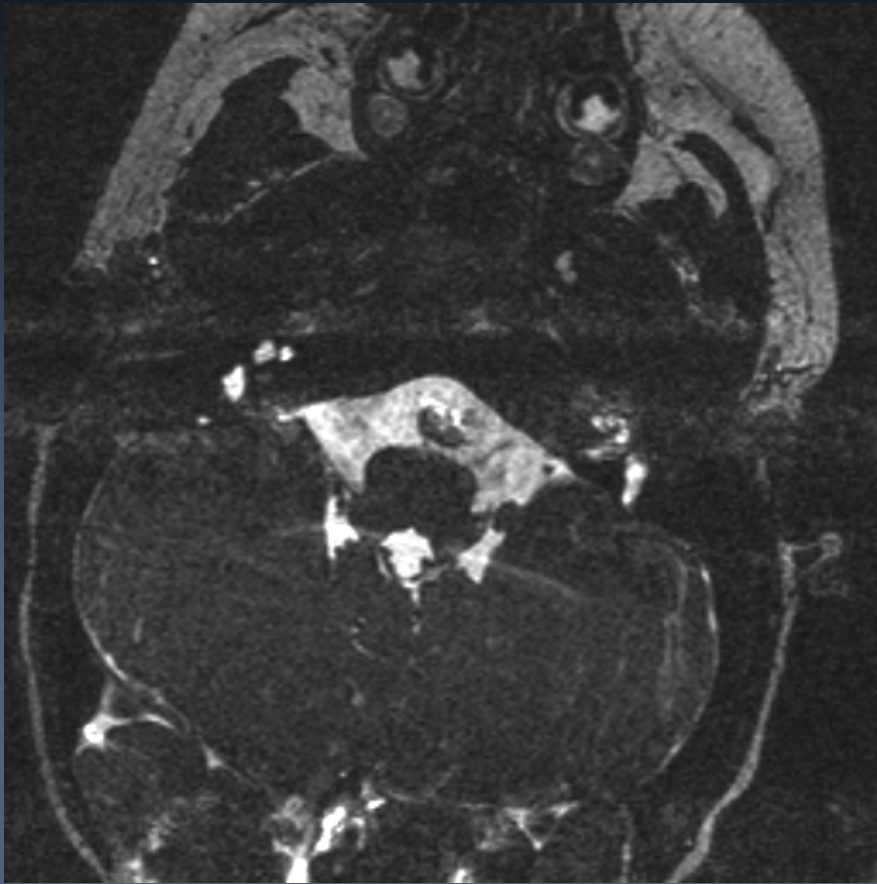
Adunka et al 2006, 2007, Buchman et al 2006

Cochlear Nerve Deficiency

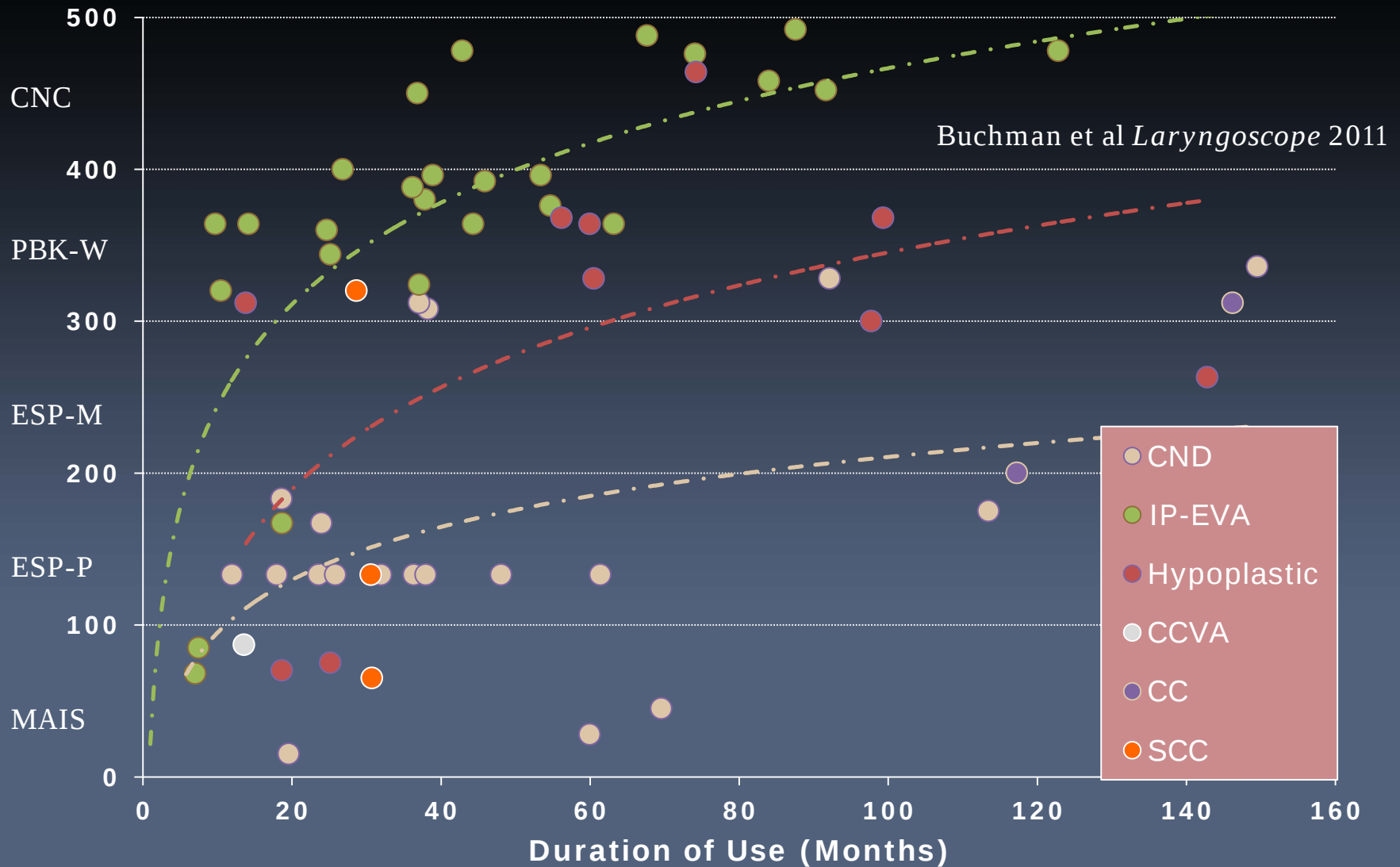


Nelson & Hinojosa, *Otology & Neurotology* (2001)

Cochlear Implant or Not?



Speech Perception (SRI-Q) by Malformation



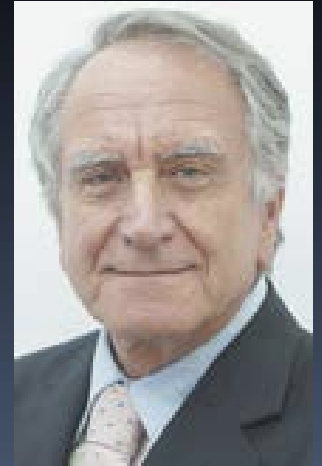
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Auditory Brainstem Implant (ABI)

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

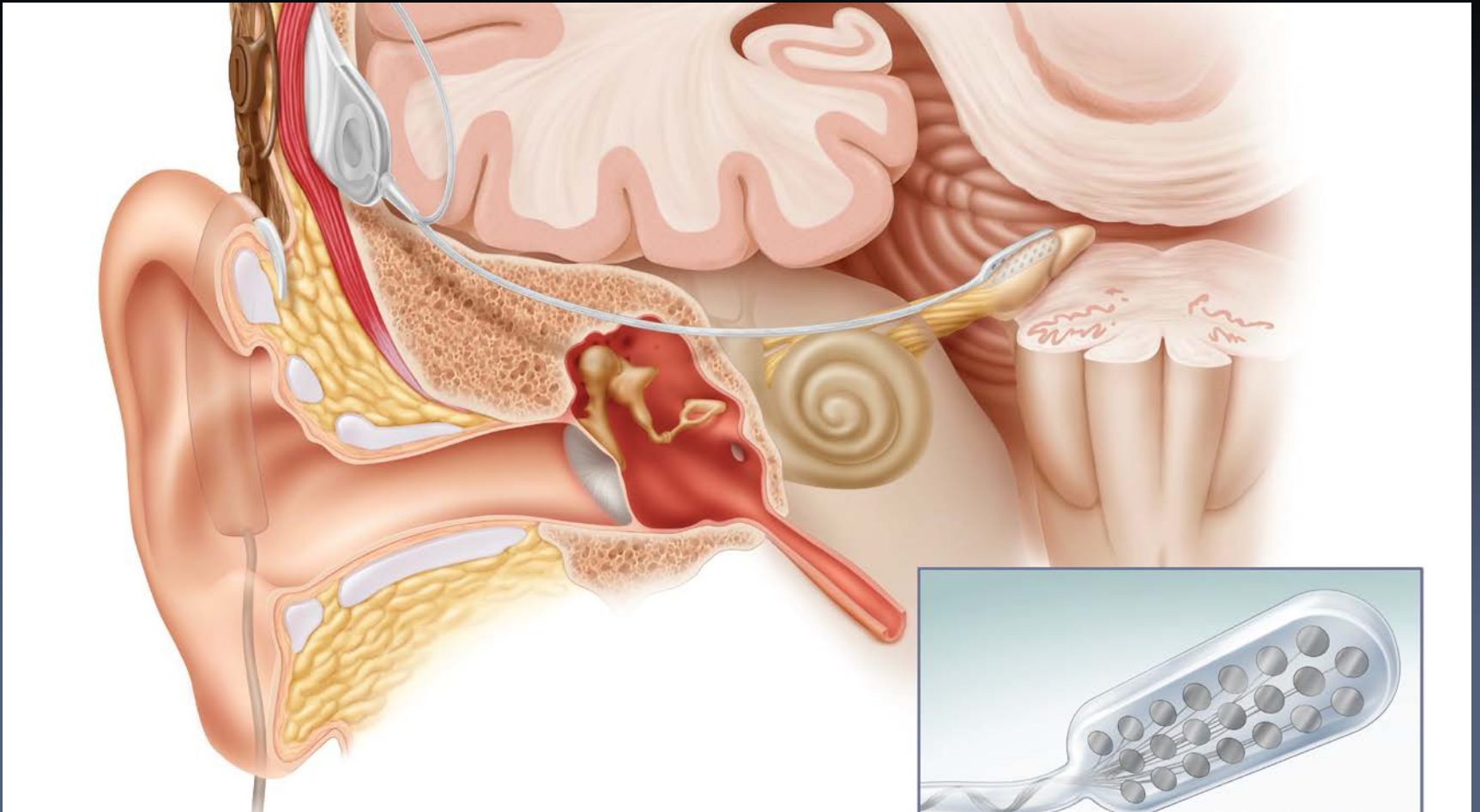


Vittorio Colletti



Levent Sennaroglu

Auditory Brainstem Implant

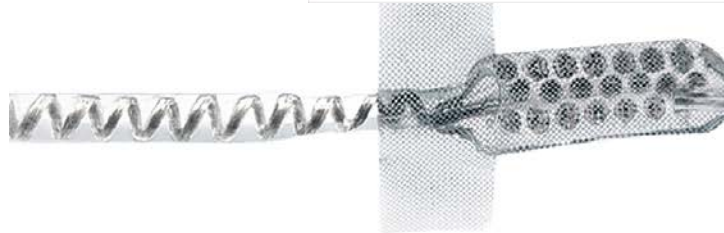


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ABI Device

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



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UNC Pediatric ABI Feasibility Study

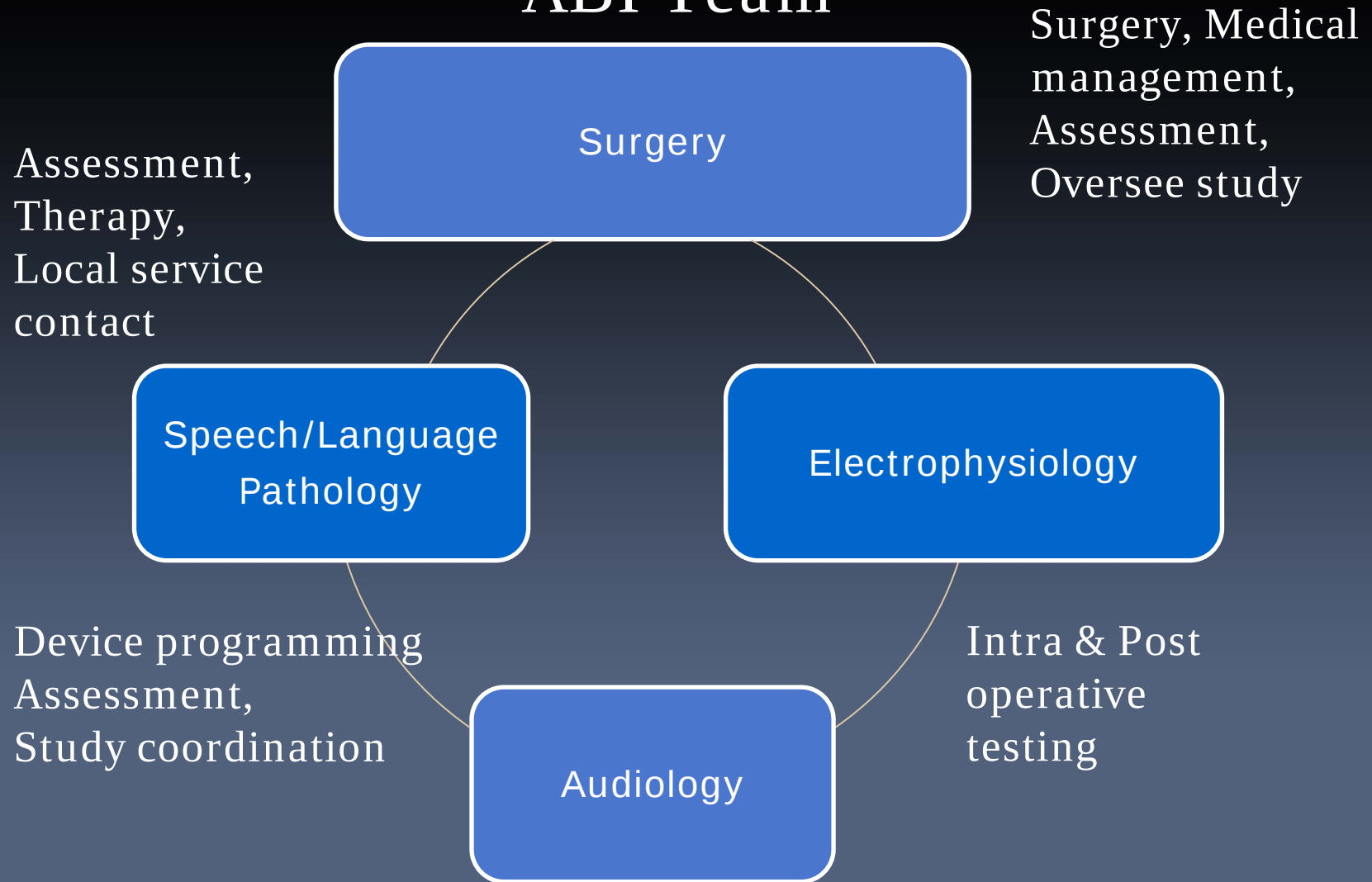
- Safety and efficacy of the Nucleus 24 Multichannel ABI:
 - to demonstrate safety of the surgical procedure
 - tolerance of device stimulation, and
 - the potential for auditory benefit beyond that experienced with their CI
- May provide the preliminary experience for a larger scale clinical trial
- 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

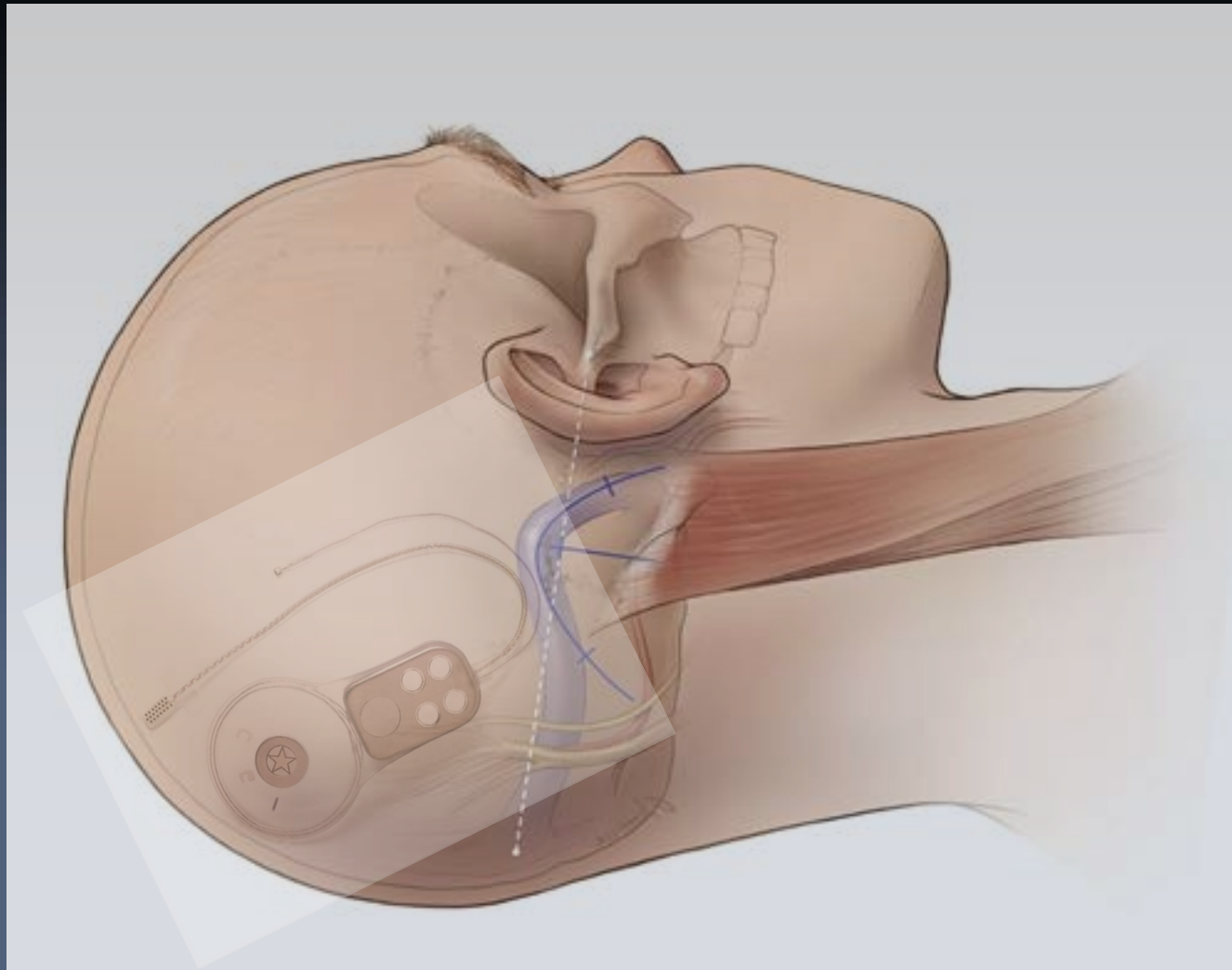
ABI Team



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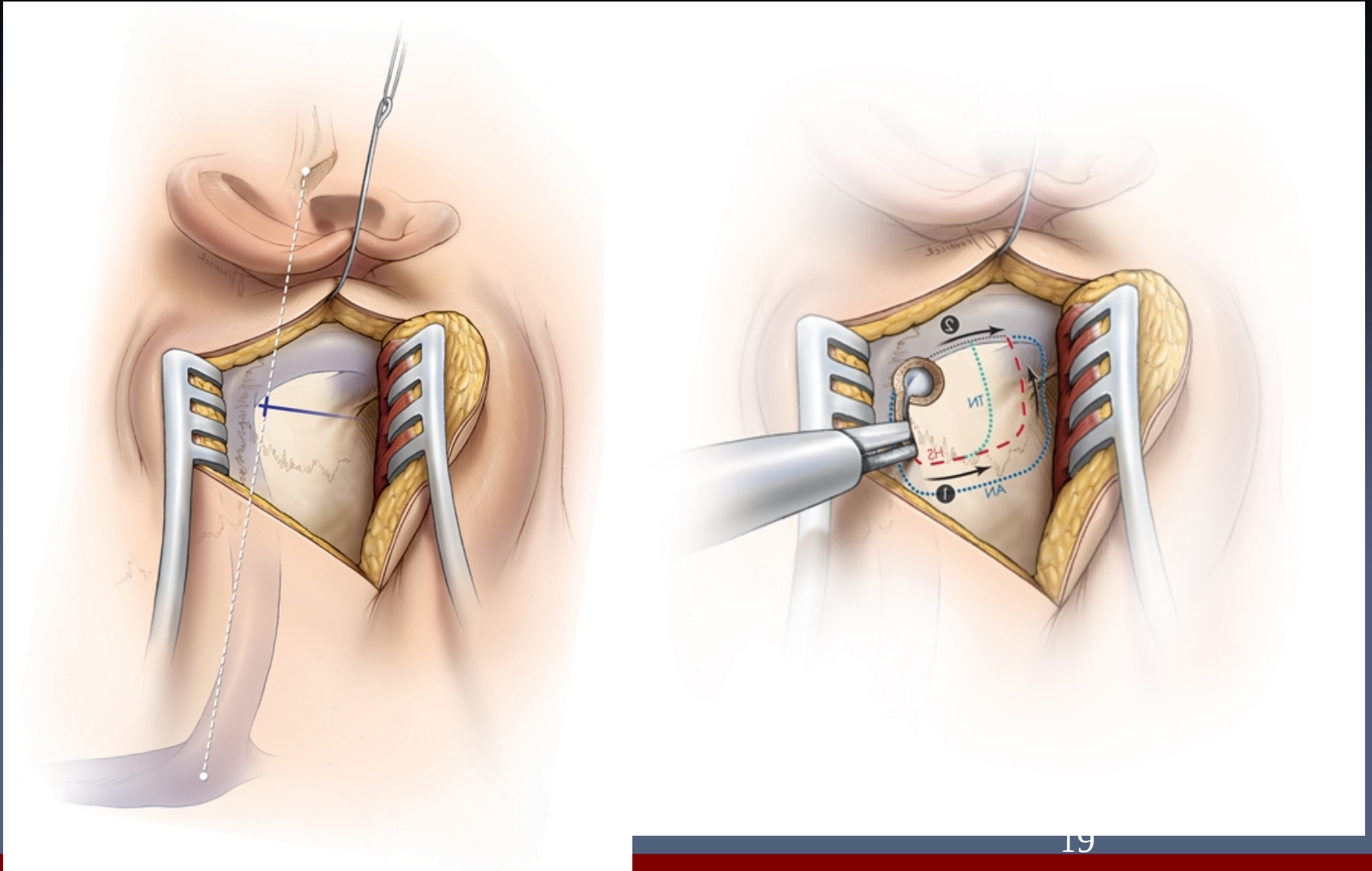
Retrosigmoid Craniotomy



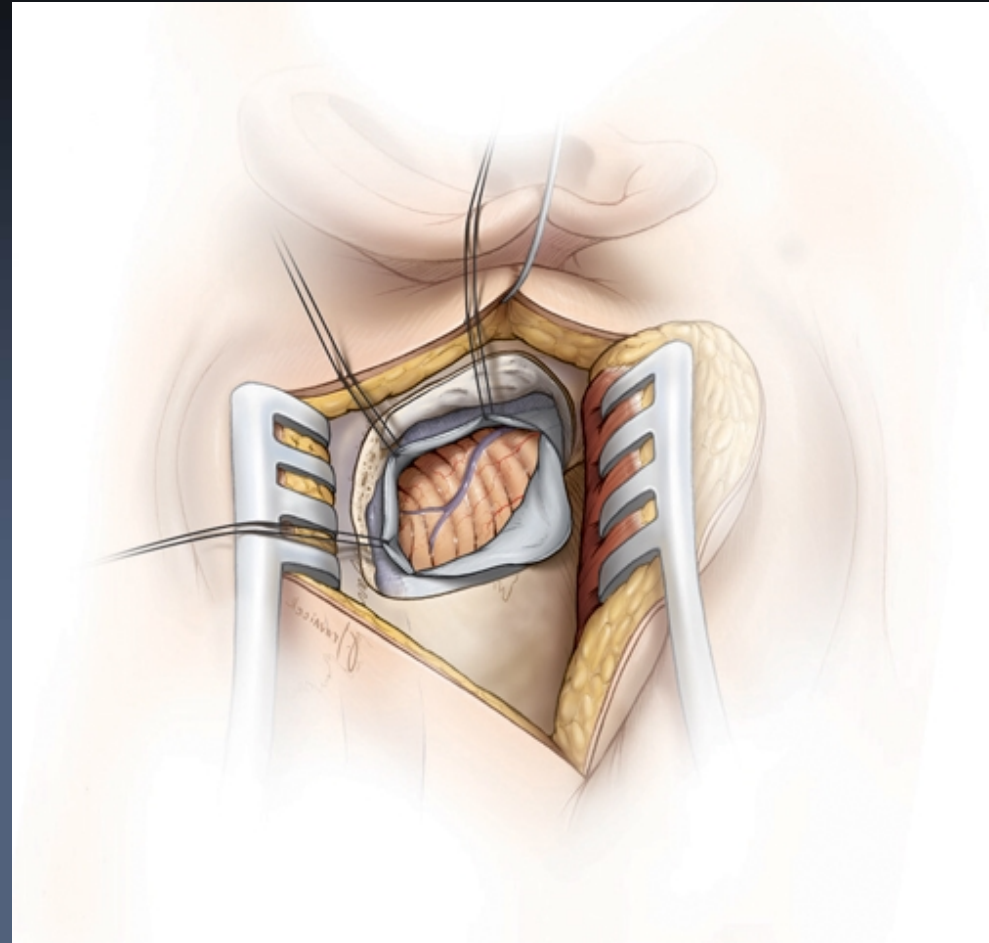
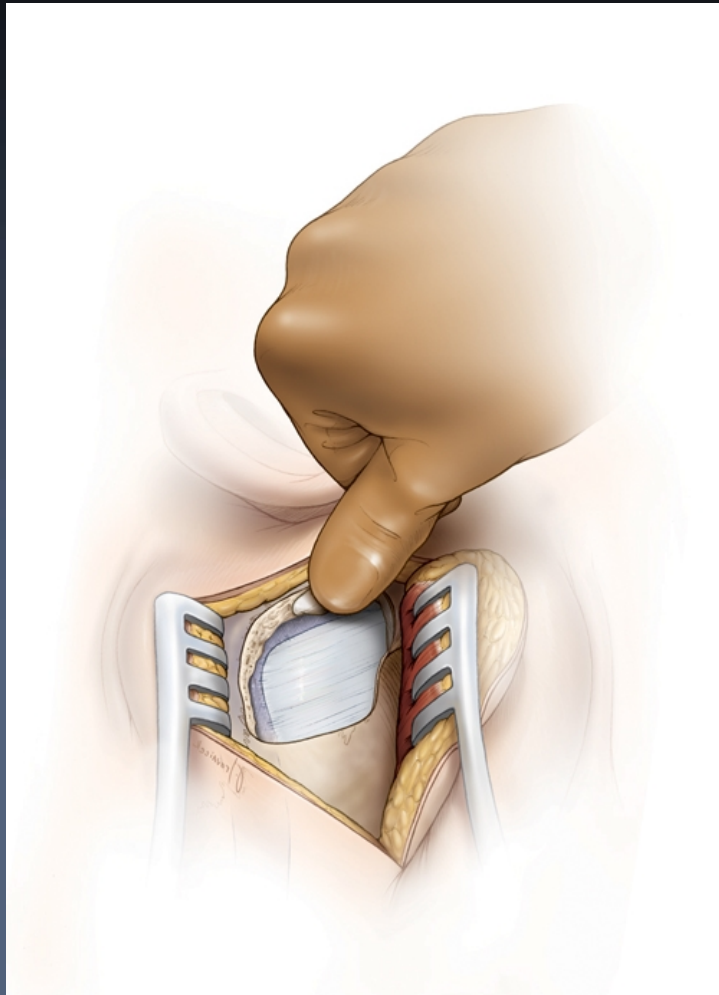
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Retrosigmoid Craniotomy

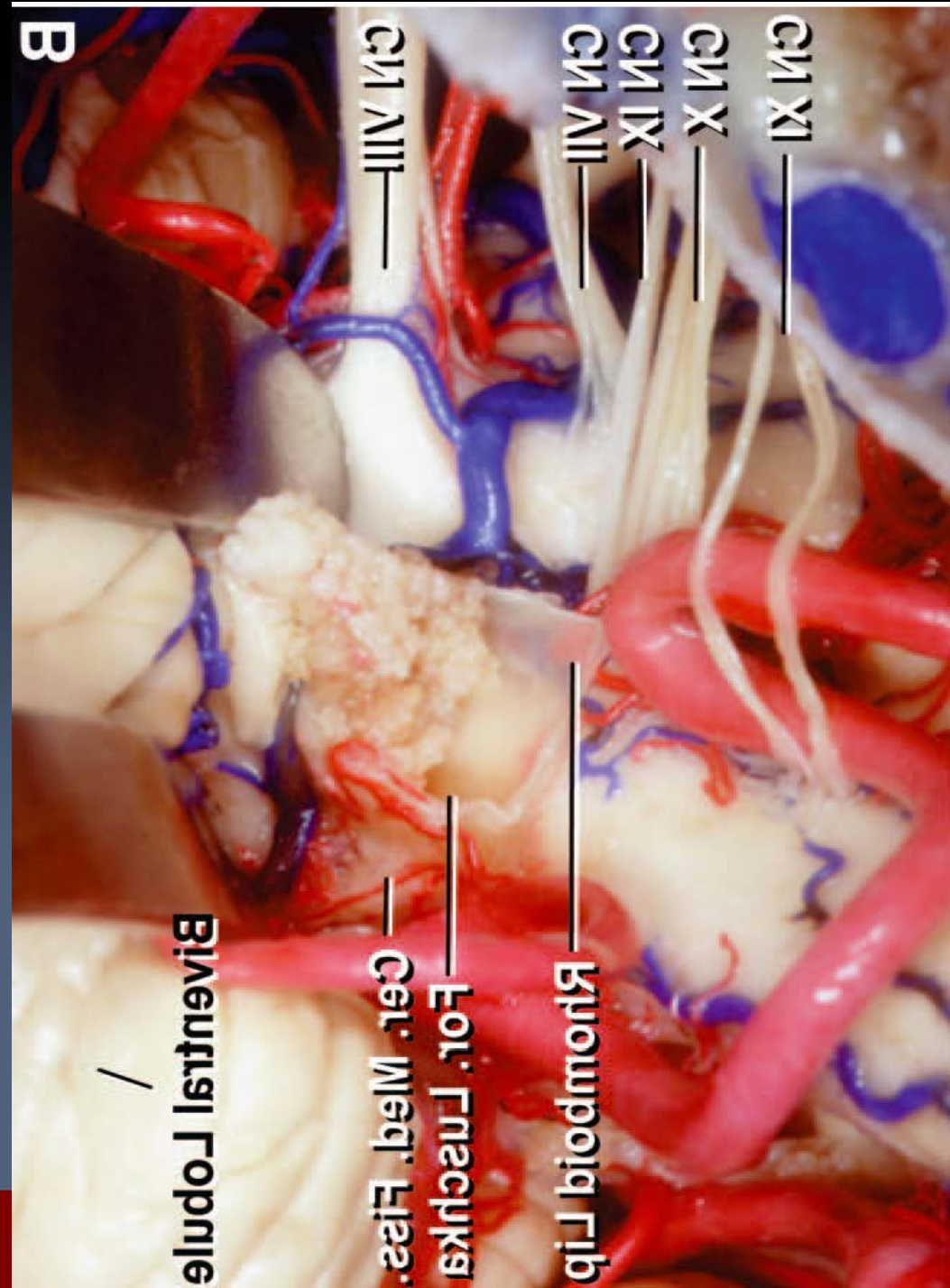


Retrosigmoid Craniotomy



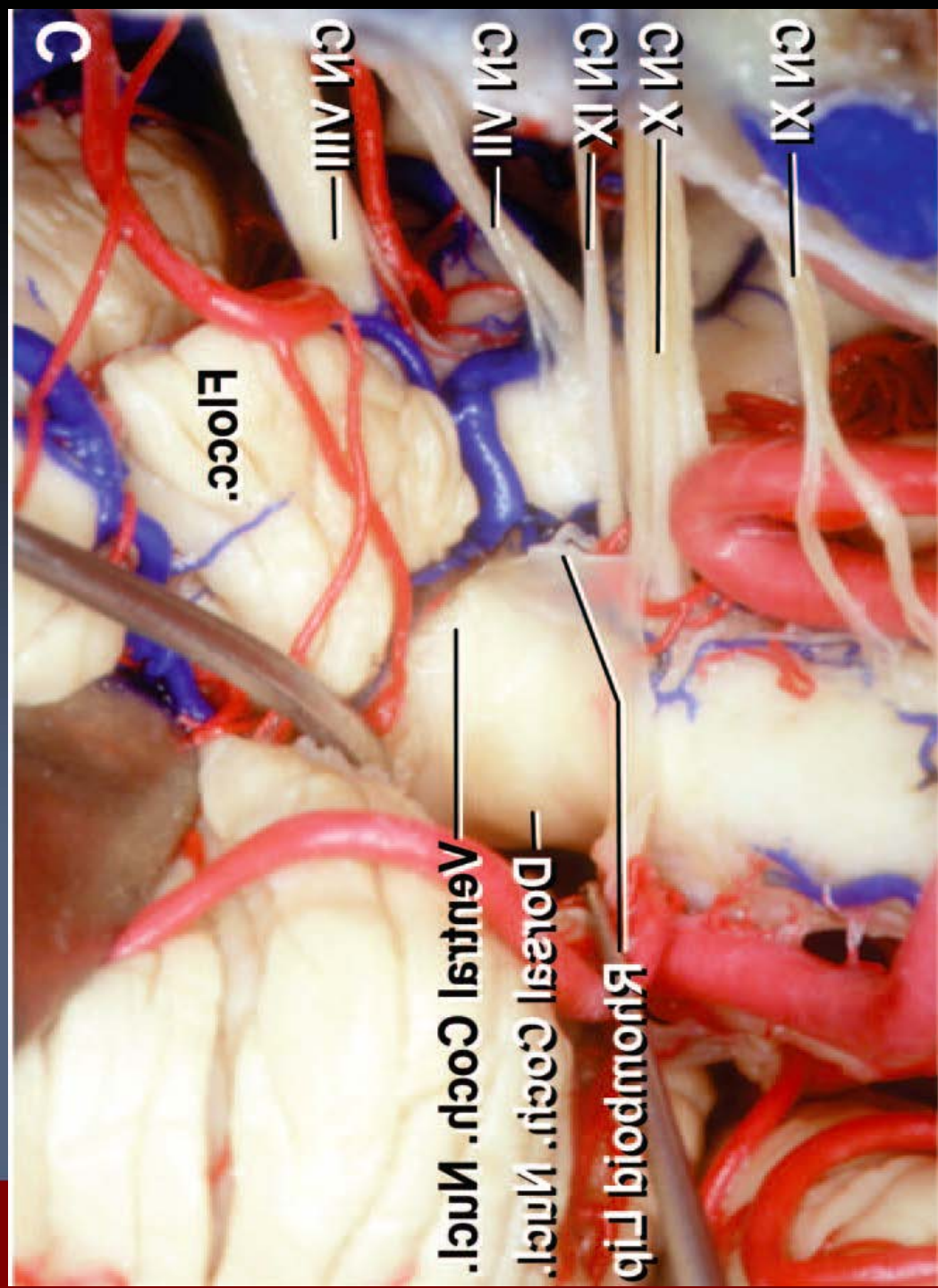
Retrosigmoid View

- 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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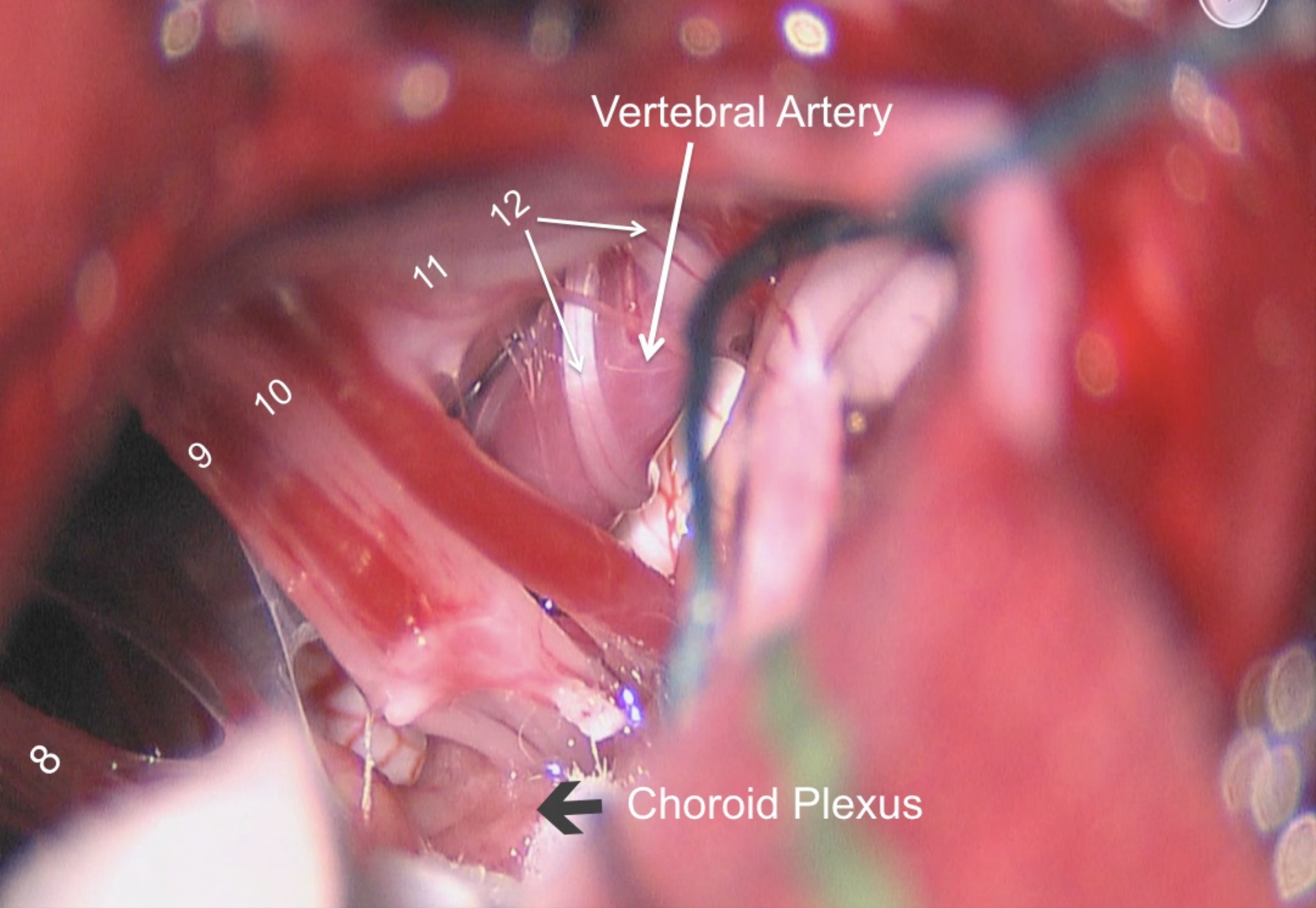
Protocol

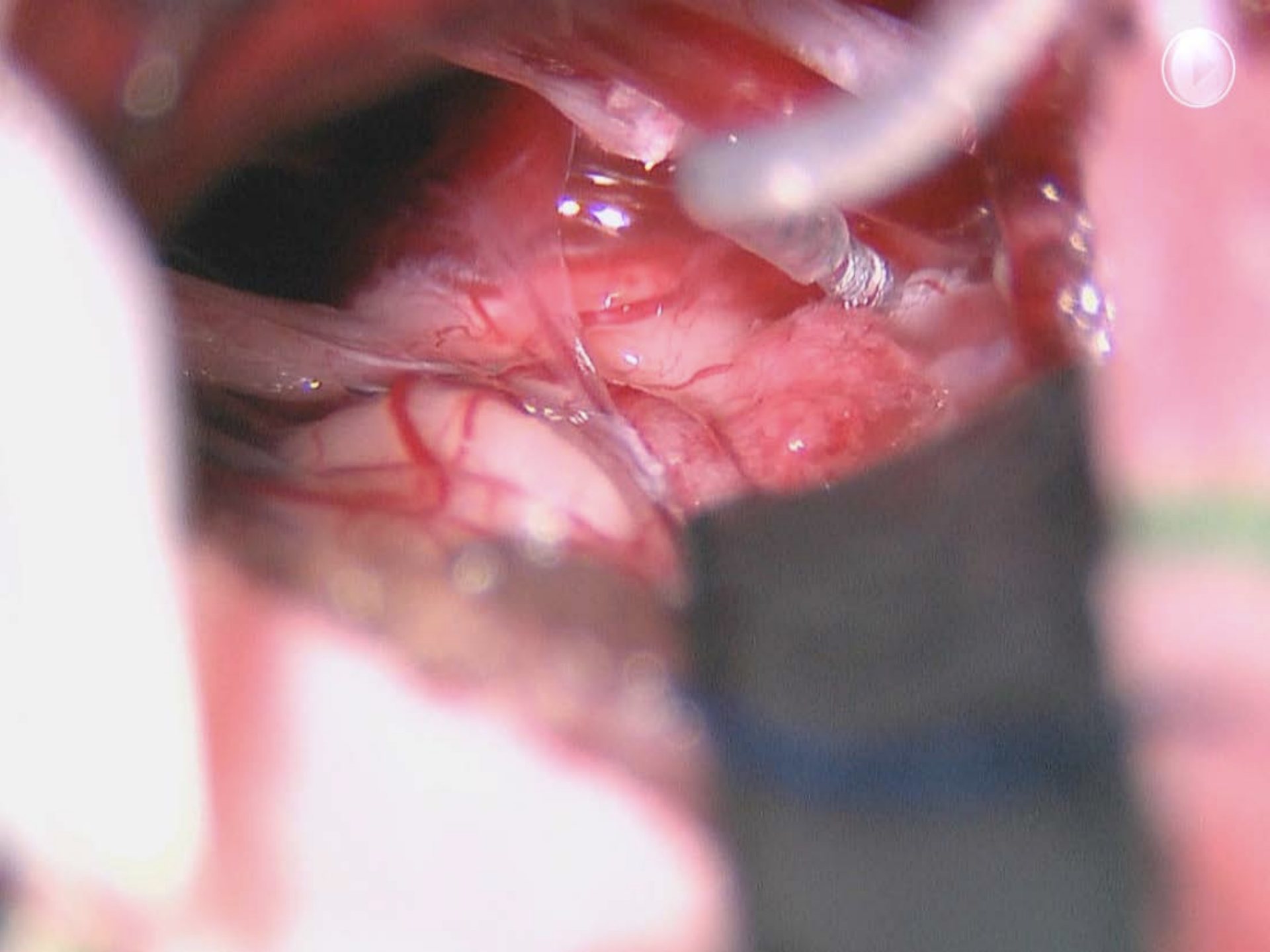
- Retrosigmoid Craniotomy
 - Nucleus 24 ABI (Cochlear Corp)
 - Monitor CN 7, 9, 10, 11
 - Implant evoked ABR
- Postop CT
- Pediatric ICU
- OR Stim prior to activate
- Activation under monitoring
- Ongoing device programming
- Speech perception/Speech & Language Assessments similar to cochlear implantation



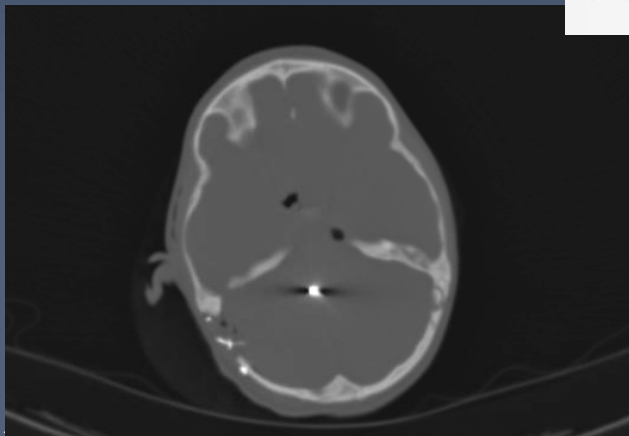
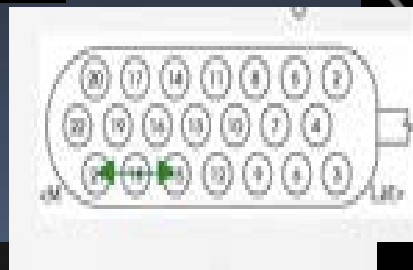
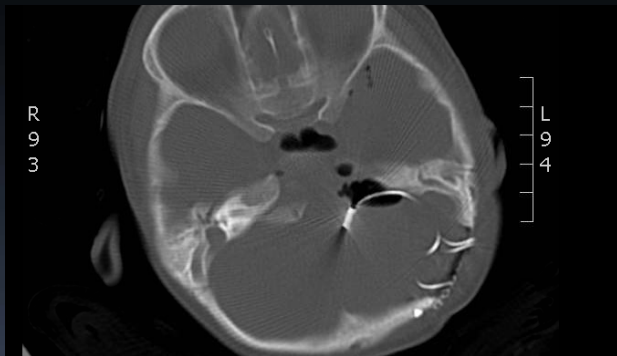
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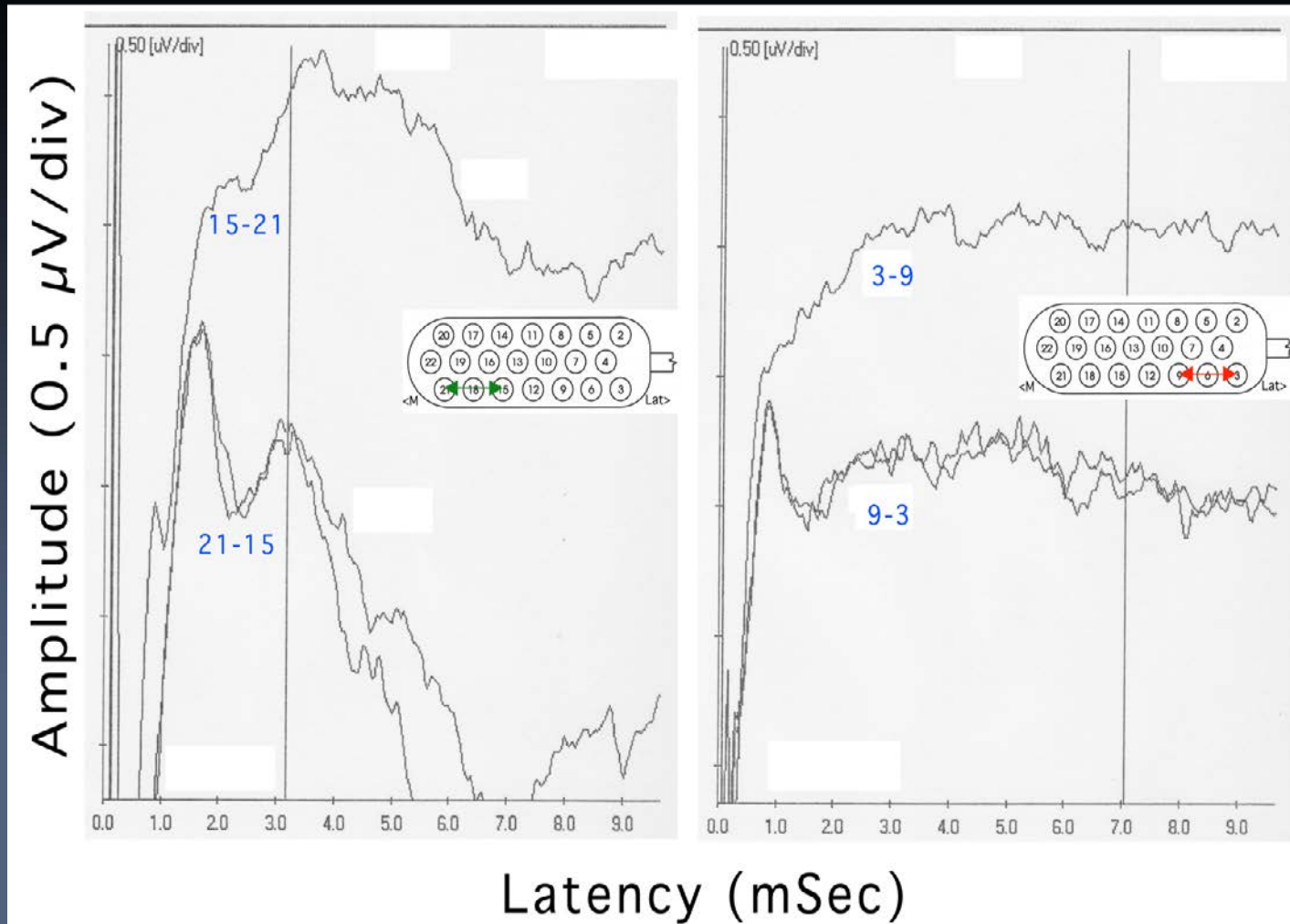
Postoperative CT Scans



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Intraop Electrically-Evoked ABR



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Audiology: Device Programming

Goal is similar to CI patient programming

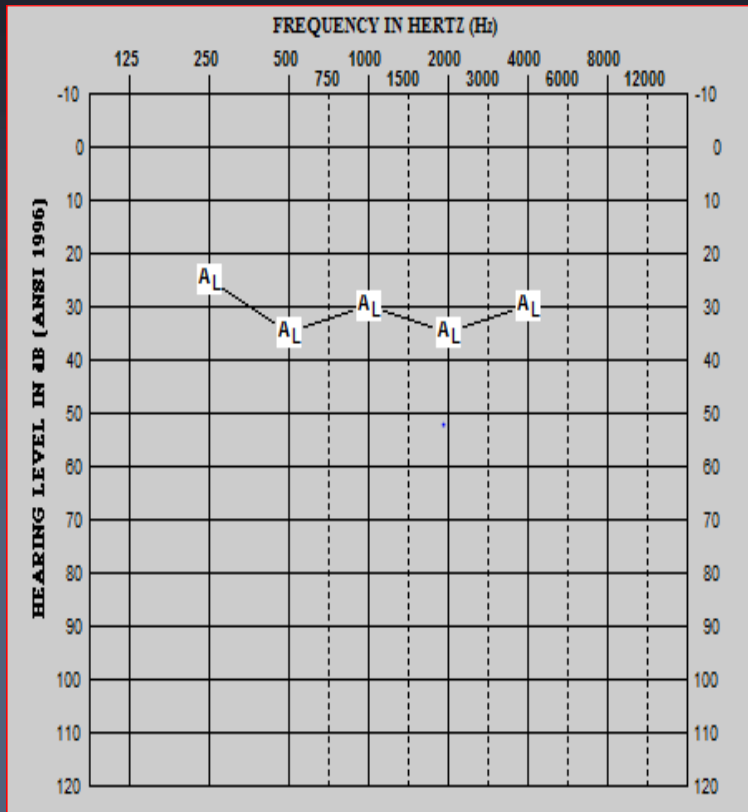
- Establish electrical threshold (T) and comfort (C) levels
- Allocation based on array design
 - Creating the auditory template that will develop with time
- Avoid stimulation of other cranial nerves (initial stim in OR)
 - Facial twitching,
 - Balance disturbance (head & trunk tilt),
 - Coughing, choking,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

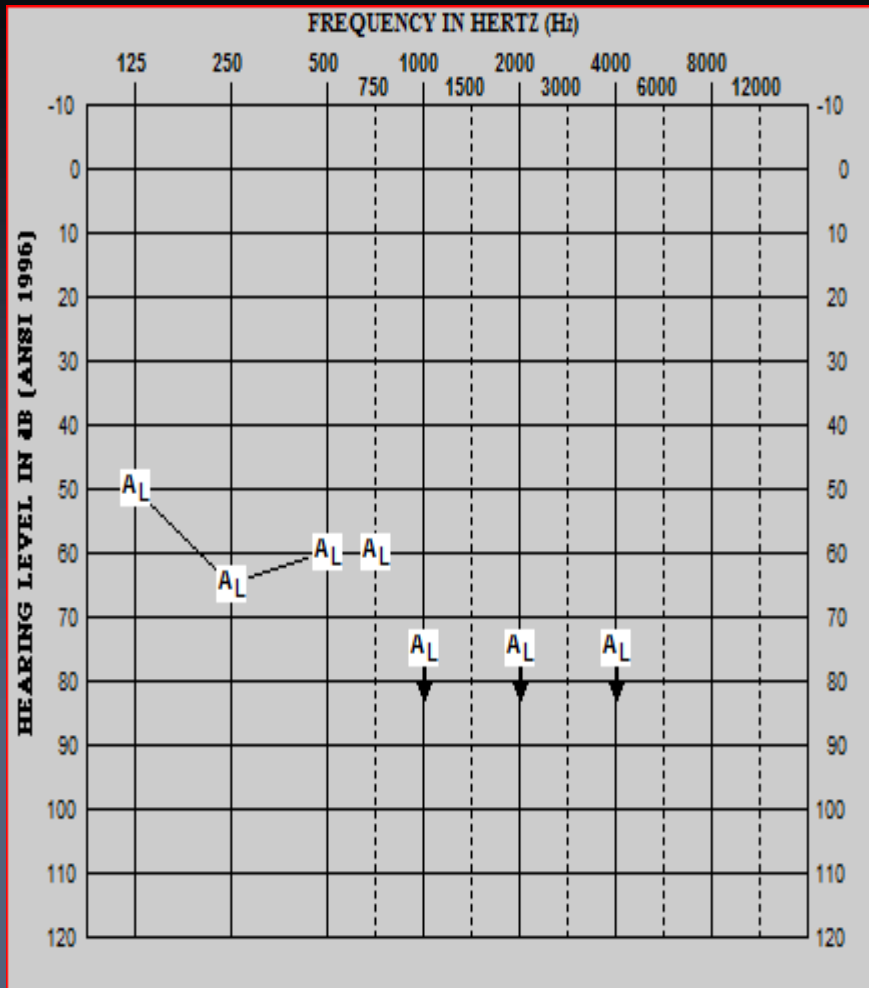
- Dx at 11 mos with HL
- CHARGE Syndrome
- Previous CI use
- Cued Speech



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Case 2



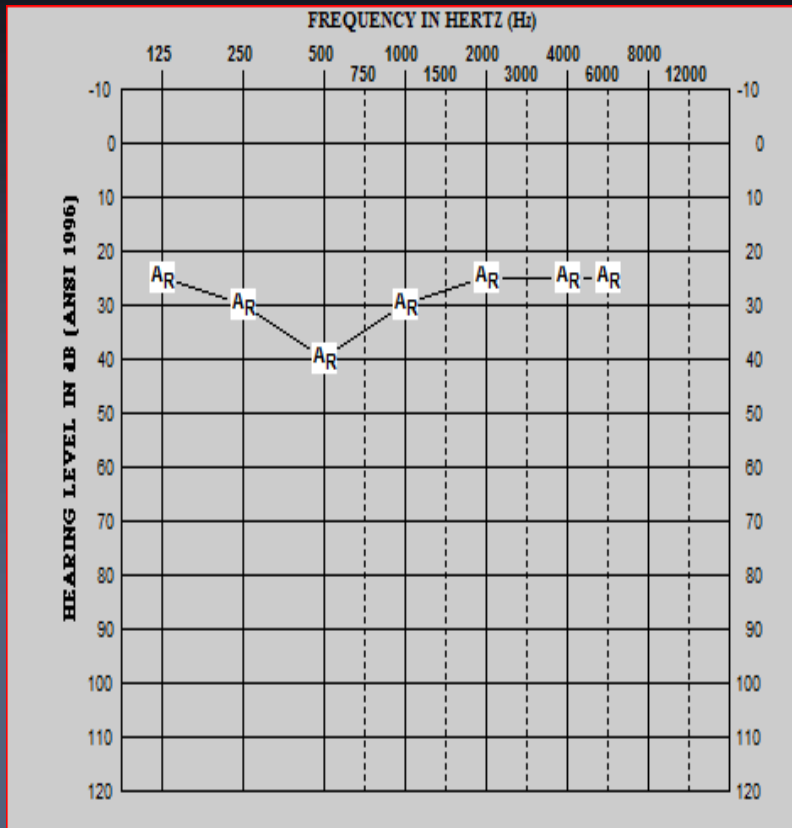
- Identified at birth
- Profound HL
- Absent cochlea and cochlear nerve
- No previous CI
- SEE



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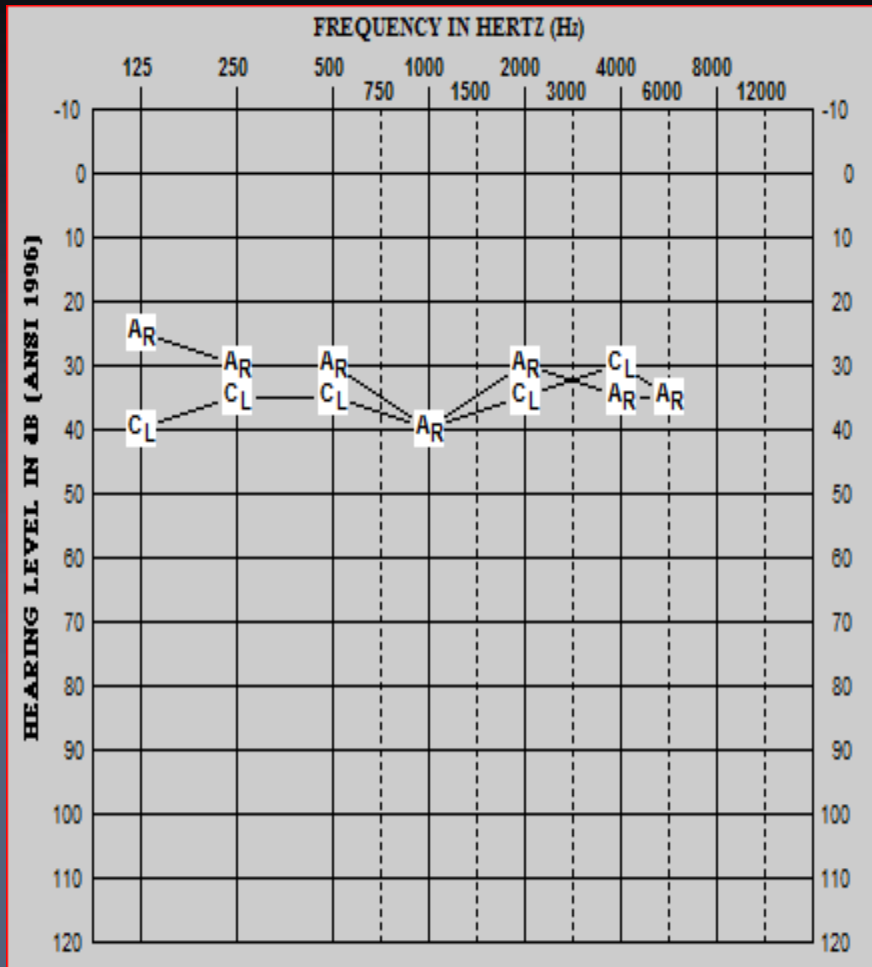
Case 3



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



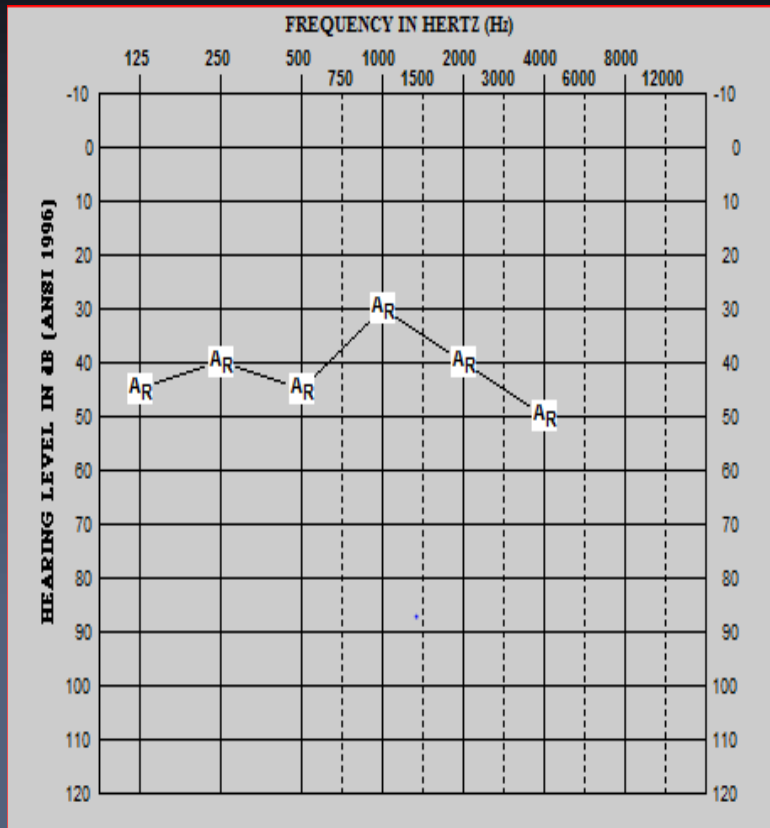
Case 4



- 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

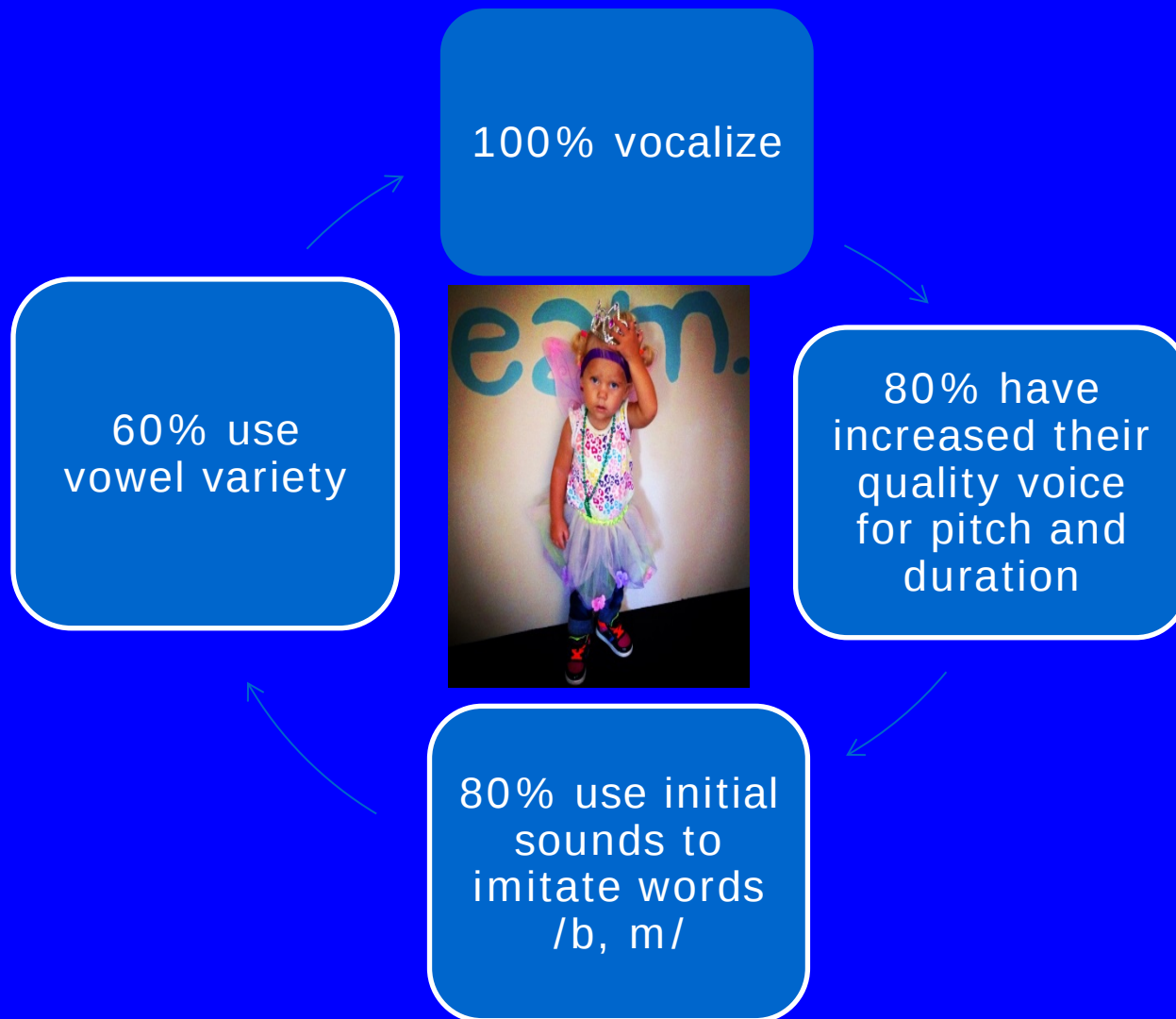
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Listening Skills and Speech Production

	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



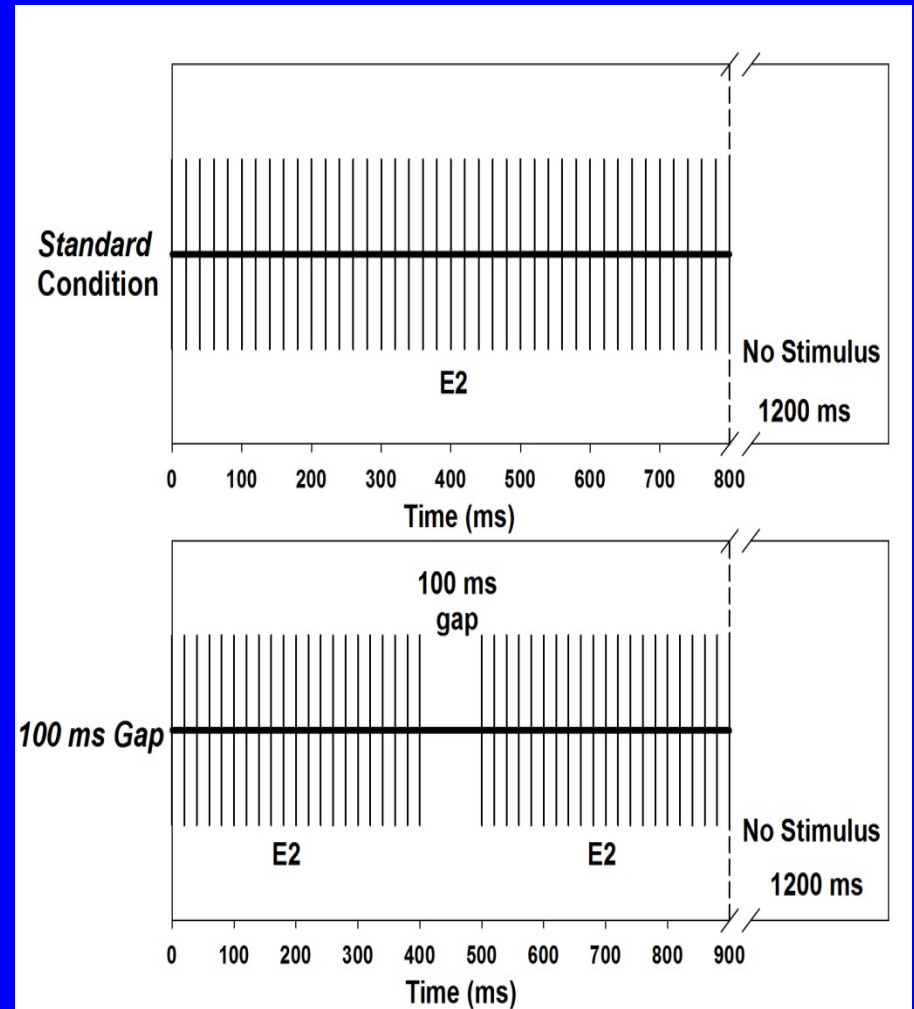
Electrophysiology (Shuman He)

- Cortical Potentials
 - Threshold Detection
 - Gap and Electrode Change

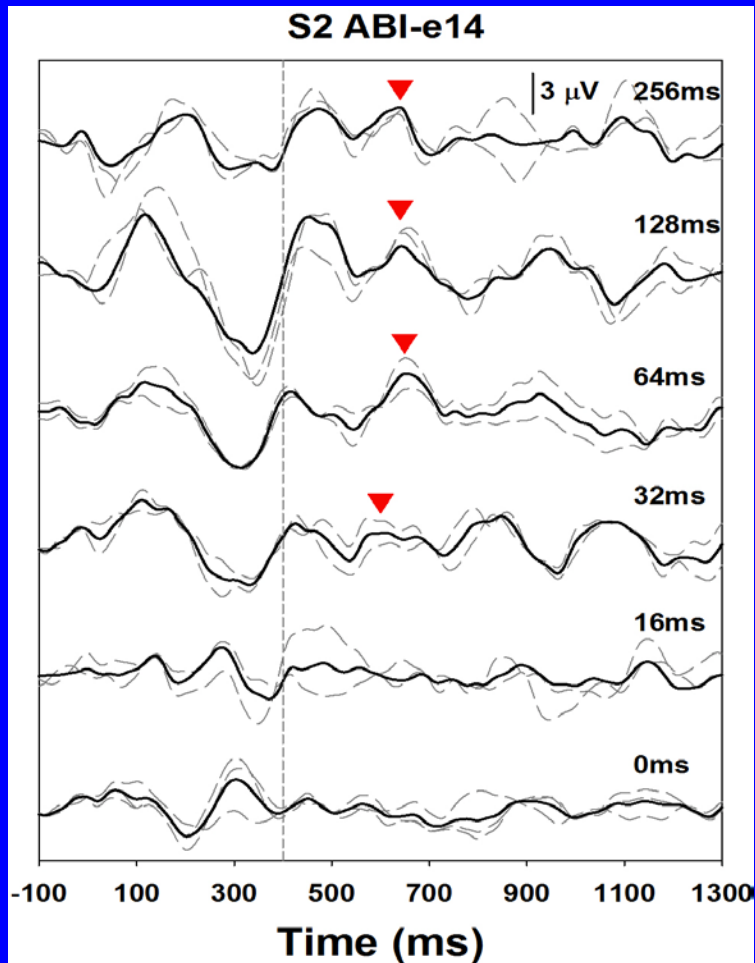
Electrophysiology

❖ *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--