Blindness separates us from things but deafness separates us from people.

~Helen Keller

Cochlear Implants

History of Cochlear Implants

- Alessandro Volta 18th century
  - Stimulated his ears with a 50V battery
  - Une recousse dans la tate
- Duchenne 1855 used alternating current
  - "the beating of fly's wings…"
- Brenner 1868 used bipolar stimulation and negative polarity to improve sound
- Jones 1940 developed theories of the mechanisms of electrical stimulation of the cochlea
- Lundberg 1950 Neurosurgeon stimulated the auditory nerve with sinusoidal current
  - The patient heard only noise

History

- 1957 Electrical stimulation *Djourno & Eyries*
- 1972 First single-channel implant *William House*
- 1984 Release of multi-channel implants
- 1985 Nucleus 22 gains FDA approval for adults
- 1990 Nucleus 22 gets FDA approval for children
- 1991 Clarion Multi-Strategy implant released
  - 1996 FDA approval for adults
  - 1997 FDA approval for children
- Bilateral implants
- Hybrid implants

Changing Criteria for Implantation

	1985	1990	1998 (Nucleus 24)	2000 (Nucleus 24 Contour)
AGE at Implantation	**Adults (18 yrs)**	**Adults & Children (2 yrs)**	**Adults & Children (18 mo)**	**Adults & Children (12 mo)**
ONSET of Hearing Loss	**Postlinguistic**	**Postlinguistic Adults Pre & Postlinguistic Children**	**Adults & Children Pre & Postlinguistic**	**Adults & Children Pre & Postlinguistic**
DEGREE of SNHL	**Profound**	**Profound**	**Severe-Profound Adults Profound Children**	**Severe-Profound Patients – 2 yrs & older Profound Children – younger than 2 yrs**
ADULT Speech Scores (open-set)	**0%**	**0%**	**40% or less (CID)**	**$\leq$ 50% (HINT) in ear to be implanted with $\leq$ 60% in the best-aided condition**
CHILD Speech Scores	**Not candidates**	**0% open-set**	**Less than 20%**	**Lack of auditory progress (MAIS, $\leq$ 30% (MLNT/LNT) (depending on age)**

How Does a Cochlear Implant Work?

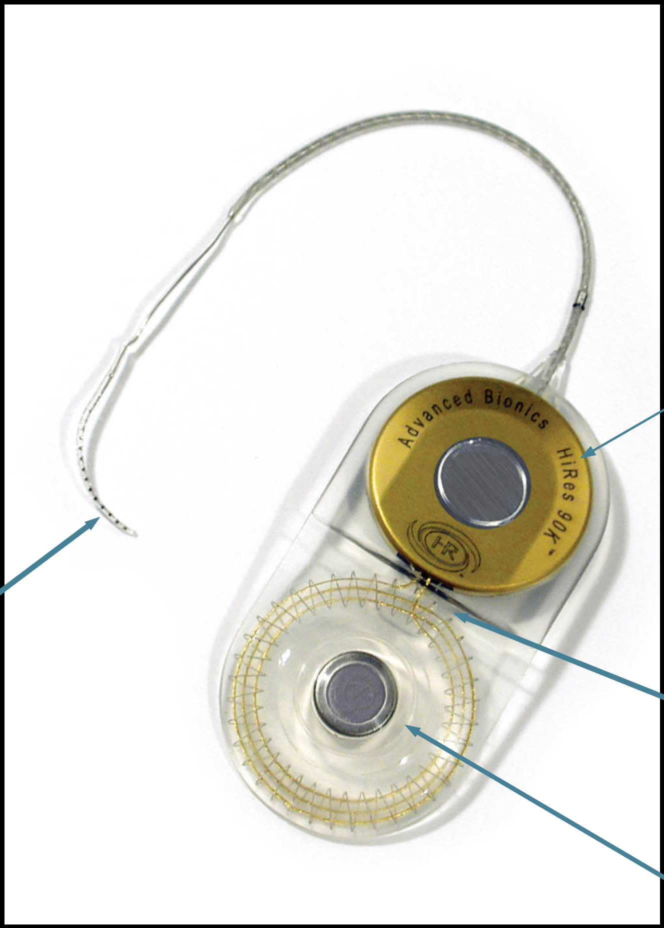
A Cochlear Implant consists of two main parts:

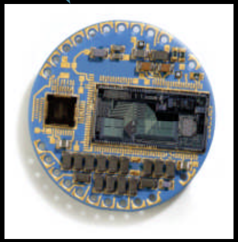
Internal Equipment

External Equipment

or

Internal Equipment
HiRes™ 90K





Internal Electronics

Electrode Array

3 turn gold wire coil

Removable Magnet

External Equipment For a Cochlear Implant

Speech Processors

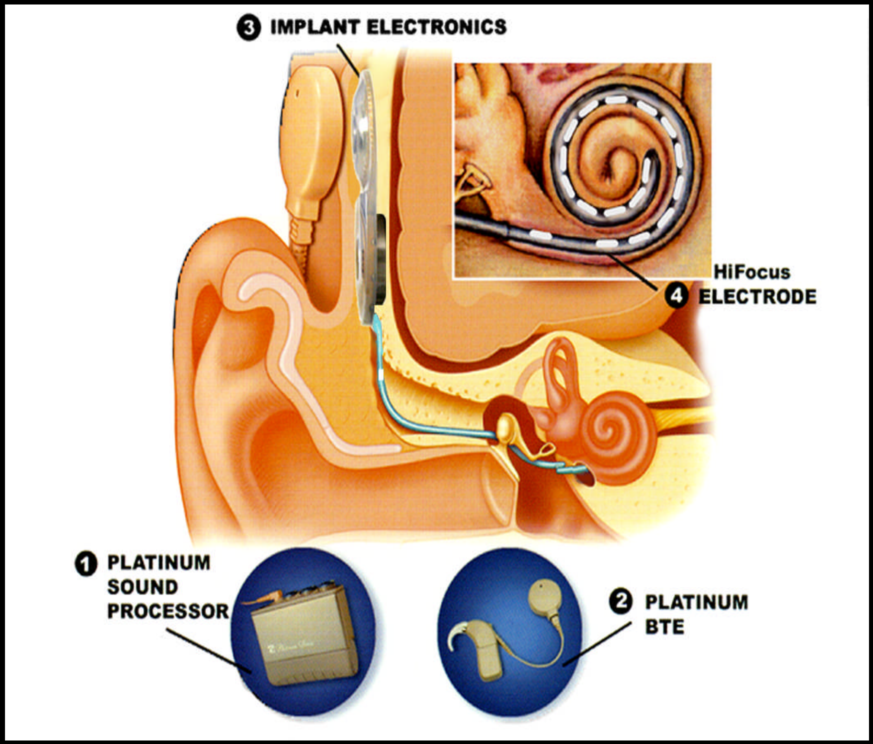
Microphone

Battery Supply

Headpiece

Cable

How Does a Cochlear Implant Work?



- Sound waves enter through the microphone.
- The sound processor converts the sound into a distinctive digital code.
- The electrically coded signal is transmitted across the skin through the headpiece to the internal portion of the device.
- The internal device delivers the sound to the electrodes.
- The electrodes stimulate the hearing nerve.
- The hearing nerve sends the signal to the brain for processing.

Prevalence

- Prelingual profound SNHL 1 – 4 per 1000 births
  - 60% genetic
    - 90% will have 2 hearing parents
    - 97% will have 1 hearing parent
  - 30% environmental
  - 9% meningitis
- By age 75
  - 360 per 1000 adults have profound SNHL

The Candidate Evaluation

Goals of Evaluation

- To determine whether an individual would perform as well with hearing aids as with a cochlear implant.
- To provide a recommendation for or against implantation
- To select an ear for implant
- To determine appropriate expectations to guide counseling

Factors that Affect Implant Performance

- Adult Implants
  - Hearing experience
    - Residual hearing
    - Length of profound hearing loss
    - Hearing history of each ear
  - Age at Onset
  - Age at Implant
  - Cognitive / Central Abilities
  - Motivation
- Childhood Implants
  - Training with Amplification
    - Consistency
  - Presence of Other Disabilities
  - Cochlear dysmorphia
  - Family Support
  - Education with Focus on Development of Spoken Language

- *Two Most Significant Factors Affecting Performance*
  - *Development of Speech*
  - *Duration of Deafness*

Audiologic Tests

- Consonant-Nucleus-Consonant   CNC
  - 50 monosyllabic words in open set format
- Hearing In Noise Test   HINT
  - Ten phonemically balanced sentences
  - Presented in quiet or noise
  - HINT-C for children
- Bamford-Koval-Bench Sentences   BKB
  - In quiet or noise
- Lexical Neighborhood Test   LNT
  - 50 monosyllabic words
    - "easy" occur frequently in the English language
    - "hard" have many lexical neighbors
- Phonetically Balanced Kindergarten Test
  - 50 words with a long duration of use.

Outcome Expectations for Children

- **Sound detection in sound fields at 25 dB for frequencies of 250 to 4000 Hz**
- **Audiometric Responses**
    - LNT-easy 48%
    - LNT-hard 44%
    - BKB sentences 57%

- **Implanted age 2 years or younger**
  - Potential for communication skills similar to normal hearing peer
  - Potential for speech to be easily understood
  - Attend local school with minimal support services
- **Implanted age 4 years or younger**
  - Vocalization at early stages after implant
  - Auditory behaviors that are evident before they can be measured
  - Speech production skills reflective of auditory abilities
  - Reduced language delays
- **Implanted age 4 to 5 years**
  - Excellent closed set performance
  - Improved speech production
  - Use of hearing to support language acquisition
  - Reduced dependence on visual cues
- **Implanted at 6 years or older**
  - Improved auditory detection
  - Improved speech perception with good closed set abilities
  - Continued dependence on visual cues for communication
- **Children with progressive loss**
  - Excellent progress with shorter duration of use
  - Similar to adults

Outcome Expectations for Adults

- Sound detection in sound fields at 25 to 30 dB for frequencies of 250 to 4000 Hz
- Audiometric Responses

	50 dB	60 dB	70 dB
CNC words	24%	39%	42%
Hint score	57%	73%	72%

- HINT at 60 dB with S/N of +10
  - 48%
- Postlingual deafened obtain good open-set speech perception as early as 1 mo
  - Can use the phone
- Prelingual deafened obtain open set speech recognition often only after 1 year of use.

Clinical Evaluation

- Audiometric Testing
  - Hearing aid use
- Imaging
- Hearing History
- Physical Exam
  - Associated syndromic features
  - Acute or Chronic ear disease
- Cognitive Abilities
- Device Selection
- Ear Selection
- Counseling
  - Meningitis
  - Commitment
  - Realistic expectations

Candidacy Criteria Adults

- Moderate hearing loss in low frequencies
- Profound hearing loss
  - >70 dB mid to high frequencies
  - <40% correct in HINT
- Little or no benefit from hearing aids
  - < 60% correct in best aided listening condition
  - < 50% correct in ear to be aided
- No medical or radiologic contraindication
- Motivated patient

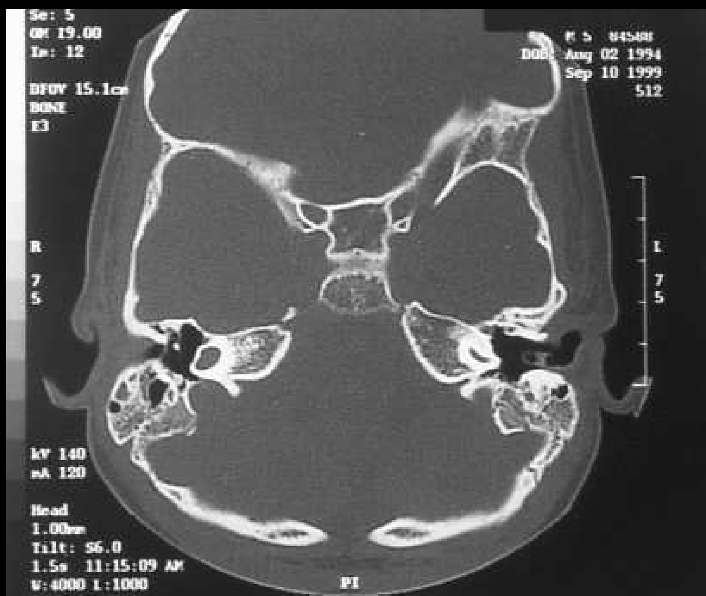
Candidacy Criteria Children

- Greater than 12 months old
- Profound hearing loss >90 dB SRT / PTA
- <20% correct in HINT or similar
  - LNT/ MLNT $\leq$ 30%
- Little or no benefit from hearing aids
  - Lack of progress in auditory skills
  - Parental responses to questionnaires MAIS
- No medical or radiological contraindication
- Placement in educational setting for concentrated auditory skills development
- Motivated family

Imaging

- High-resolution CT of the Temporal Bones
  - **Anatomy of the cochlea**
    - Most dysplasia's (Mondini's) obtain excellent results
    - Aplasia (Michel's) is an absolute contraindication
  - **Intact internal auditory canal**
    - Canal height should be greater than 1.5 mm
    - Consider MRI T2-weighted
    - Consider vestibular testing
  - **Primary of secondary bone diseases**
    - Cochlear otosclerosis
    - Padgets disease
    - Camurati-Engelmann disease
  - **Wide vestibular aqueduct**
    - CSF gusher
      - Elevate head
      - Seal cochleostomy
      - Lumbar drain
  - **Labyrinthitis ossificans**
    - 90% are limited to the area around the round window
    - Consider MRI

Imaging


Se: 5
Im: 12
DFOV 15.1cm
BONE
E3
DOB: Aug 02 1994
Sep 10 1999
512
R
7
5
L
7
5
kV 140
mA 120
Head
1.00mm
Tilt: S6.0
1.5s 11:15:09 AM
W:4000 L:1000
PI


Ex:6827
Se:3
Im:3
S29.2
AS
DEC 10, 1996
R
L
W:4002/L:803
PI

Nucleus 5

- Flexible silicone housing with a titanium case
- Thinnest receiver on the market
- MRI safe up to 1.5 T with magnet removed
- Electrode array
  - 22 half banded platinum electrodes
  - Variably spaced over 15mm
  - Overall length is 24mm
- Perimodiolar design
  - Comes loaded on a stylet
  - 10 stiffening rings
  - Easiest to insert

HiRes 90K

- Flexible silicone housing around a titanium case
- 2.5mm thick on top of skull
- MRI safe to 1.5 T with magnet removed
- HiFocus Ij electrode is “banana-shaped”
  - Comes in a reloadable insertion tube
- Electrode array
  - 16 contacts spaced 1.1 mm apart
  - Contacts spaced over 17mm
  - Overall length 24mm
  - Thickness is 0.5 to 0.8 mm

Med-El C40+

- Housed in a ceramic case
- MRI safe at .2T with magnet
  - In Europe testing has exceeded 1.0T
- Electrode array
  - 12 pairs of electrodes
  - Tapered design
  - Spaced over 26.4mm with 2.4mm between each
  - Overall length is 31.5mm
  - Compressed array is spaced over 12.1mm overall length is 18mm.

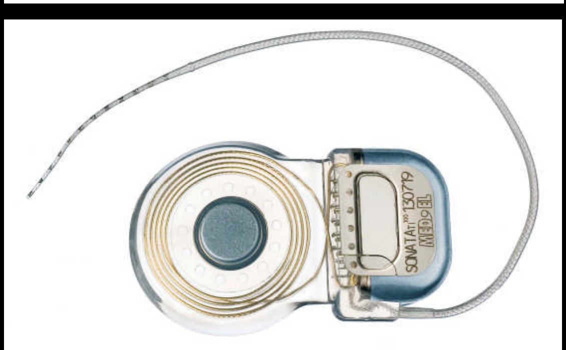
External Processors

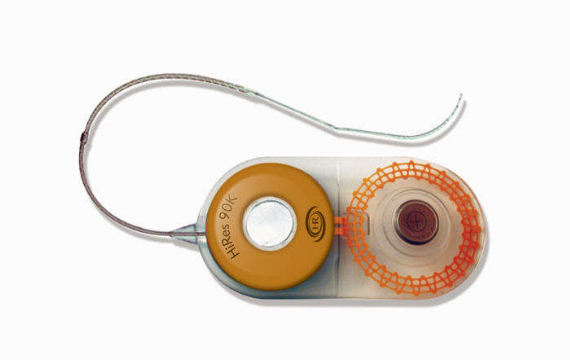
- BTE or Body-worn
  - Battery life
  - Auxiliary microphones
  - Telephone adaptors
  - FM Systems
  - Cosmetic considerations
    - Weight and size
    - Design
    - Colors

- Processor Strategies
  - CIS
  - N of M
  - Spectral peak extraction SPEAK
  - Advanced combination encoder ACE

Device Selection

- Cochlear Corporation
  - Available devices
    - Nucleus Freedom Contour Advance
    - Split Array
    - ABI
  - Quality, Reliability, NRT
- Med-El
  - Available devices
    - Standard
    - Compressed
    - Split Array
    - ABI
  - Size, Battery life, electronic features
- Advanced Bionics
  - Available device
    - Hi Res 90K
  - Aesthetics





Migirov 2009

Ear Selection

- Best results are obtained from the ear with
  - the shortest duration of deafness
  - better word recognition
  - better acoustic thresholds at more frequencies.
- If one ear has never had acoustic stimulus the other ear is chosen
- If either ear is acceptable, implant the ear that will benefit least from traditional amplification
- If either ear can be amplified the choice is made on nonaudiologic basis
  - Handedness
  - Patient preference
- All things equal select the right ear to capture the contralateral left-hemisphere specialization for speech recognition

Meningitis

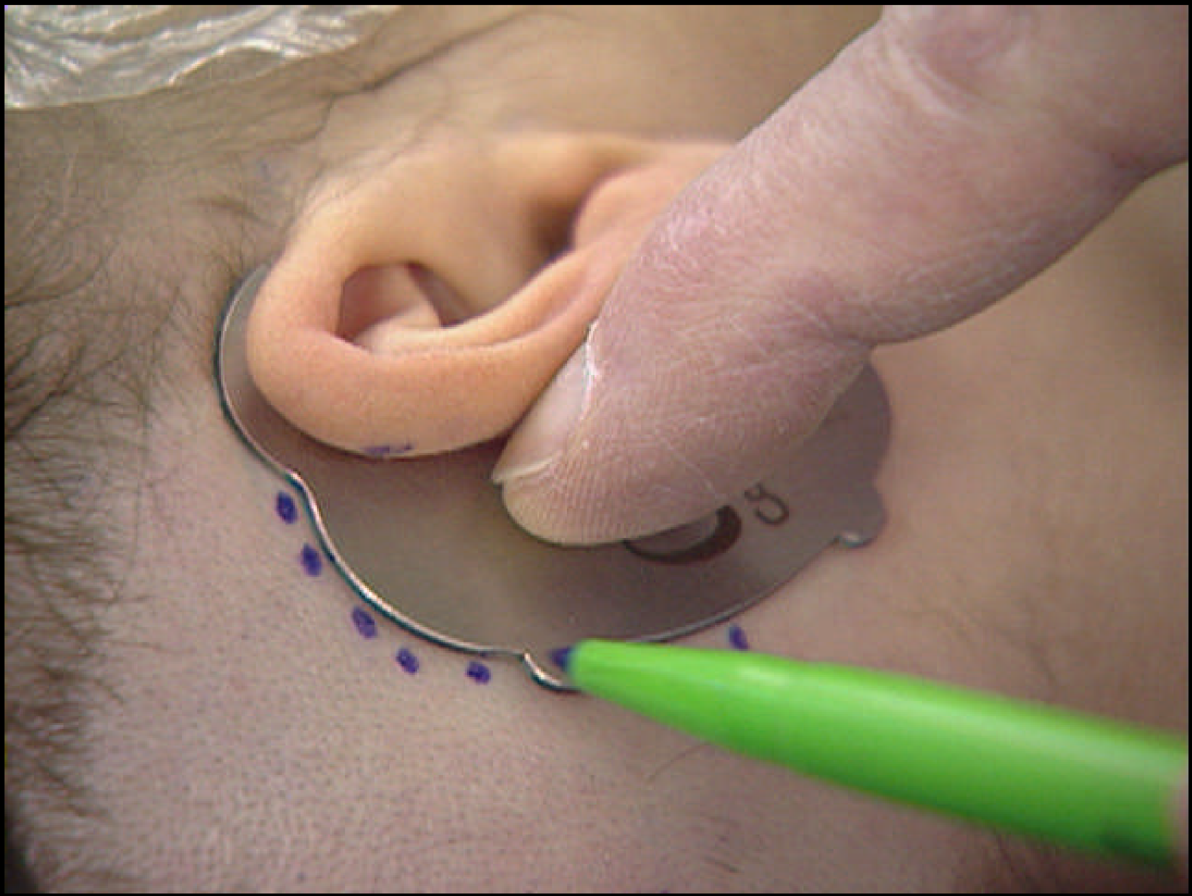
- Reefhuis et al 2003 evaluated 4264 children and found .5% risk of meningitis
- CDC recommends immunization for pneumococcus and H. influenzae
- Hearing impaired are at increased risk for meningitis without implant
- Advanced Bionics “positioner” has been removed from the market

Counseling : Risks and Benefits

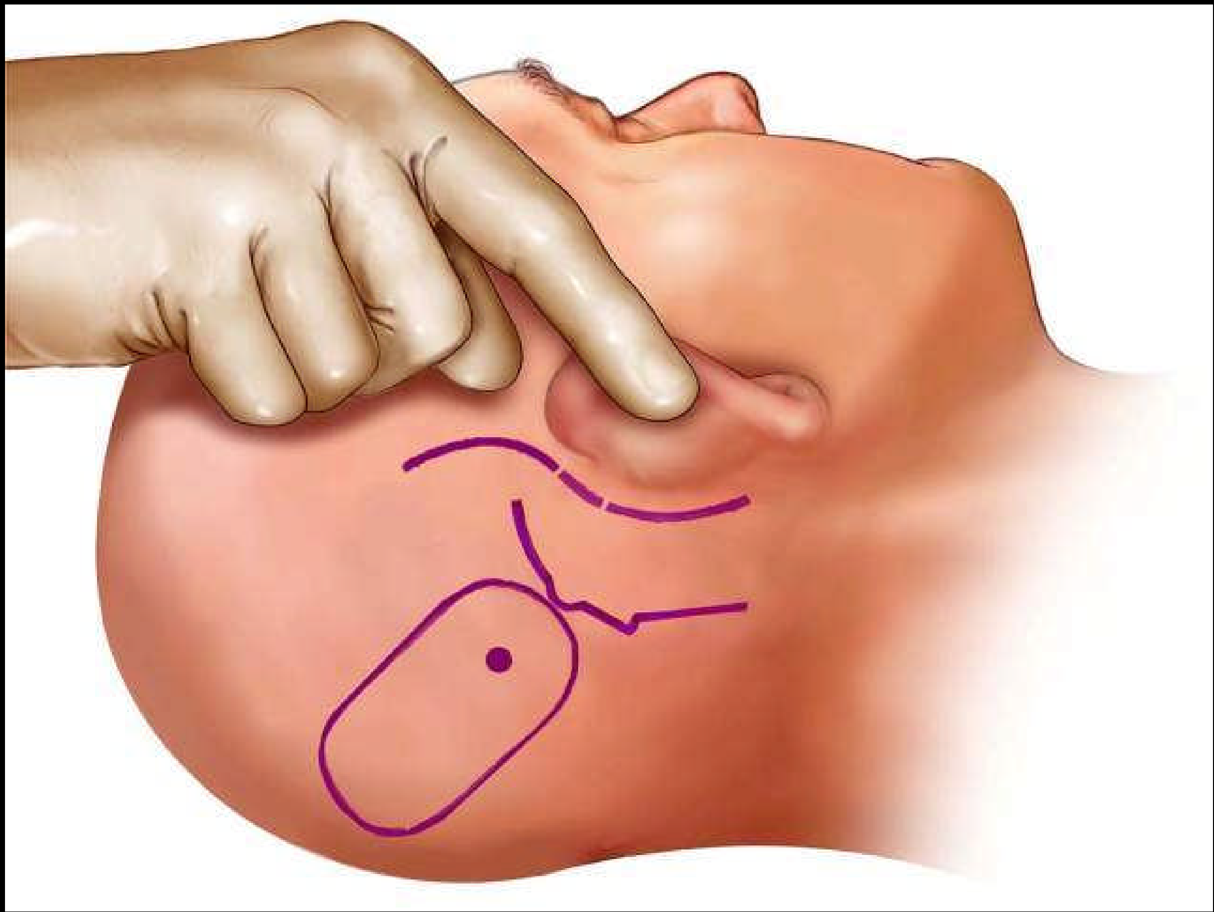
- Bleeding
- Pain
- Infection (Vaccination!!)
- Scar
- Taste disruption
- Facial nerve paralysis
- Dizziness (test pre-operatively)
- Delayed failure
- Delayed infection
- Wound breakdown
- Surgical risk of anesthesia

1. Environmental awareness and responsiveness
2. *Potential for significantly better speech understanding than with a hearing aid*
3. Less reliance on visual cues and speech reading
4. Less reliance on others for communication success
5. Potential to use a voice phone
6. Potential to appreciate music
7. Increased independence in daily living
8. Increased confidence; social, vocational, communicative

Surgical planning

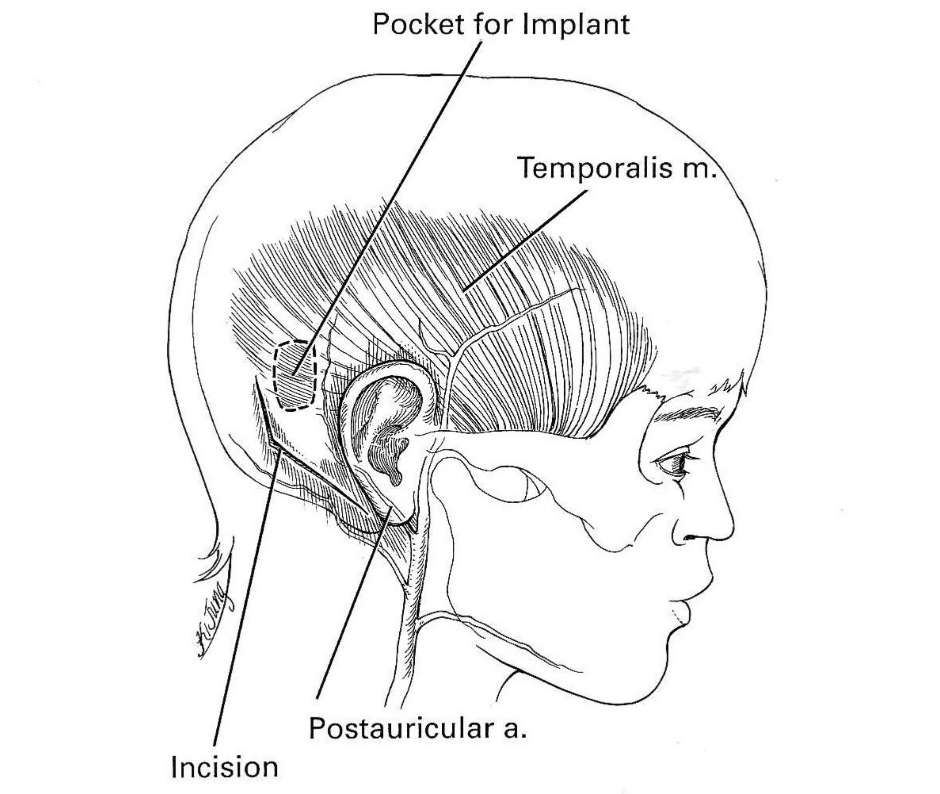


Typical incision lines

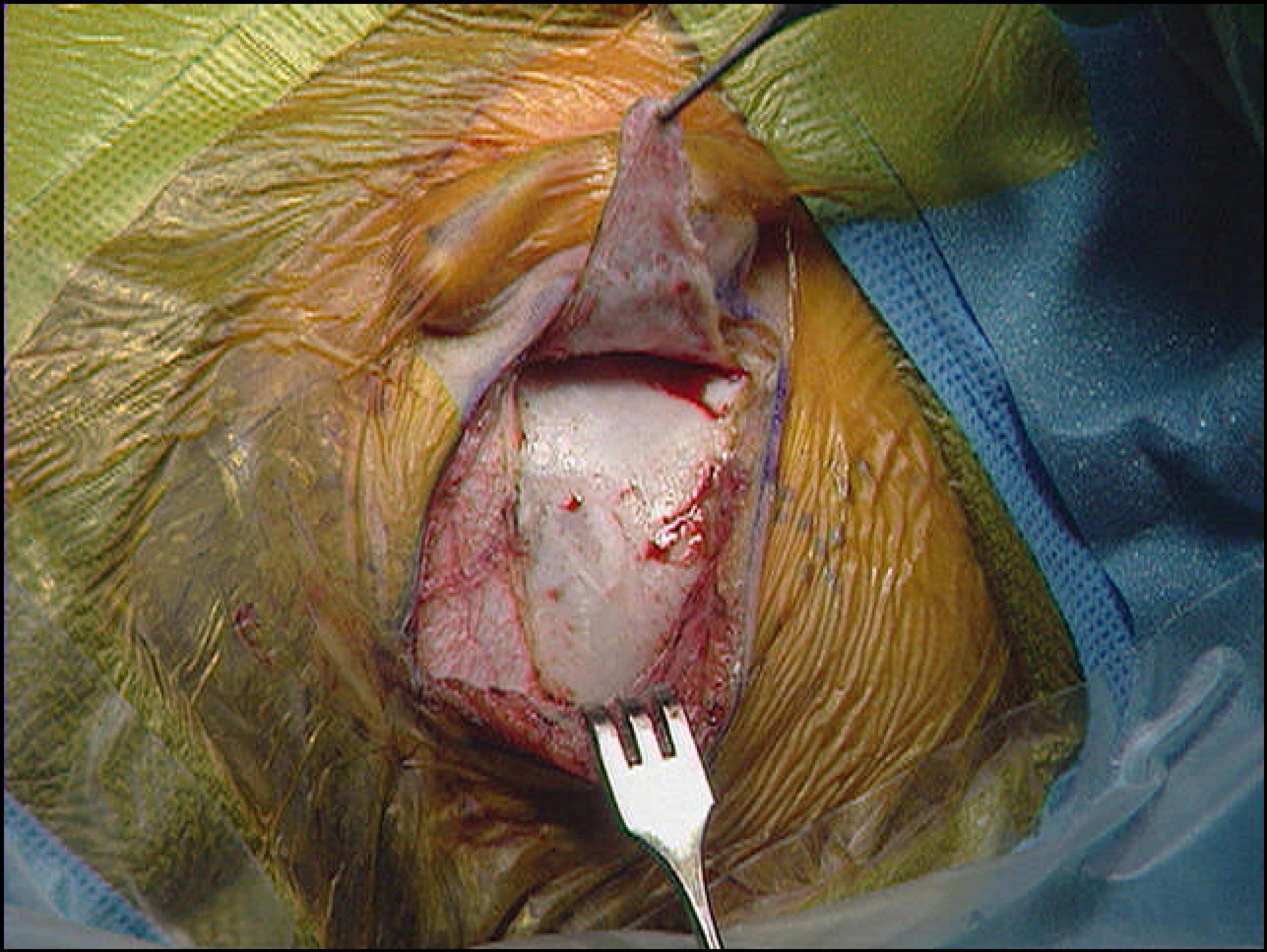


Surgical Planning

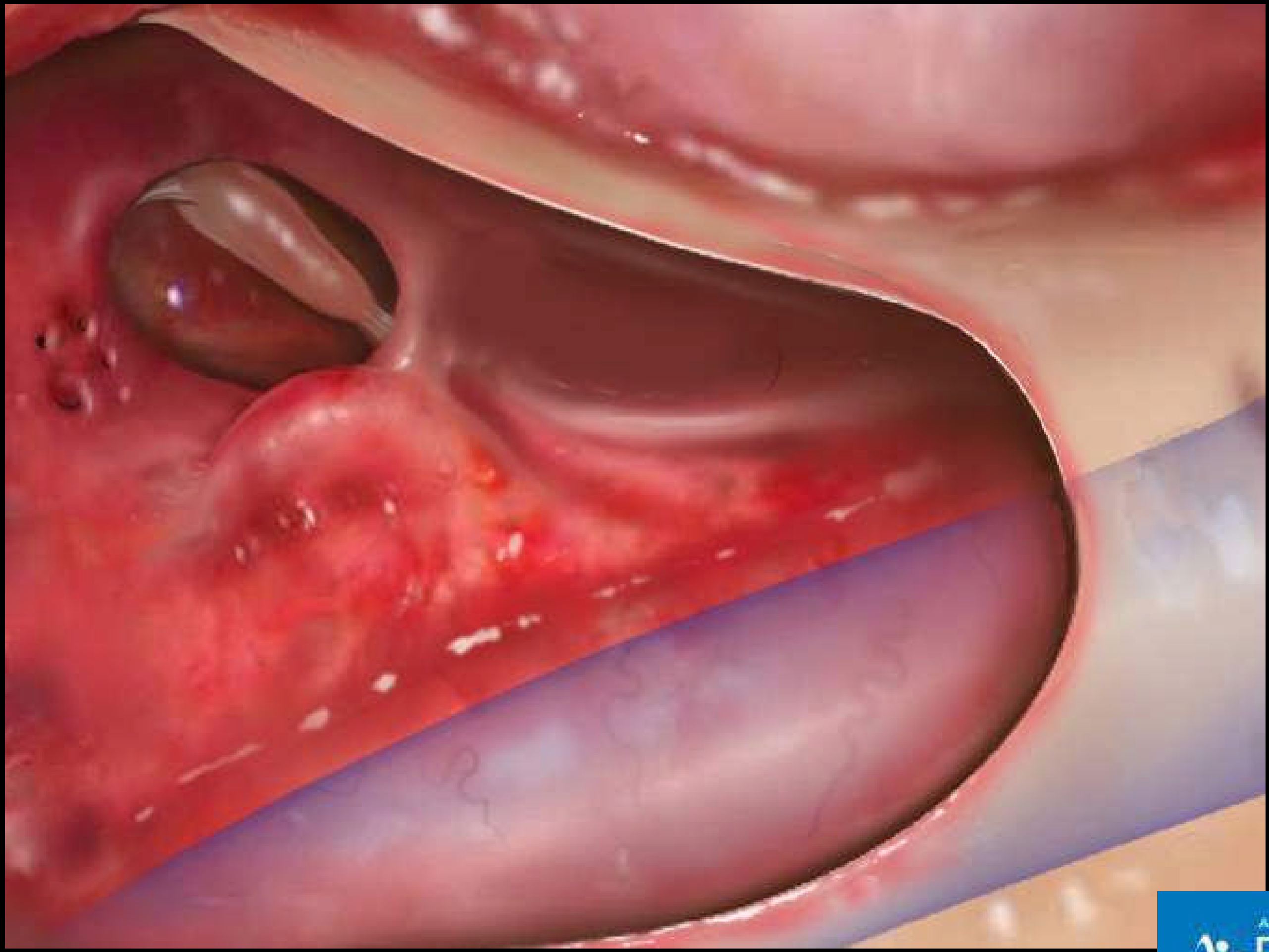
Incisions



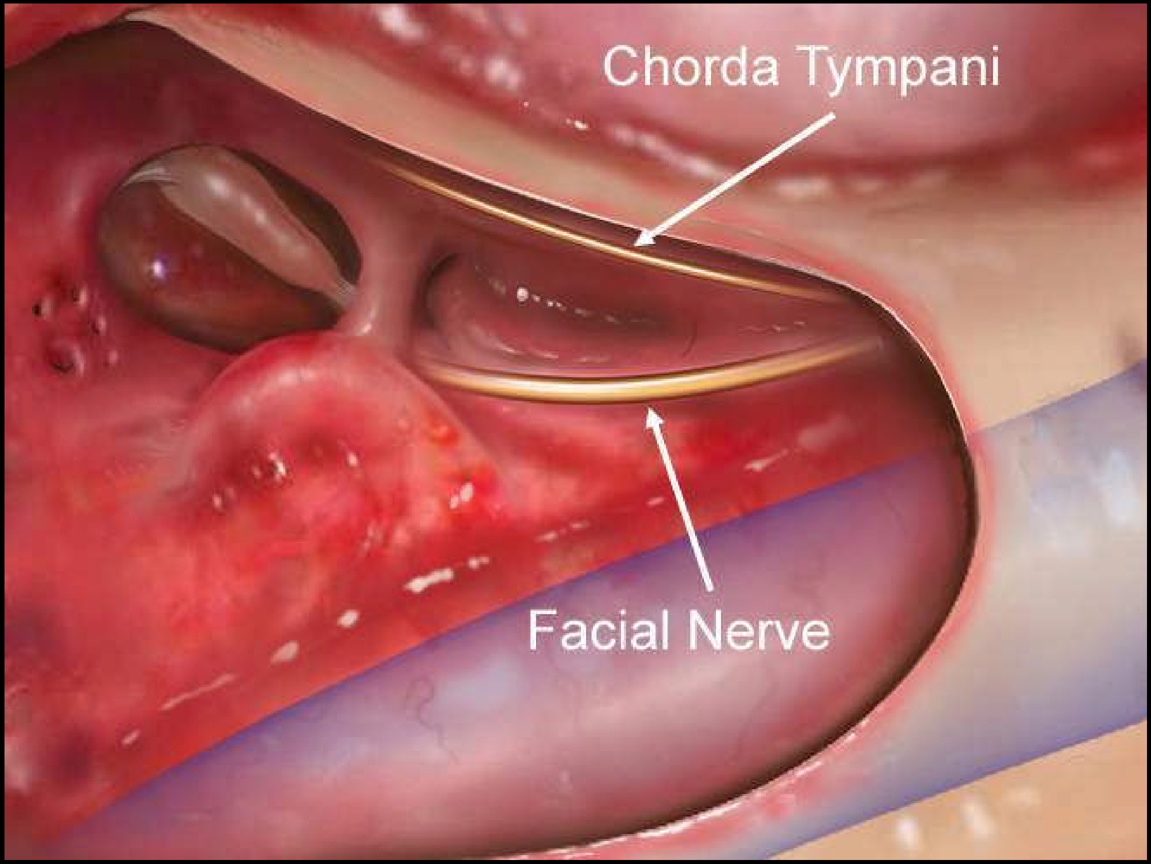
Surgical Dissection



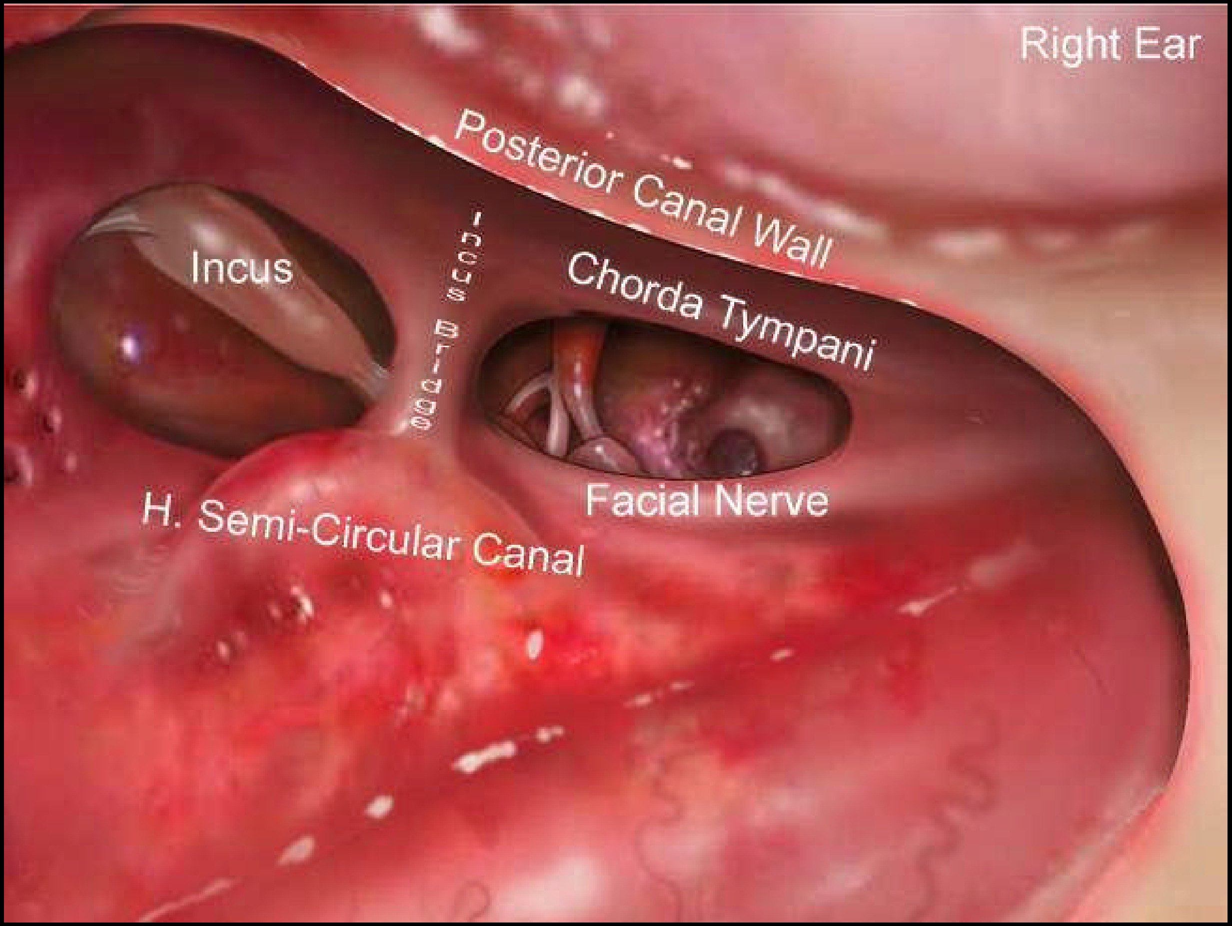
Completed mastoidectomy



2. Develop the facial recess

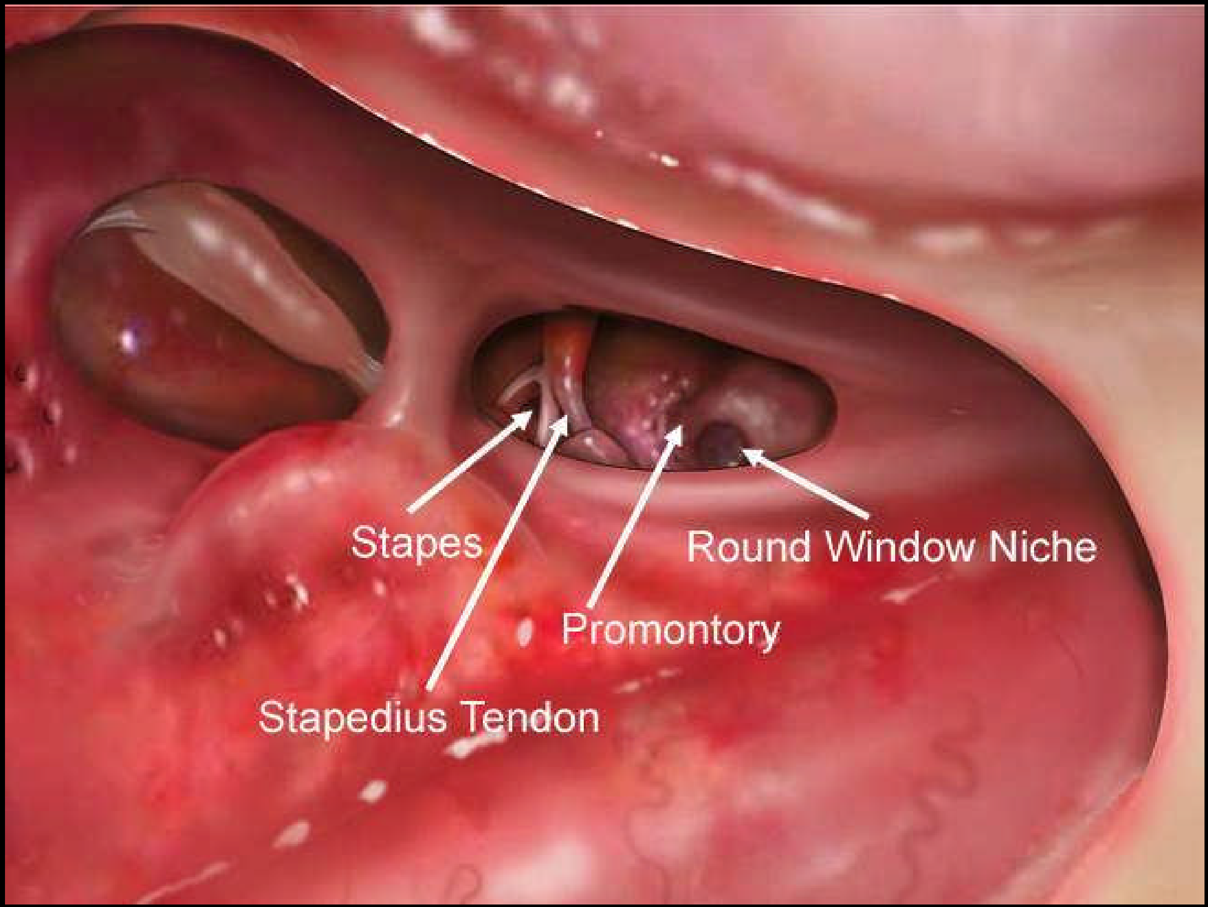


Right Ear Facial Recess



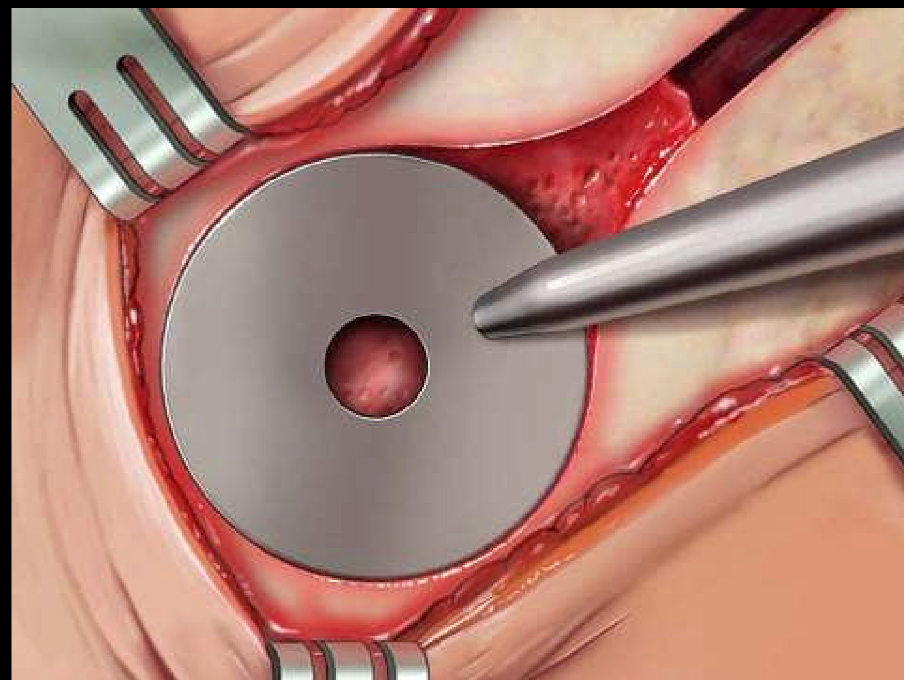
Develop the facial recess

The middle ear seen through the facial recess.

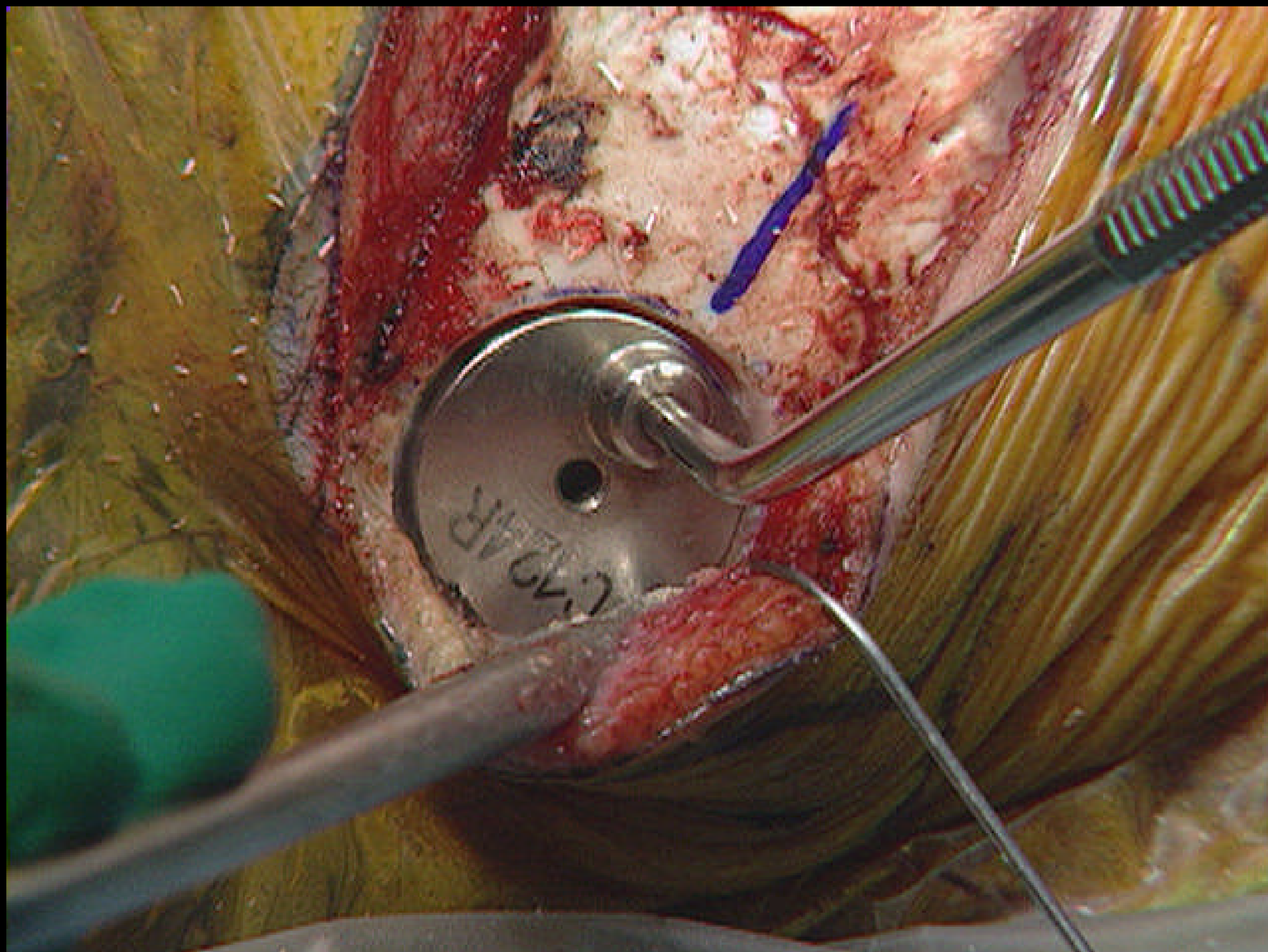


Drill the bony well

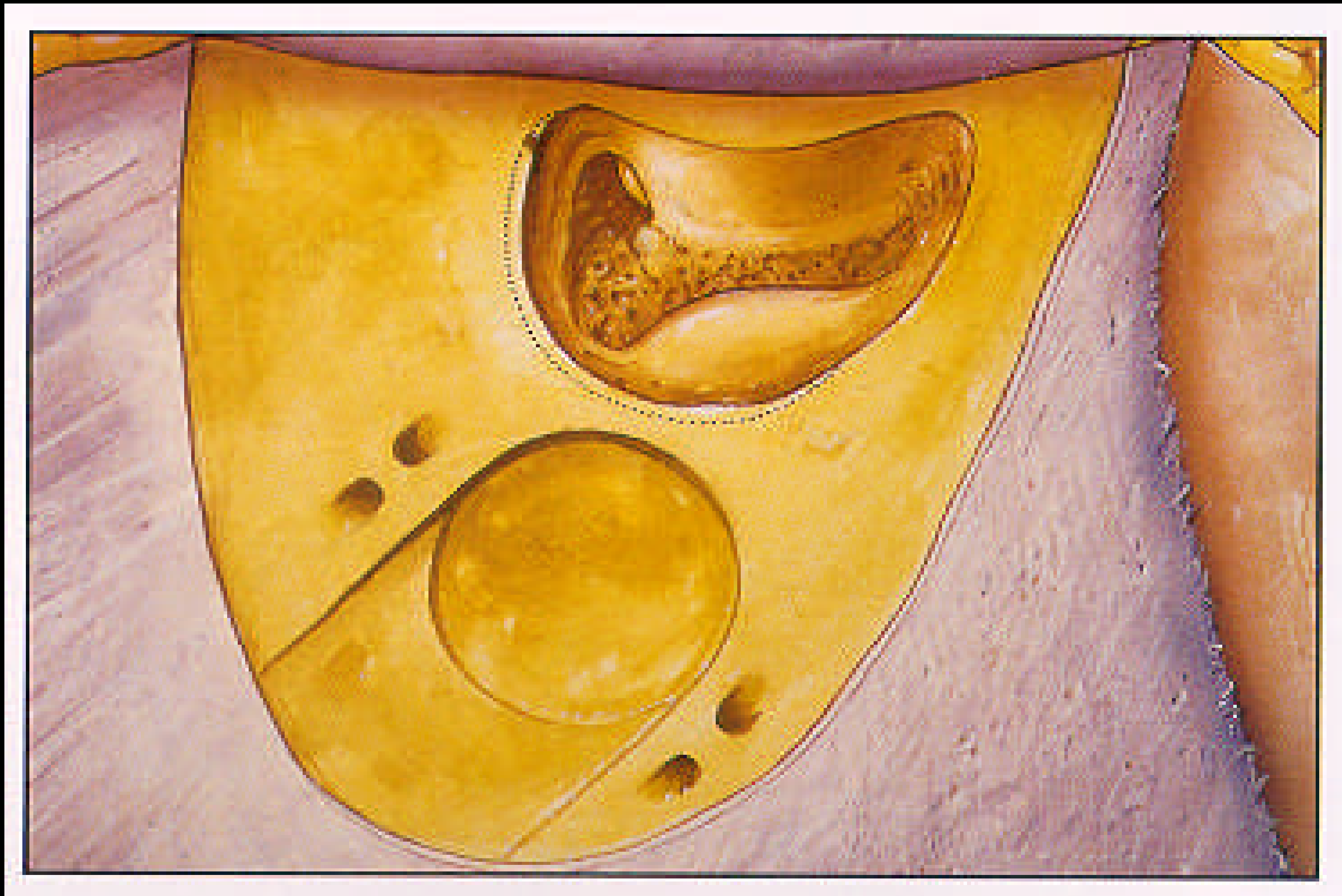
Use the recess gauge to check the size of the bone bed. The gauge should fit loosely into the well.



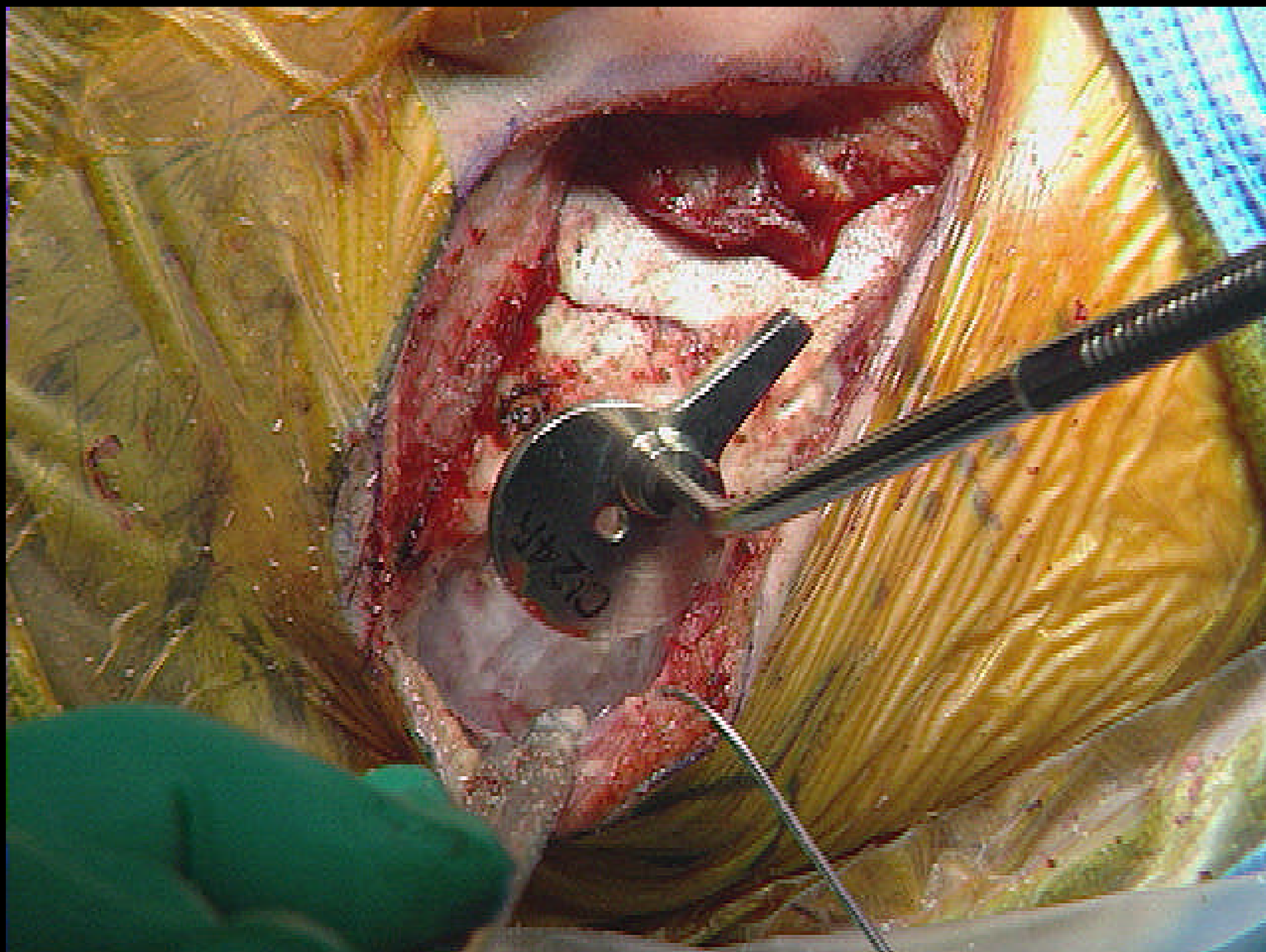
Marking the Well



Receiver-Stimulator Well Bed



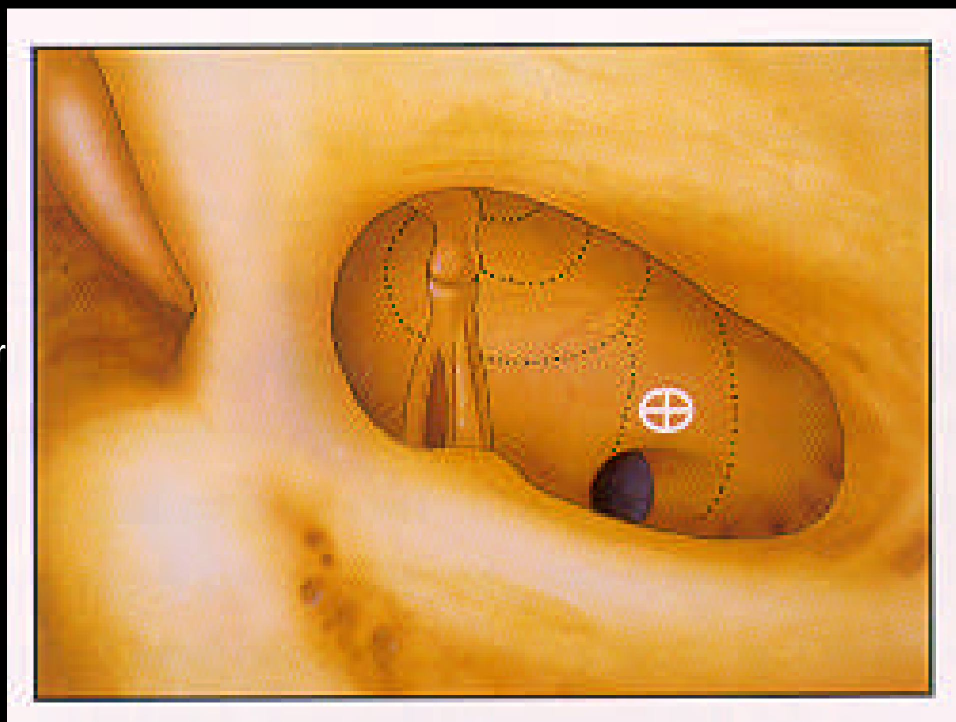
Positioning the Array



Cochleostomy

Anterior (nose)

Superior (head)



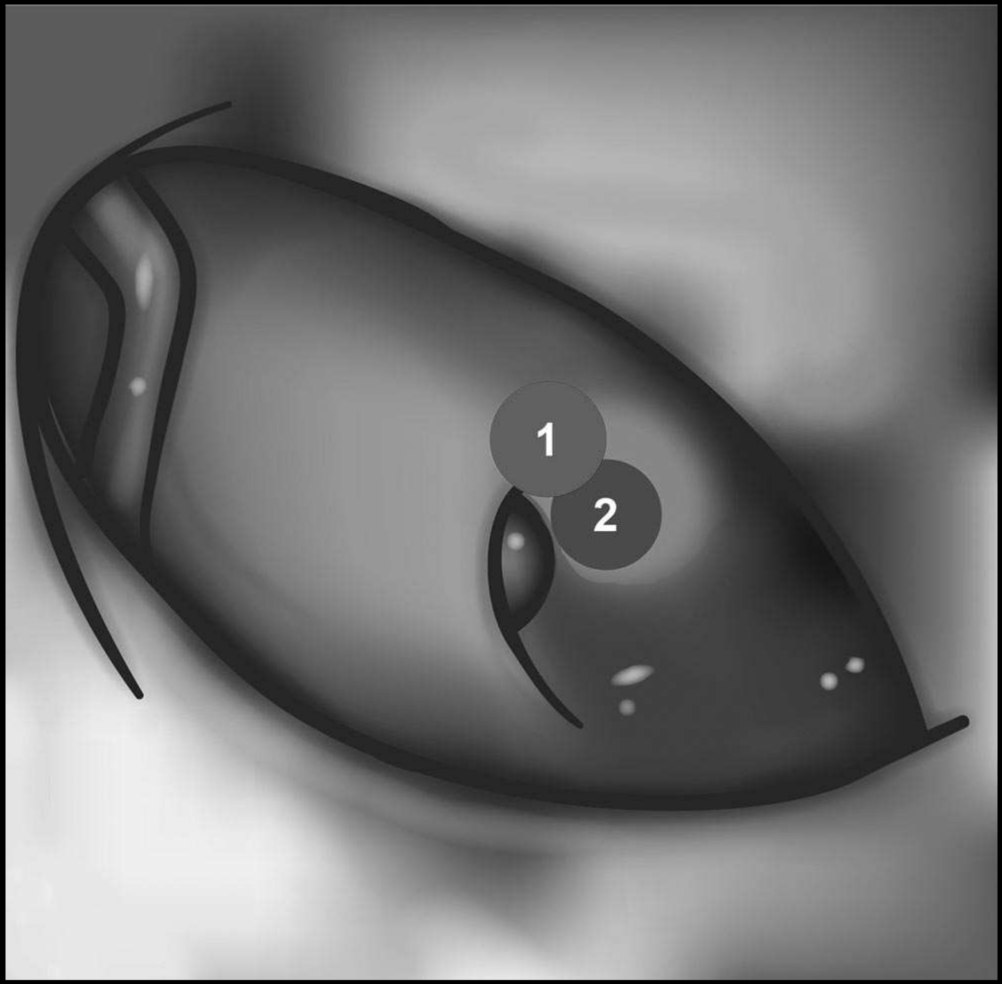
Inferior (feet)

Posterior (table)

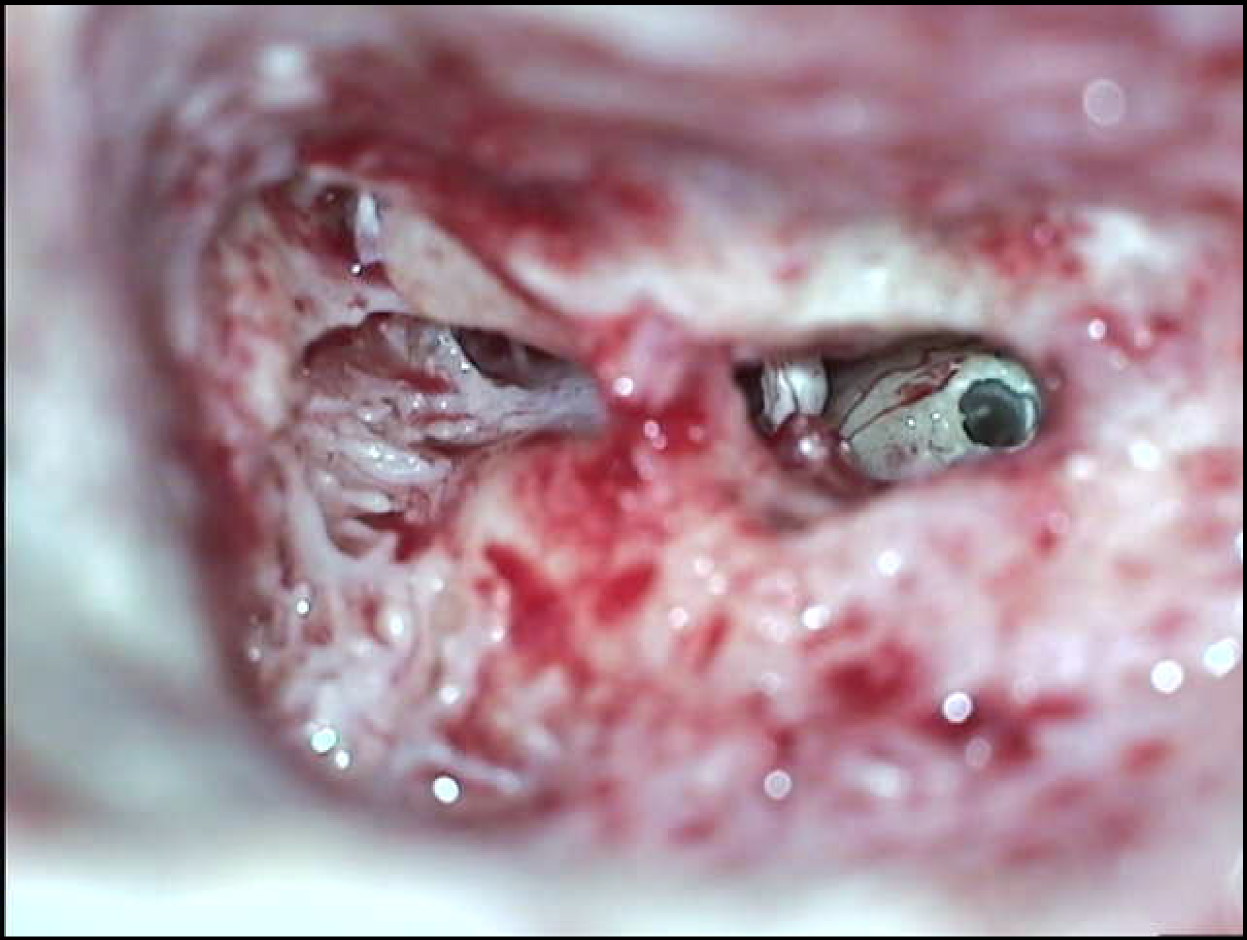
Right Ear

- The cochleostomy is created anterior and inferior to the round window. (1.5 and 1.0 diamond burrs.)

Cochleostomy

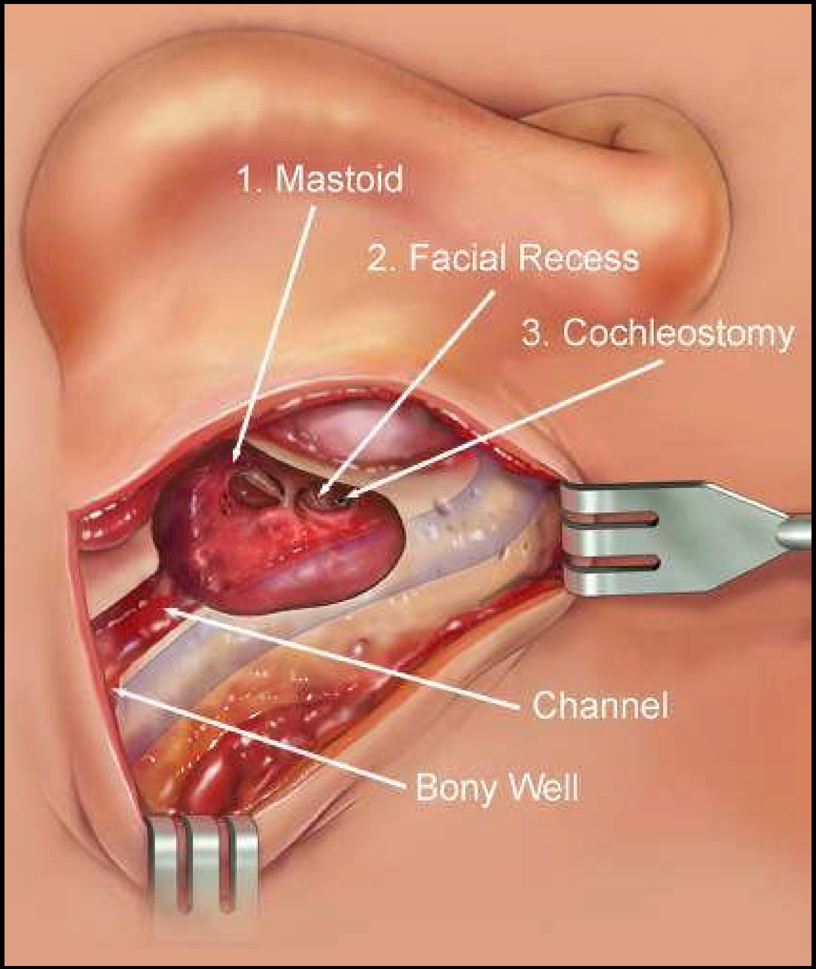


Cochleostomy



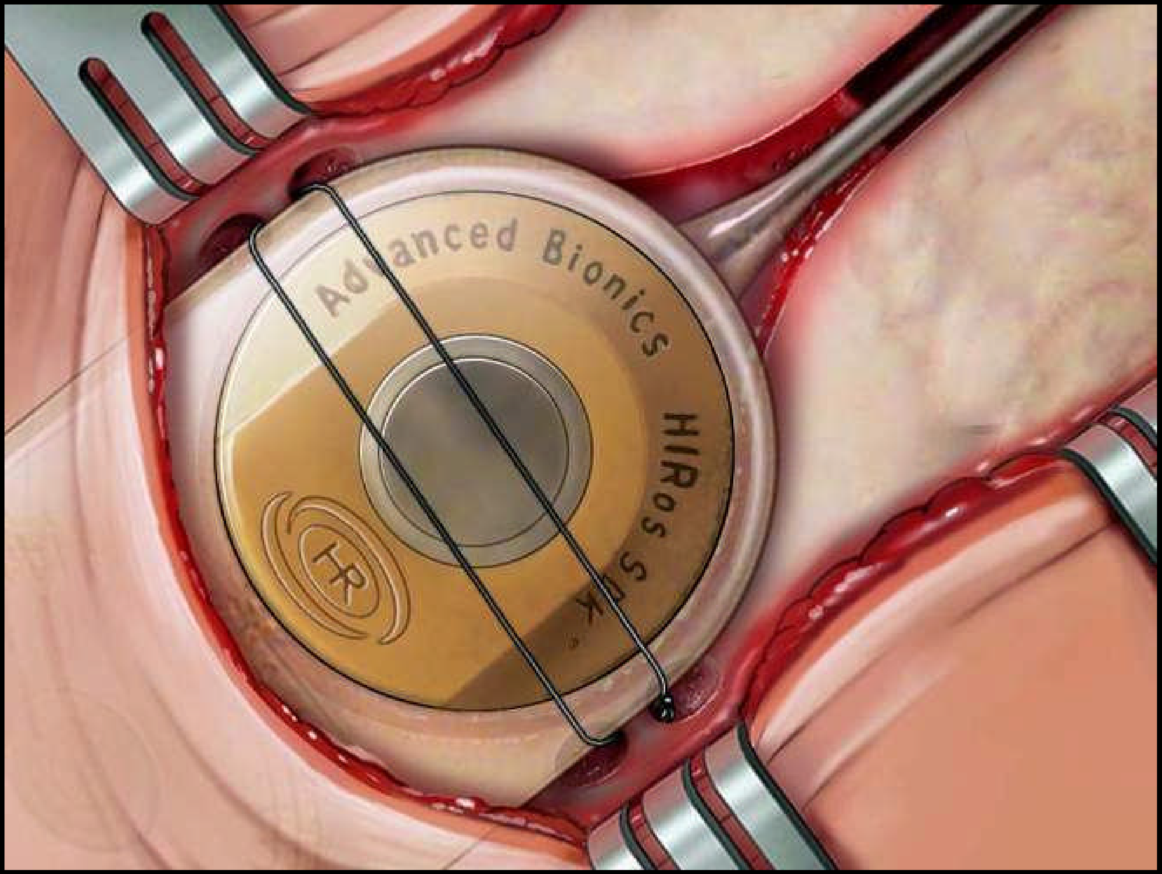
Right Ear

Cochlear implant surgery: three holes, a well and a channel



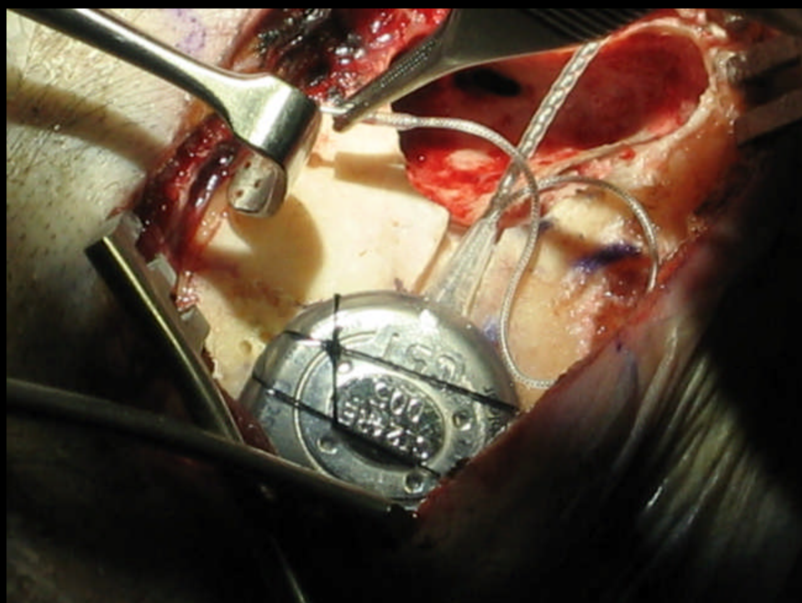
Verify placement of the implant

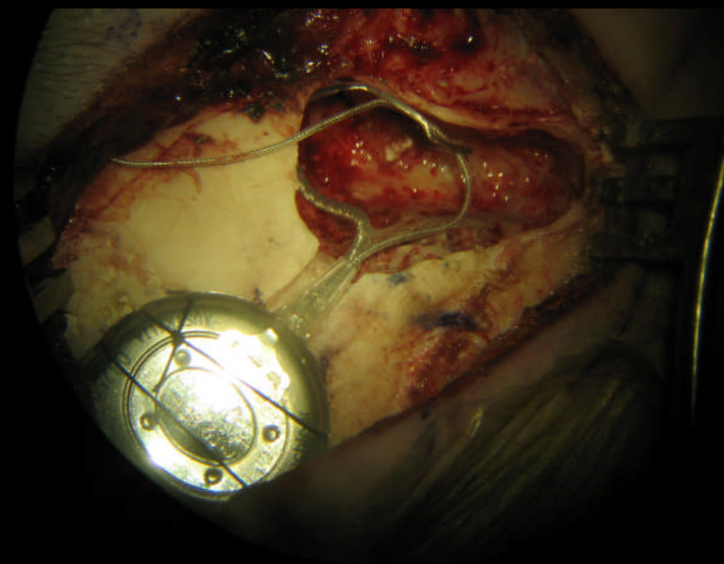
Ensure that the titanium case seats correctly and lies flat in the previously created bony well.
Place the suture knot at the side of the implant.



ADVANCED BIONICS

Securing the Implant

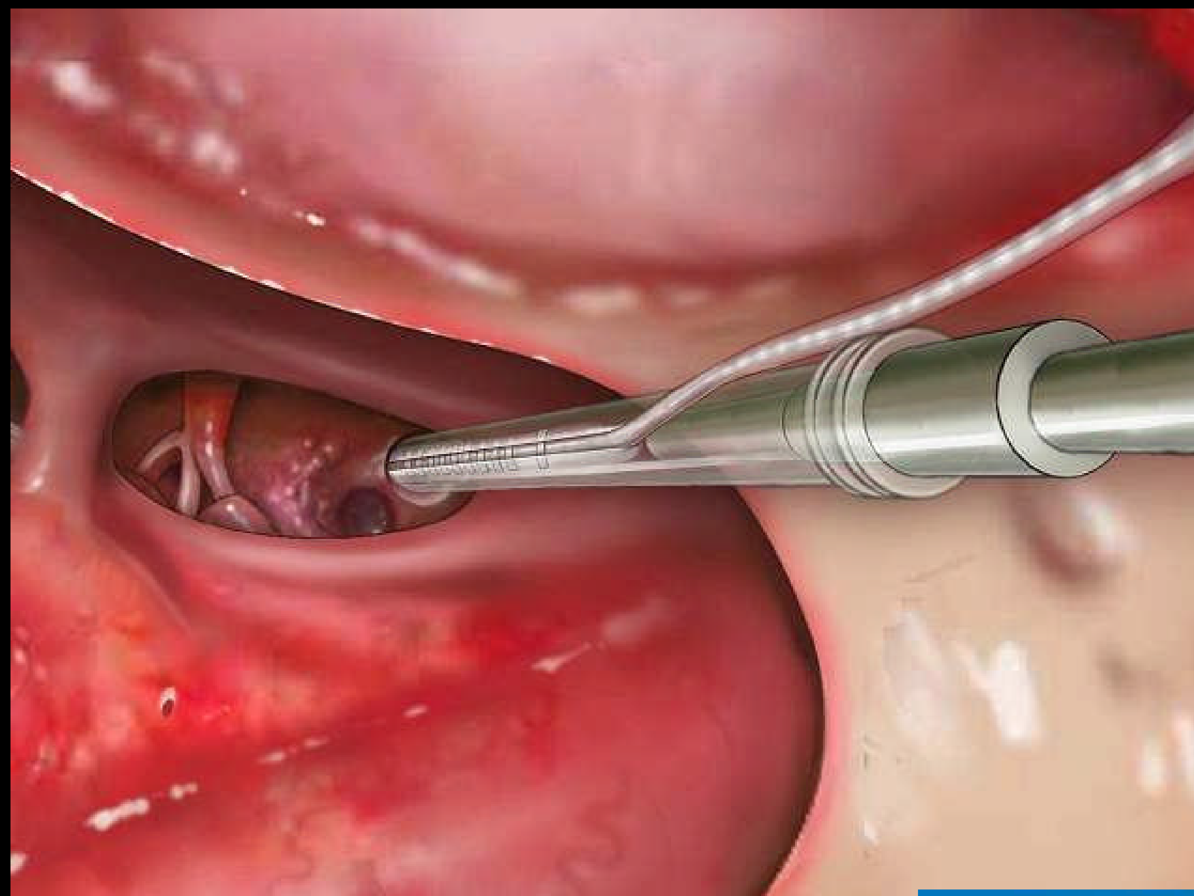


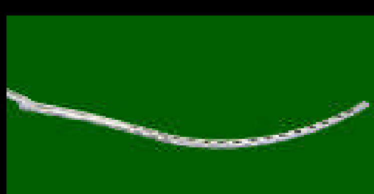


Electrode insertion

Place the insertion tube at the cochleostomy toward the basal turn of the scala tympani.

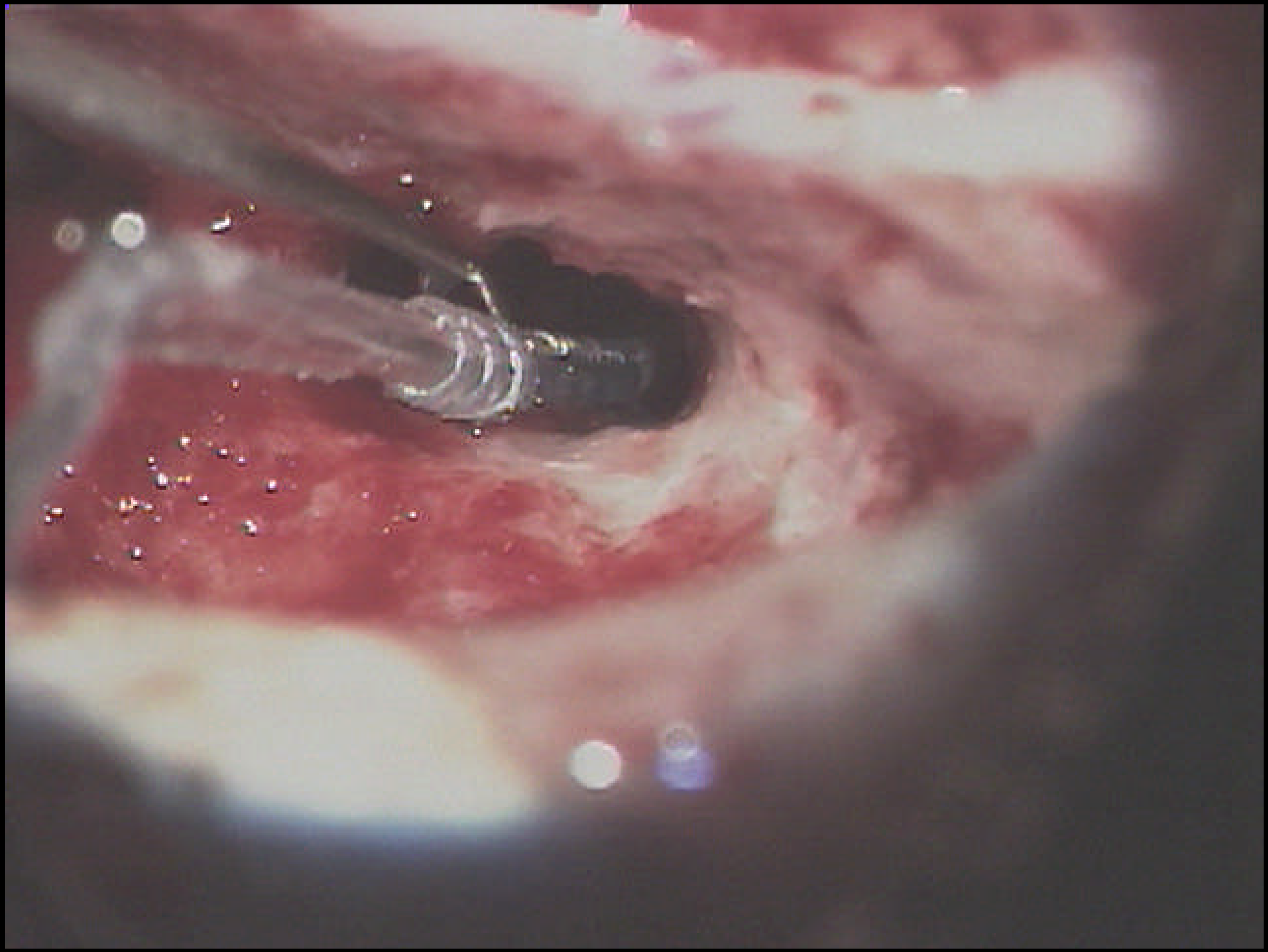
Direct insertion tube slot *superiorly* toward the modiolar (or inner) wall.



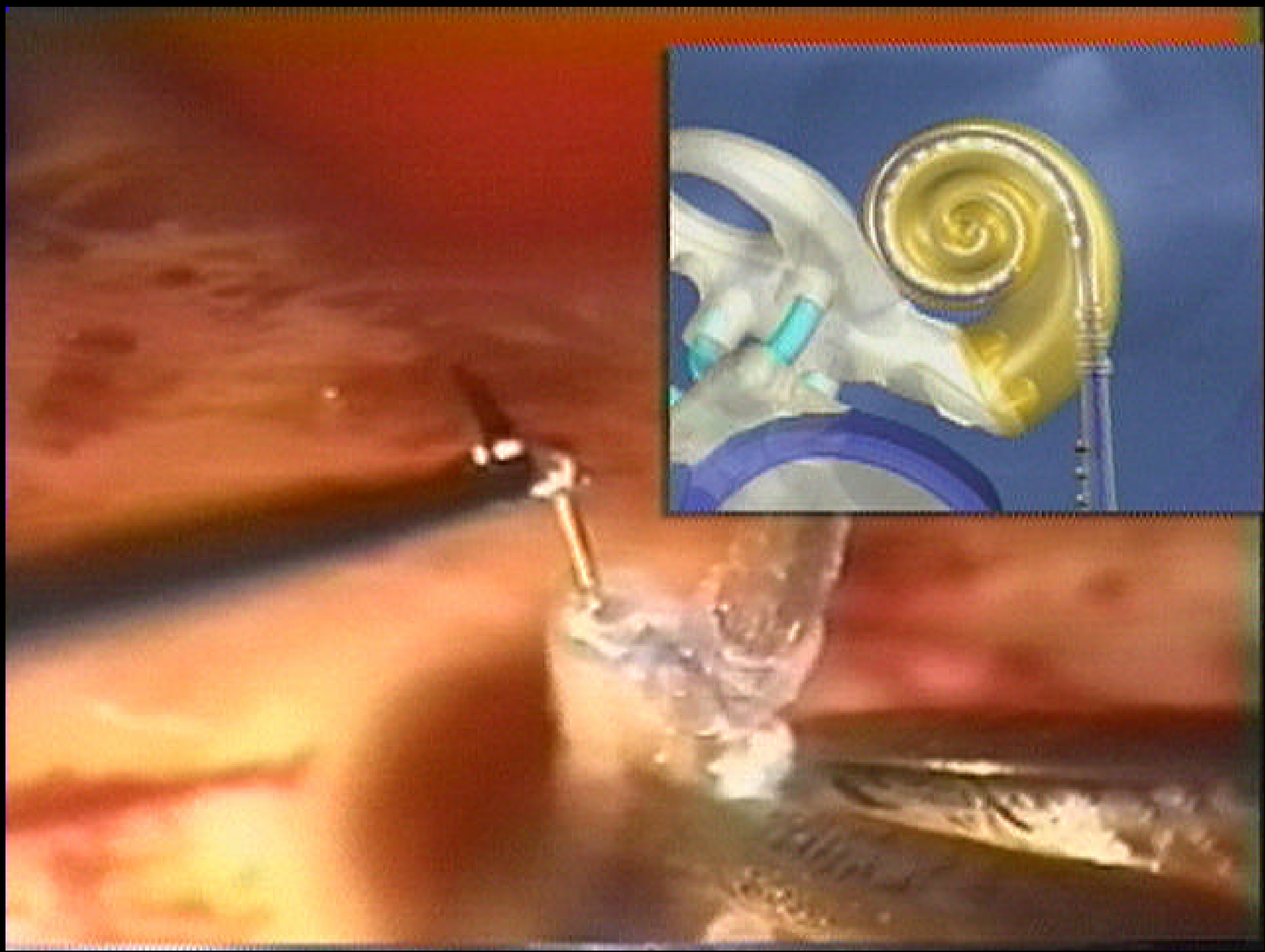


ADVANCED BIONICS

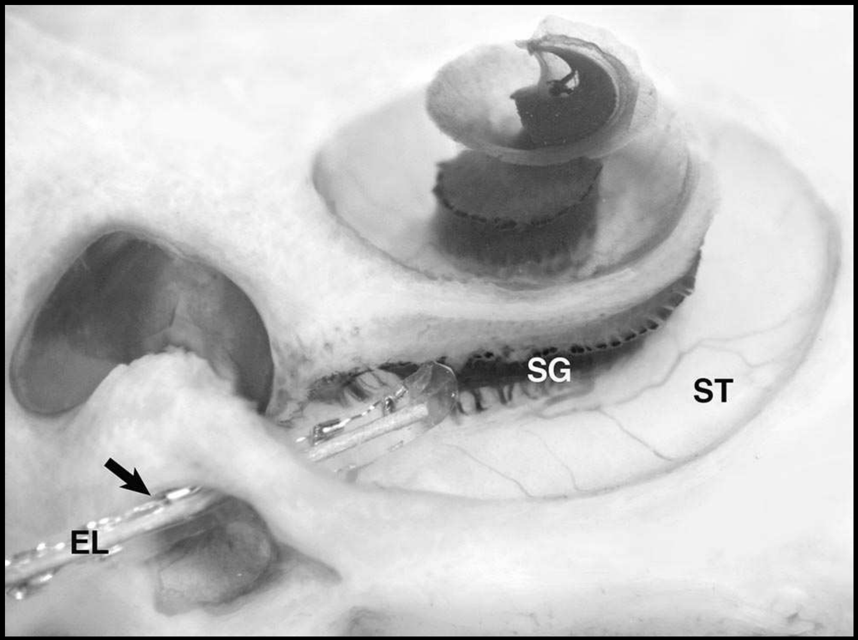
Inserting the Electrode



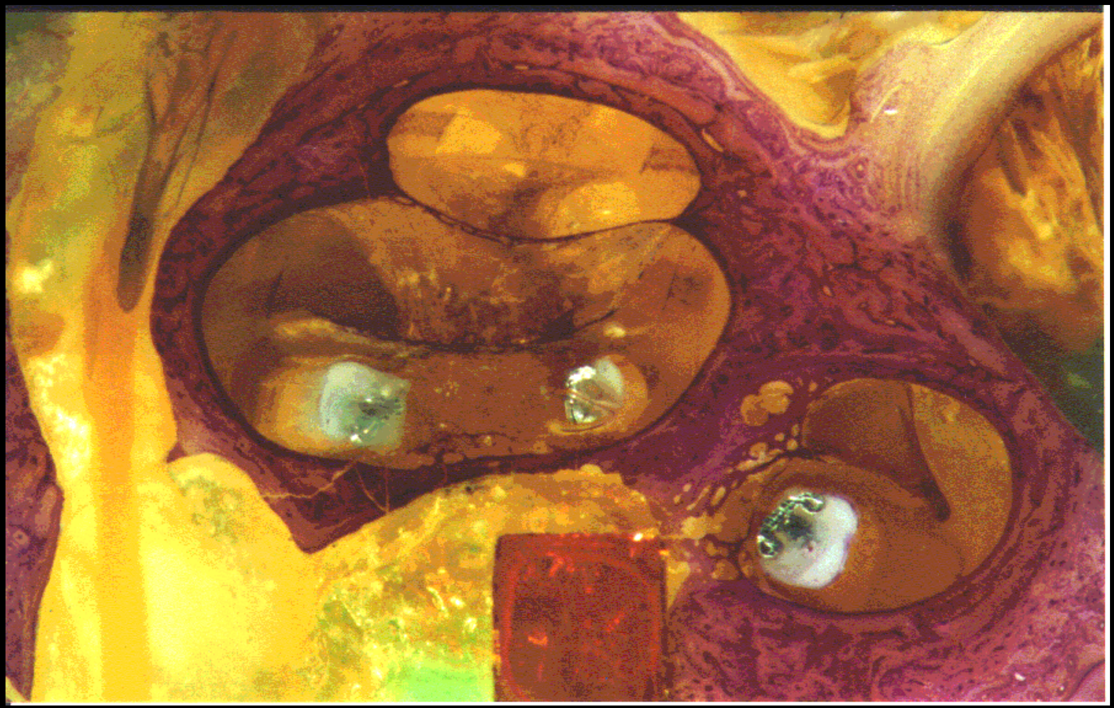
Stylet Withdrawal



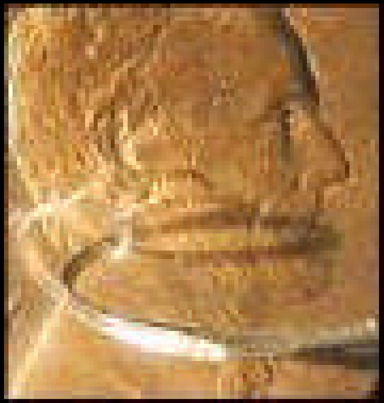
Electrode Insertion



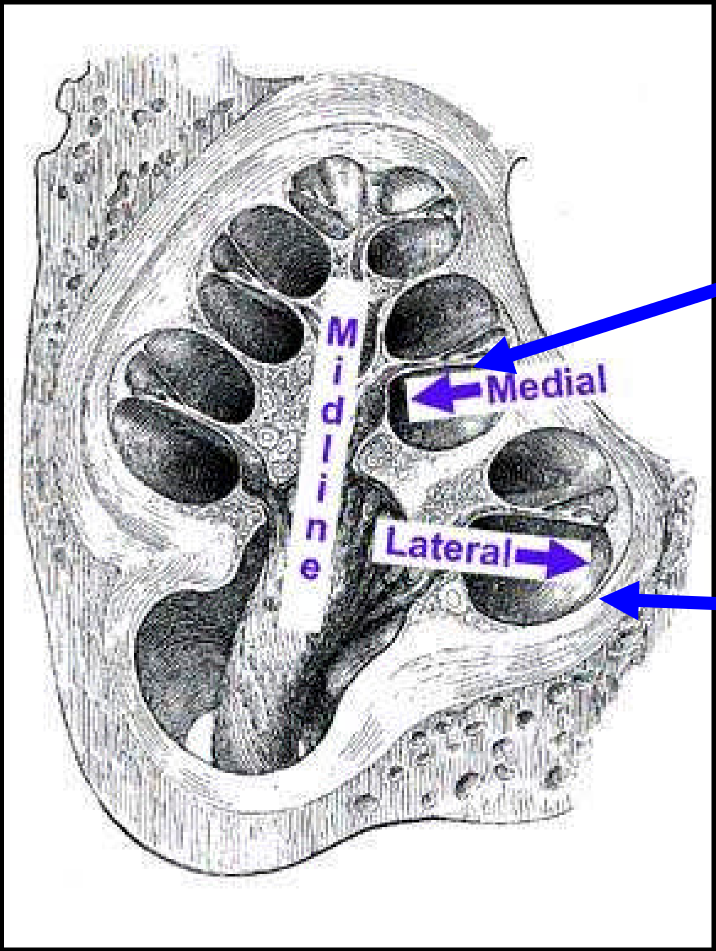
Contour Design

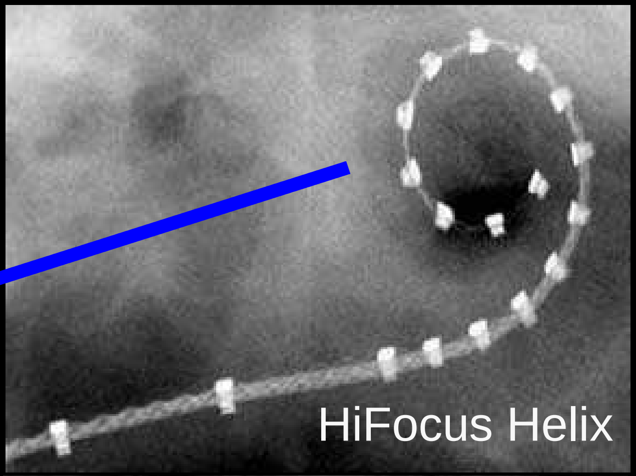


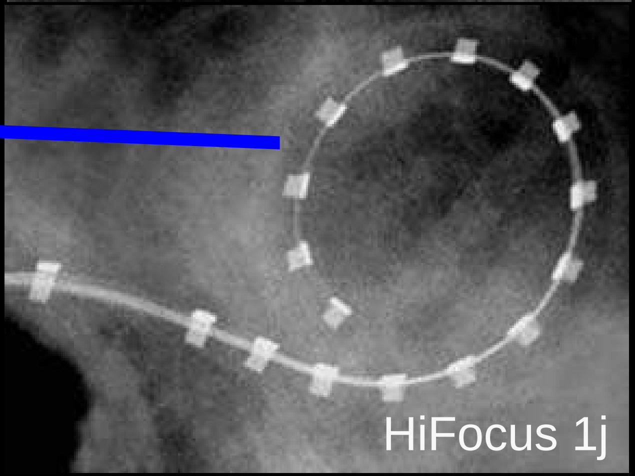
Occupies < ½ cross section of scala tympani



Confirm Placement

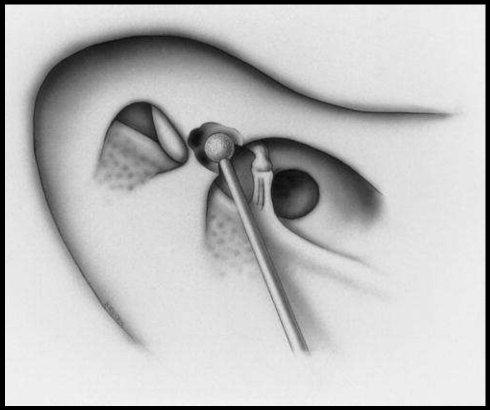


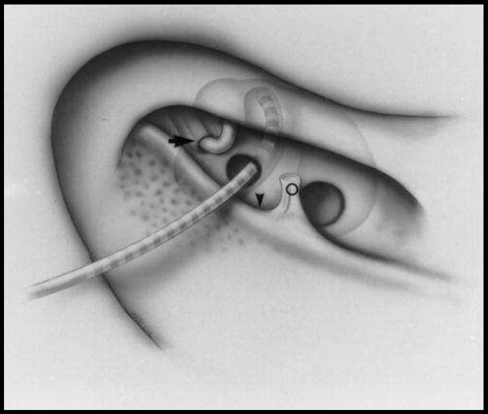


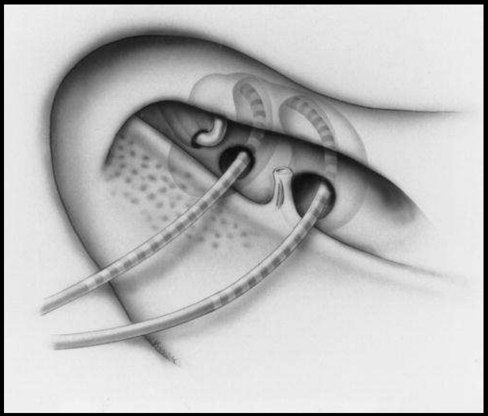


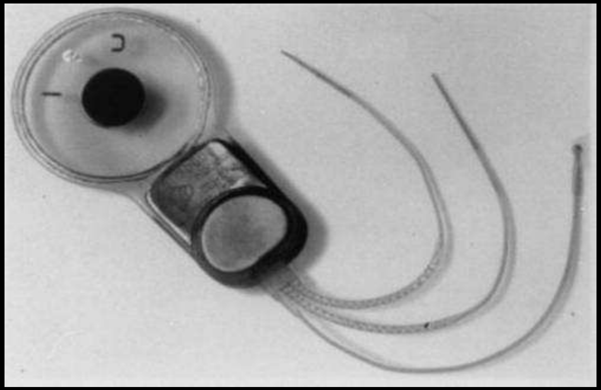
ADVANCED BIONICS

Double Array Implant

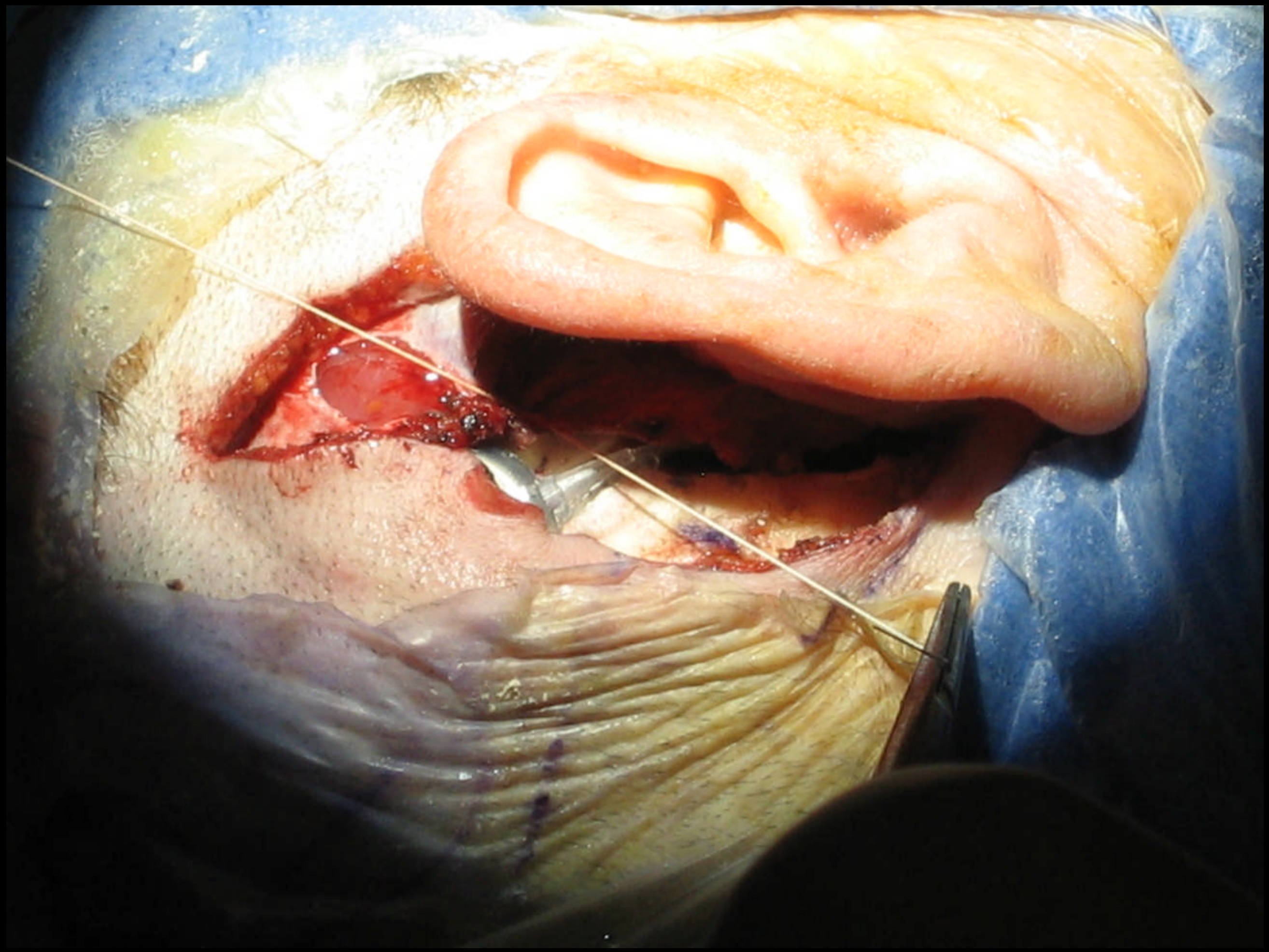




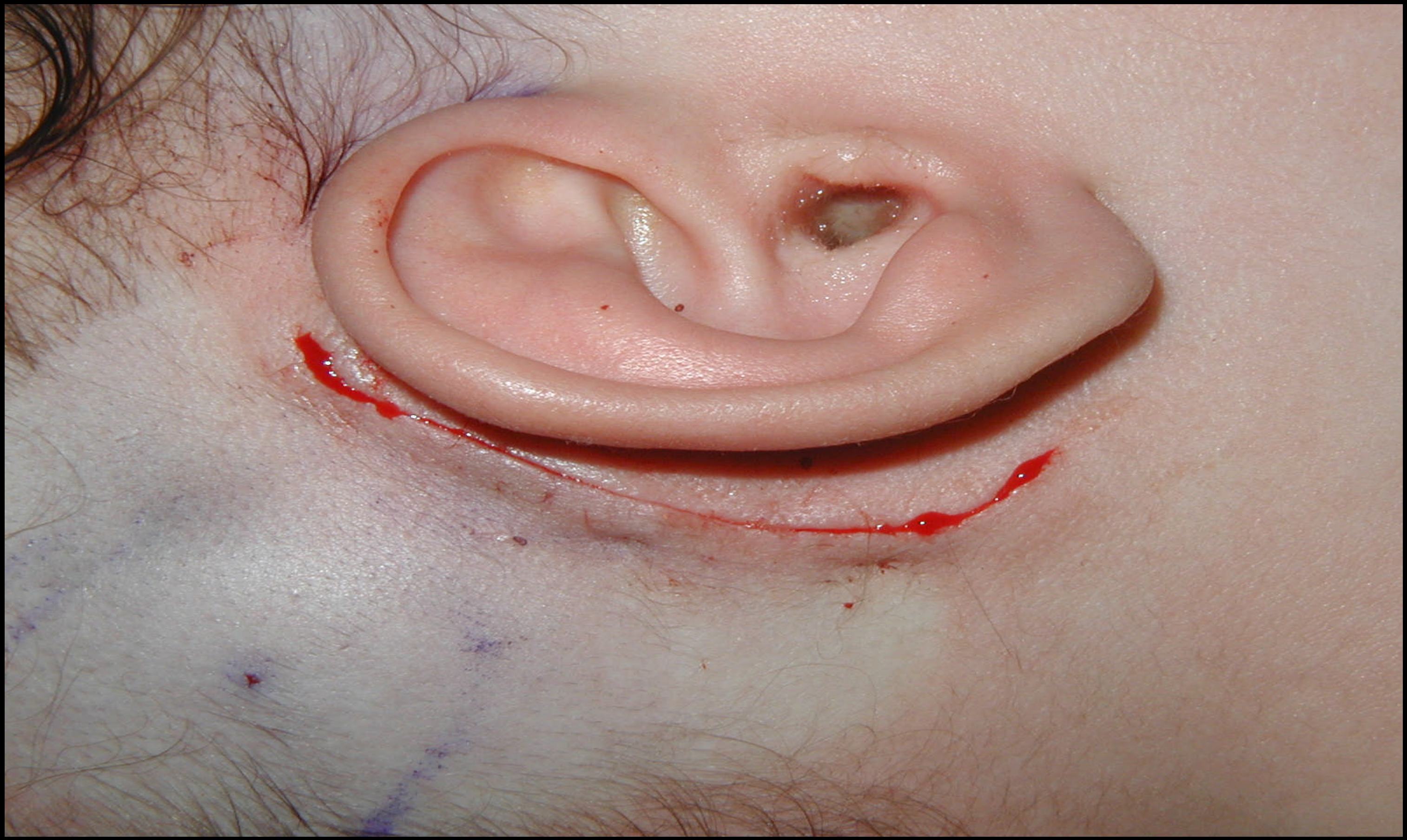




Closing with NRT

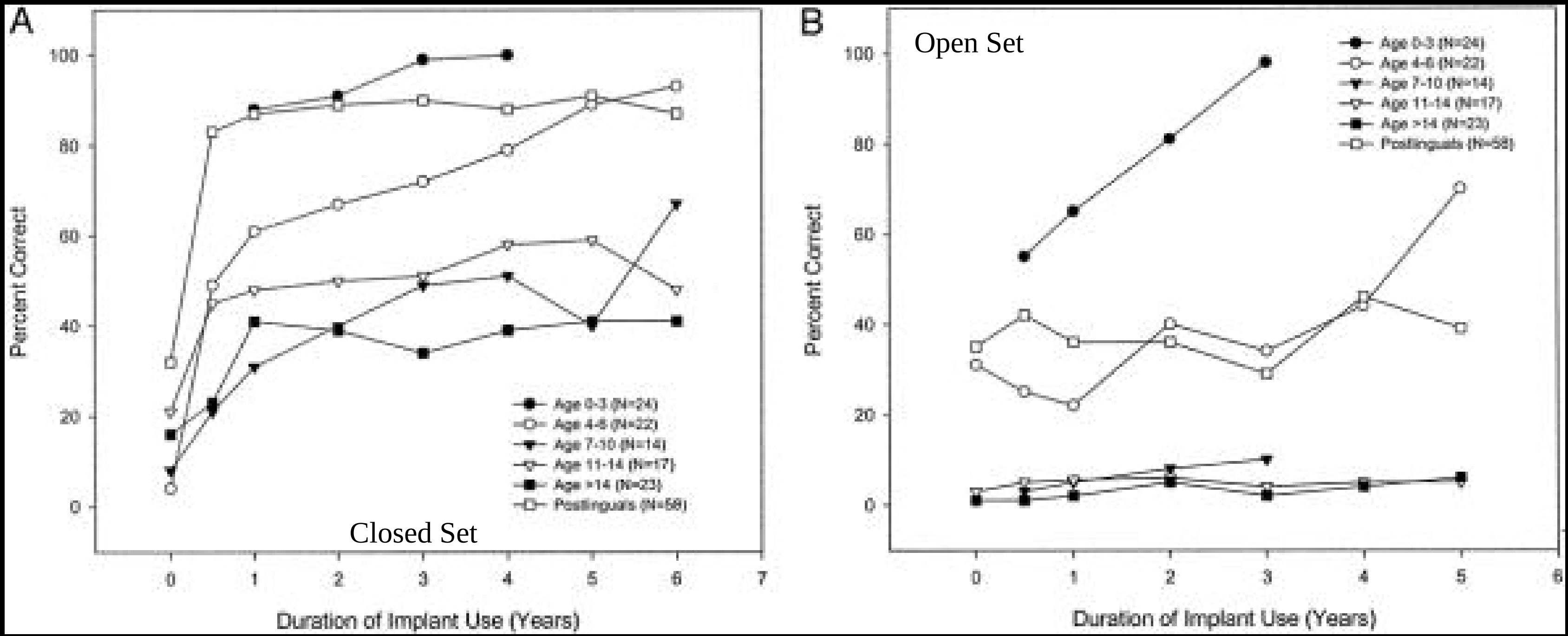


Incision closed



Results

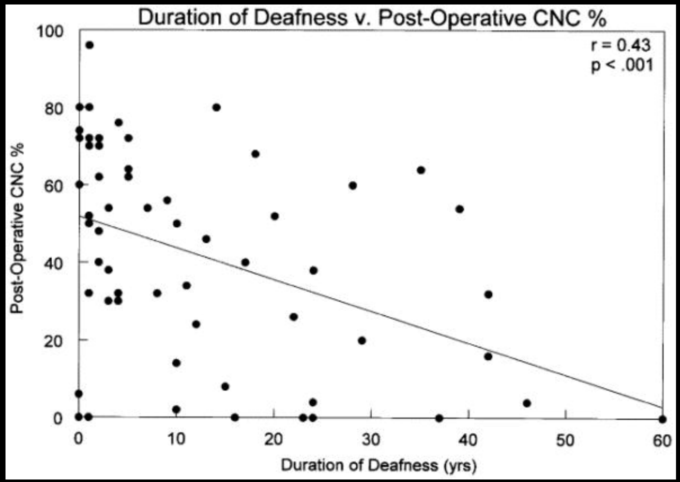
- Prelingually deafened children and Postlingually deafened adults

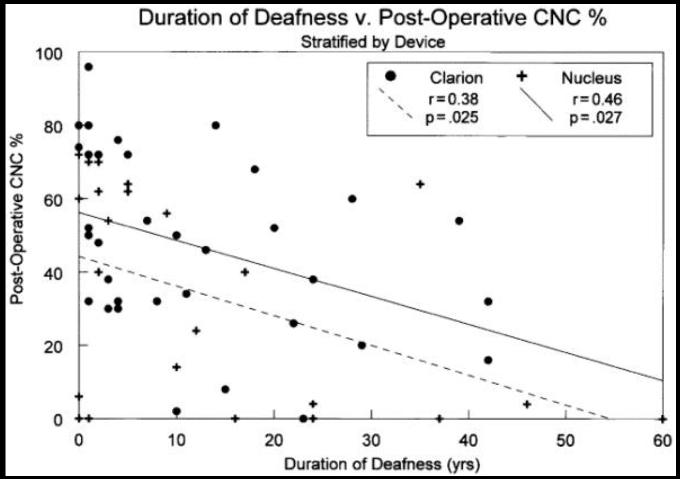


Wooi Teoh 2004

Duration of Deafness

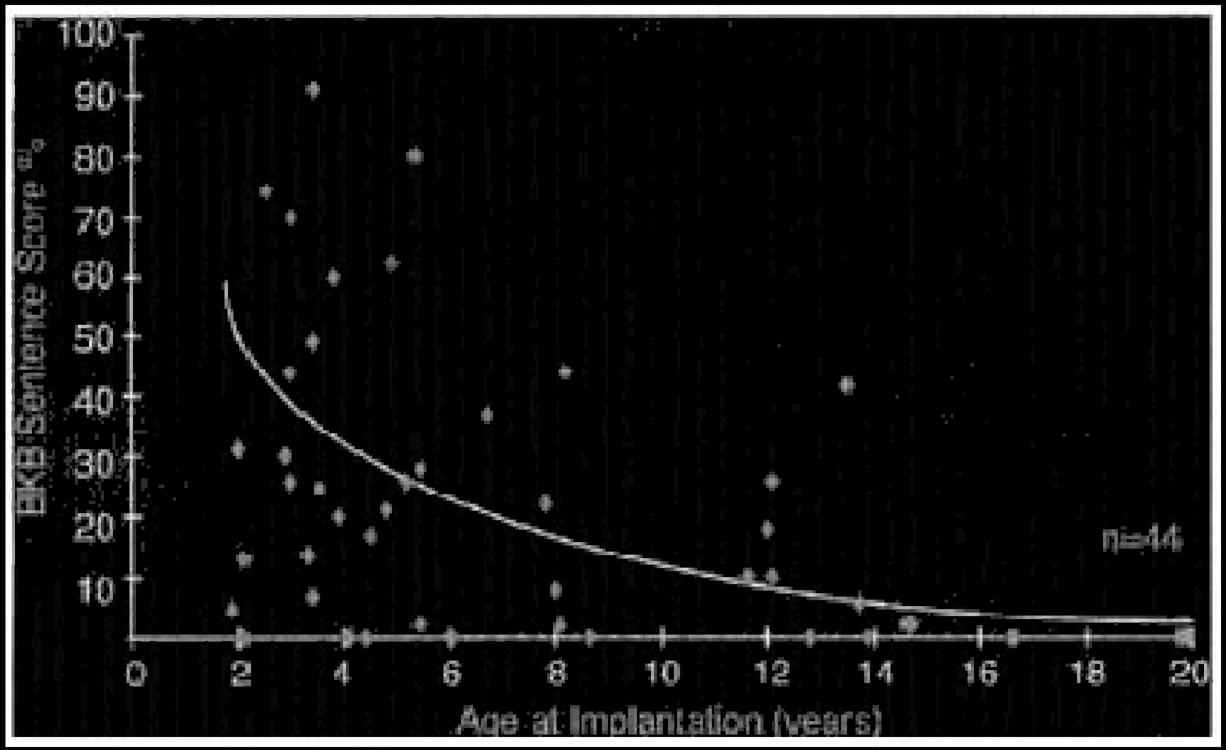
- Friedland 2003
- 58 postlingually deafened adults





Results

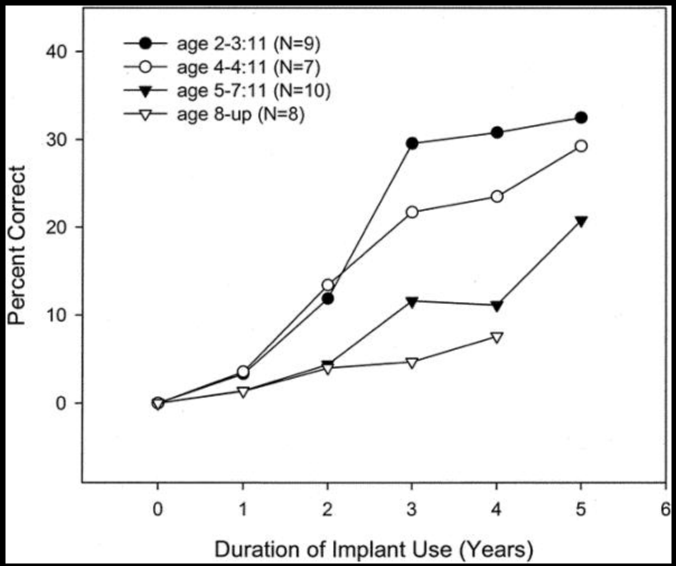
- Earlier implantation appears to provide better results
- Open Set Prelingual Deafness

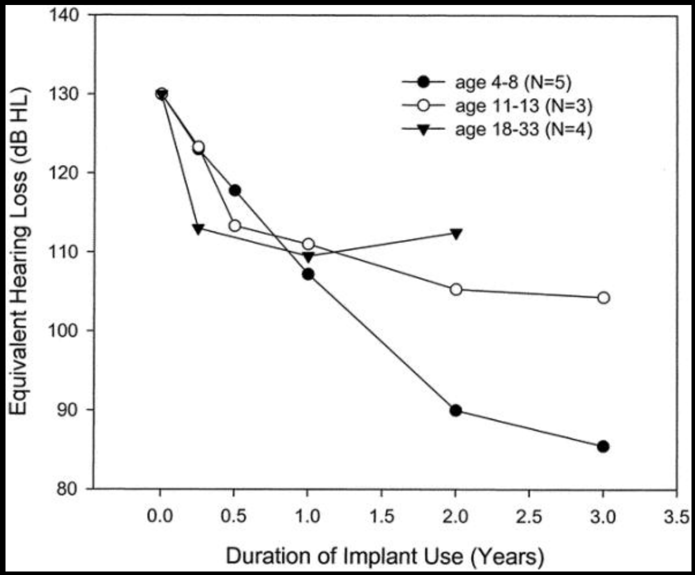


Clark 1999

Age at Implant

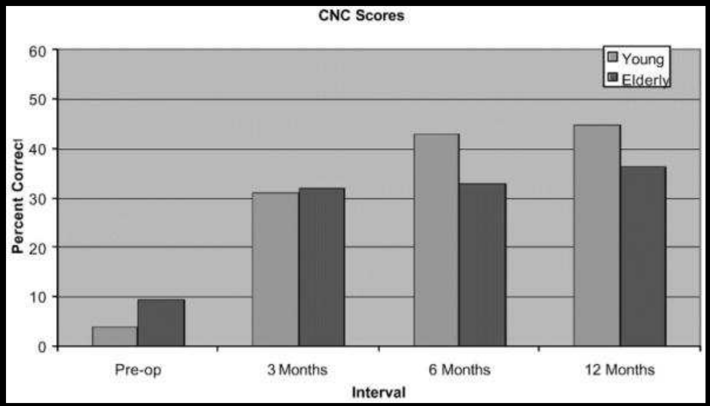
- Wooi Teoh 2004
- Prelingually deafened children
- PB-K test scores
  - Open set word recognition
- Significant differences after age 5
- Only minimal results after age 8

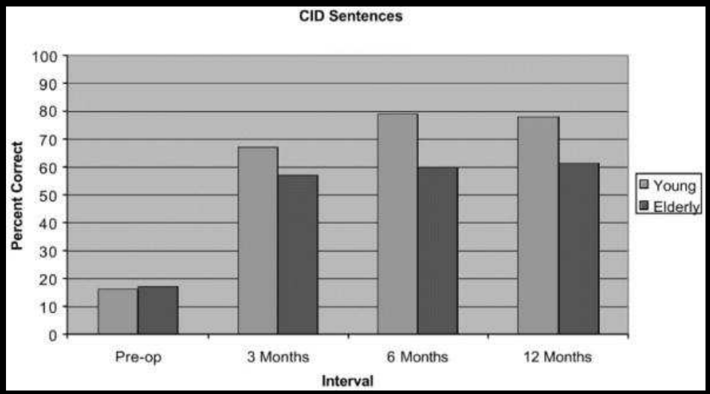


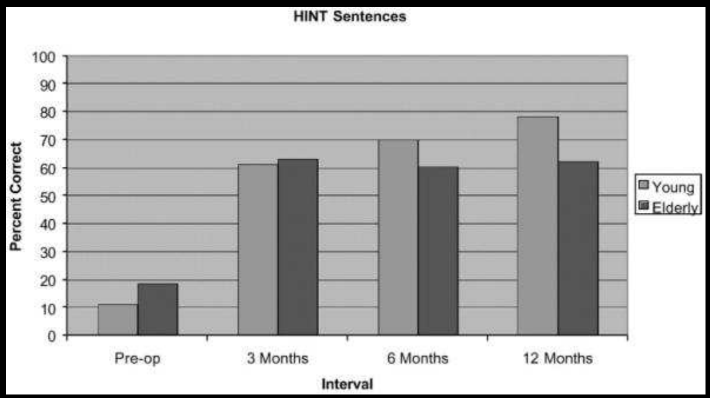


Adult Implants

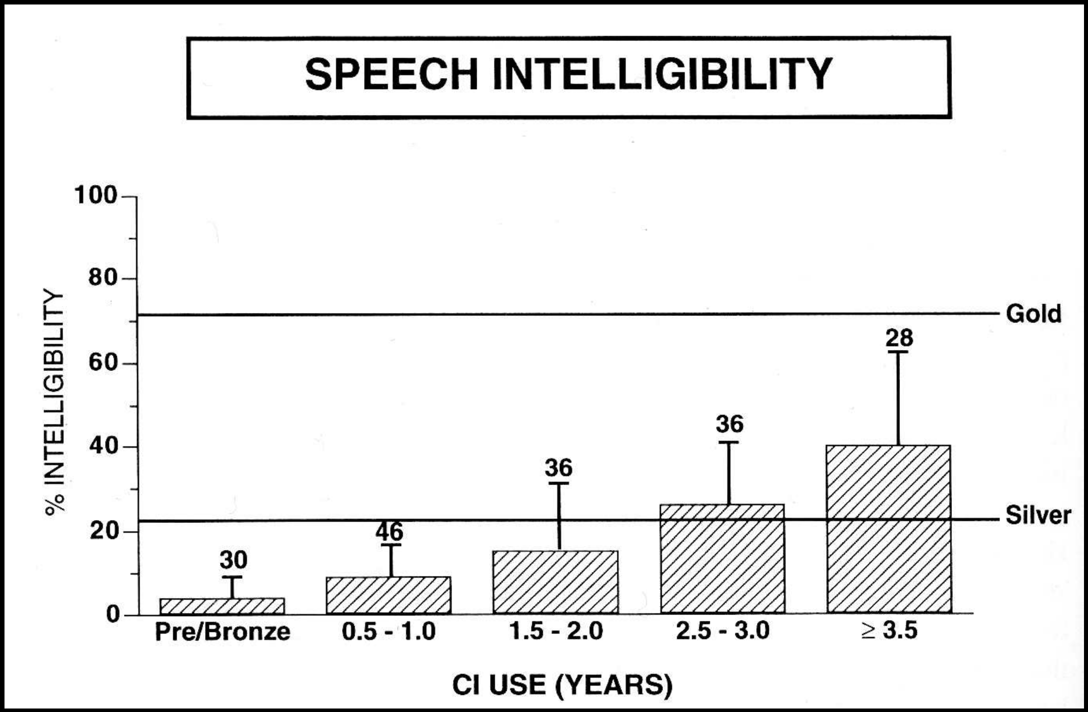
- Chatelin 2004
  - 65 adults >70
  - 101 adults <70
- Significant differences in performance
- Reasons for difference
  - Age related loss of spiral ganglion cell
  - "Central Presbyacusis"
  - General cognitive deficits impede rehabilitation







Non Hearing Benefits



Bilateral Implants

- Bilateral implantation ensures implantation of the ear with the best performance
- There is unanimous support for binaural amplification in the literature
  - Benefits of normal binaural hearing may include
    - Sound localization
    - Improved understanding in background noise
    - Improved understanding in quiet due to increase in perceived intensity

Bilateral Implants – Acoustic Principles

- Binaural Squelch
  - Definition: Squelch excludes undesired lower-power input signals that may be present at or near the frequency of the desired signal
  - Auditory brainstem nuclei process timing, amplitude and spectral differences between ears for clearer separation of signal and noise
  - A combination of CNS and brainstem function to provide a better central representation of the sound

Bilateral Implants – Acoustic Principles

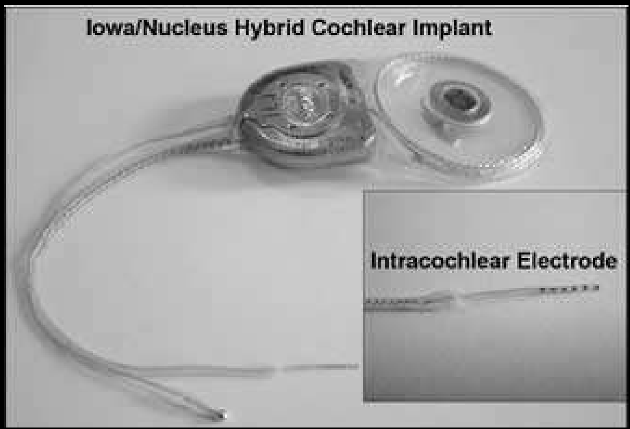
- Shadow Effect
  - There is always one ear with a more favorable SNR depending on the location of the signal and the background noise
  - The attenuation of the skull contributes to attenuation of the background noise to one ear
  - The CNS can choose to “attend” to the better performing ear
  - Unilateral hearing will always try to turn there head to the best position missing some direction of sound

Bilateral Implants – Acoustic Principles

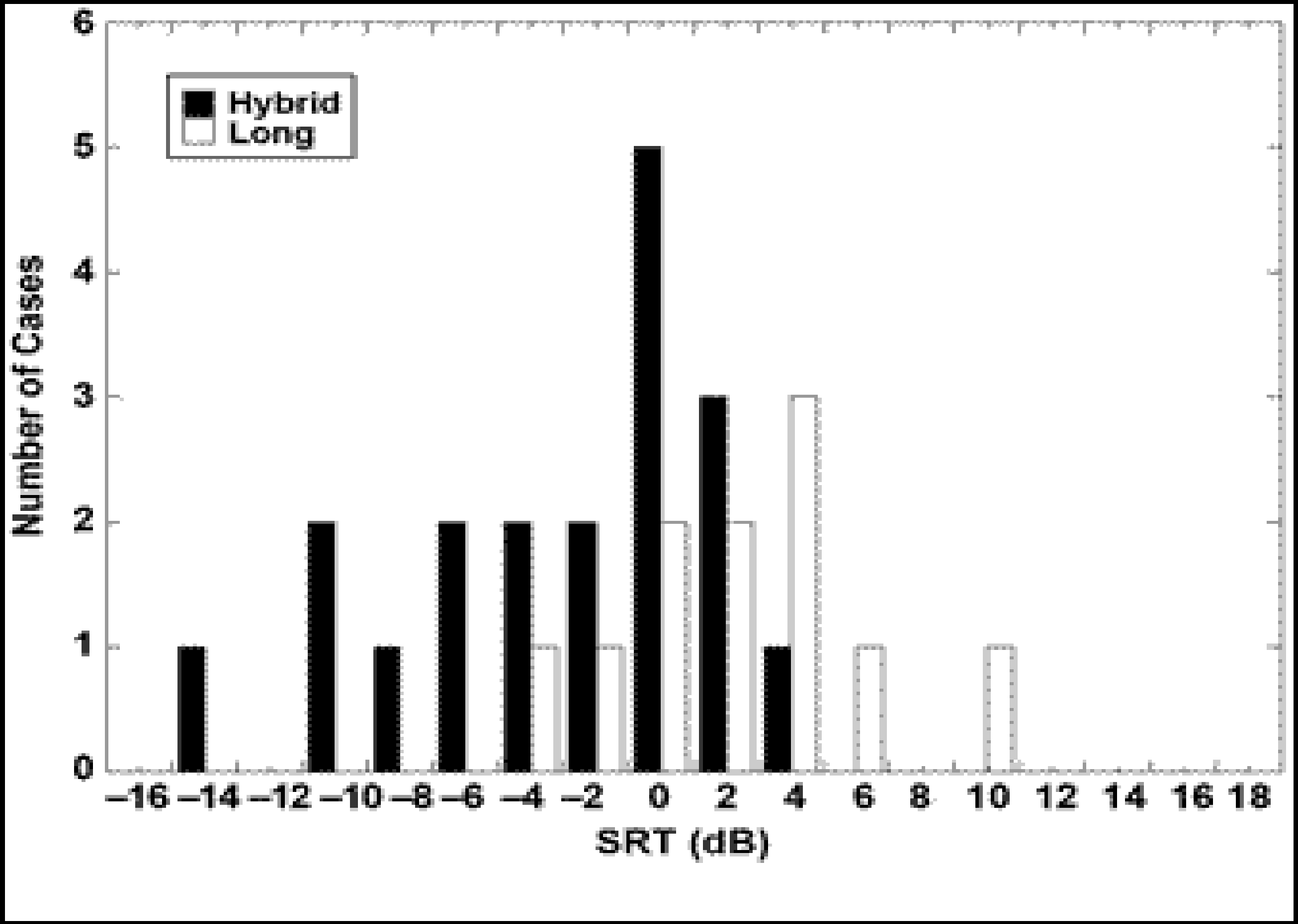
- Binaural Summation
  - Stereo effect; sounds presented to two ears will seem louder
  - Summation improves loudness perception by 3 dB
  - Redundancy in cues improves sensitivity to fine differences and improved speech perception

Hybrid Implants

- Implant users have poor frequency resolution
  - Accounts for poor hearing in noise
  - Accounts for poor appreciation of music
  - Tone discrimination
    - Normal hearing can discriminate semitones 1 key on piano
    - Implant users need ½ to 2 octaves
- Short implant allows preservation of low frequency inner hair cell function and electric stimulation of high frequency ganglia
  - Frequency resolution is improved.



Hybrid Implant Results



Complications

- Flap breakdown
- Misplacement of electrode
- Facial nerve paresis
- Foreign body reaction (rejection)
- Perforated tympanic membrane
- CSF leak
- Disequilibrium
- Magnet or receiver displacement

Board Question

- 60 % are Genetic
  - 70% are nonsyndromic
    - 50% are due to mutation of a single gene (21% of all congenital hearing loss)
    - Which one??

- *CONNEXIN 26*
  - What does Cx26 code for?

- *Gap Junction Protein Beta 2*
  - GJPB2

Board Question

- 60 % are Genetic
  - 30% are syndromic
    - 400 genetic syndrome associated with hearing loss
    - Only 2 are common, which ones?
      - *Pendred syndrome*
      - *Usher syndrome*
- *Jervell and Lange-Nielson syndrome*
- *NF II*

Board Question

- Three most common dominant syndromes.
  - Typically diagnosed at birth

- *Stickler syndrome*
- *Branchiootorenal syndrome*
- *Waardenburg syndrome*

Board Question

- How would you diagnose Auditory Neuropathy

- *Severe to profound SNHL*
- *Thresholds fluctuate*
- *SRT does not match PTA*

- *ABR abnormal*
- *OAE normal*

Board Question

- Congenital deafness is often due to prenatal infections with what organisms?

- *TORCH*

Wayne K. Robbins, D.O.

3455 Regency Park Dr
Grand Blanc, MI 48439
810 694-0600