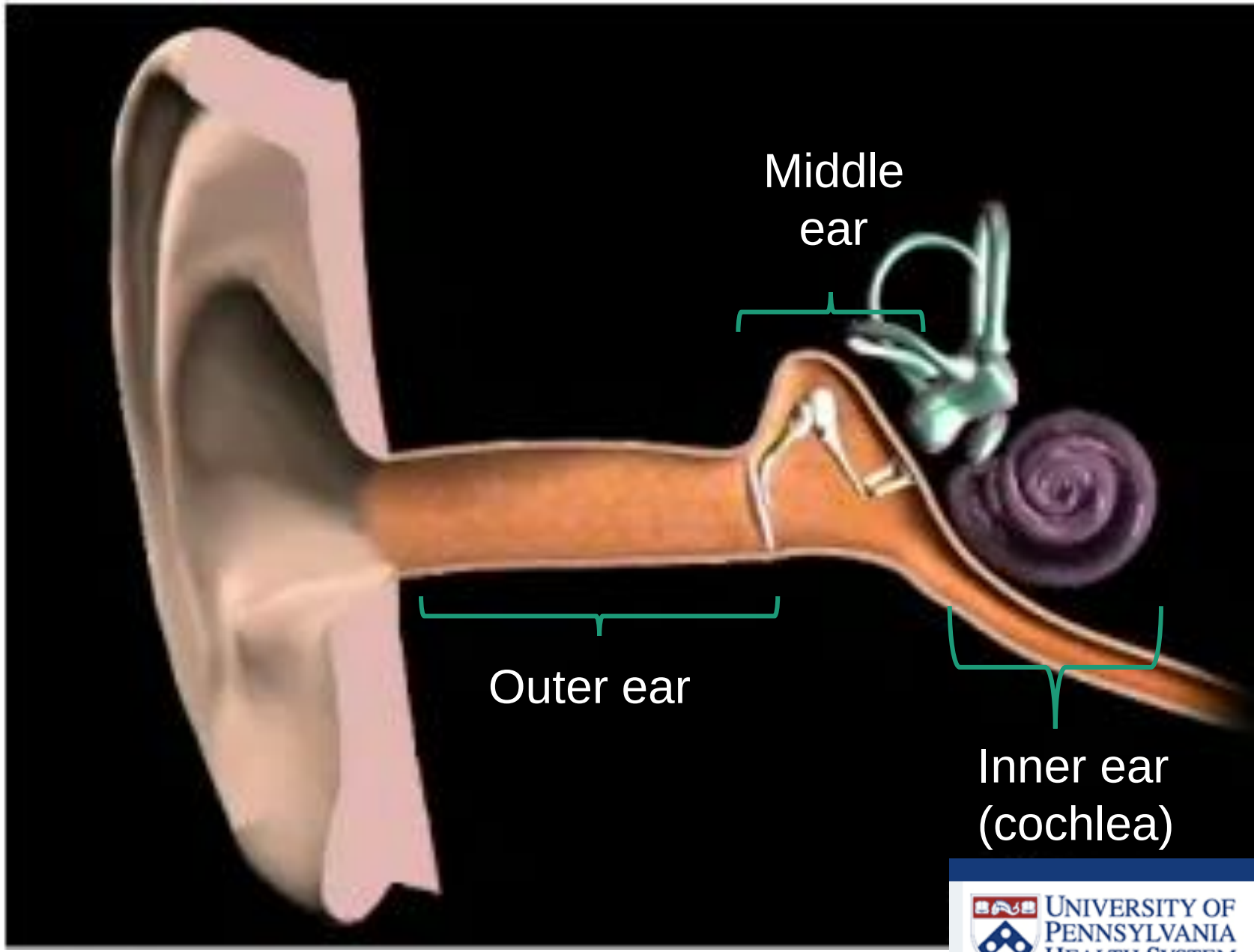
Cochlear Implants:
Auditory & Developmental Neuroscience

Dan Ofer

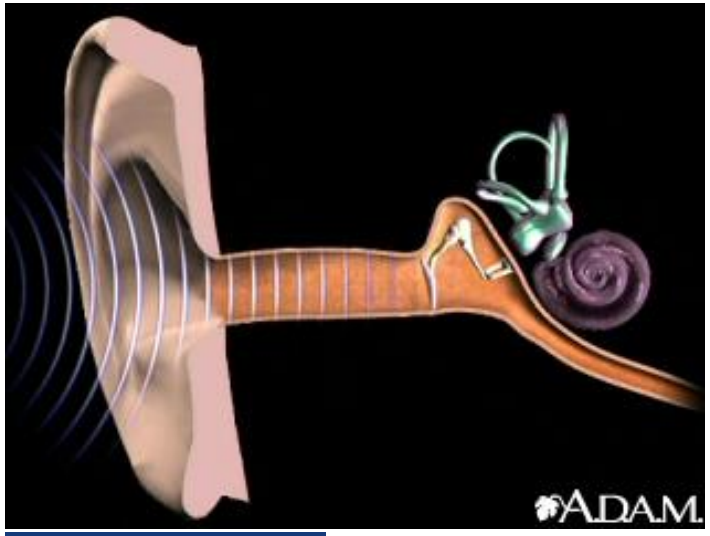
Normal Hearing - Anatomy



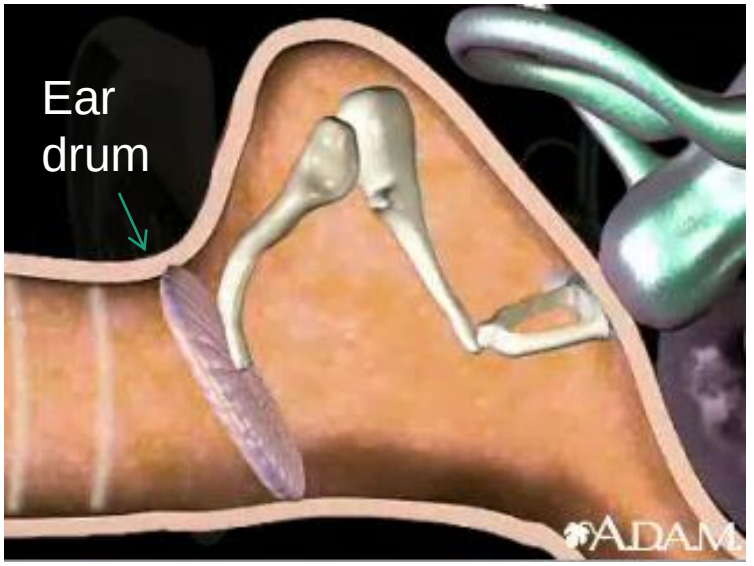
UNIVERSITY OF PENNSYLVANIA HEALTH SYSTEM

How Do We Normally Hear?

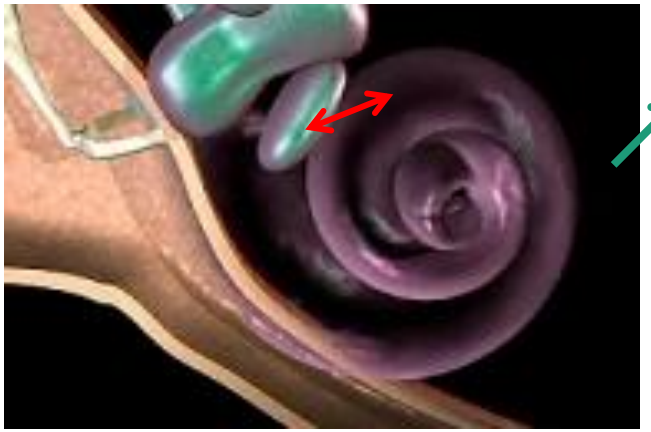
Sound enters ear…



The eardrum and middle ear vibrate…



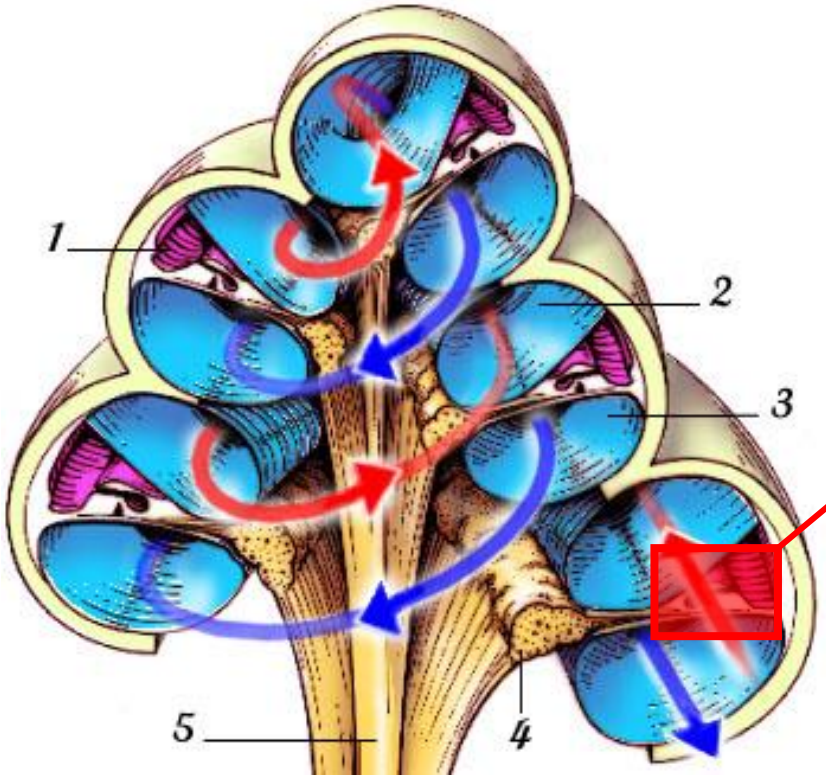
Creating pressure within the **cochlea**…



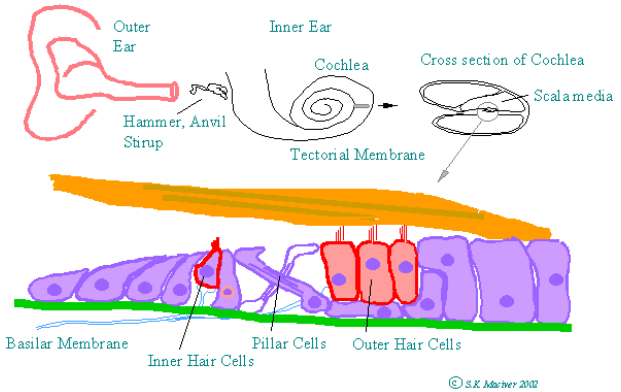
Transduced to neural impulses, then brain via auditory nerve.

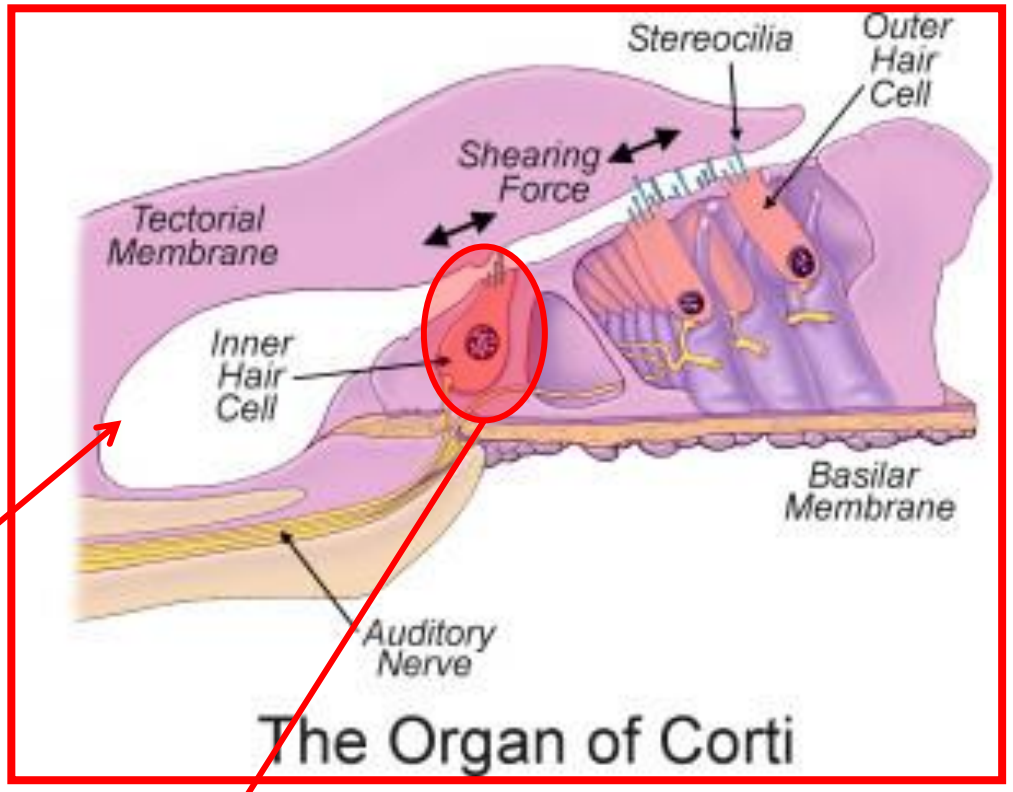
The Cochlea: Sound to Neural Impulses

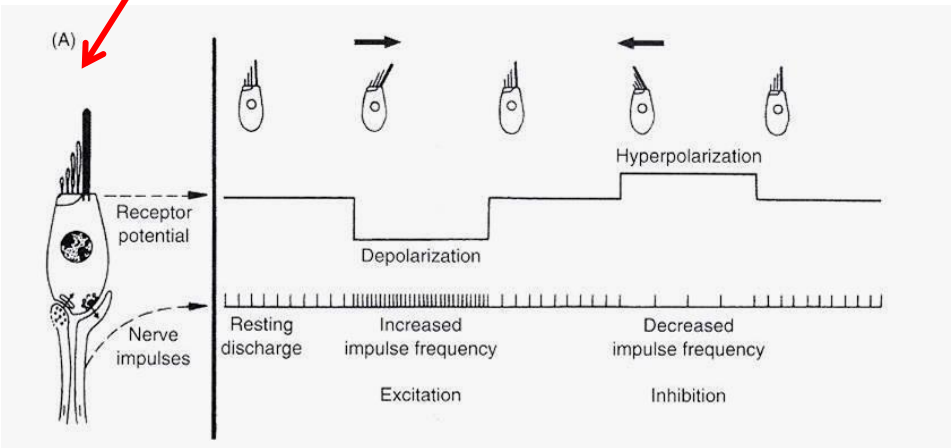
Cochlea Cross-section



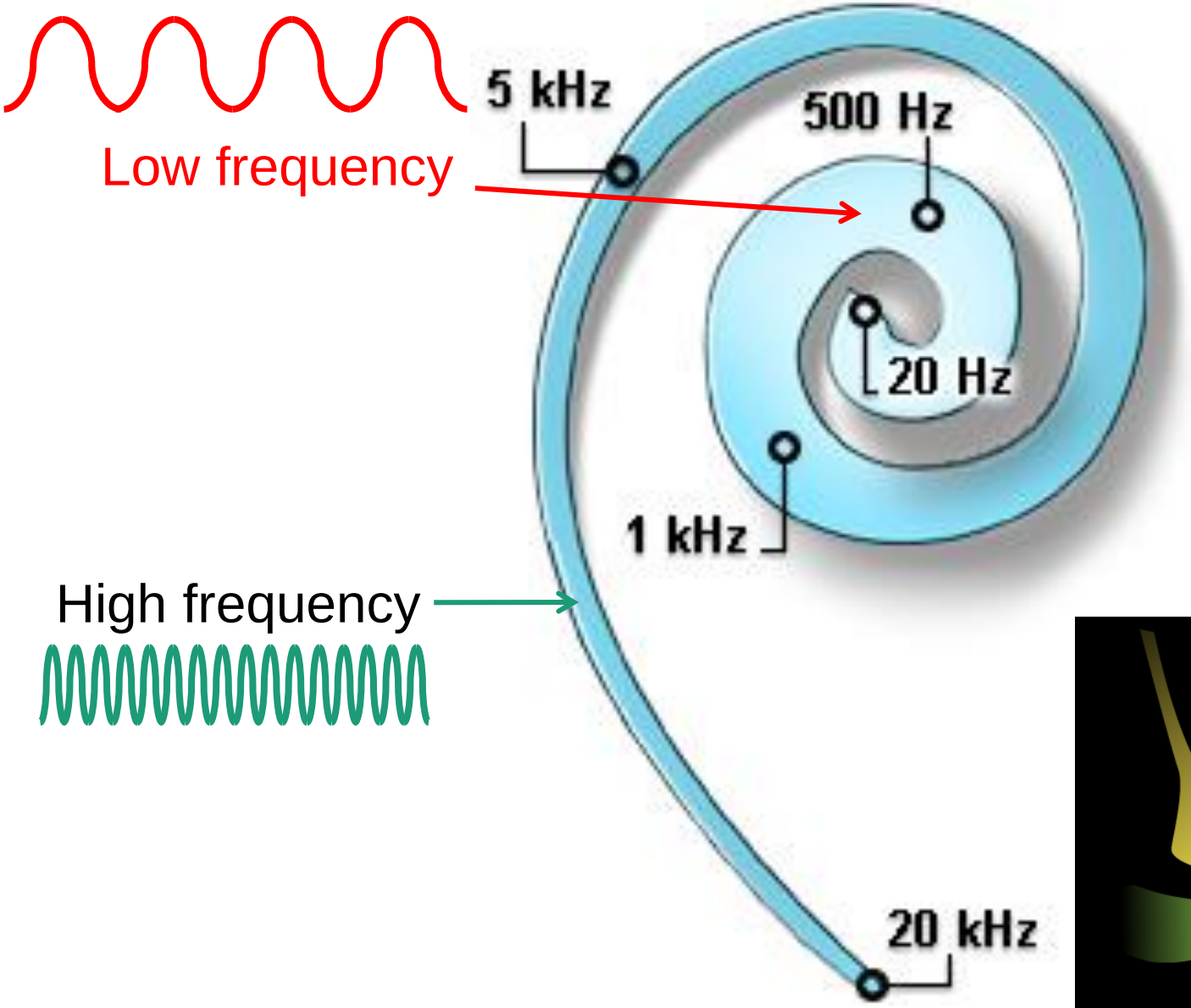
http://www.iurc.montp.inserm.fr/cric/audition/

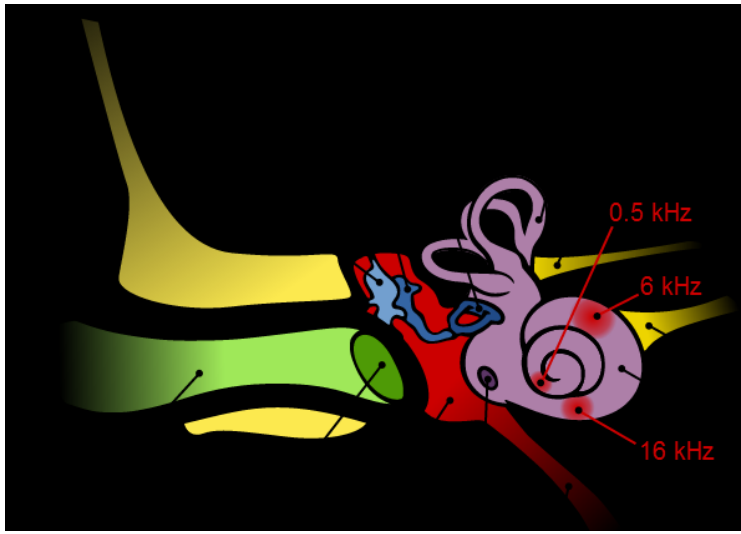






Tones and the Cochlea: Tonotopic!





Hearing Loss

Can happen due to disease, genetics, damage, age..

Common causes :

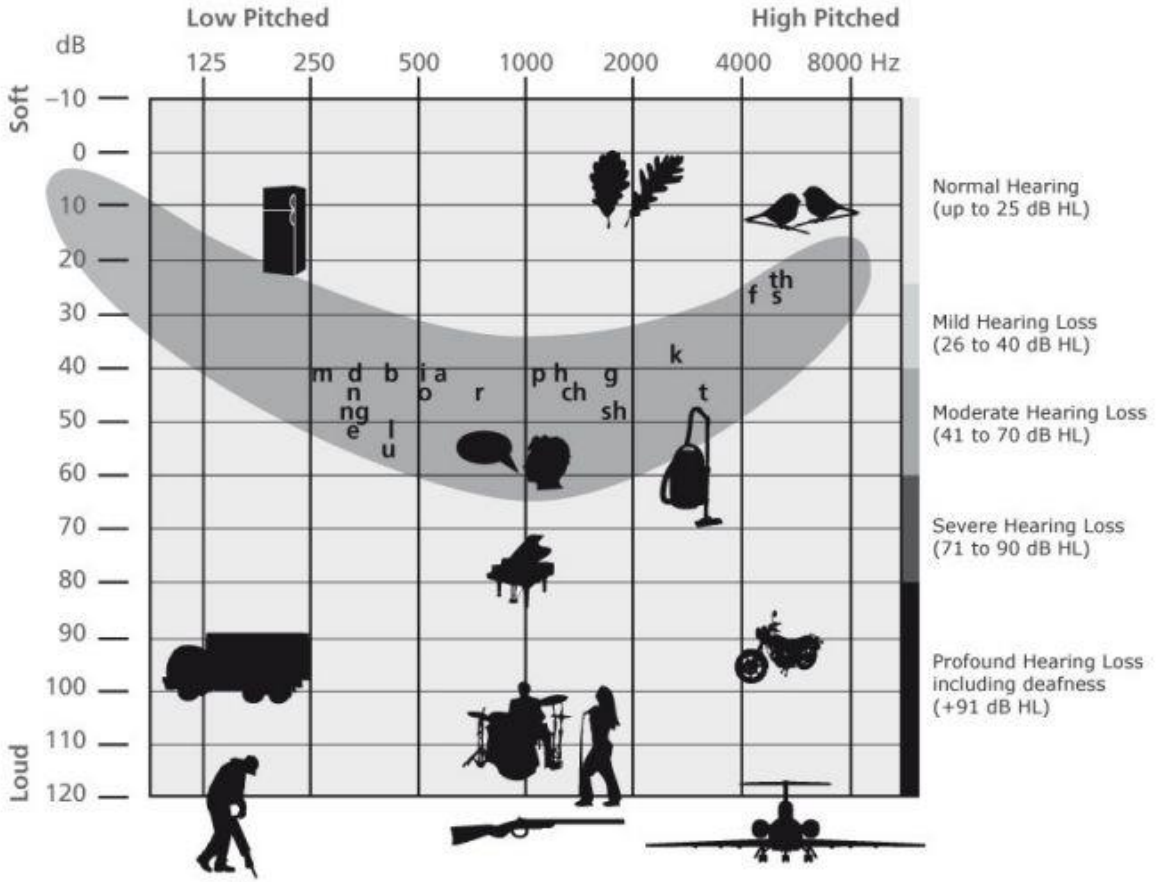
- Damage to hair cells (**Inner** and/or **outer**) due to innate vulnerability.
  - Partial damage to auditory nerve.
  - Damage to components in parts of ear.
    - Etc..

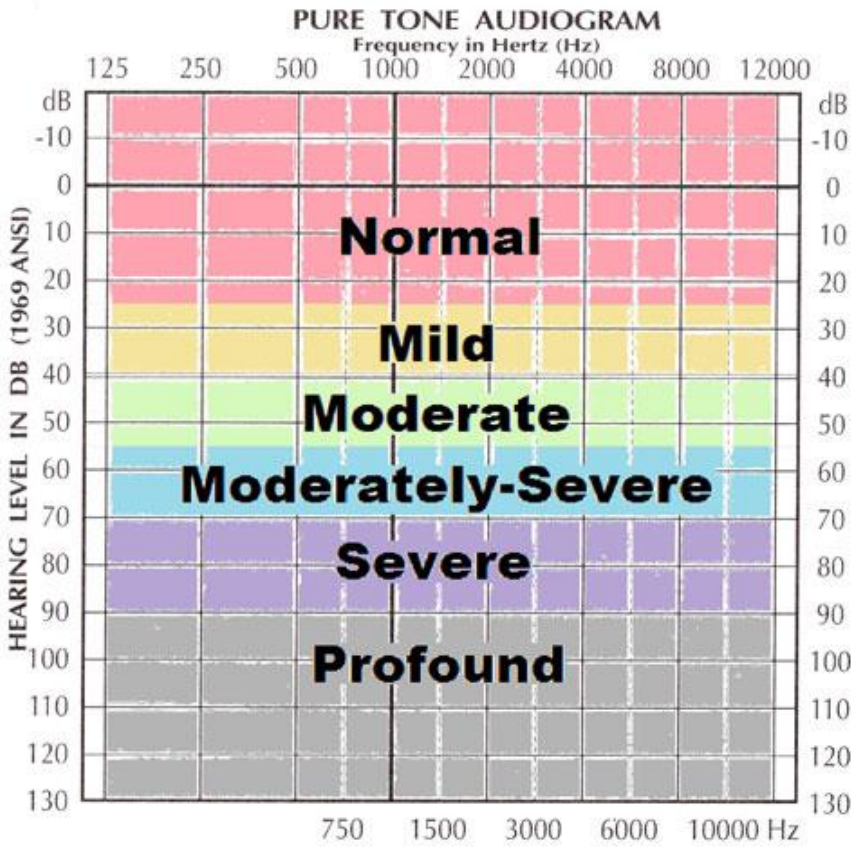
Hearing Loss Levels

-10 - 25 dB HL: normal hearing

> 90 dB HL: **profound hearing** loss

Reminder: (dB is a LOG scale! 10^x ***!!)***





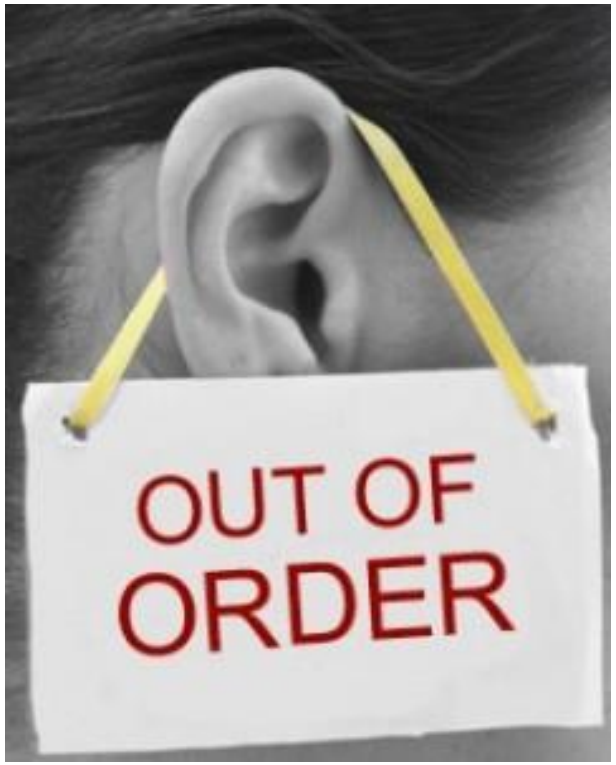
http://auditoryneuroscience.com/acoustics/clinical_audiograms
http://www.phonak.com/com/b2c/en/hearing/understanding_hearingloss/types_of_hearing_loss.html

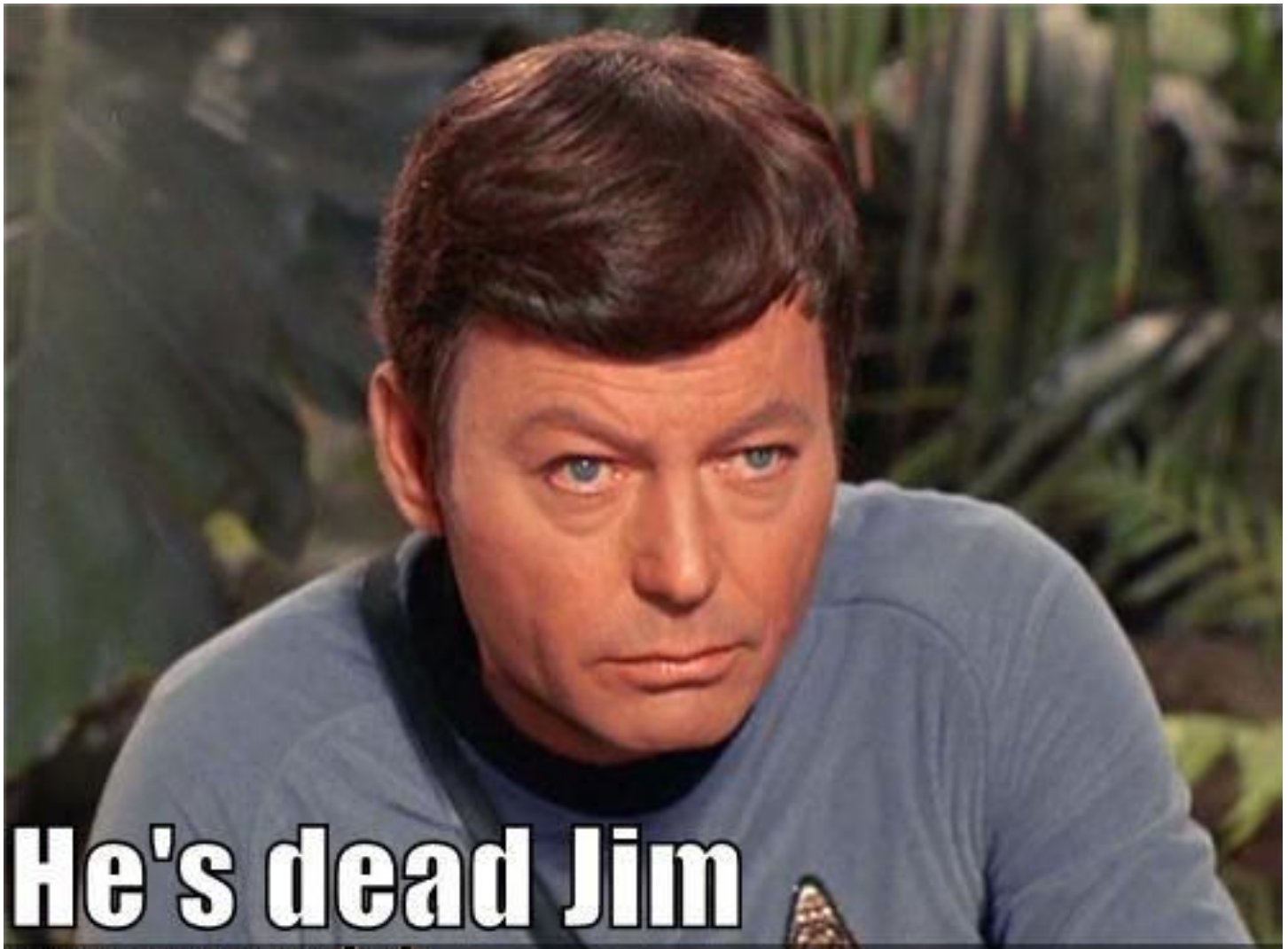
Hearing Aids = Amplify

Horns? Yes, to help him hear; they are attached to his eye-glasses

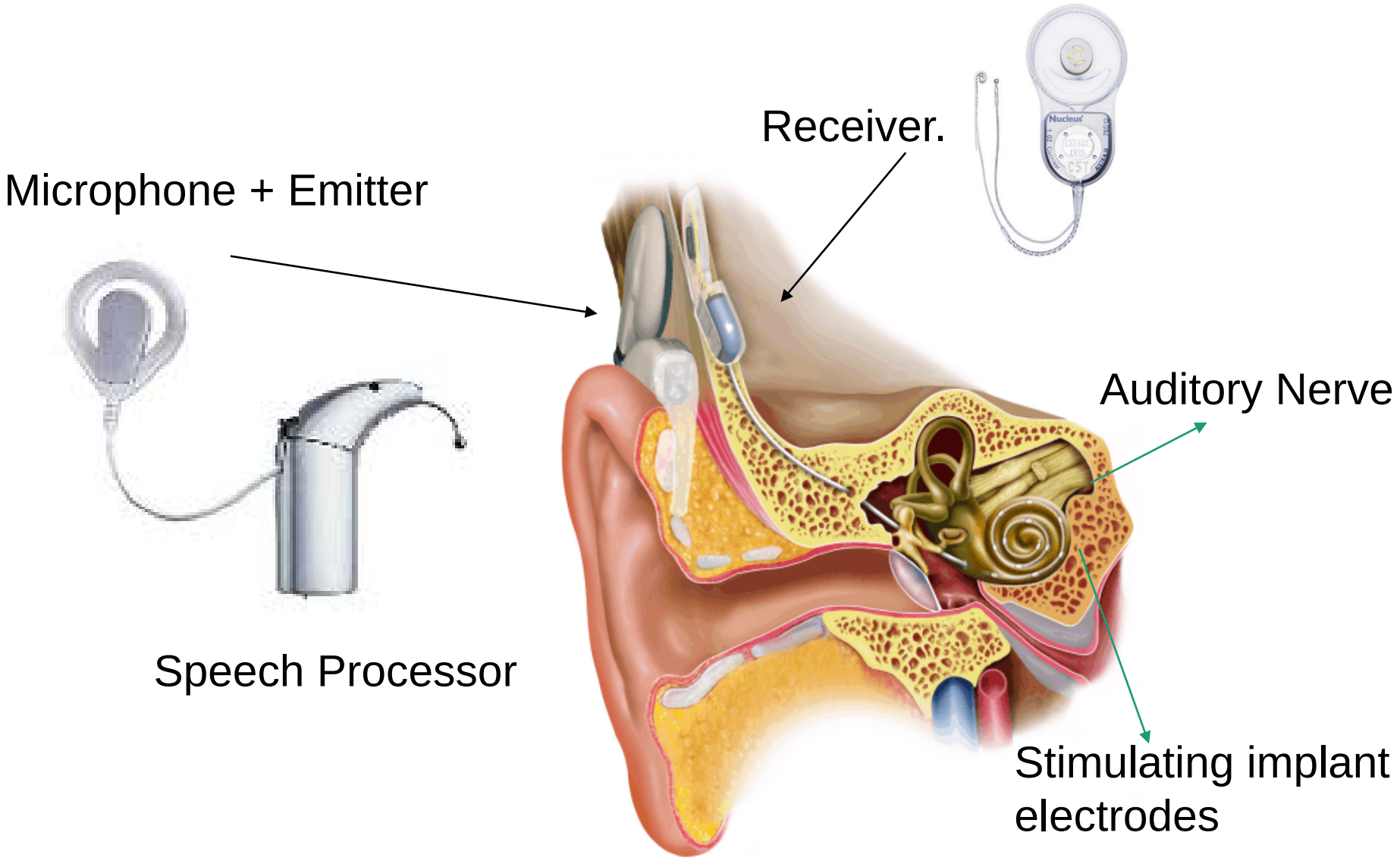
What happens when “no” residual hearing left to amplify???

PROFOUND-Severe Hearing Loss

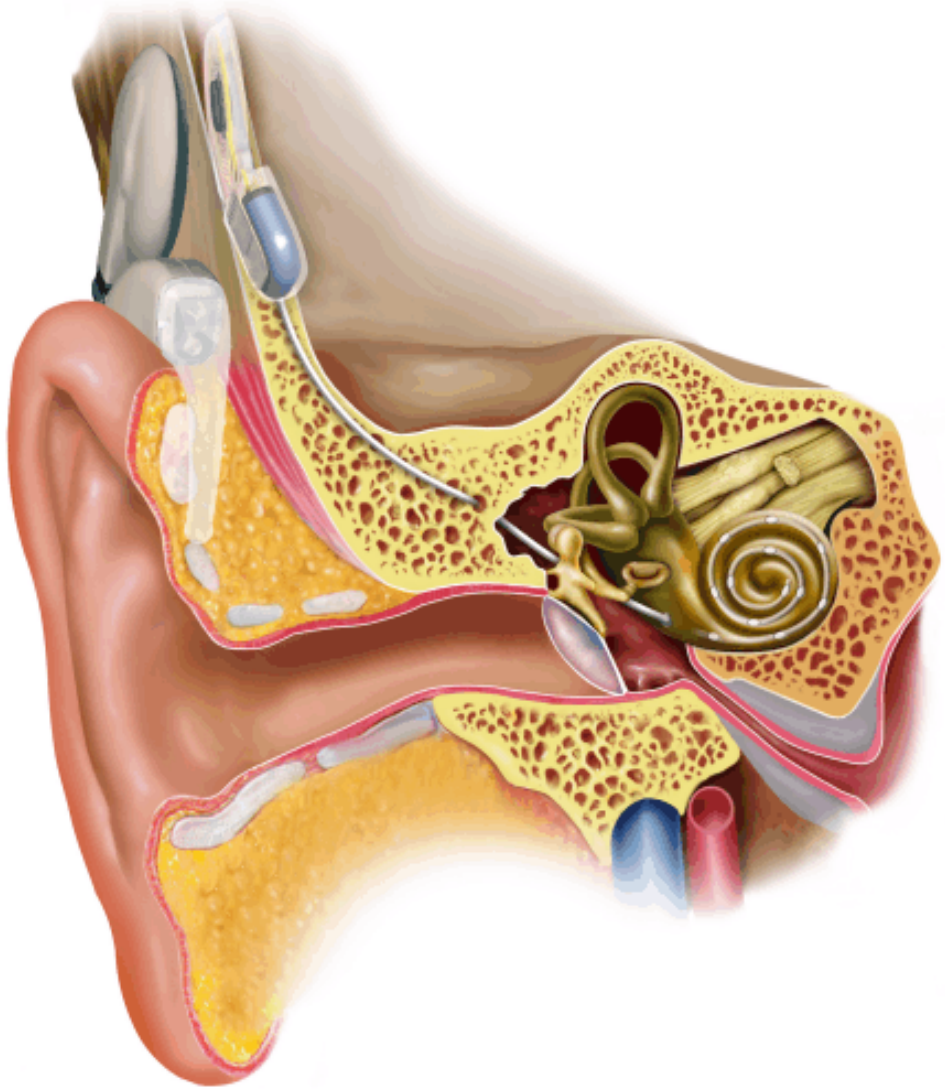


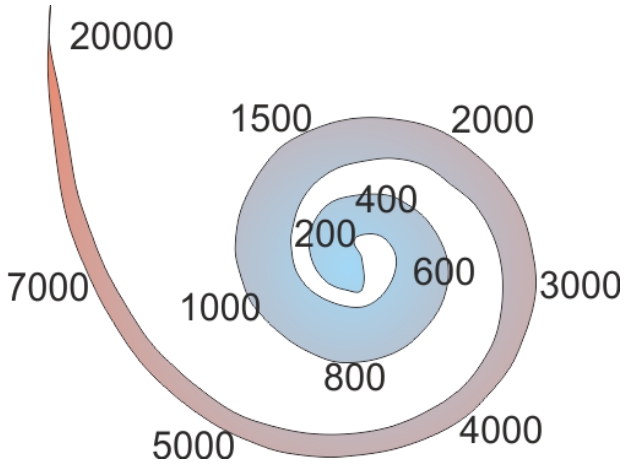


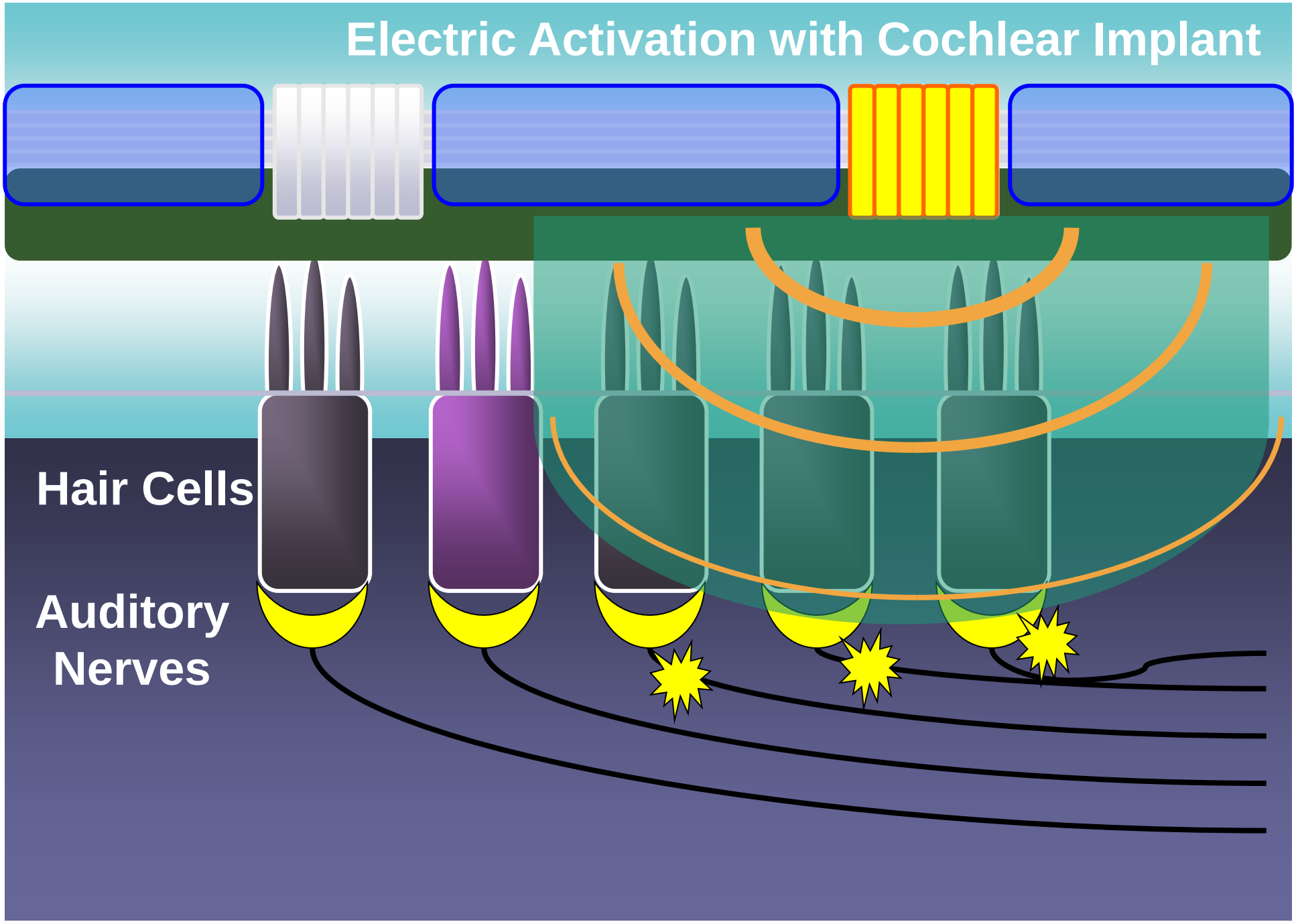
Cochlear Implants



Cochlear Implants: Stimulating Electrodes inside the Organ of Corti



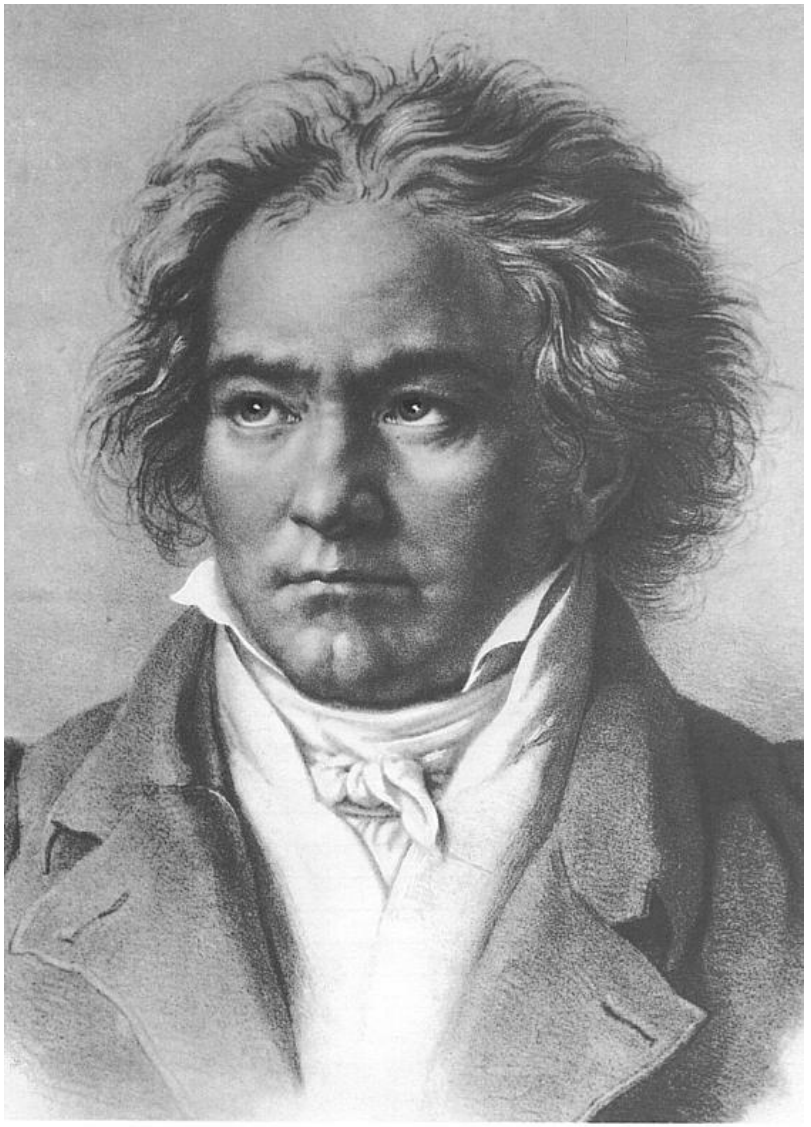



Electric Activation with Cochlear Implant
Hair Cells
Auditory Nerves

Limitations of Cochlear Implants

- The electrode array does not reach all the way in.
- Modern implants = ~20 electrode channels, but lots of electrical "NOISE", because of the highly conductive perilympthatic fluid.
  - # "Effective" separate frequency channels is even lower (~ 9).
  - Compare to ~3,500 natural Inner Hair Cells (with amplification + compression by 12,000 Outer hair cells!)

Cochlear Implants
Music in your Ears?



LUDWIG VAN BEETHOVEN
KREIDEZEICHNUNG VON AUGUST VON KLÖBER AUS DEM JAHR 1818

Normal Ludwig

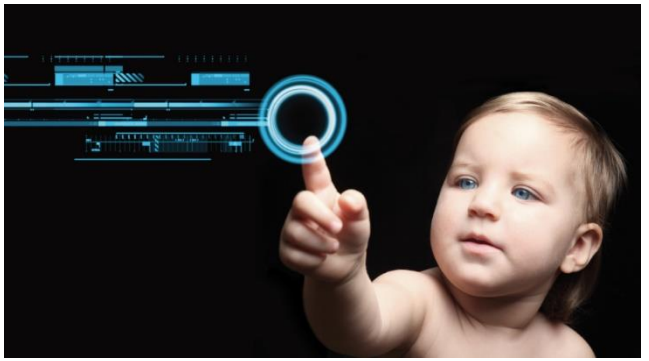
CI Ludwig

http://auditoryneuroscience.com/prosthetics/music
http://auditoryneuroscience.com/prosthetics/noise_vocoded_speech

Factors Effecting Cochlear Implant Performance

- **Age at time of implant**
- Age of onset of deafness
- Duration of deafness
- Others:
  - Commitment!! (family and individual)
  - Therapy, training..

The Problem with Age: The Early brain:

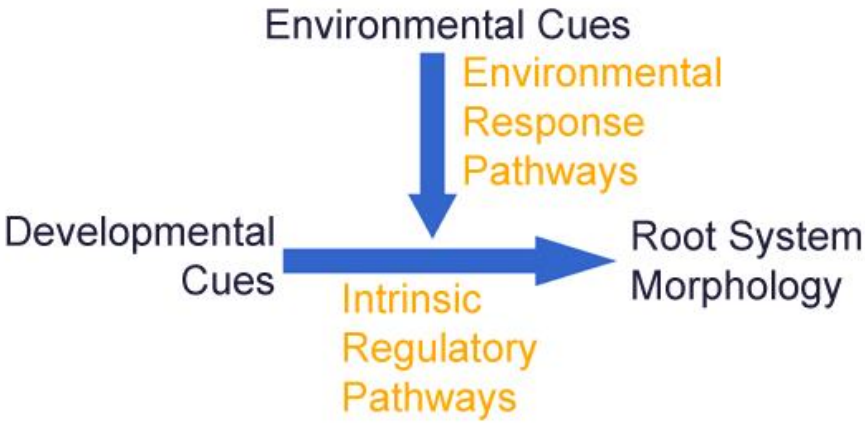


Young	Old
Learning is Easy	Learning is HARD!
Totally new abilities acquired (walking, talking..)	No new “tricks”.

*** Bavelier, & Hensch et al.. (2010). Removing brakes on adult brain plasticity: From molecular to behavioral interventions. Journal of Neuroscience,**

Experience and Neurodevelopmental periods

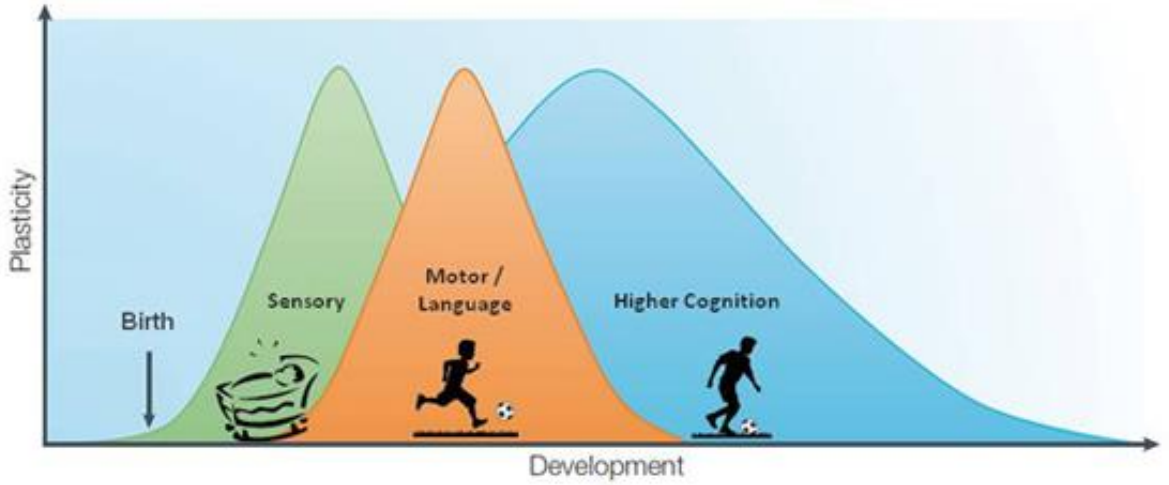
- **Experience Dependent** Plasticity ~= Environmental input modulates.
  - Doesn't apply for all cortical circuits!
  - Different types, circuits, **periods**.
  - Depends on organism, even for the same senses ..



Sensitive periods

- Periods of increased sensitivity during development.
- Effects of experience are **enhanced**.
- **Quantitative Difference**

Fig 1: Windows of plasticity in brain development



Adapted from Hensch, T. K. (2005). Critical period plasticity in local cortical circuits. *Nature Reviews Neuroscience, 6*(11), 877–888.

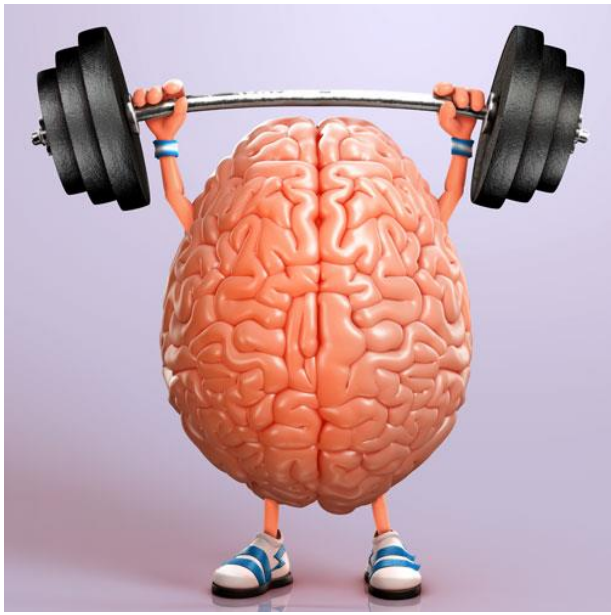
Sensitive periods (II)

LeBlanc JJ, Fagiolini M. Autism: a "critical period" disorder? Neural Plast. 2011
Montessori, Maria (1949). *The Absorbent Mind*

Critical periods

A strict time window during which environmental experience provides information essential for a function's development and "fixates" it, permanently.

Qualitative Difference



CRITICAL PERIOD PLASTICITY IN LOCAL CORTICAL CIRCUITS. Hensch. NATURE REVIEWS NEUROSCIENCE. (2005)

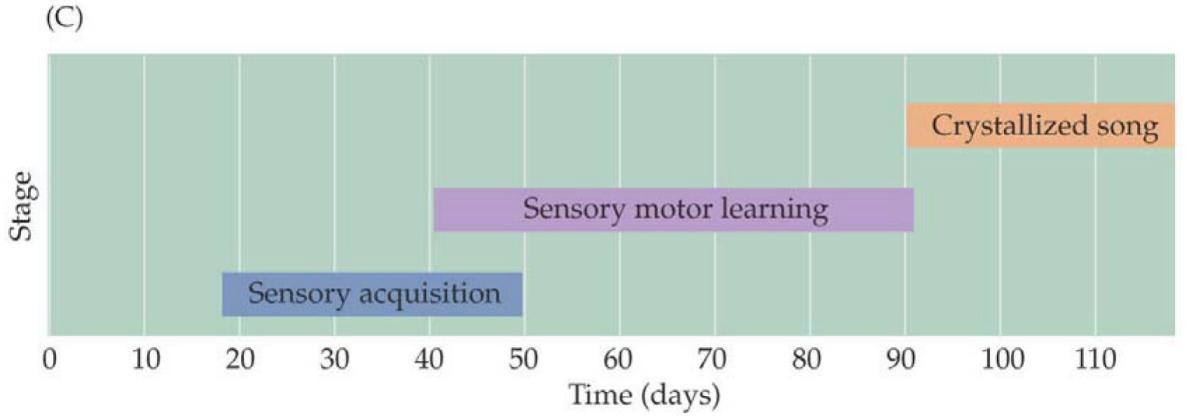
Critical period: Parental Imprinting

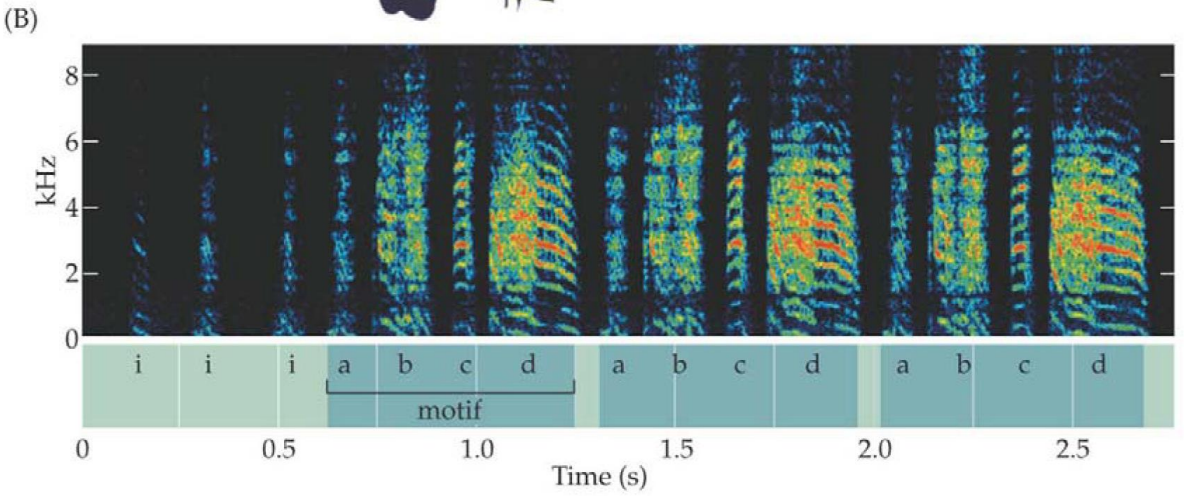
Critical Period for Song Acquisition

Selectivity for songs of own species.

In the lack of own species' song ….adaptation.

Critical period ends at puberty





Critical period: ocular dominance monocular deprivation

- Hubel & Wiesel.
- Showed that ocular deprivation during CP causes monocular dominance, *Ambylopia*.
- Only if done during *critical period*.

Wiesel, Hubel (1963). "Effects of visual deprivation on morphology and physiology of cell in the cat's lateral geniculate body". Journal of Neurophysiology

Spoken Language Development in Children Following Cochlear Implantation

Goal: "To assess spoken language acquisition following cochlear implantation in young children."

- Examined over 3 Years, with children under 5.

Metric: Performance on measures of spoken language comprehension and expression (Reynell Developmental Language Scales).

JAMA. 2010 Apr 21;303(15):1498-506. doi: 10.1001/jama.2010.451.
Niparko JK, Tobey EA, Thal DJ, Eisenberg LS, Wang NY, Quittner AL, Fink NE; CDaCI Team.

Spoken Language Development in Children Following Cochlear Implantation

- Research Question: Effects of age at time of cochlear implantation on improvement in deaf children.
- "The younger the better" ?
- + Effects of other co-variables. (Deaf parents, home, prior level of hearing, etc').

- Expression & Comprehension scores:
- Vs other age groups.
- Vs Comparable hearing peers.

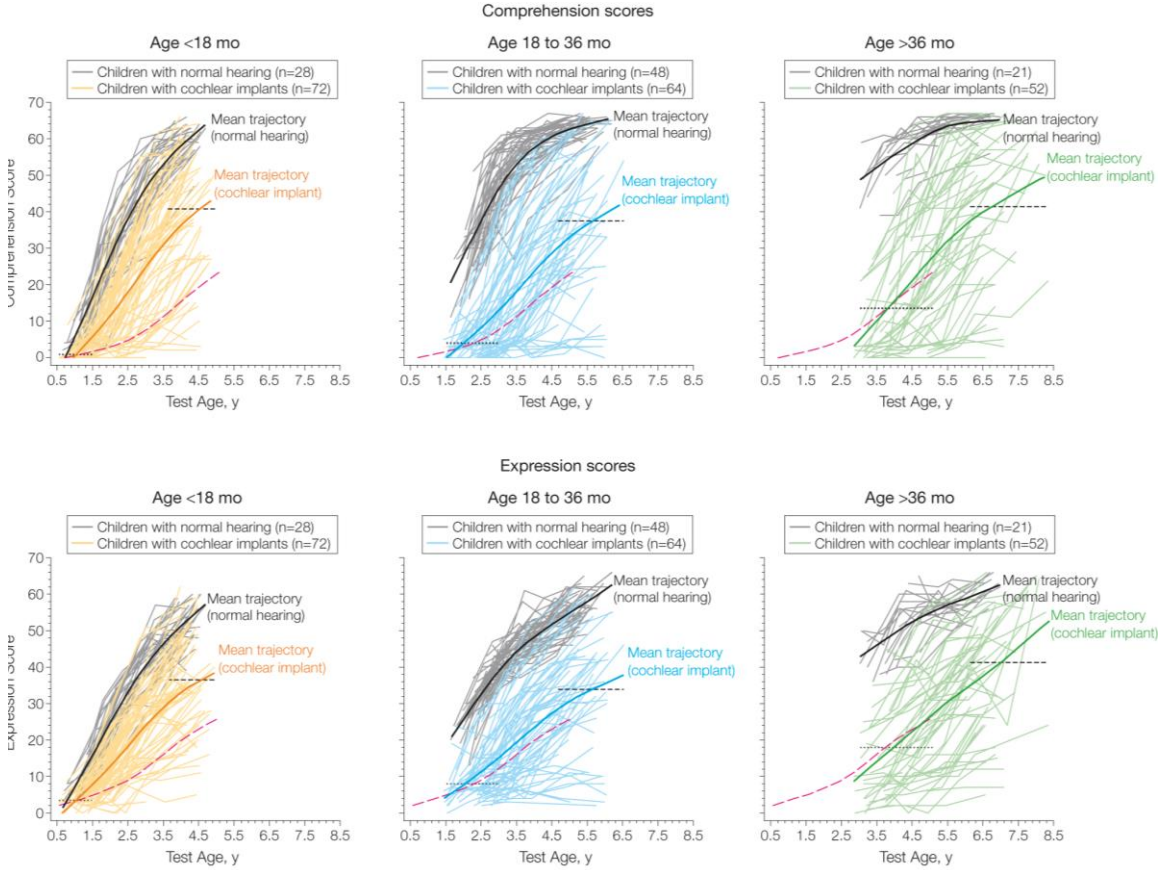
JAMA. 2010 Apr 21;303(15):1498-506. doi: 10.1001/jama.2010.451.
Niparko JK, Tobey EA, Thal DJ, Eisenberg LS, Wang NY, Quittner AL, Fink NE; CDaCI Team.

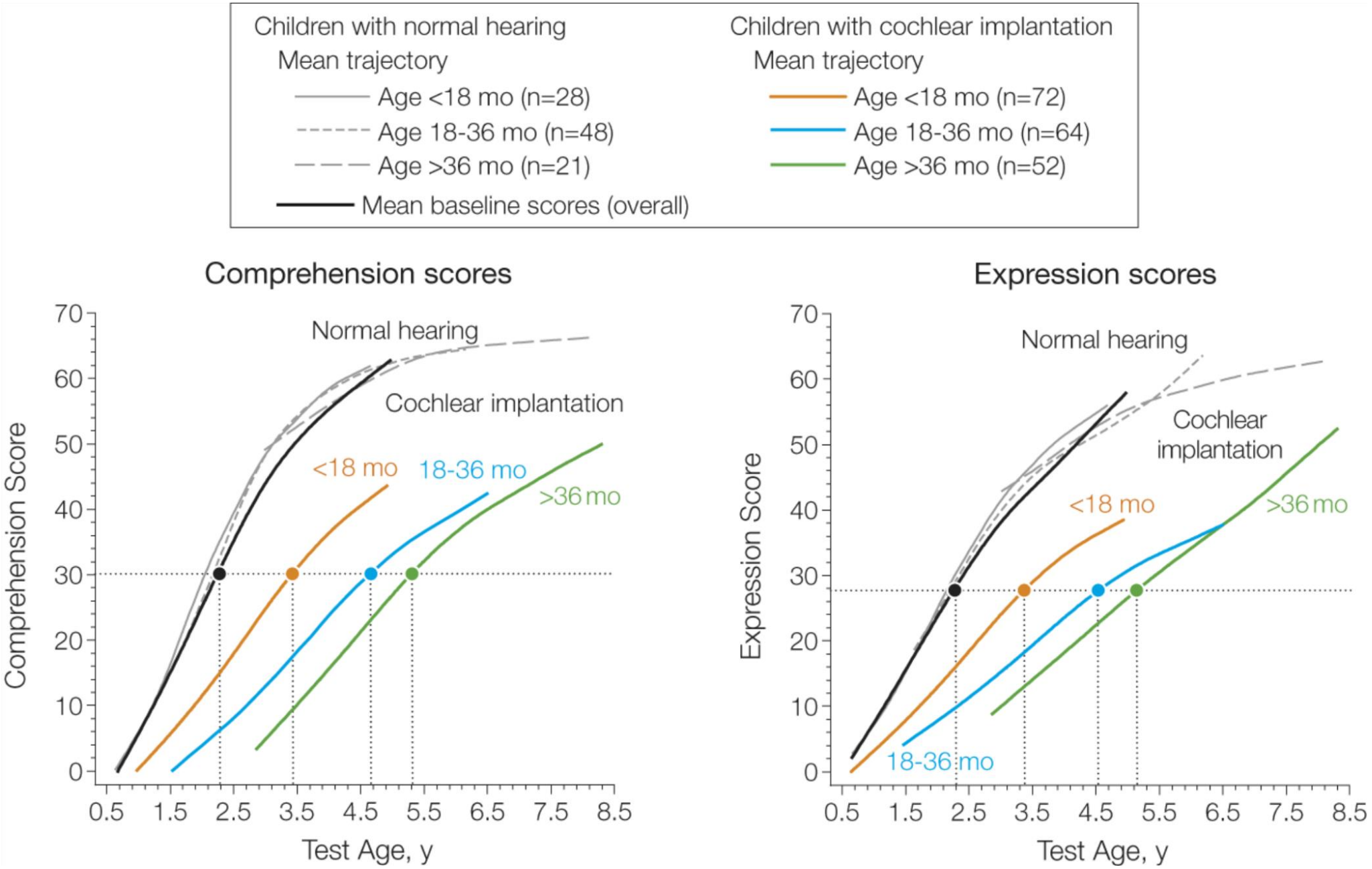
Spoken Language Development in Children Following Cochlear Implantation

Groups: •

Children with normal hearing
Mean trajectory
- Age <18 mo (n=28)
- Age 18-36 mo (n=48)
- Age >36 mo (n=21)

Children with cochlear implantation
Mean trajectory
- Age <18 mo (n=72)
- Age 18-36 mo (n=64)
- Age >36 mo (n=52)





From: **Spoken Language Development in Children Following Cochlear Implantation**
JAMA. 2010;303(15):1498-1506.

Table 3. Multivariable-Adjusted Mixed-Effects Modeling Analyses for Children Undergoing Cochlear Implantation[a]

	Comprehension		Expression	
	Estimate (95% CI)	*P* Value	Estimate (95% CI)	*P* Value
Factors Associated With RDLS Raw Scores at Baseline				
Child characteristic				
Mean hearing threshold, per 20-dB increase	−0.31 (−1.11 to 0.49)	.45	−0.17 (−0.75 to −0.41)	.57
Family characteristics				
Mean rating of parent-child interactions, per point increase	0.51 (−0.15 to 1.16)	.13	0.59 (0.06 to 1.12)	.03
Self-reported income <$50 000[b]	0.28 (−0.41 to 0.96)	.43	−0.24 (−0.72 to 0.24)	.32
Intervention characteristics				
Time with hearing, per 6-mo increase	0.71 (0.05 to 1.38)	.04	0.62 (0.24 to 0.99)	.001
Time with hearing loss (prior to amplification), per 6-mo increase	0.01 (−0.42 to 0.44)	.97	0.07 (−0.25 to 0.39)	.68
Time with amplification, per 6-mo increase	−0.14 (−0.49 to 0.22)	.44	−0.02 (−0.32 to 0.28)	.89
Spoken communication mode[b]	0.08 (−0.52 to 0.69)	.78	−0.14 (−0.53 to 0.26)	.50
Factors Associated With Differential Rate Increase in RDLS Raw Scores Over 3 Years				
Child characteristics				
Mean hearing threshold, per 20-dB increase	−2.28 (−4.31 to −0.25)	.03	−2.24 (−4.20 to −0.27)	.03
SRI score, per 100-point increase[c]	3.76 (3.07 to 4.45)	<.001	1.98 (1.41 to 2.54)	<.001
Family characteristics				
Mean rating of parent-child interactions, per point increase	3.75 (1.77 to 5.73)	<.001	3.45 (1.60 to 5.29)	<.001
Self-reported income <$50 000[b]	−2.20 (−3.93 to −0.47)	.01	−0.82 (−2.45 to 0.82)	.33
Intervention characteristics				
Time with hearing, per 6-mo increase	−1.45 (−2.70 to −0.19)	.02	−2.20 (−3.60 to −0.81)	.002
Time with hearing loss (prior to amplification), per 6-mo increase	−0.74 (−1.91 to 0.43)	.21	−0.51 (−1.63 to 0.60)	.37
Time with amplification, per 6-mo increase	−1.31 (−2.11 to −0.52)	.001	−1.12 (−1.79 to −0.44)	.001
Spoken communication mode[b]	1.57 (−0.22 to 3.35)	.09	1.69 (−0.02 to 3.41)	.05
Bilateral cochlear implantation status, time-dependent	2.19 (−0.86 to 2.55)	.11	1.67 (−0.94 to 4.27)	.21

Abbreviations: CI, confidence interval; RDLS, Reynell Developmental Language Scales; SRI, Speech Recognition Index.
[a] Each variable is adjusted for study center, sex, race, ethnicity, maternal education (high school graduate or not), hearing onset, cognition, RDLS raw scores at baseline, time to implant activation, time-dependent bilateral implant status, and all other variables listed in the table.
[b] As measured at baseline.
[c] Speech Recognition Index score was included as a time-dependent variable. The mean increase in SRI scores over the 3-year follow-up period was 349 points, moving from a mean of 45 points at baseline to 395 points after 3 years.

- Time spent Deaf. (Age)
- Prior level of hearing.
- Parents.

Clear Improvement!

From: **Spoken Language Development in Children Following Cochlear Implantation**
JAMA. 2010;303(15):1498-1506.

Conclusions:

- Severe-Profoundly deaf children should get a unilateral cochlear implant as young as possible (~12 months) *
- Less time deaf = better and faster outcomes.
- Cochlear implants improve expression and comprehension (for children and post-lingually deaf adults).
- Lots of room for improvement!
- Technology, resolution, biological cures (regeneration).
- Reopening of critical/sensitive periods.. (Valproate!)

***** "Cochlear Implants for Children with Severe-to-Profound Hearing Loss". Blake C. Papsin, and Karen A. Gordon. N Engl J Med 2007; DOI: 10.1056

ALL GOOD THINGS COME TO AN END

THIS IS THE END