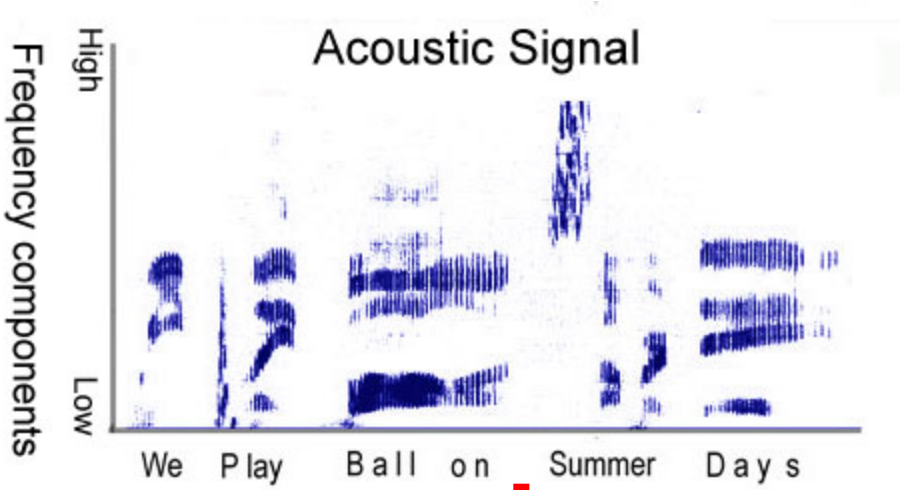
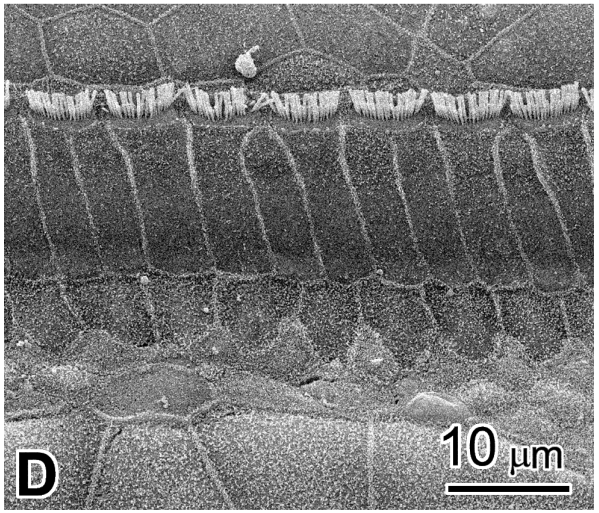
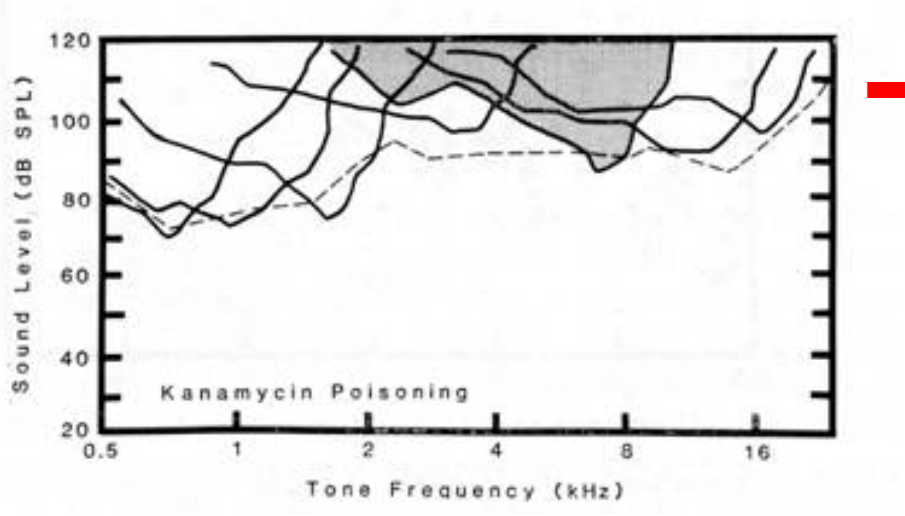
Coding of Speech Signal in Cochlea with OHC degeneration

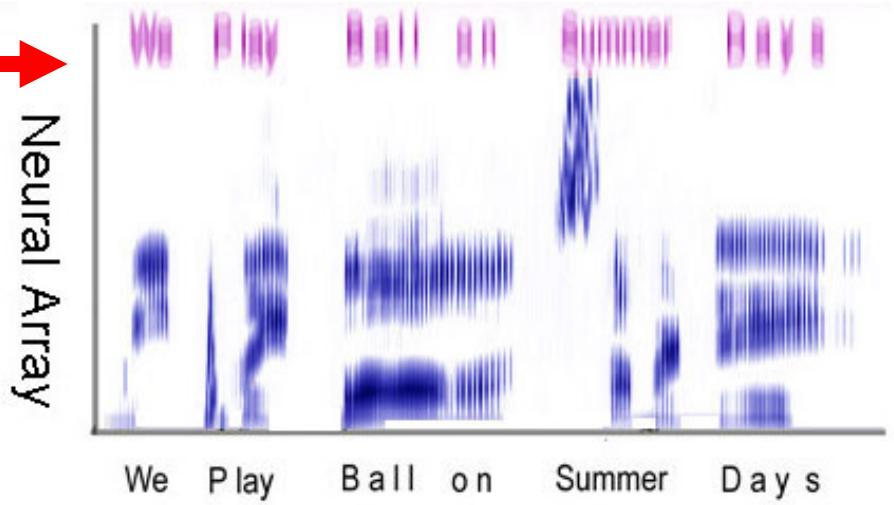




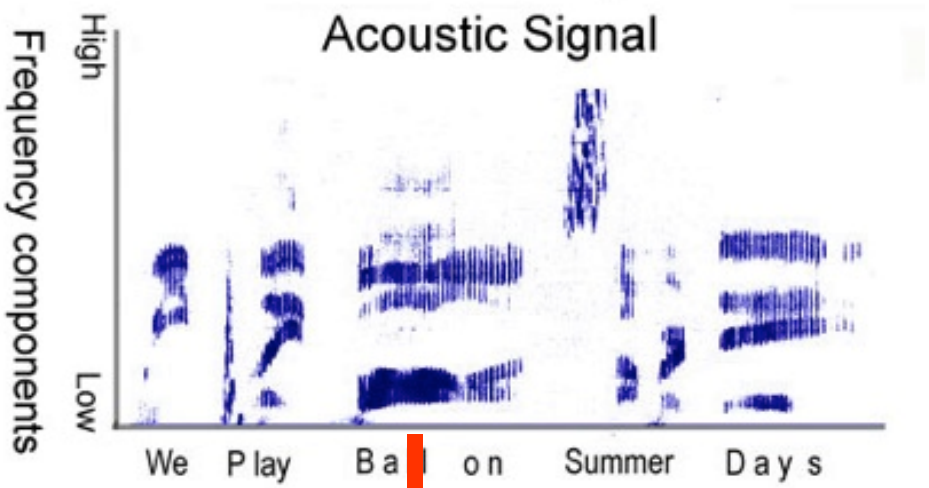
Degraded neural representation

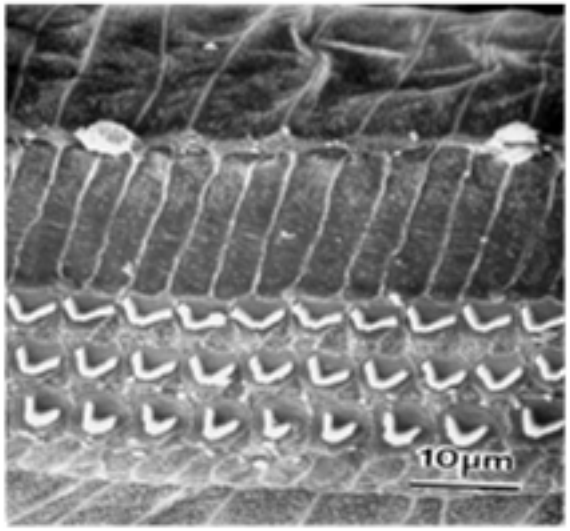


Cochlear Analysis Filters

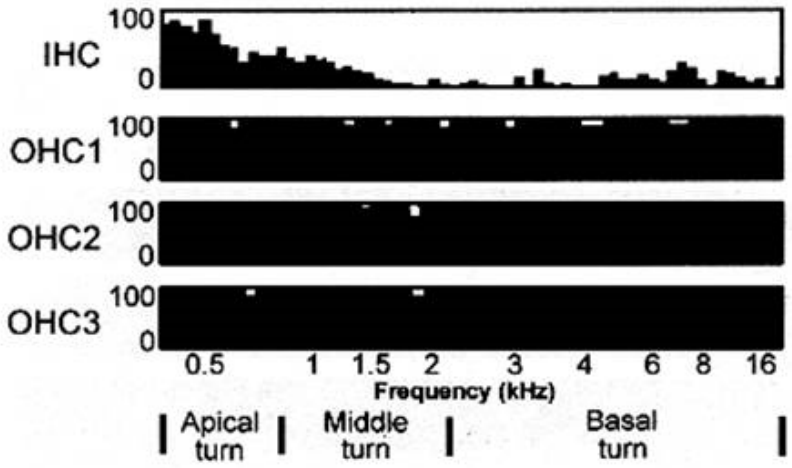


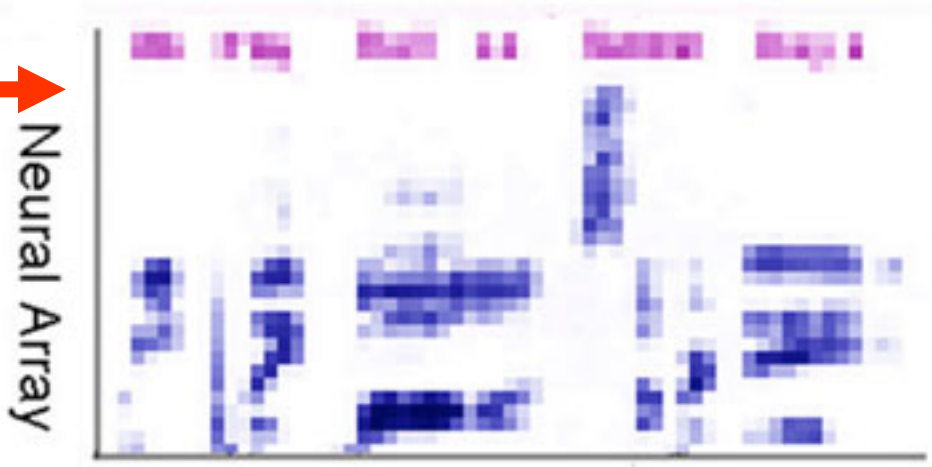
Coding of speech signals with (sub-total) IHC degeneration (as in auditory neuropathy)



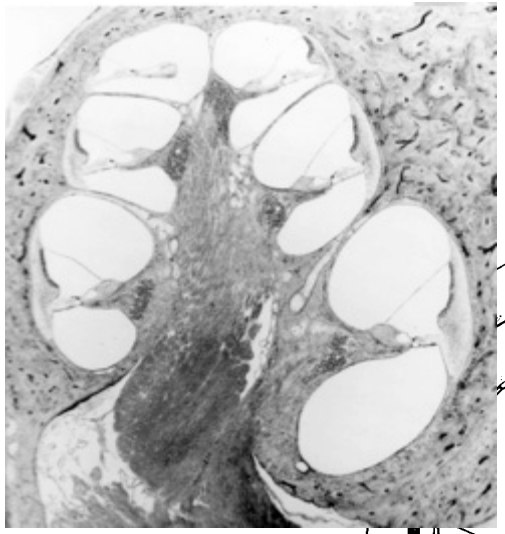


poor neural representation because of reduction in channel capacity





Cochlear areas of maximum vulnerability



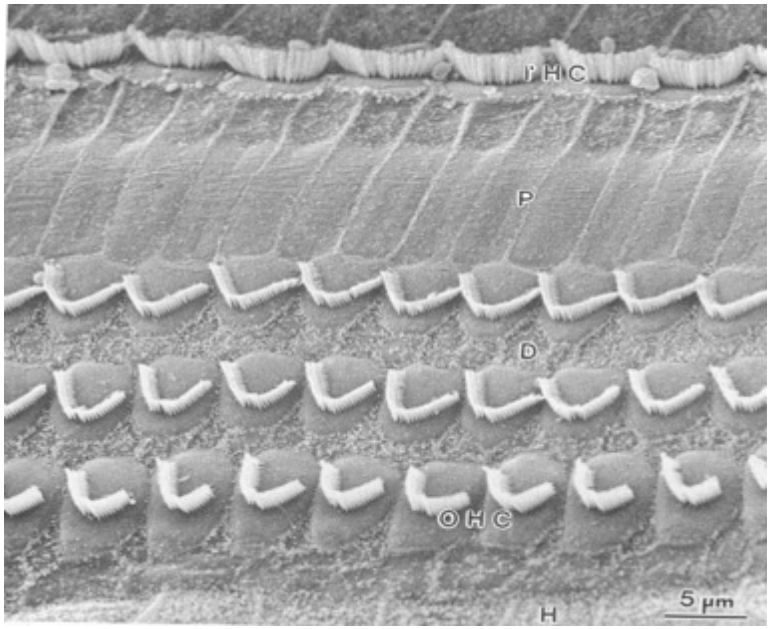
Stria vascularis
hypoxia, ischemia
loop diuretics (Lasix)
metabolic inhibitors
old age
viral infection
genetic mutation
(sometimes reversible)

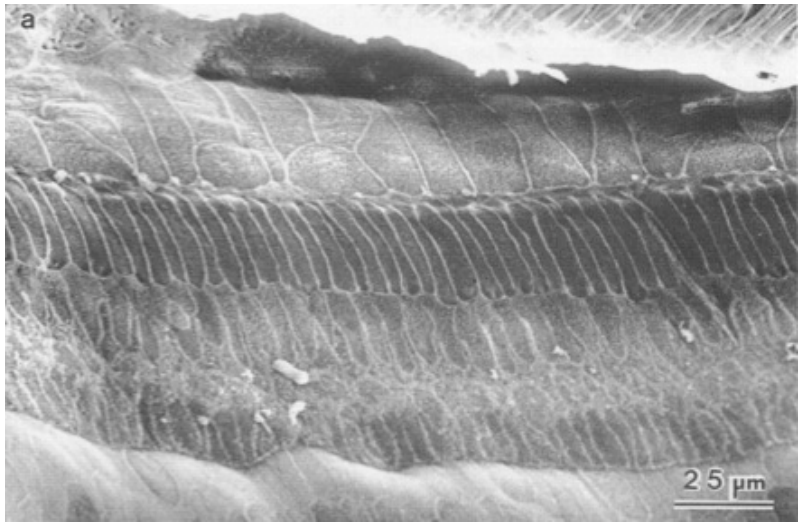
Haircells
ototoxic drugs
e.g. aminoglycosides
old age
acoustic trauma
genetic mutation
(not reversible)

Inner haircell synapse
some drugs e.g. aspirin
chronic hypoxia
anti cancer drugs – carboplatin
noise exposure
(sometimes there is recovery)

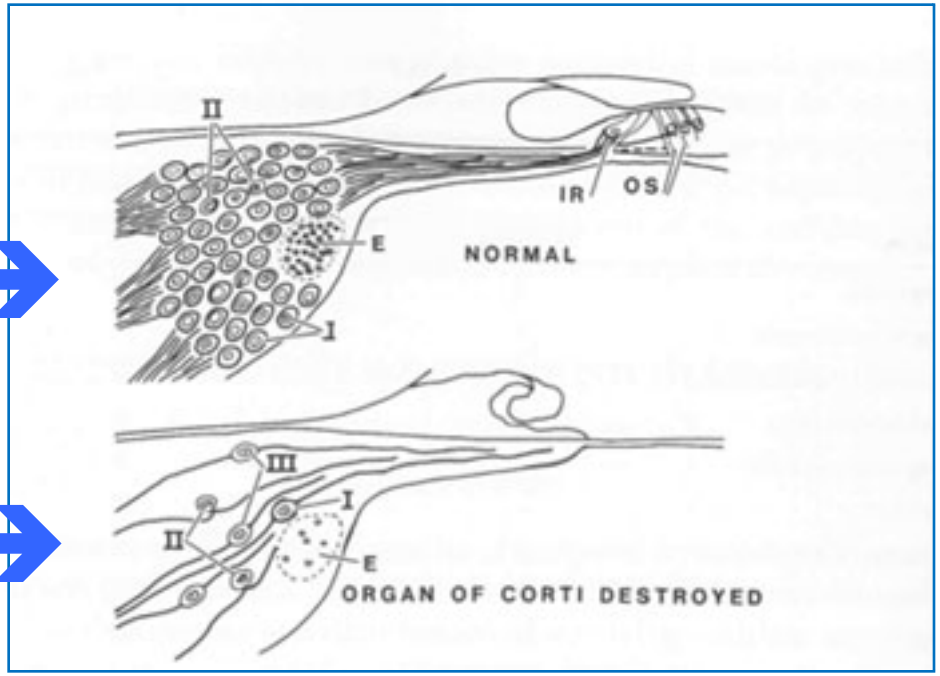
Spiral ganglion cells
loss of inner haircells
Glutamate excitotoxicity
Sensorimotor neuropathy
Hidden hearing loss?

When inner hair cells are damaged, spiral ganglion cells degenerate





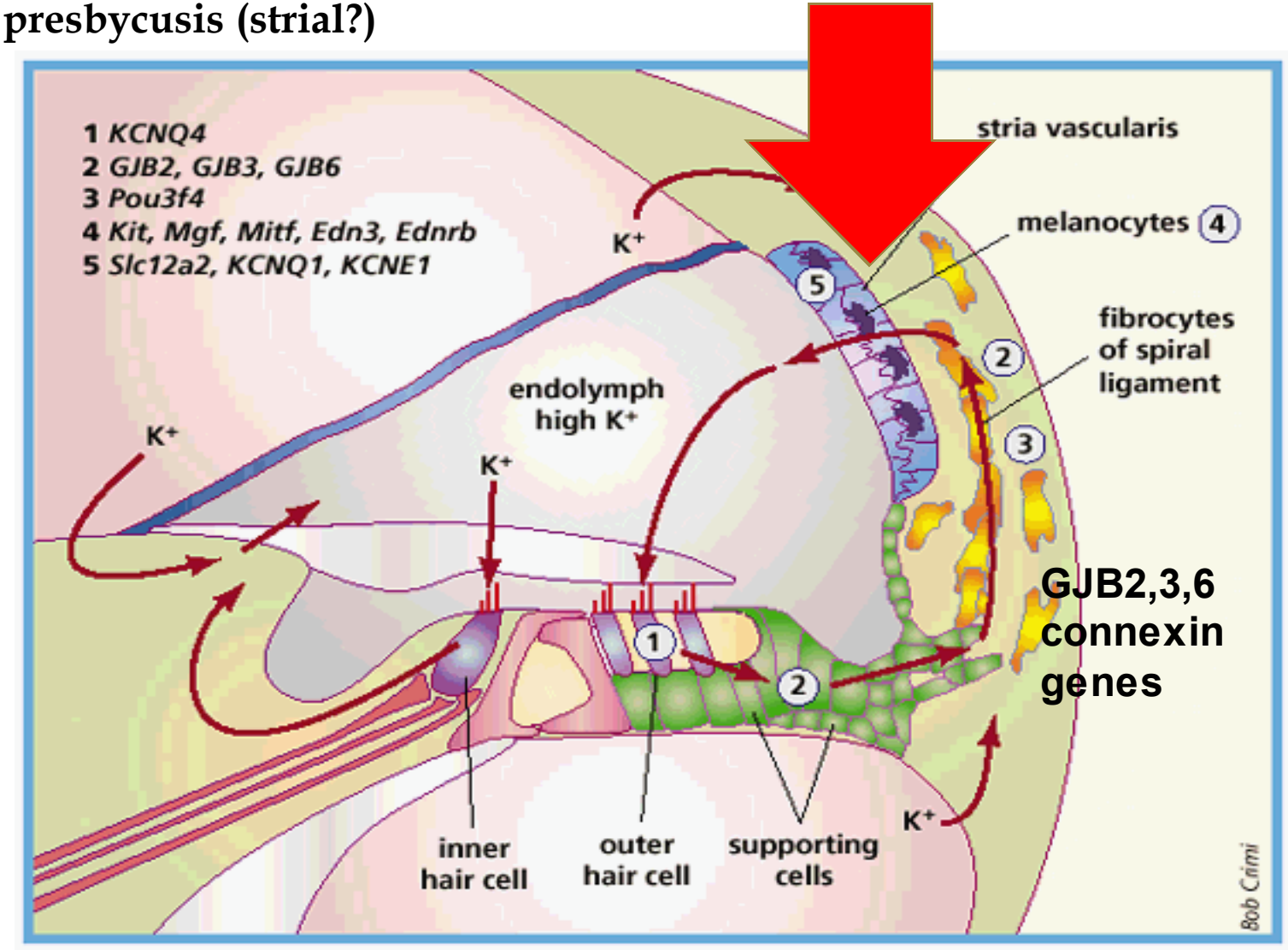
From studies by Dr. H. Spoendlin



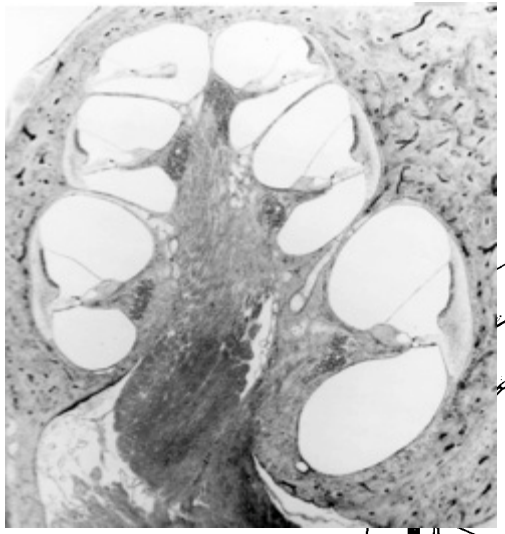
Inner haircell loss causes spiral ganglion cell degeneration AND degenerative change in second and third order central auditory neurons

THIS IS, OF COURSE, A MAJOR ISSUE IN COCHLEAR IMPLANTATION

Progressive loss has many causes, some of which start at the level scala media and stria vascularis, e.g. connexin (and many other) gene mutations, enlarged vestibular aqueduct, CMV infection, strial presbycusis (strial?)



Cochlear damage CASCADE effect



Strial dysfunction

Loss of endocochlear potential

Outer haircell dysfunction

Loss of cochlear amplifier
Degeneration inner haircells

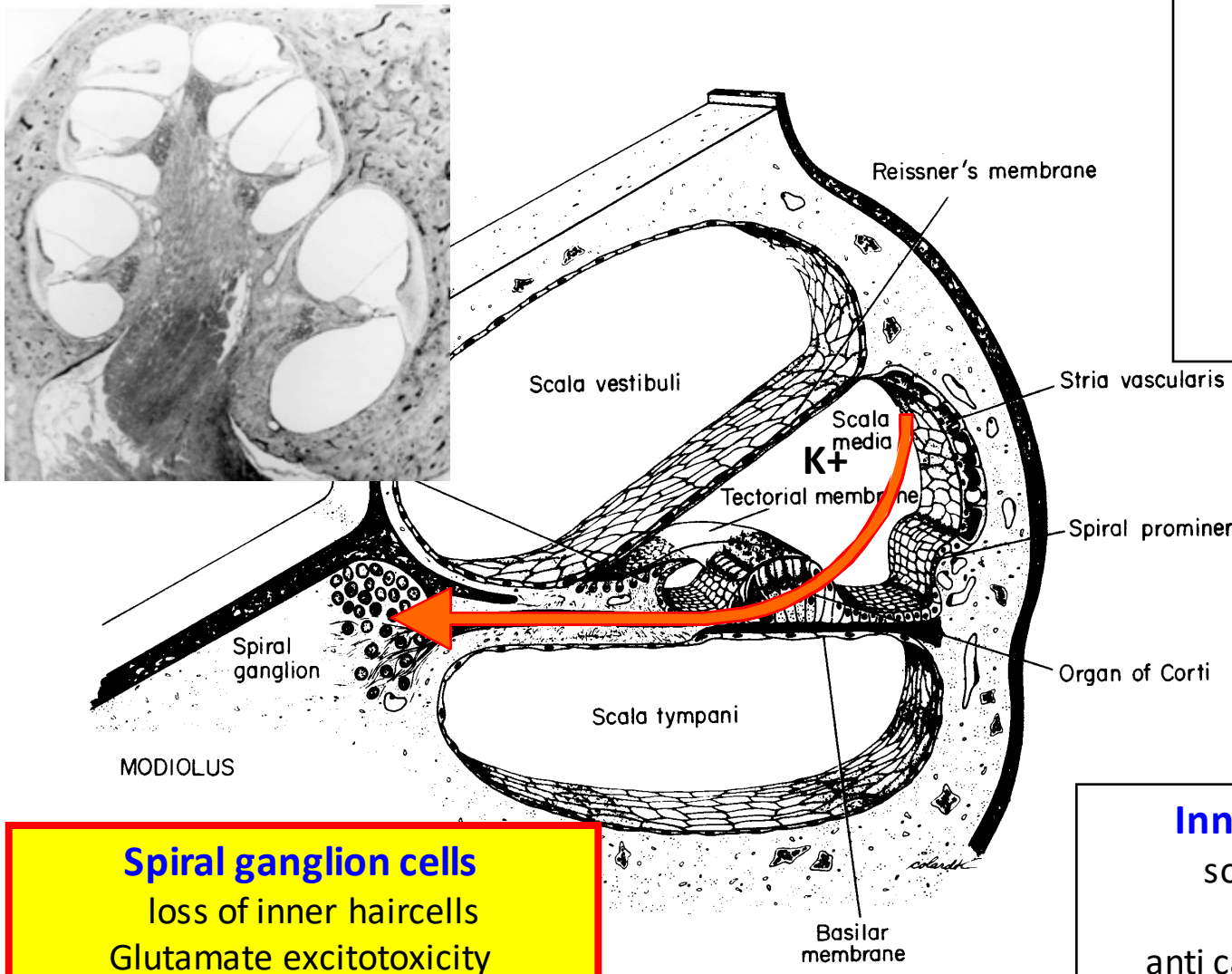
Inner haircell synaptopathy

Degeneration of neural connections with spiral ganglion cells

Spiral ganglion cell loss

progressive neuropathy in second and third order neurons

Cascading progression of cochlear degeneration



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some drugs e.g. aspirin
chronic hypoxia
anti cancer drugs – carboplatin
noise exposure
(sometimes there is recovery)

Take home message #2

Many ototoxic insults can cause haircell degeneration.

The pattern of haircell damage can take many forms depending on etiology.

For some ototoxic agents the initial damage is to OHC, for others IHC degeneration, for some causes both.

Functional deficits depend on type and pattern of haircell loss.

Perhaps we could define some distinct sub-classes:

"outer haircell SNHL"; "inner haircell SNHL"

"inner haircell synaptopathy" (Liberman and Kujawa)

Towards an improved classification of SNHL

We have studied the pathogenesis of sensorineural hearing loss In a range of animal models. Some of my own work presented today.

SNHL has many different causes, many sites, patterns and sequences of cochlear damage, resulting in many forms of hearing dysfunction

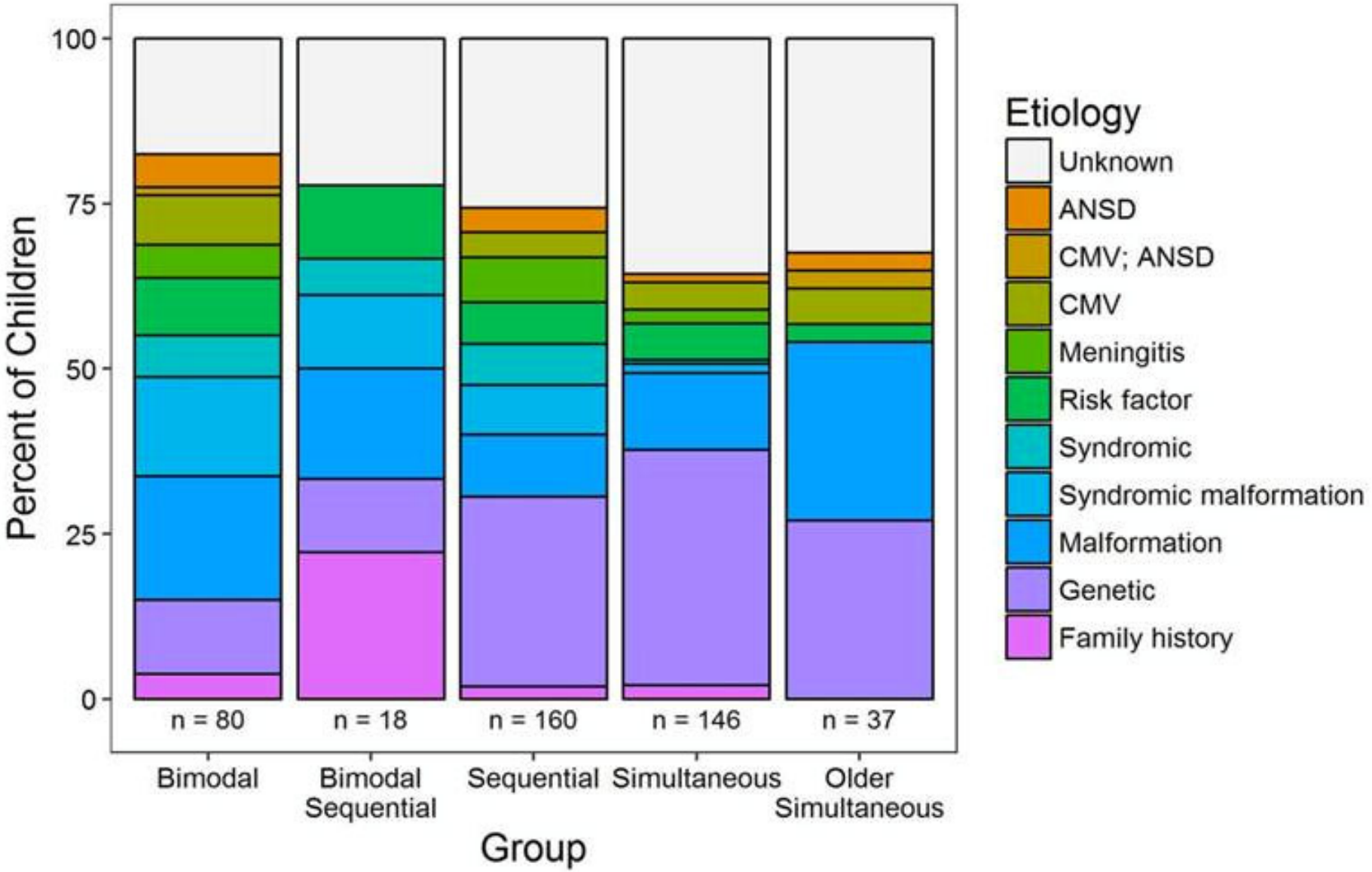
Sensorineural hearing loss is a SPECTUM DISORDER!

We need to pay more attention to etiology, and from there understand the pattern or sequence of cochlear damage. Then we can consider the very DIFFERENT functional consequences of each lesion type.

OVERALL THESIS:

If we pay more attention to etiology we can distinguish classes or types of SNHL. This in turn will narrow our focus on treatment and rehabilitation strategy and provide more accurate prognosis.

Classification of children with cochlear implants at the Hospital for Sick Children, Toronto



A Comprehensive Study on the Etiology of Patients Receiving Cochlear Implantation With Special Emphasis on Genetic Epidemiology

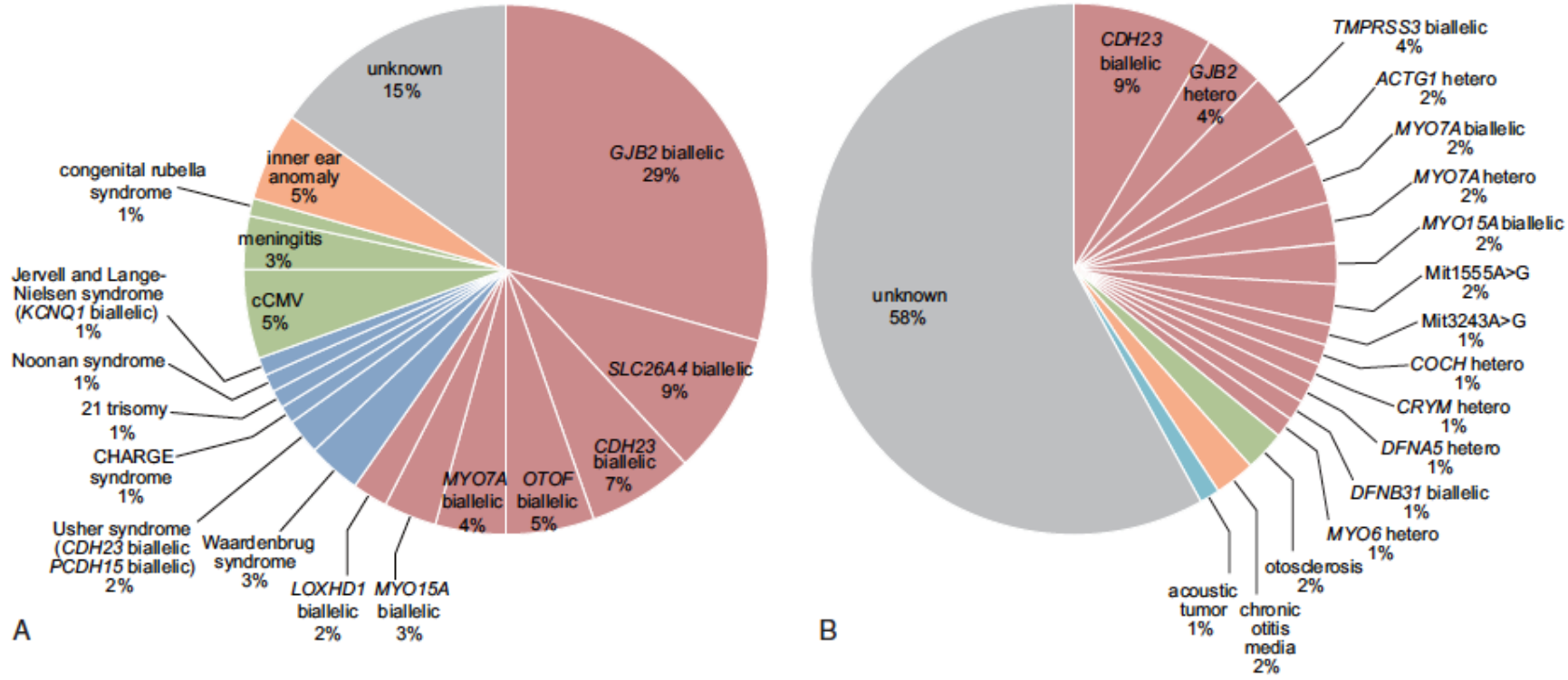


FIG. 1. Etiology of CI/EAS patients. (*A*) Prelingual CI/EAS patients (n = 88). Blue indicates syndromic hearing loss; gray, unkown; green, infection-induced hearing loss; orange, inner ear anomaly; pink, nonsyndromic hearing loss associated with specific gene mutations. (*B*) Postlingual CI/EAS patients (n = 77). Blue indicates acoustic tumor; gray, unkown; green, otosclerosis; orange, otitis media; pink, nonsyndromic hearing loss associated with specific gene mutations.

Maiko Miyagawa, Shin-Ya Nishio, and Shin-Ichi Usami (Matsumoto Japan)
Otol Neurotol. 2016 Feb;37(2):e126-34.

Annual meeting CAA 2018. Niagara Falls. October 2018

Towards an Improved Sub-Classification of SNHL Spectrum Disorder

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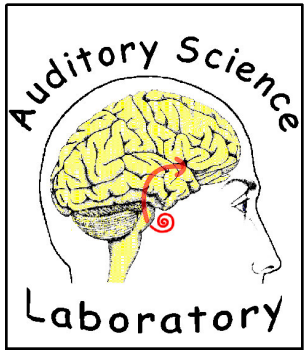
University of Toronto

Director, Auditory Science Laboratory,
Program in Neuroscience and Mental Health
The Hospital for Sick Children, Toronto



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Acknowledgements

Residents and fellows

Peter White
Mark Wake
Michael Ruckenstein
Jafri Kuthubutheen
David Clinkard
Amoodi H,
Kansandamy T,
Grewal AS,
Stephen Chen
Qian W
Noam Harel
Makoto Shirane,
Noriyuki Fukushima,
Akira Nagasawa,
Sachio Takeno,
Akinobu Kakigi,
Haruo Hirakawa
Shoichi Sawada.
Naoki Mori
Uijimoto Konomi

Graduate trainees

Daniel Ibrahim
Hormoz Hamrahi
Mattia Carraro
Ali Almishaal
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Technologists:

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Tracy Stockley
Blake Papsin
Vincent Lin
Joe Chen
Vera Prijs (Leiden)
George Tavarkiladze (Moscow)
Albert Park (Utah)
Matthew Firpo (Utah)
Ted Evans (Keele, UK)
Michael Portmann (Bordeaux)
Jean-Marie Aran (Bordeaux)
Rene Dauman (Bordeaux)

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