

Hearing and Balance: Impact and Outcomes of Dual Sensory Impairments in the Pediatric Population With Sharon Cushing – 7 pm ET

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Hearing and Balance: Impact and Outcomes of Dual Sensory Impairments in the Pediatric Population

Sharon L. Cushing MD MSc, FRCSC,
Otolaryngologist, Hospital for Sick
Children

Oct 20th, 2020

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Canadian Academy of Audiology is a professional association dedicated to enhancing the role of audiologists as primary hearing health care providers through advocacy, education and research.

Salima Jiwani - Host



Dr. Salima Jiwani is a Past President of CAA, and is the clinical and research Director of Audiology at AudioSense Hearing & Balance, a centre of excellence in Hearing and Balance Healthcare.

Salima earned her Ph.D. in auditory neurophysiology with the Institute of Medical Science at the University of Toronto.

Speaker: Sharon L. Cushing MD MSc



Dr. Sharon Cushing is a full time paediatric otolaryngologist at The Hospital for Sick Children in Toronto, and an Associate Professor and Clinician Investigator at the University of Toronto.

She is the Director of the Cochlear Implant Program at the Hospital for Sick Children. Dr. Cushing has a clinical and surgical interest in disorders of the external, middle and inner ear, including hearing loss and vestibular dysfunction. Her research interest include vestibular and balance function and dysfunction in children, and its association with hearing loss and cochlear implantation.

Hearing and Balance: Impact and Outcomes of Dual Sensory Impairments in the Pediatric Population

Sharon L. Cushing



Disclosures

- Speaker's Bureau
 - Interacoustics, Cochlear Corporation
- Royalties
 - Plural Publishing
 - Editor: Balance Disorders in the Pediatric Population
- Patent Holder:
 - Patents #: 7041-0: Systems And Methods For Balance Stabilization
- Sponsored Research Agreement
 - Cochlear Americas

Cochlear Implant Research Team

SickKids®



DIRECTORS

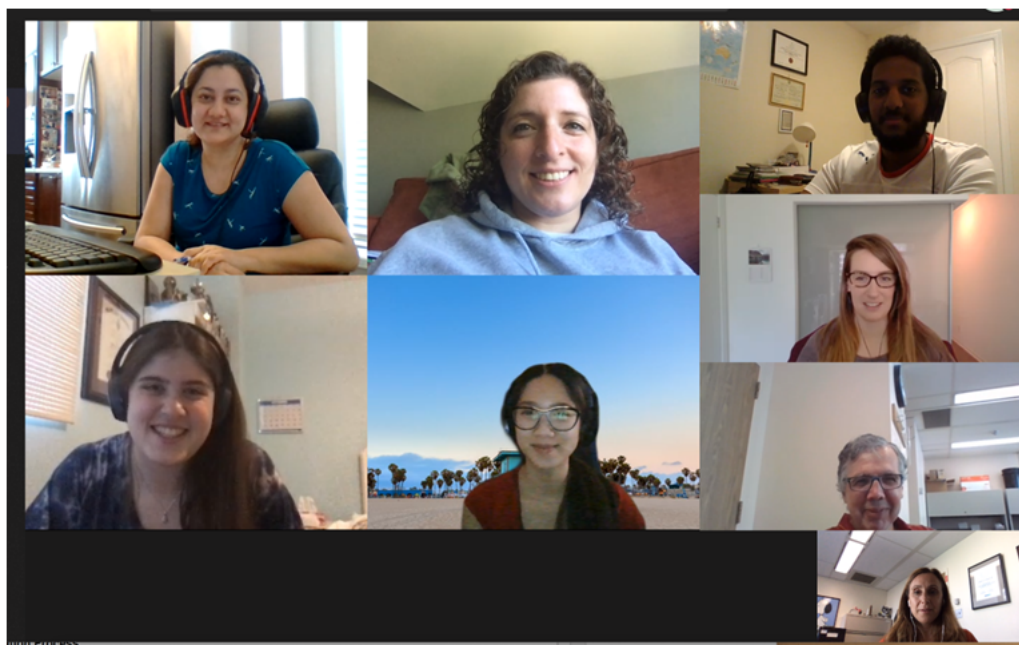
- Karen Gordon
- Blake Papsin
- Sharon Cushing

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- Carmen McKnight
- Christina Lavallee

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- Salima Jiwani
- Melissa Polonenko
- Nikolaus Wolter
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FUNDING

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- Susan Blaser
- Adrian James
- Sam Doesburg
- Vicky Papaioannou

Local - External

- Sandra Trehub
- Frank Russo

International

- Robert Cowen
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Cochlear Implant Team



- | | | | |
|---------------------|--------------------|--------------------|---------------------|
| • Blake Papsin | • Gina Goulding | • Mary Lynn Feness | • Valerie Simard |
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Making Sense of the World



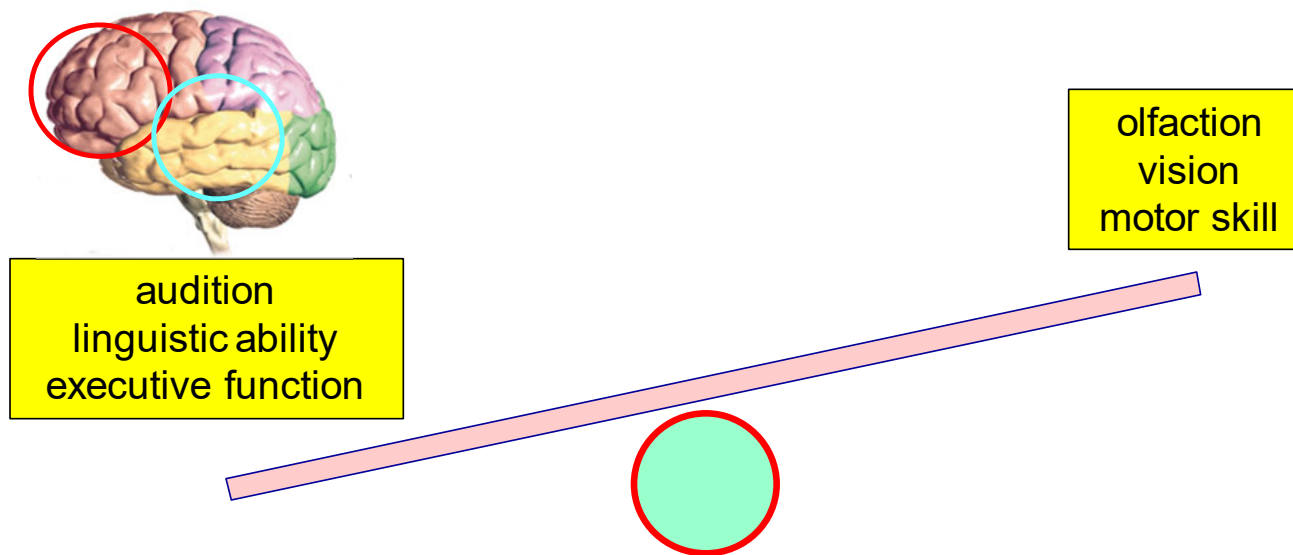
Making Sense of the World - Perspective



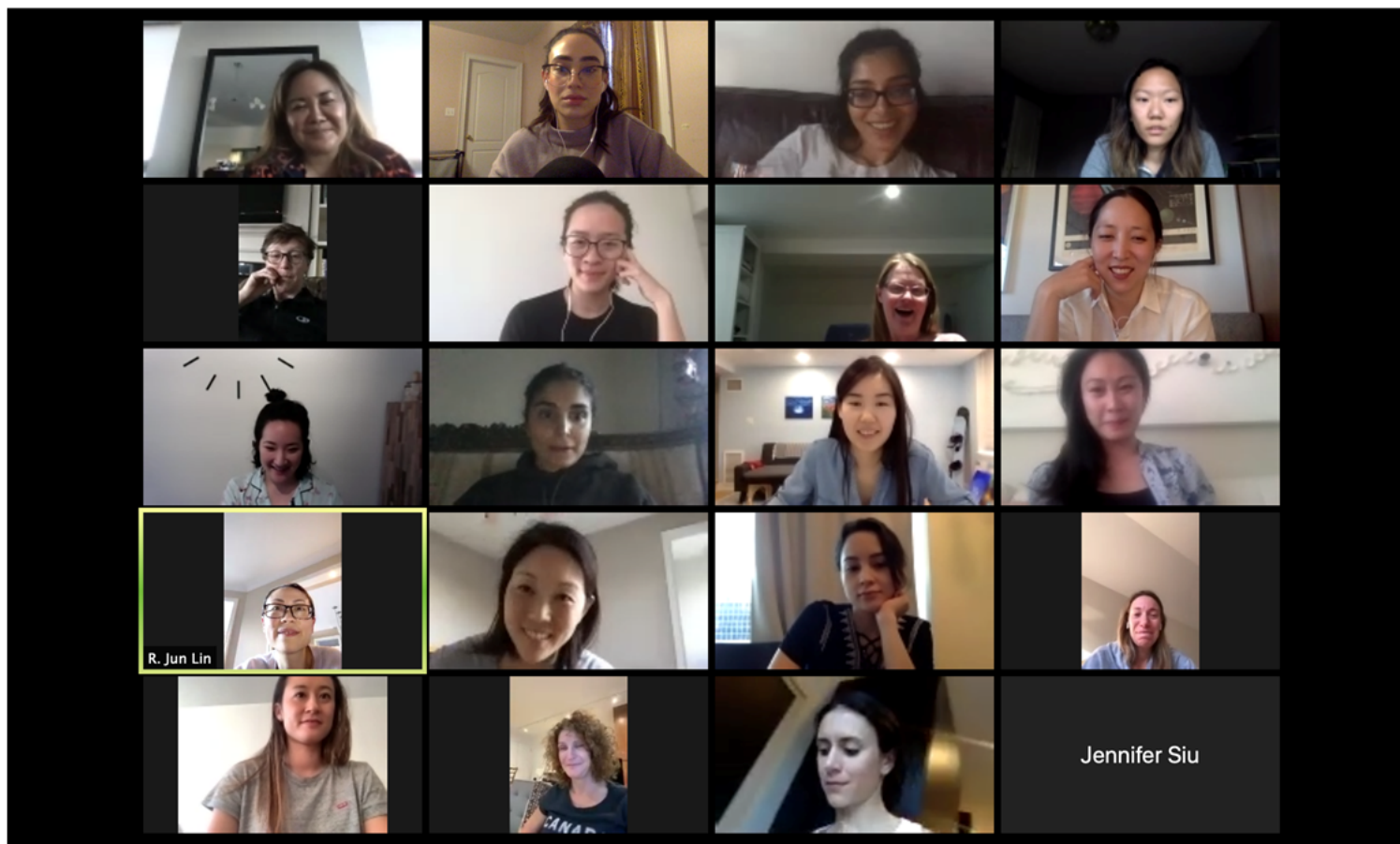
Sensory Interaction / Integration



Human = Language Predators

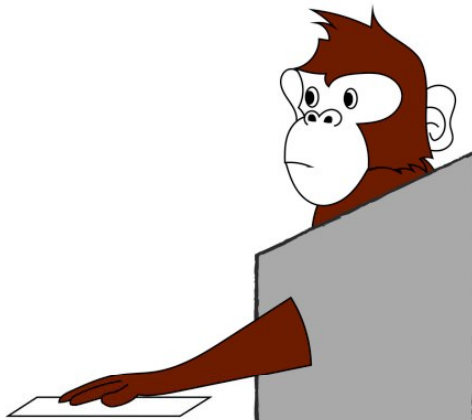


Connecting During a Pandemic

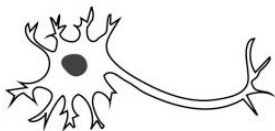


Wired for Connection – Mirror Neurons

(A) Monkey at rest

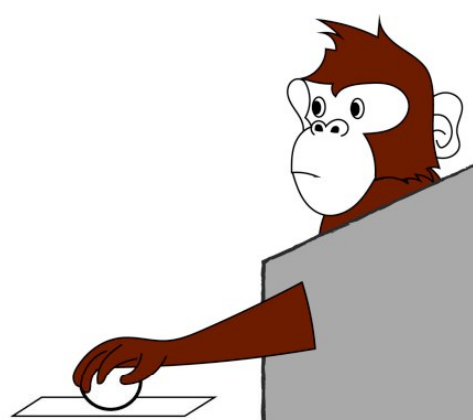


A mirror neuron is in a **resting state**

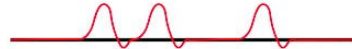


(No electrical signals)

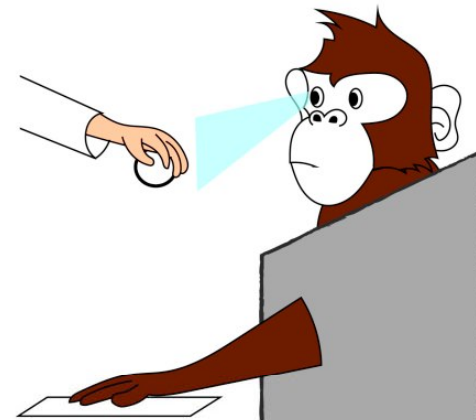
(B) Grasping execution



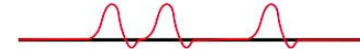
A mirror neuron **fires**



(C) Observation of grasping movements



A mirror neuron **fires**



Restoring Connection with Cochlear Implants: What are we missing?



Sharon L. Cushing

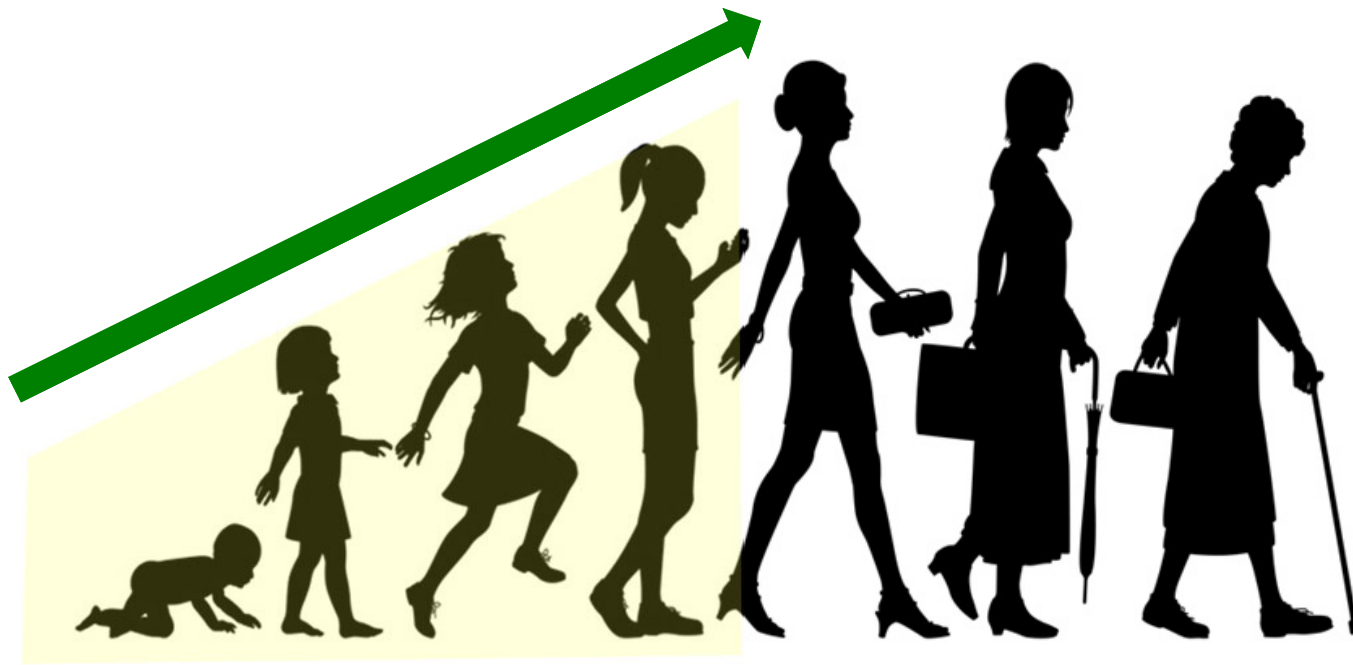




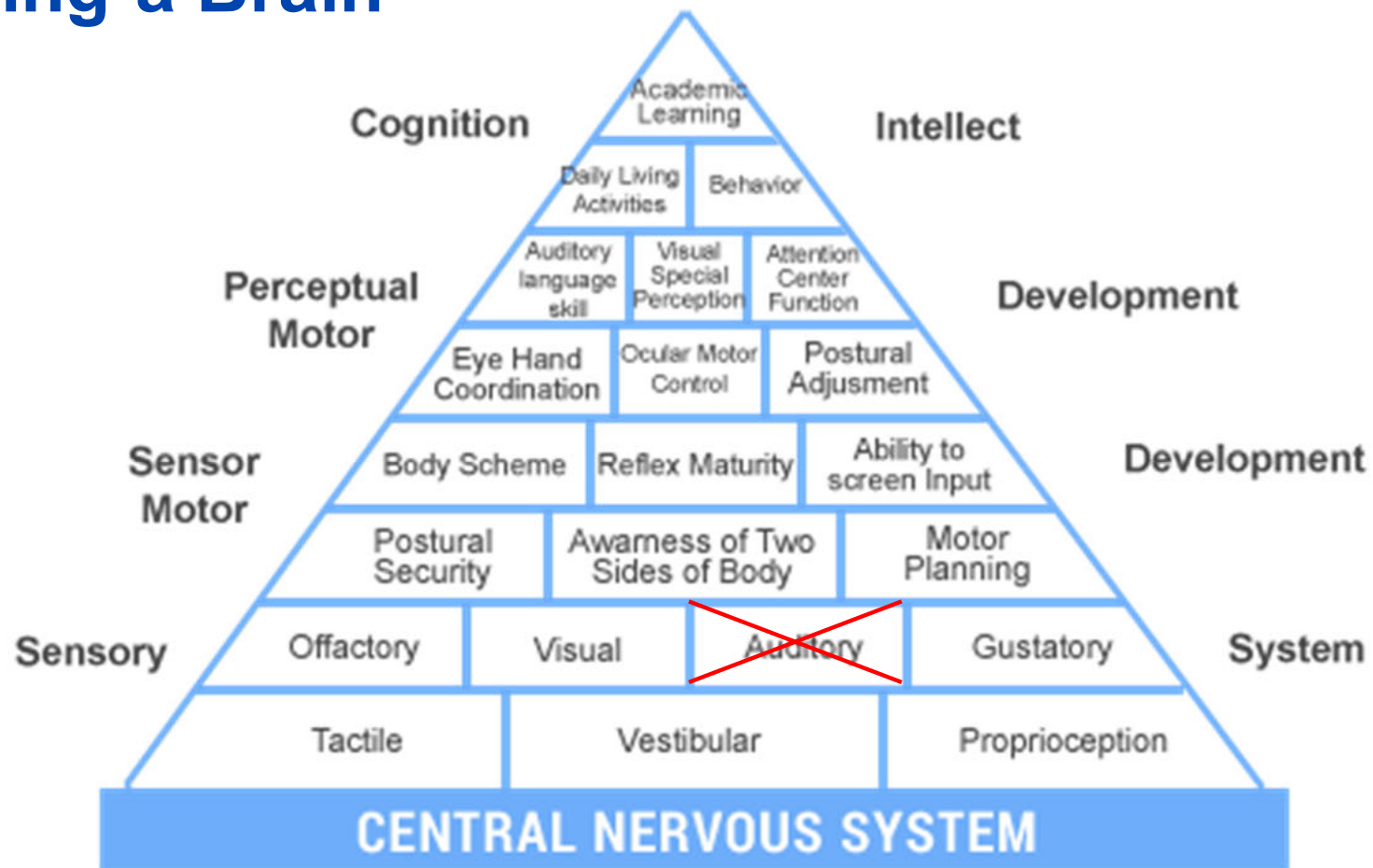
Cognitive Decline



Cognitive Incline

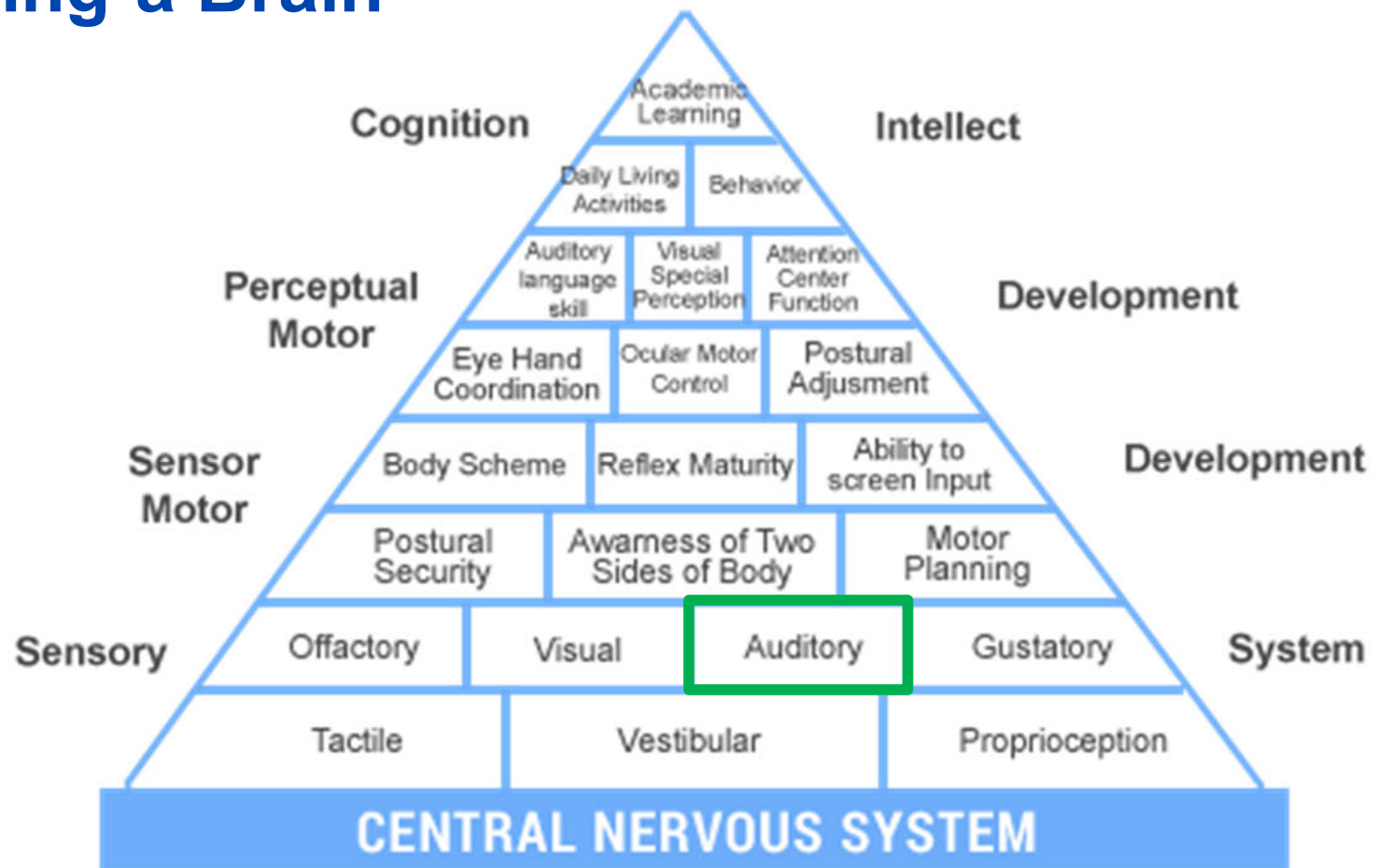


Building a Brain



Pyramid of Learning. (Williams & Shellenbeger, 1-4)

Building a Brain



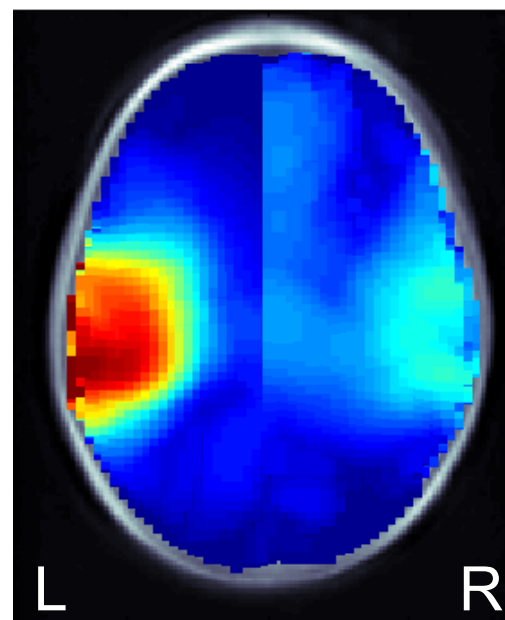
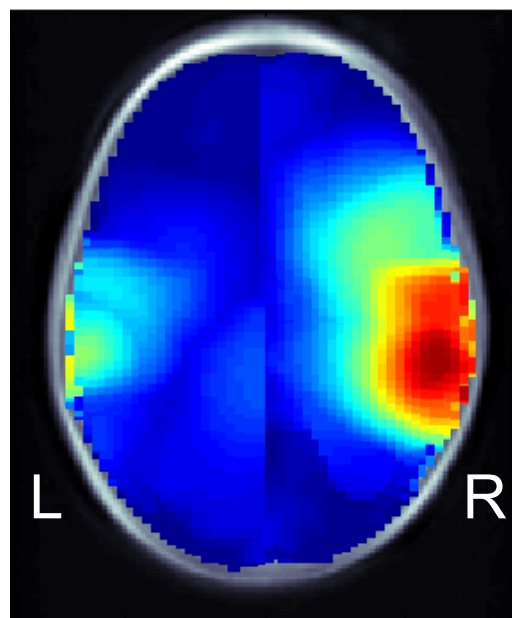
Pyramid of Learning. (Williams & Shellenbeger, 1-4)

Normal Aural Preference

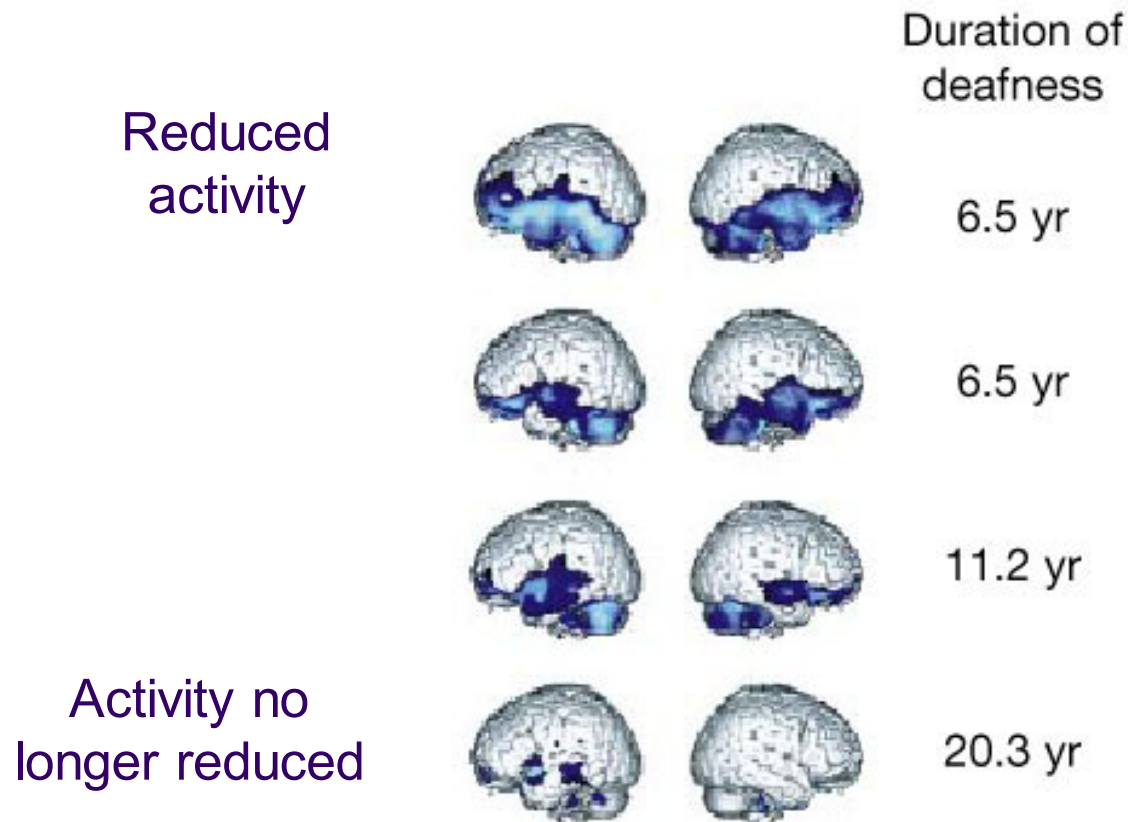
Pi



Click
(n=5)

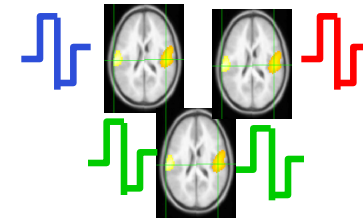
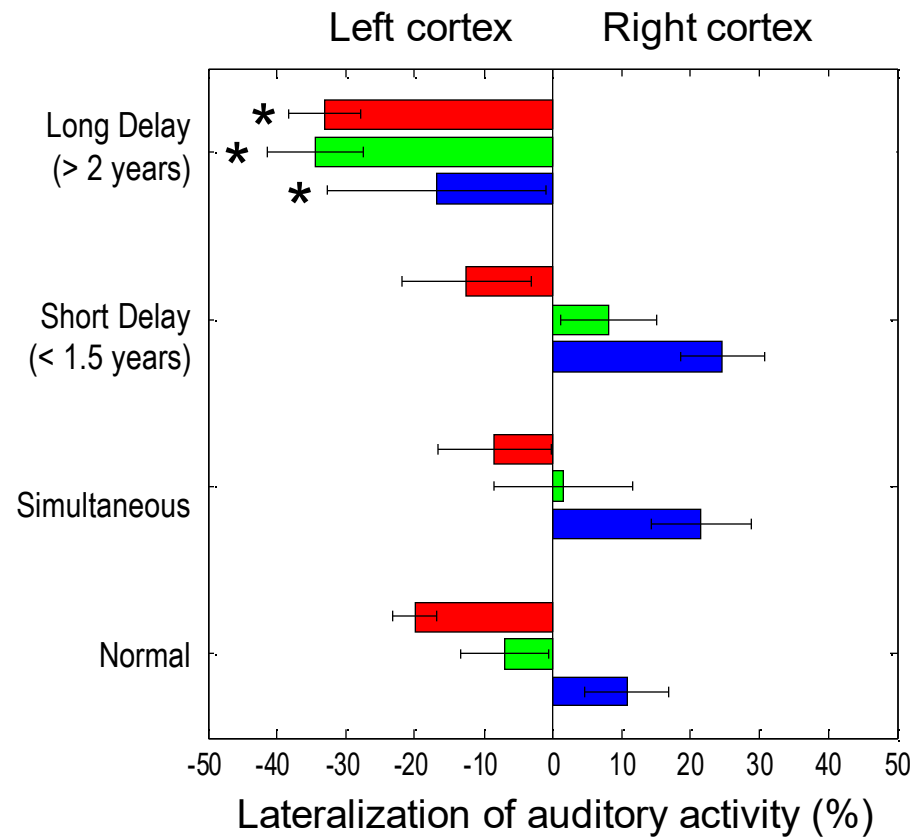


Auditory Deprivation



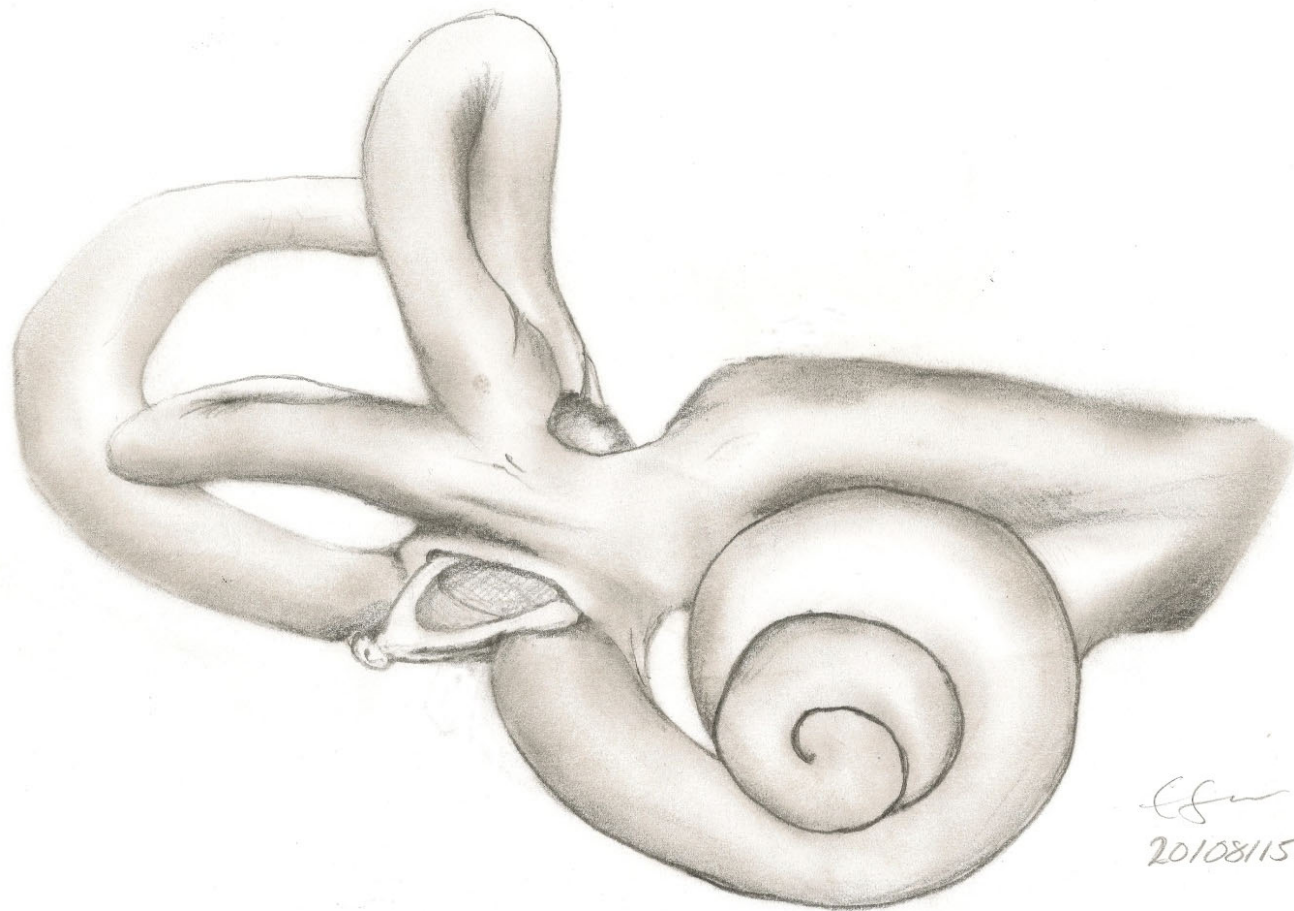
Lee, et al., Nature, 2001

Abnormal Aural Preference



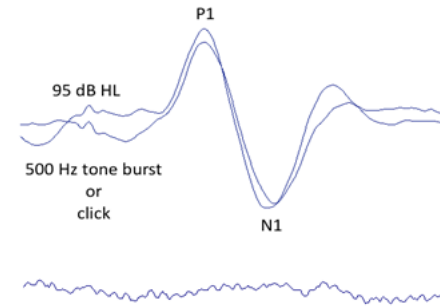
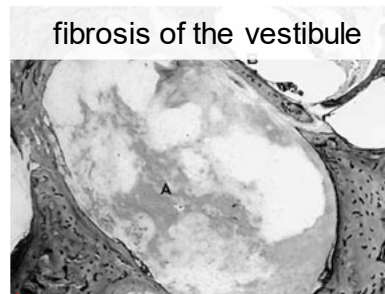
* P<0.05



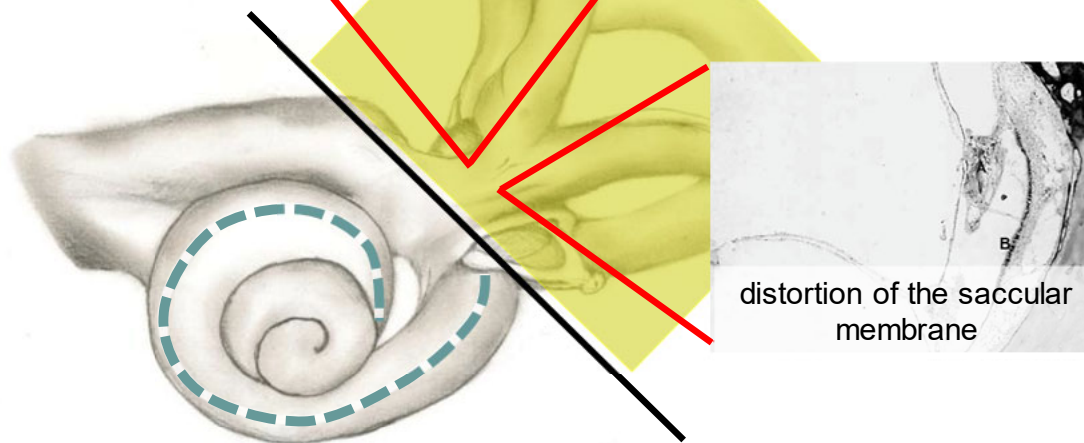


efw
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Vestibular Effects of Cochlear Implantation (*negative*)

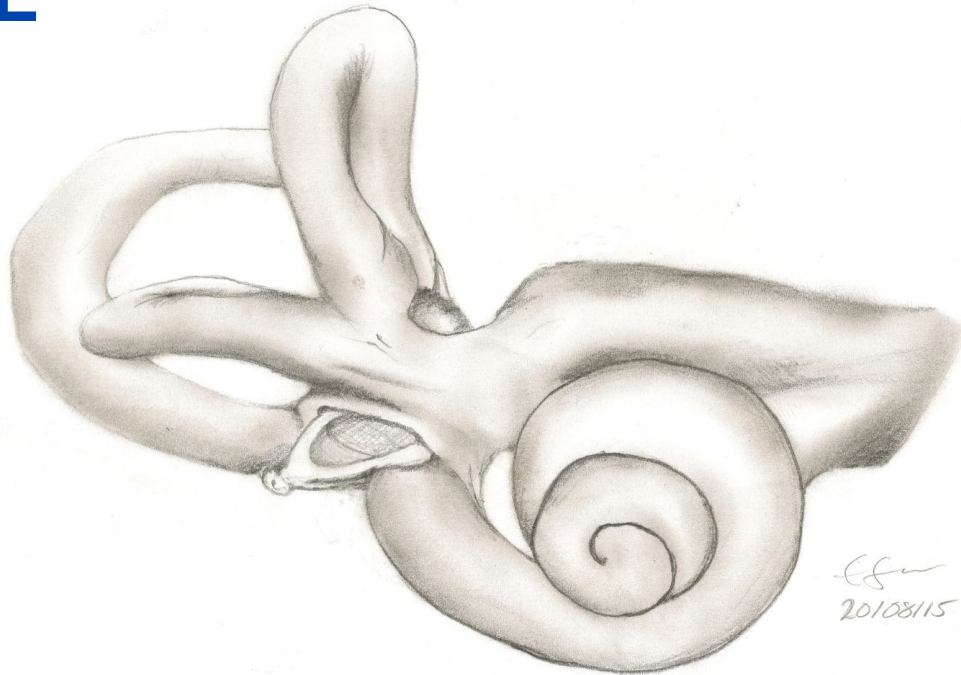


20-100% lose cVEMP
responses after CI



Vestibular Dysfunction is Common in Children with SNHL

70% dysfunction
35-40% complete
areflexia



vestibular impairment is the single
most common associated feature of SNHL

Otol Neurotol. 2013

Measuring Balance

Bruininks-Oseretsky (BOT2) Balance Subtest

Balance subtest Items		Max. Score
1. Standing with feet apart on a line	Eyes Open	10 sec.
	Eyes Closed	10 sec.
2. Walking forward on a line		6 steps
3. Standing on one leg on a line	Eyes Open	10 sec.
	Eyes Closed	10 sec.
4. Walking forward heel to toe on a line		6 steps
5. Standing on one leg on a balance beam	Eyes Open	10 sec.
	Eyes Closed	10 sec.
6. Standing heel-to-toe on a balance beam		10 sec.

N= 190 children with CI tested



Vestibular Loss Impacts Balance

balance deficits are not always apparent

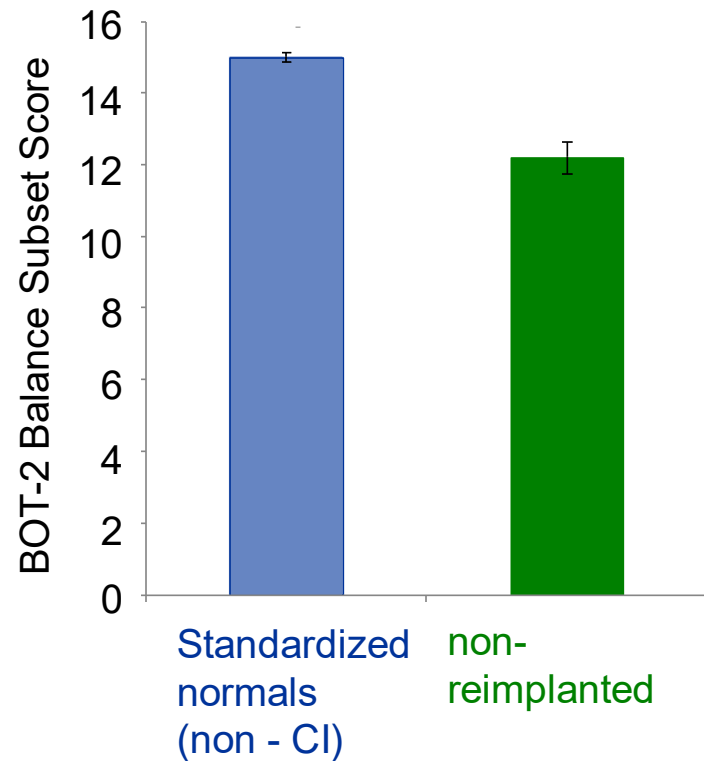




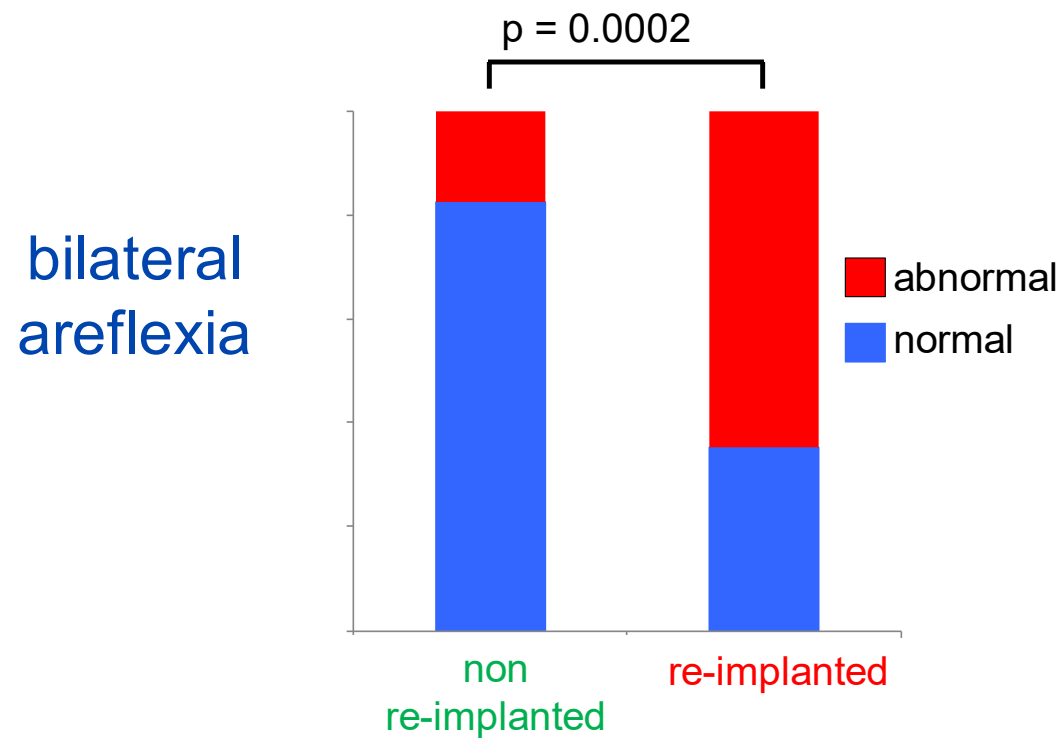


When Balance is Poor – Implants Fail

- static and dynamic balance – BOT-2



When Vestibular Function is Poor – Implants Fail



When Vestibular Function is Poor – Implants Fail

vestibular & balance
dysfunction

α

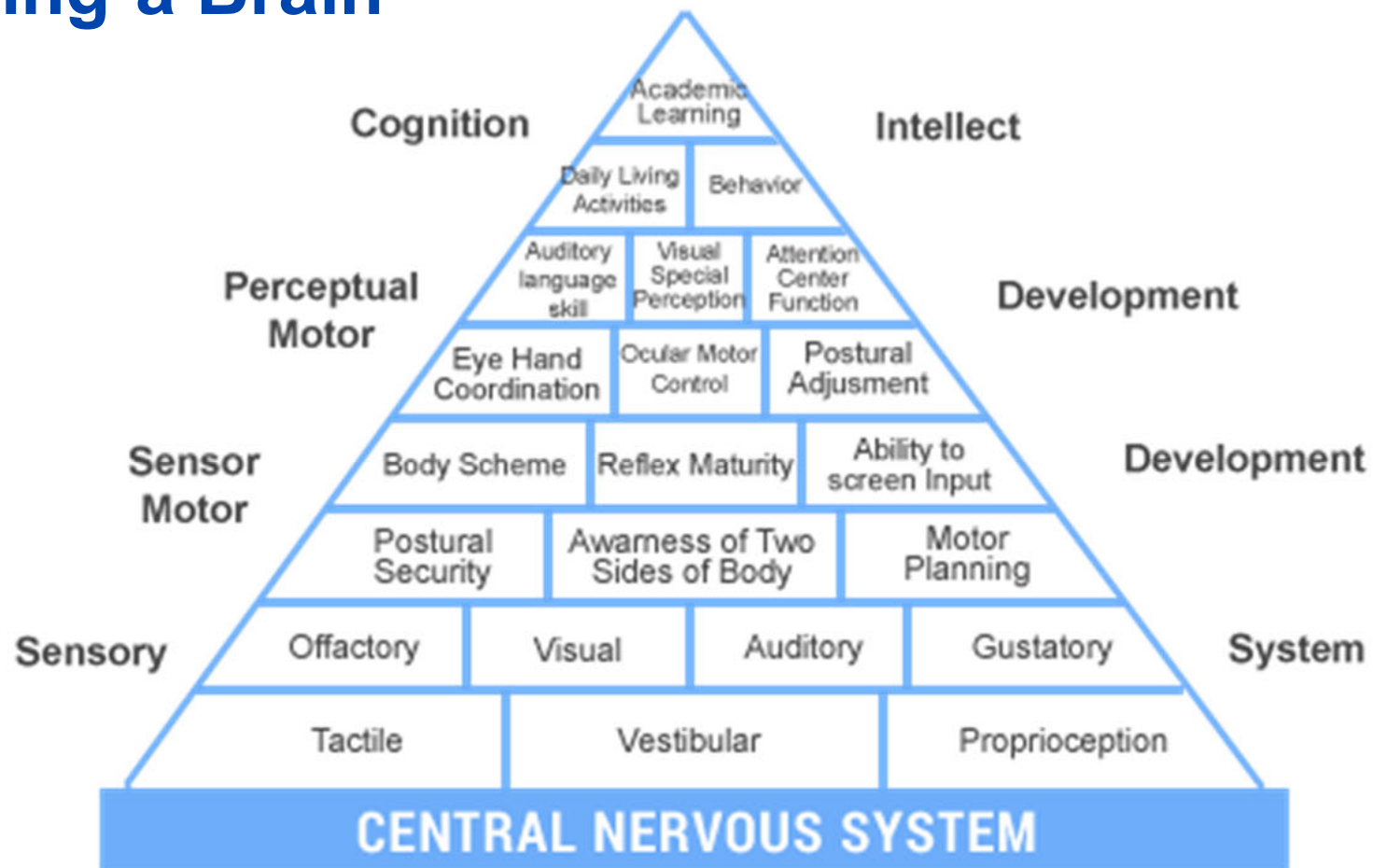
risk of CI failure

Bilateral Areflexia

Odds ratio: 8.6

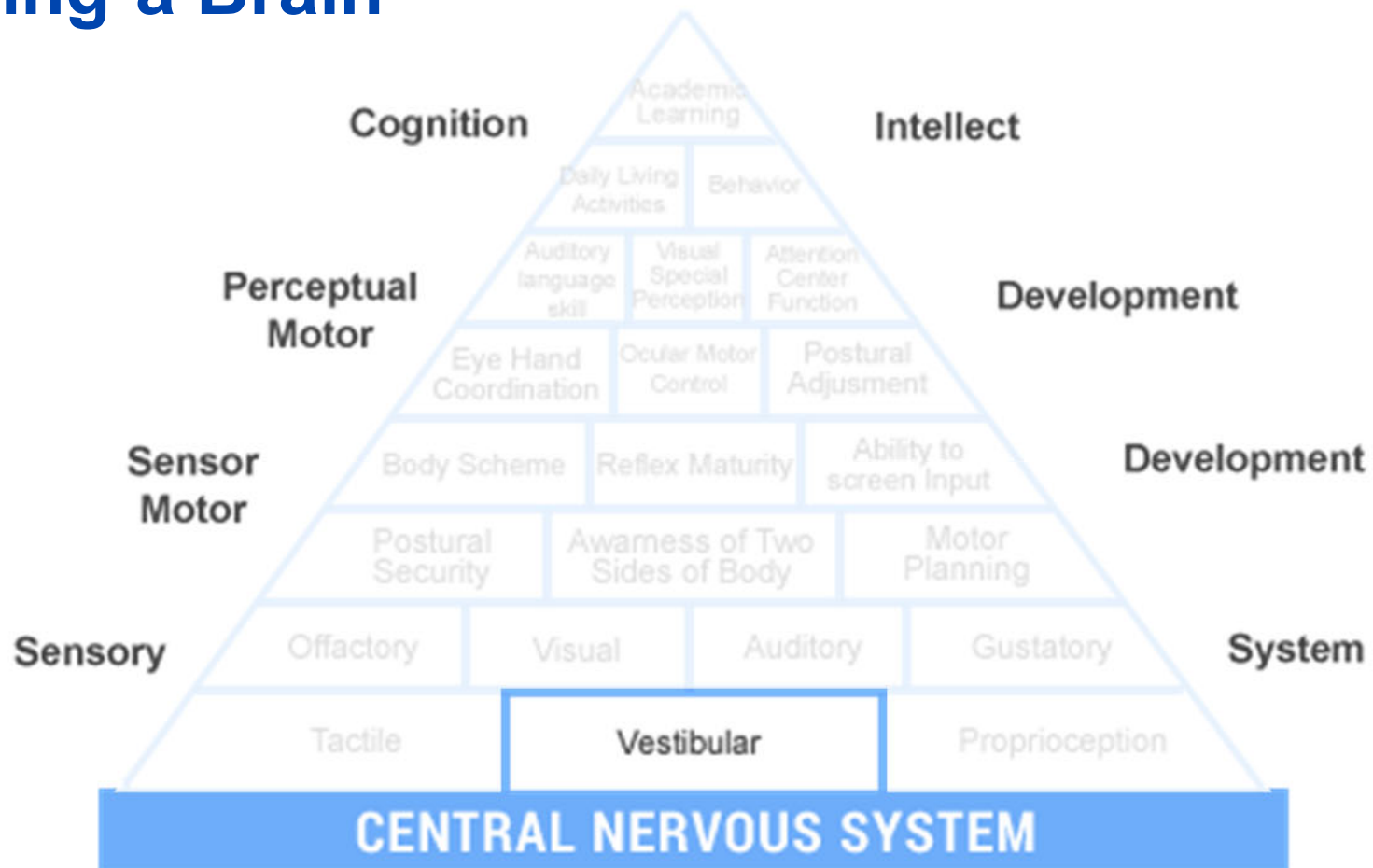
Implant failure

Building a Brain



Pyramid of Learning. (Williams & Shellenbeger, 1-4)

Building a Brain

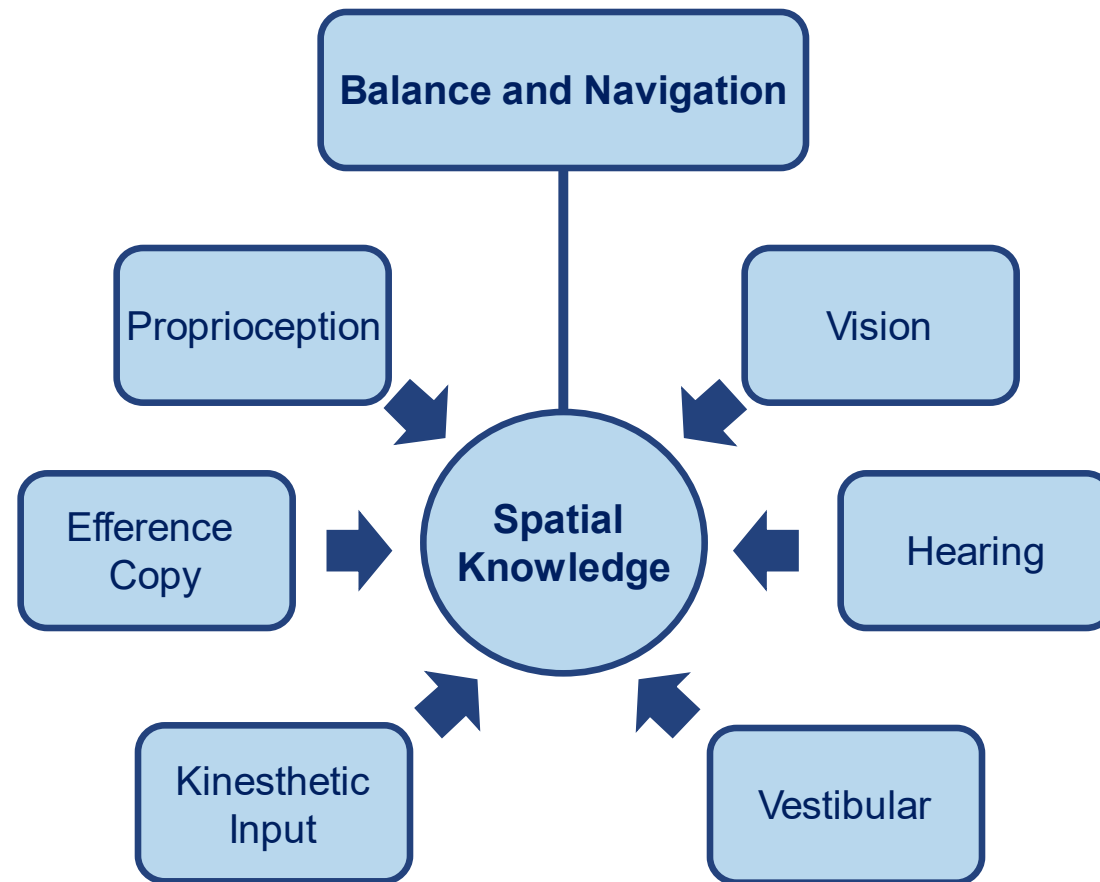


Pyramid of Learning. (Williams & Shellenbeger, 1-4)

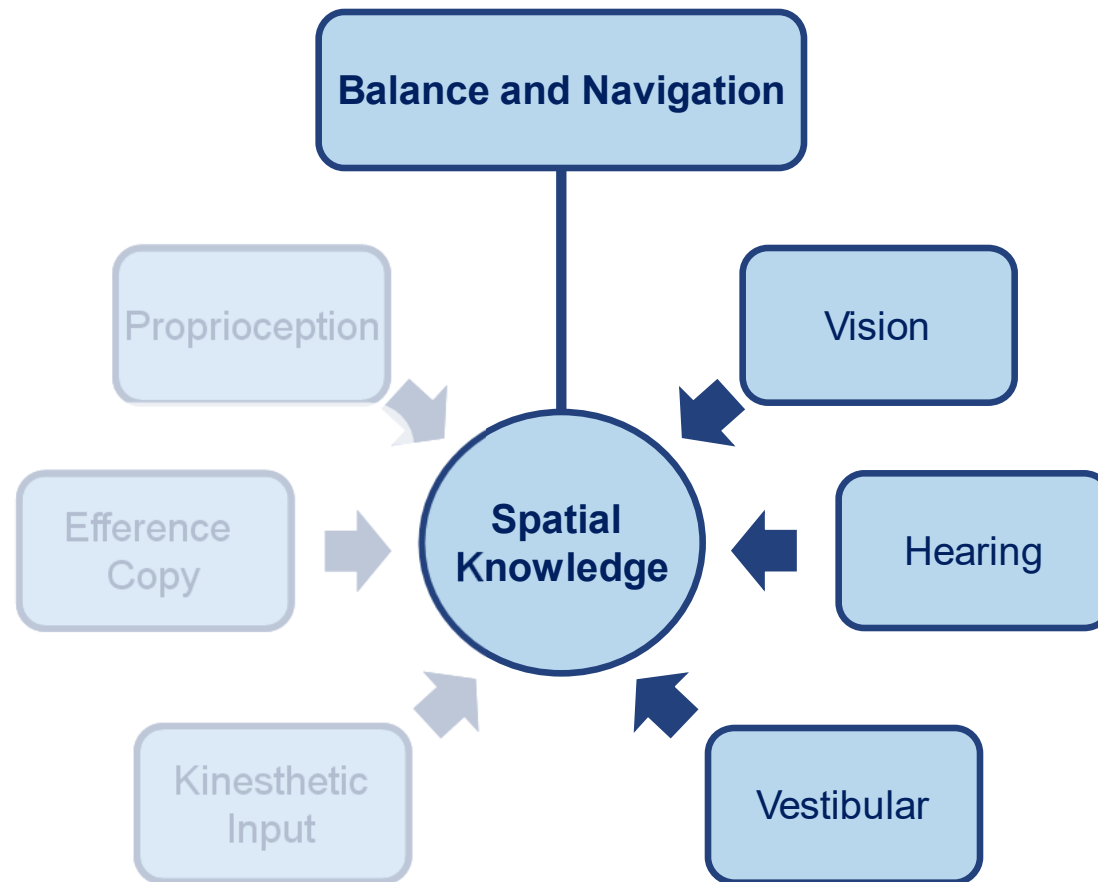
Balance and Navigation



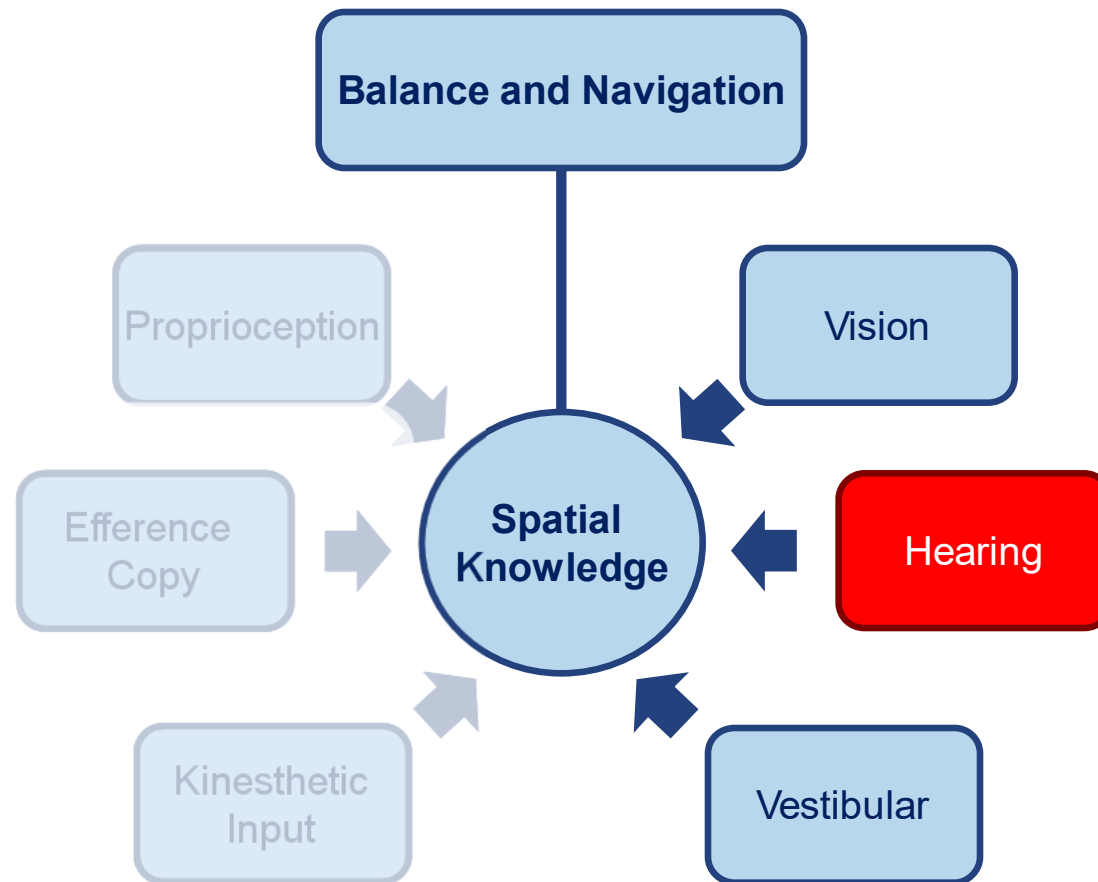
Development of Spatial Knowledge



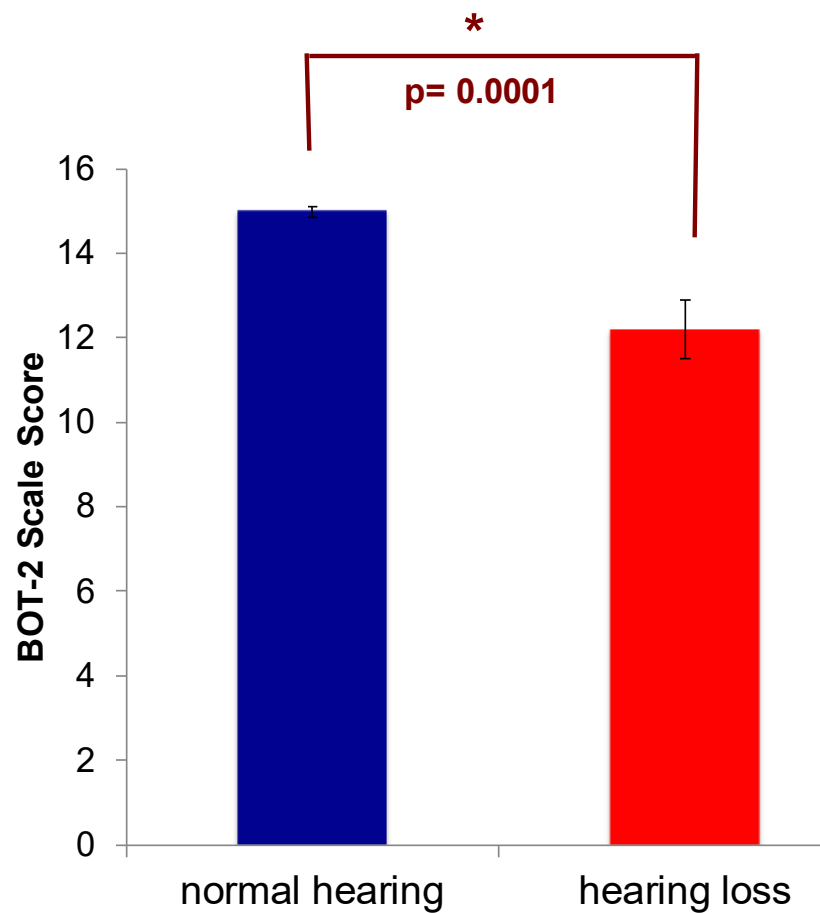
Development of Spatial Knowledge



Development of Spatial Knowledge



Hearing Loss Impacts Balance





Unilateral



Bilateral



Effects of Poor Spatial Hearing

Normal binaural hearing



Normal hearing (n=36)
 10.0 ± 3.3 years of age

Abnormal binaural hearing



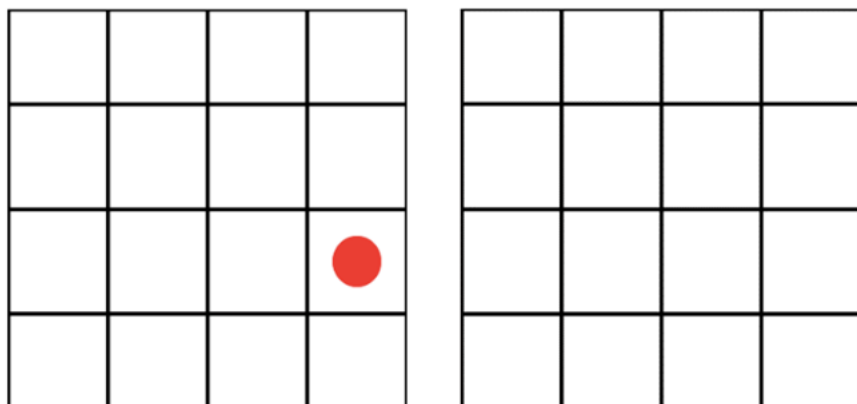
Bilateral CI (n=32)
 9.1 ± 3.0 years of age

No binaural hearing

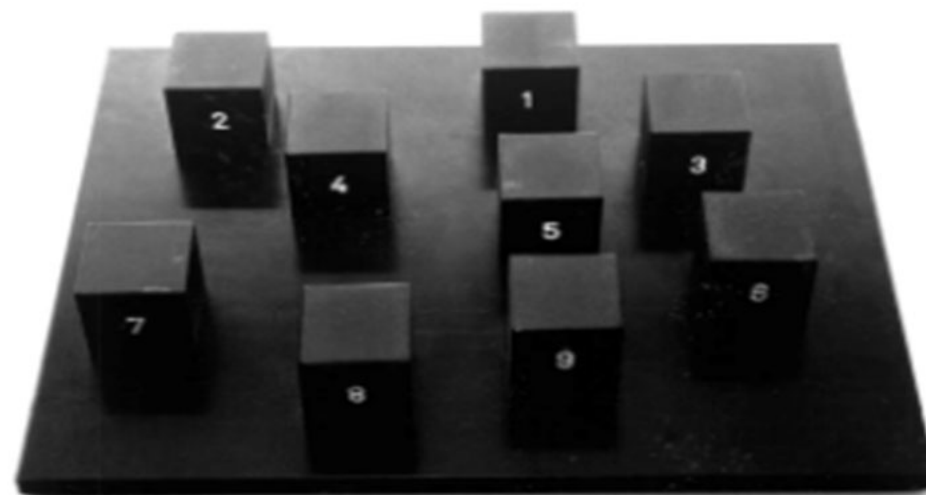


Untreated SSD (n=20)
 10.8 ± 4.1 years of age

Memory Deficits are Visuospatial



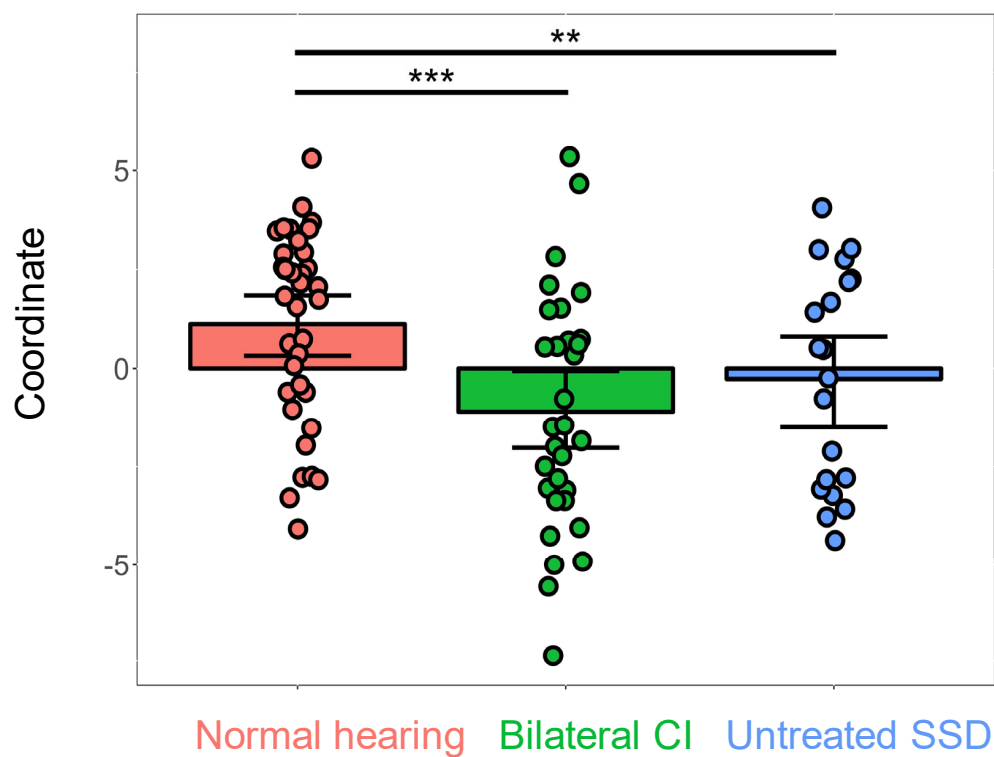
Dot Matrix Test



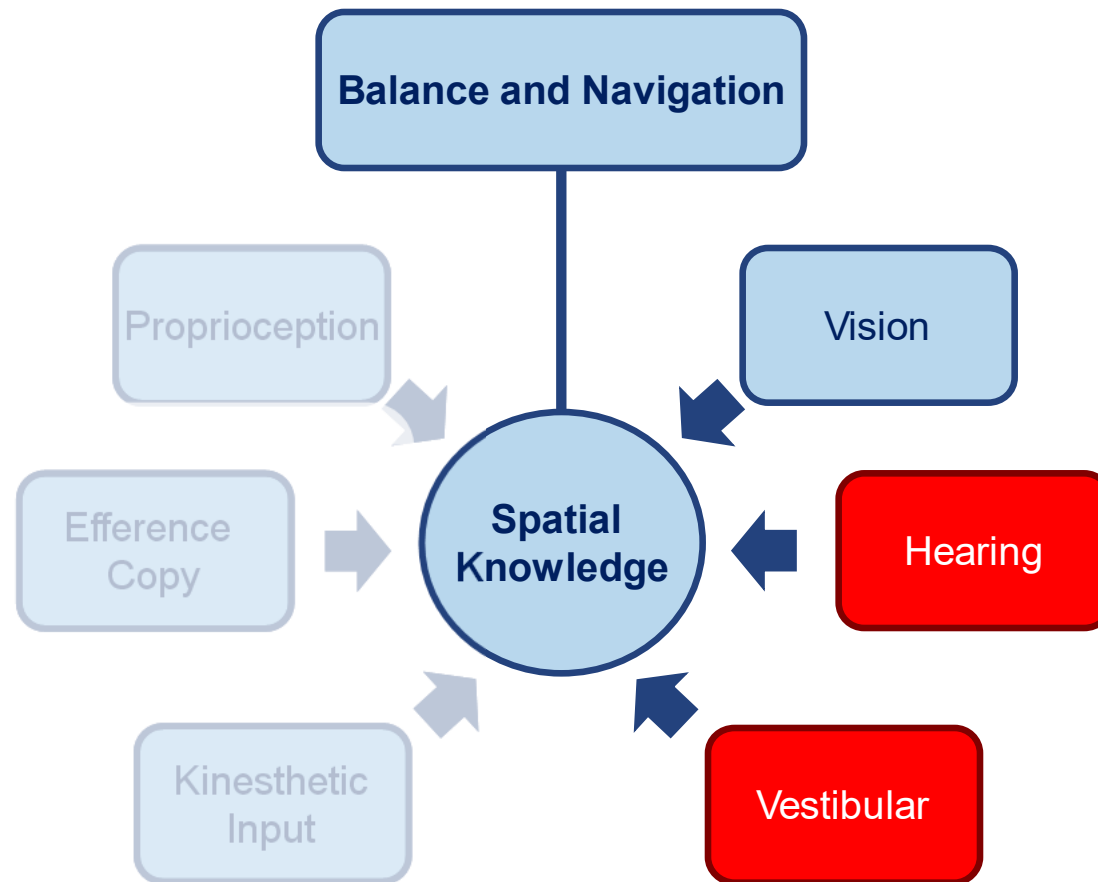
Corsi Block Test

Memory and Learning Deficits

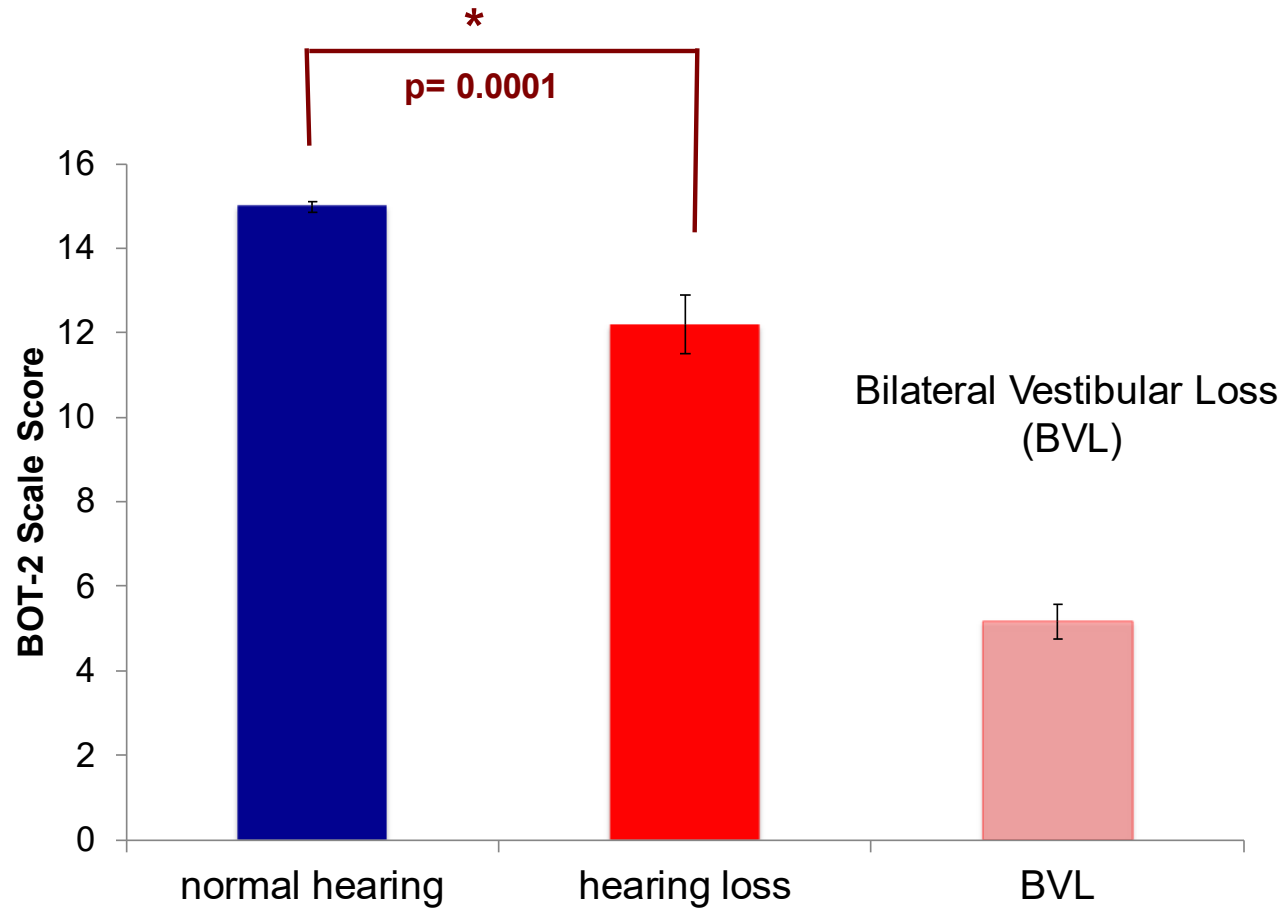
Component 1: Memory and Learning



Development of Spatial Knowledge

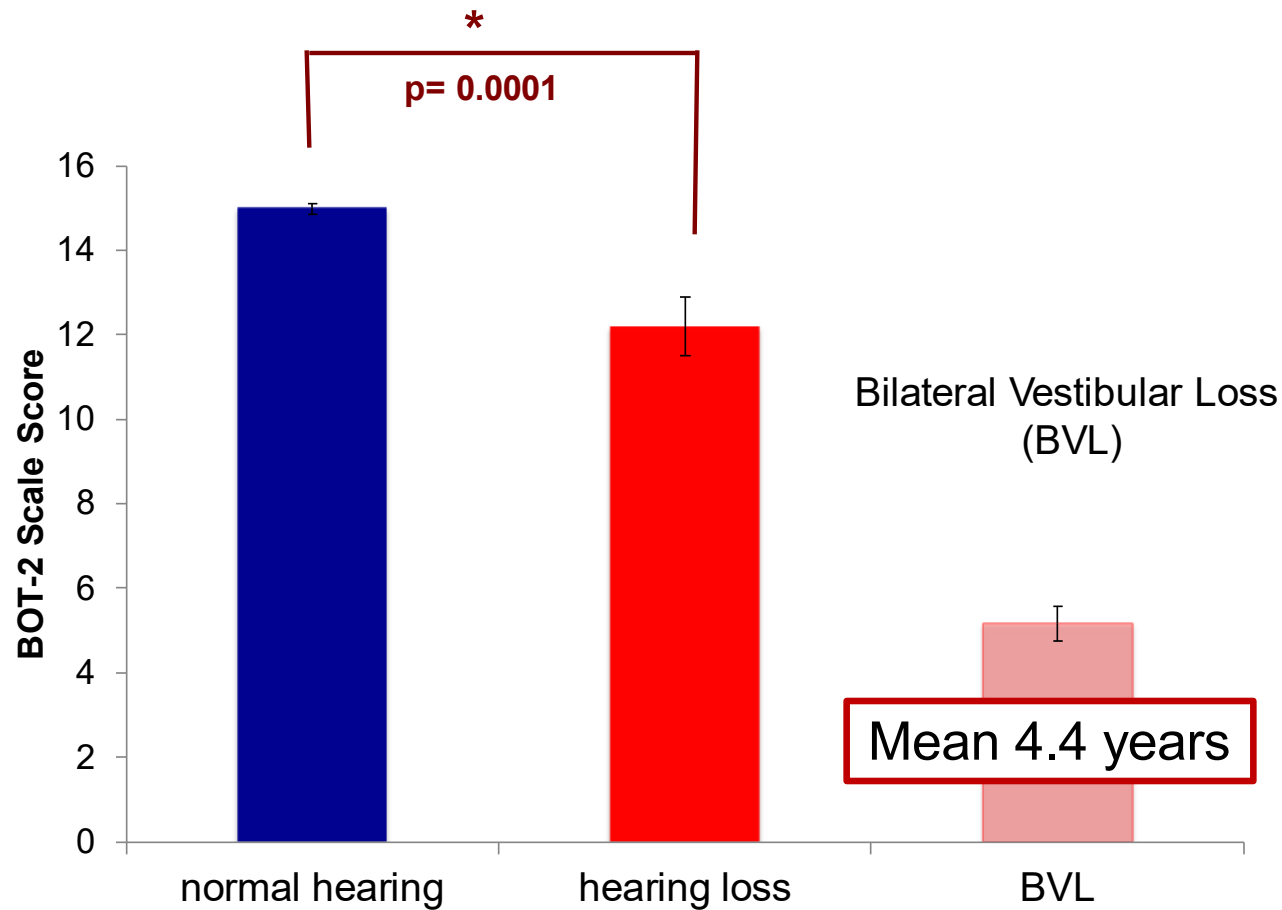


Sensory Loss Impacts Balance



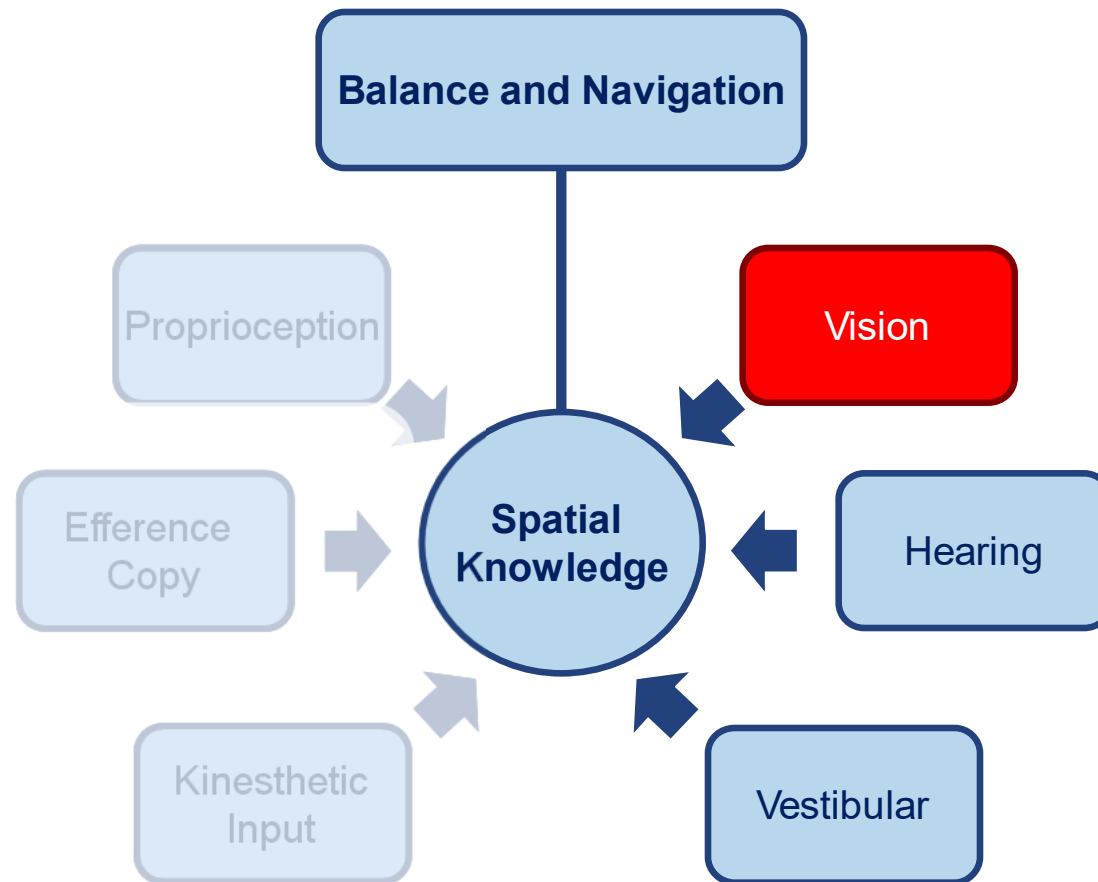
Otol Neurotol. 2009

Sensory Loss Impacts Balance

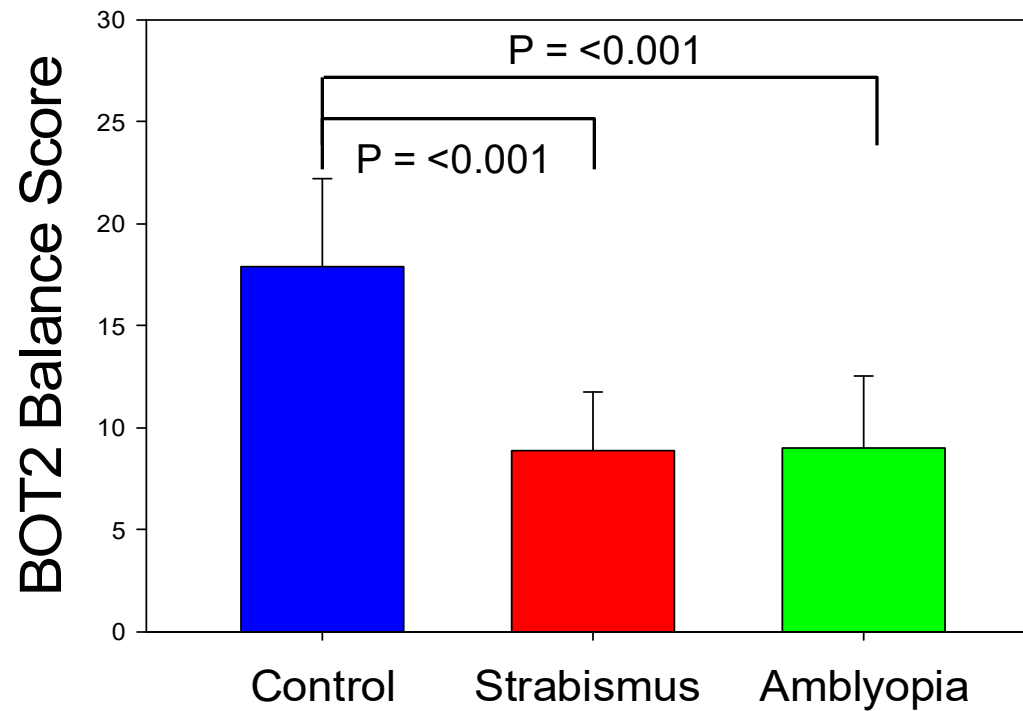




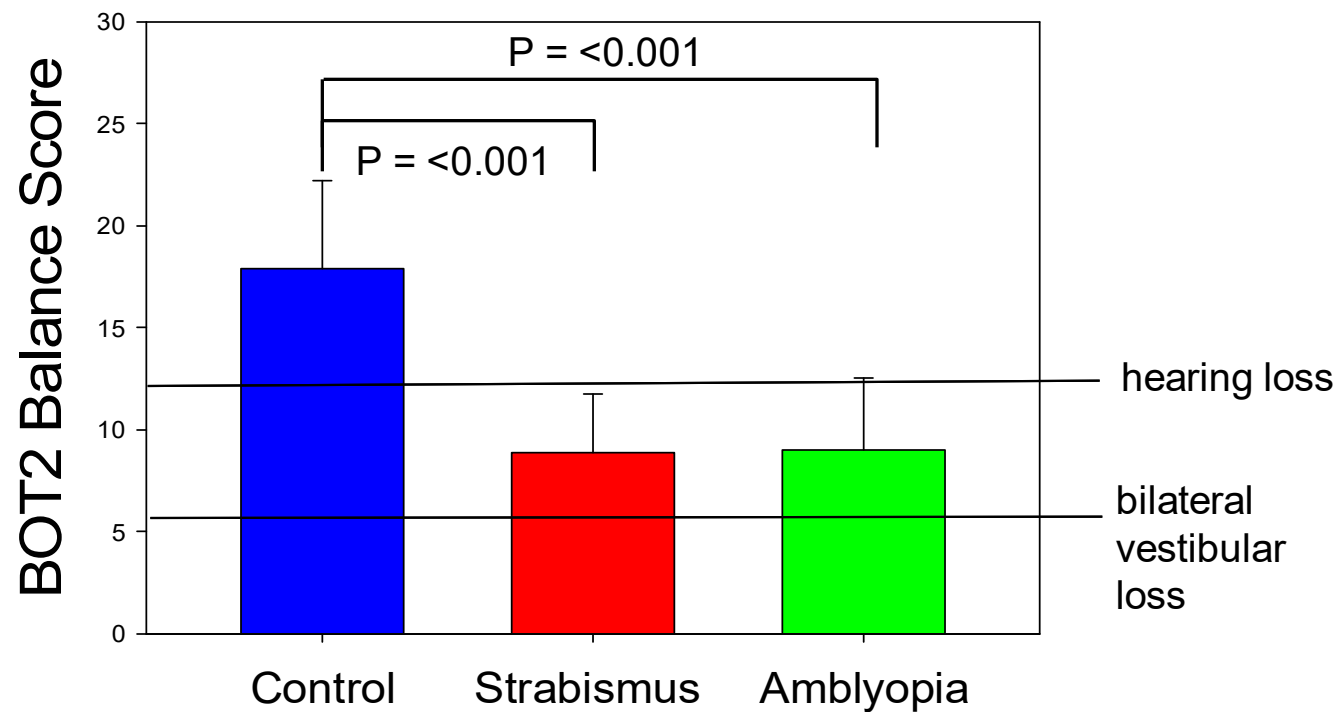
Development of Spatial Knowledge



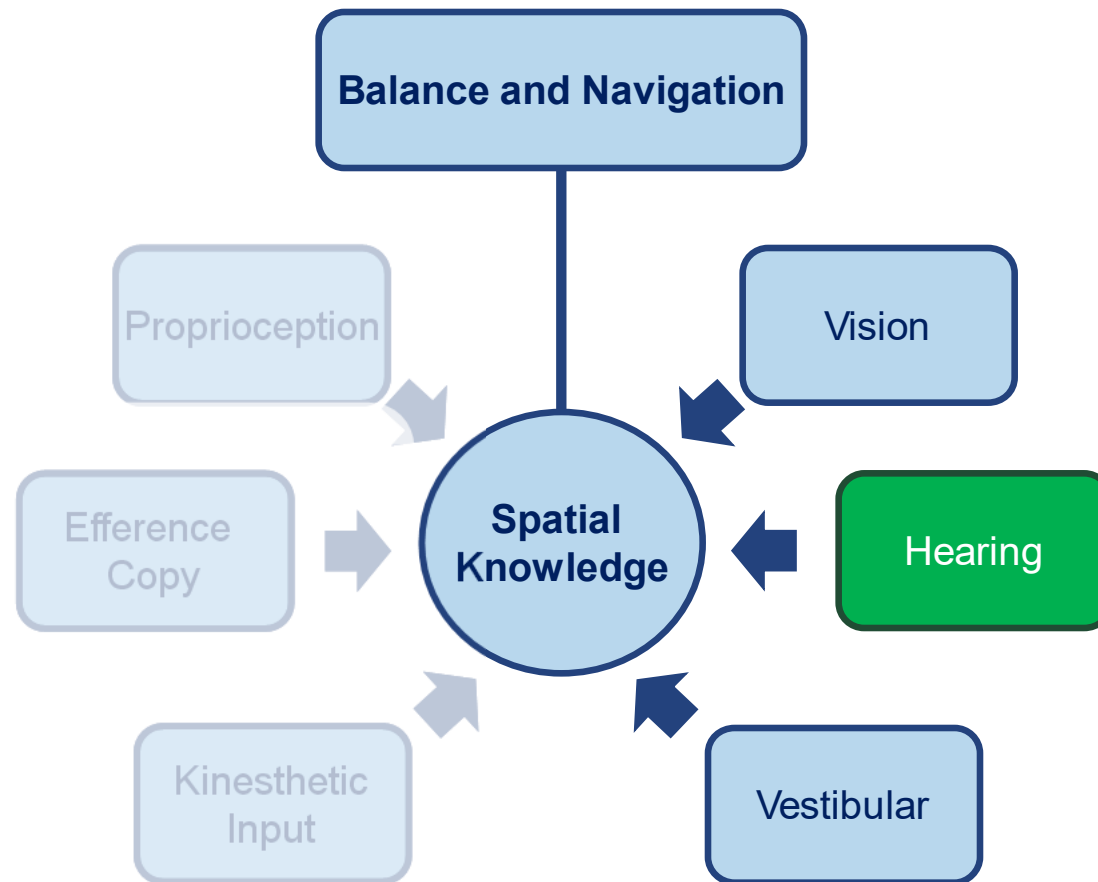
Balance is Impaired in Developmental Visual Anomalies



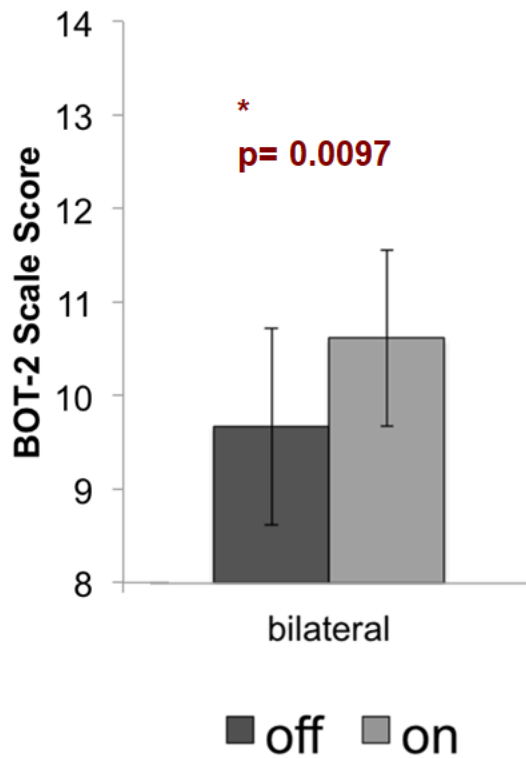
Balance is Impaired in Developmental Visual Anomalies



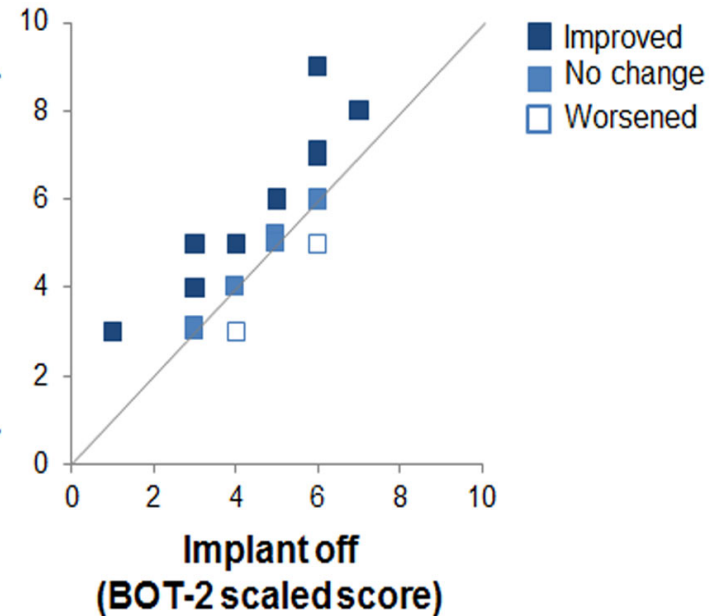
Development of Spatial Knowledge



Vestibular Effects of Cochlear Implantation (*positive*)

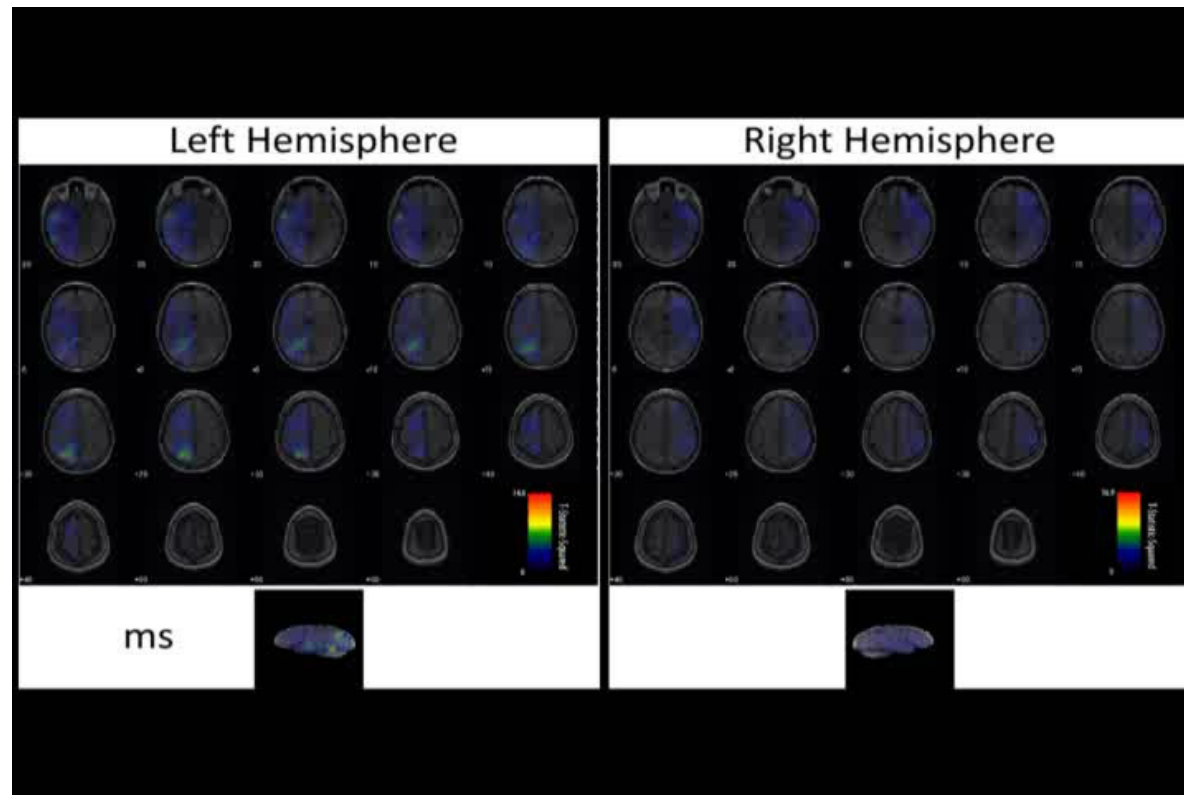


Laryngoscope. 2008



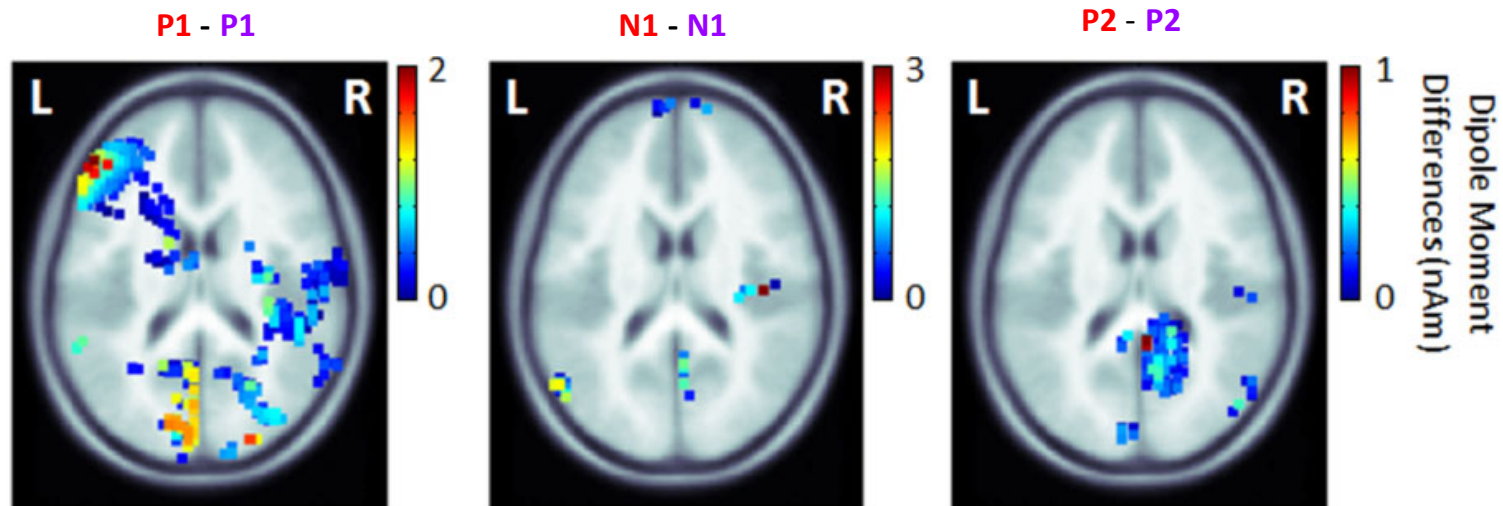
Audiol Neurotol. 2019

Measuring Plasticity



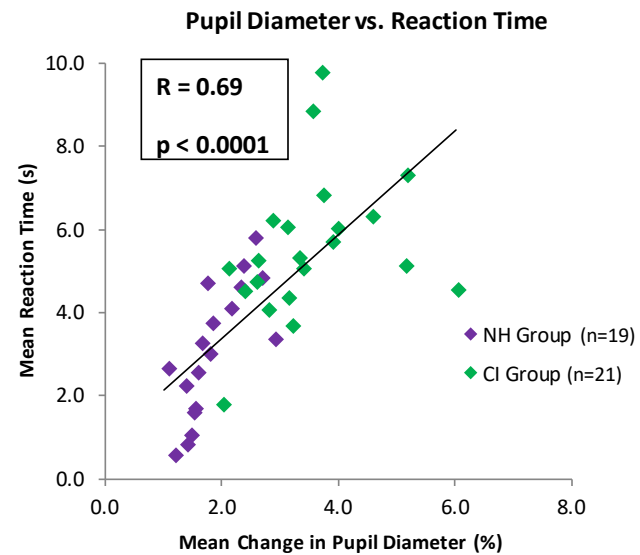
Plasticity is Required for Listening

Right Stimulation
CI Experienced – Normal Hearing Dipole Differences



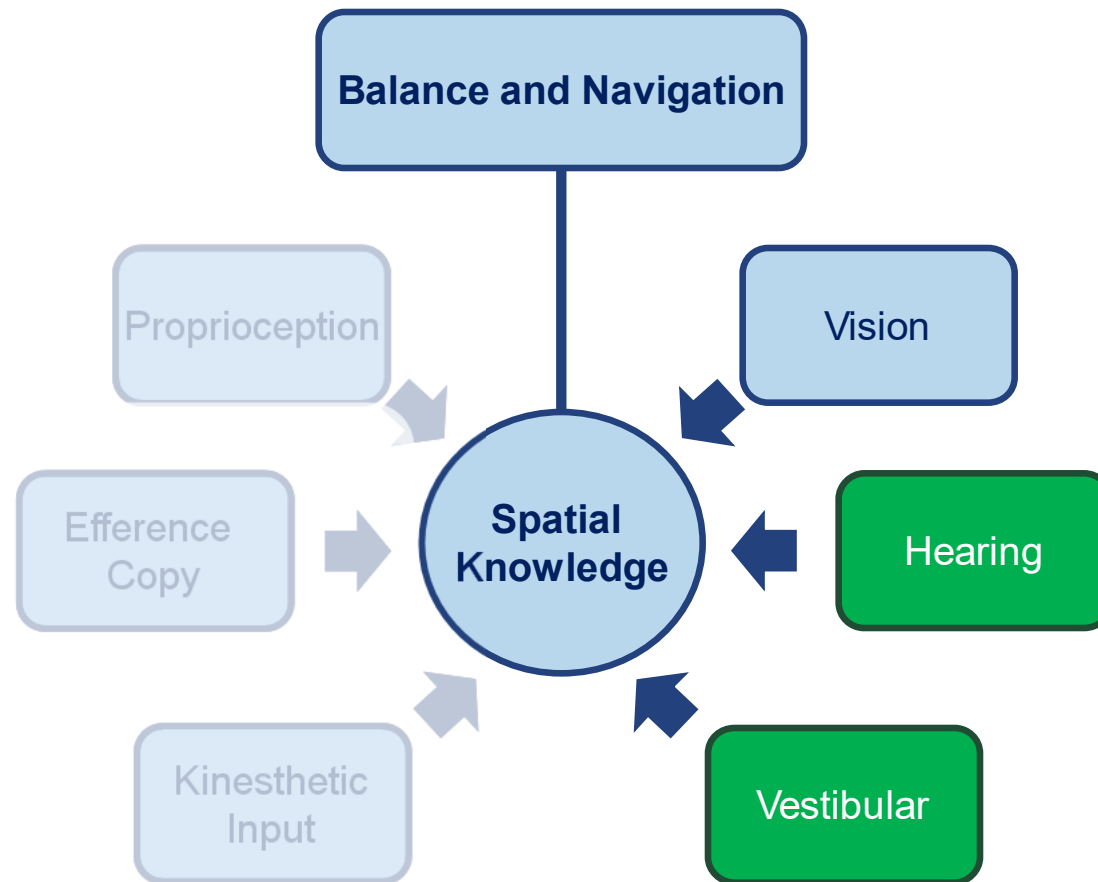
The Effort of Processing

Binaural fusion & listening effort

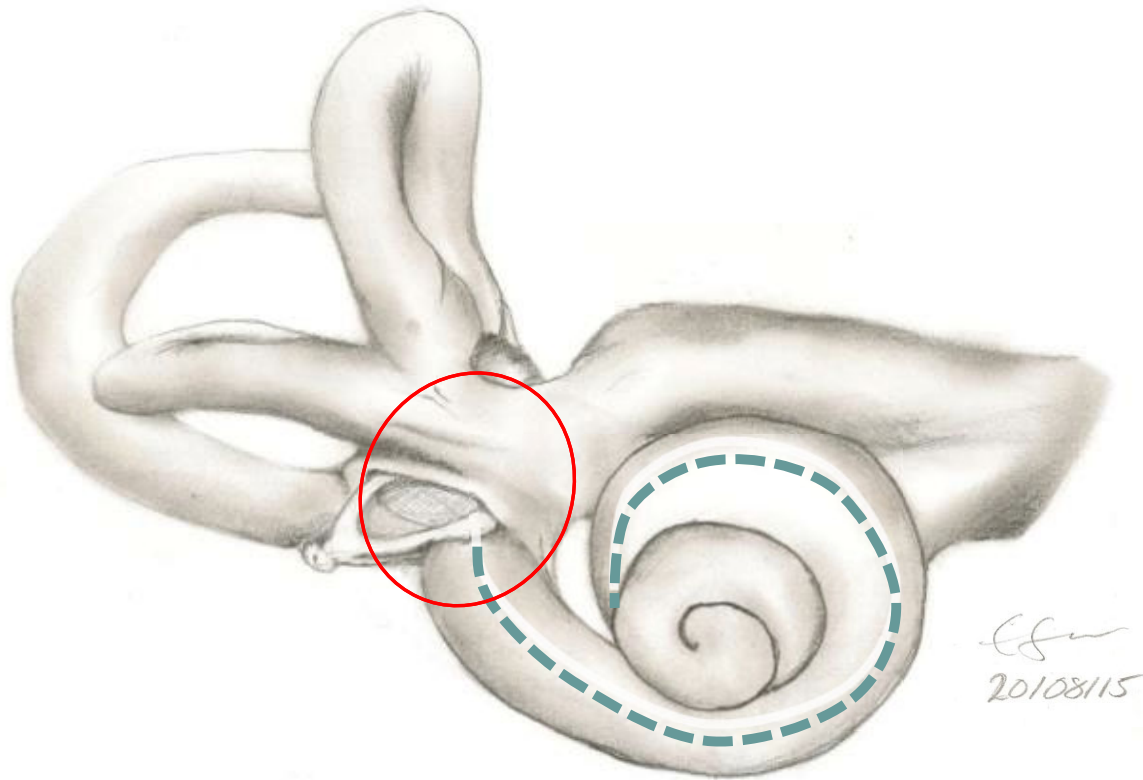




Development of Spatial Knowledge



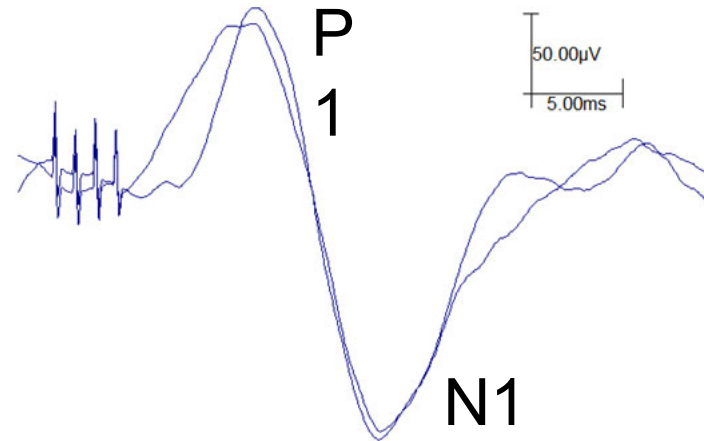
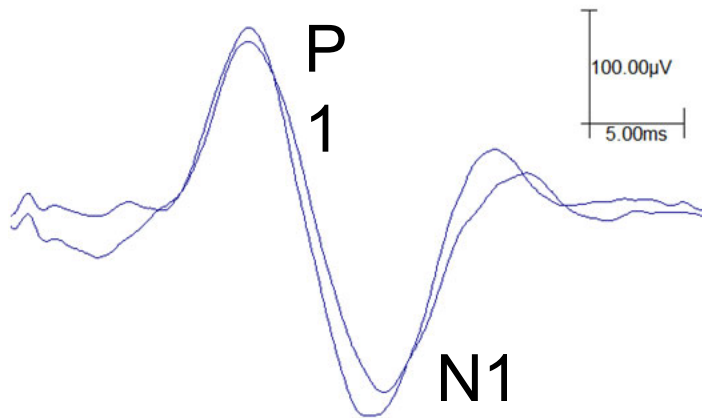
Can Electrical Current from a Cochlear Implant Spread to the Vestibular System ?



VEMP Waveforms (same ear)

acoustic

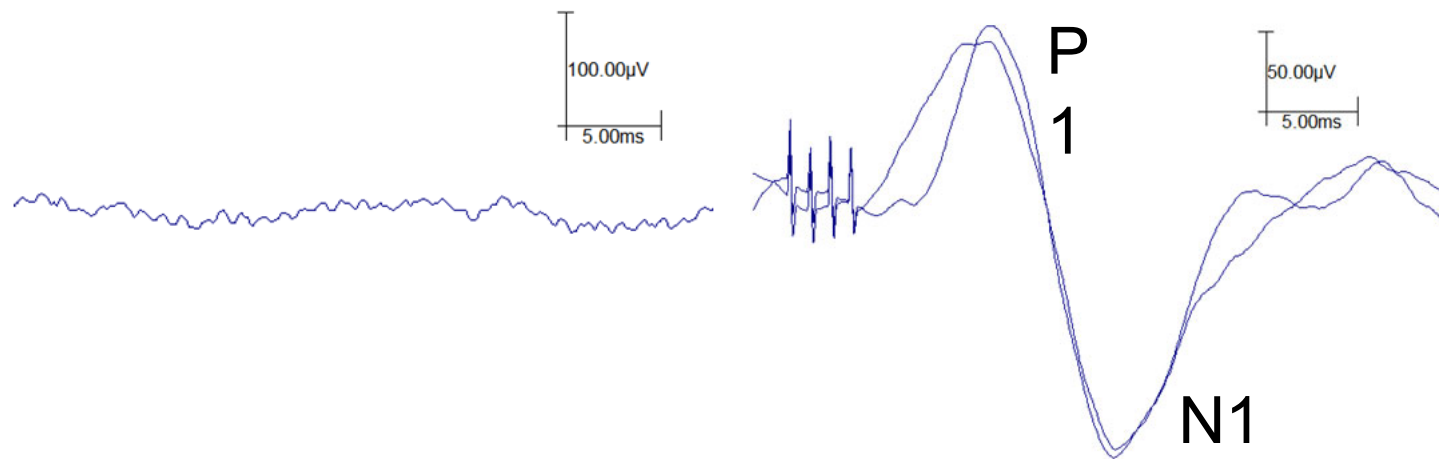
electric



VEMP Waveforms (same ear)

acoustic

electric

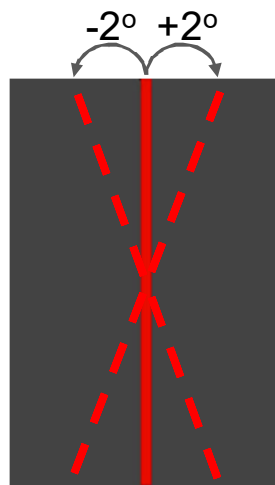


Vestibular Abnormalities Cause Functional Deficits



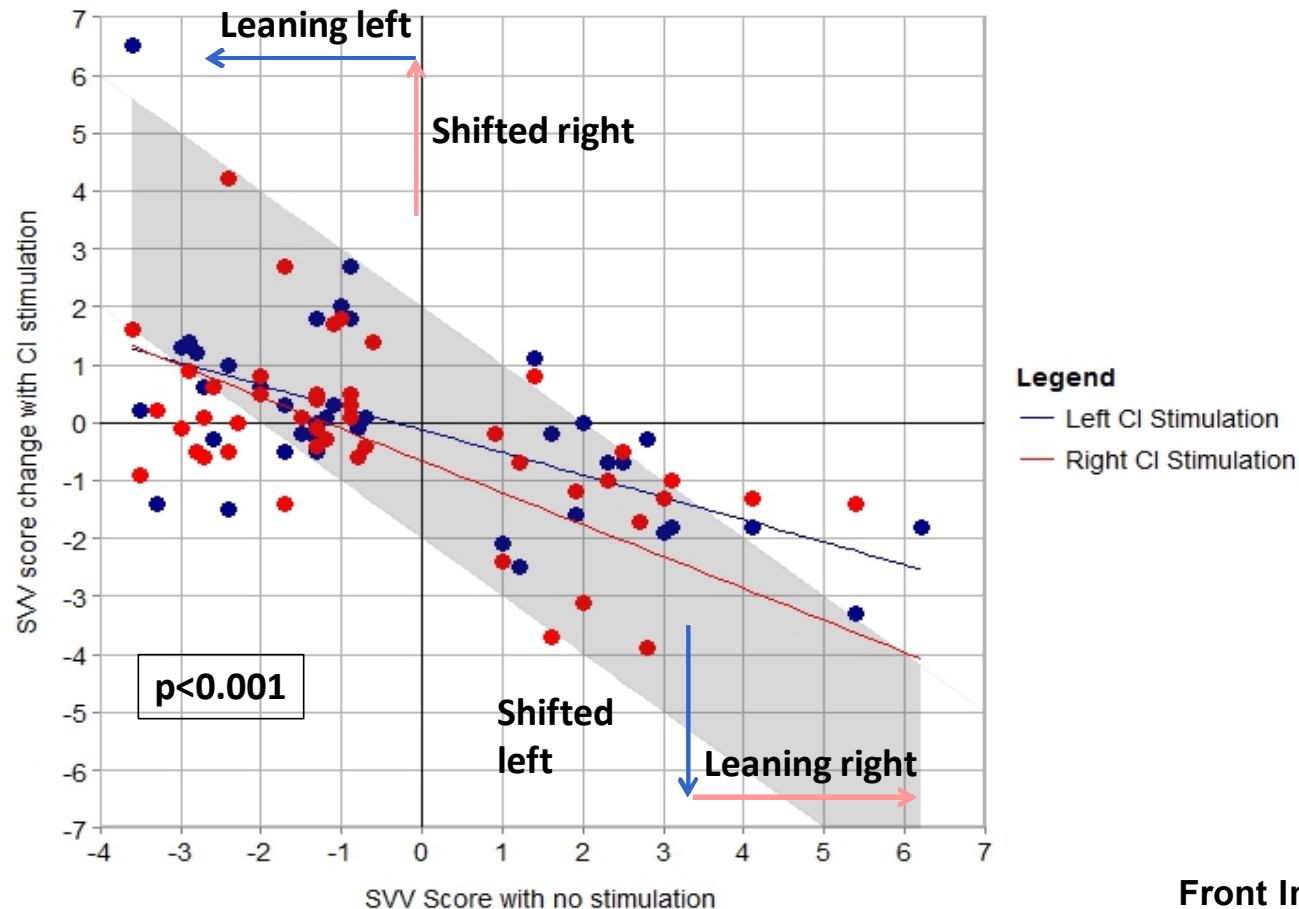
by Stephanie Jewell

Subjective Visual Vertical (SVV)



- Positive → tilt to the right
- Negative → tilt to the left
- Normal range: $\pm 2^\circ$
(Brodsky 2015, 2016)
- Bias → initial direction of linear marker
(Pagarkar 2008; Toupet 2015)

Electrical Stimulation Shifts the Perception of Vertical Back to Center



TransCochlear Stabilization of Balance

- accelerometer and gyroscopic sensor
- conversion to CI stimulation
 - head referenced
 - deviation signalled
 - transferred electrically to implant
 - 16 patients



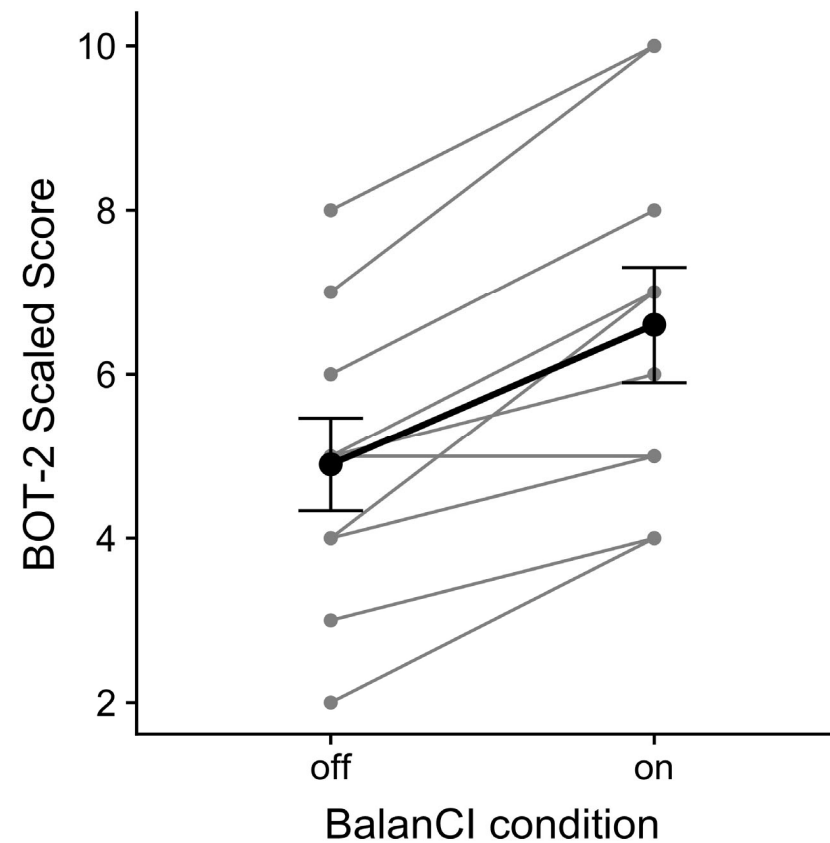
OFF



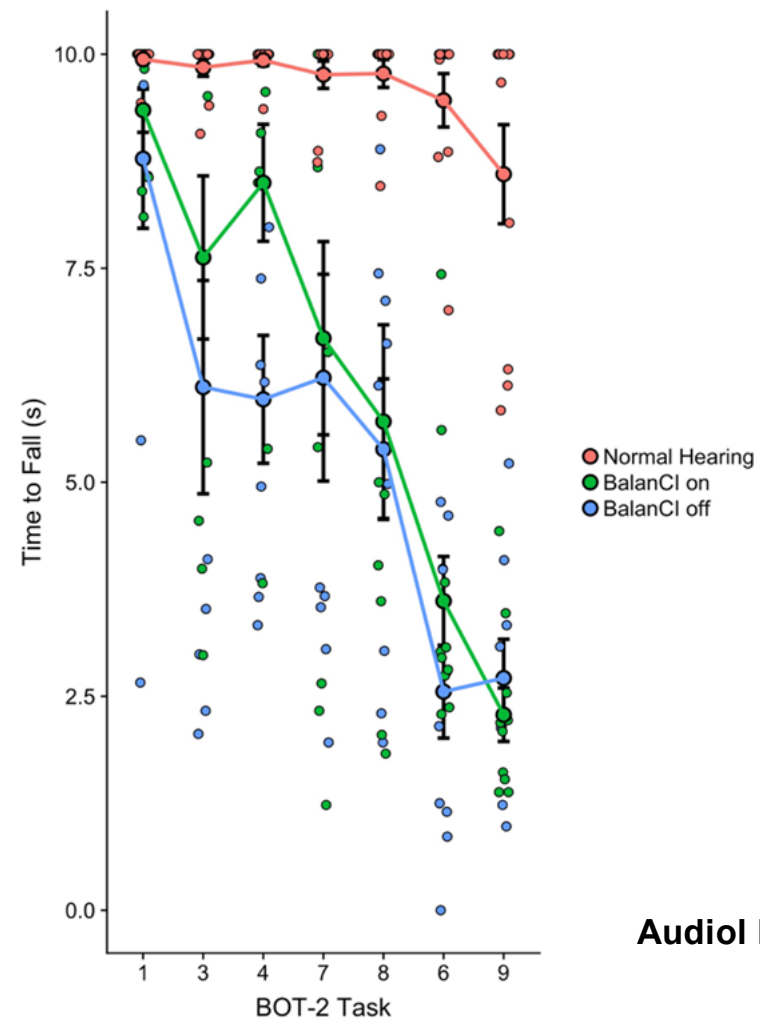
ON



Meaningful Implant Stimulation Improves Balance

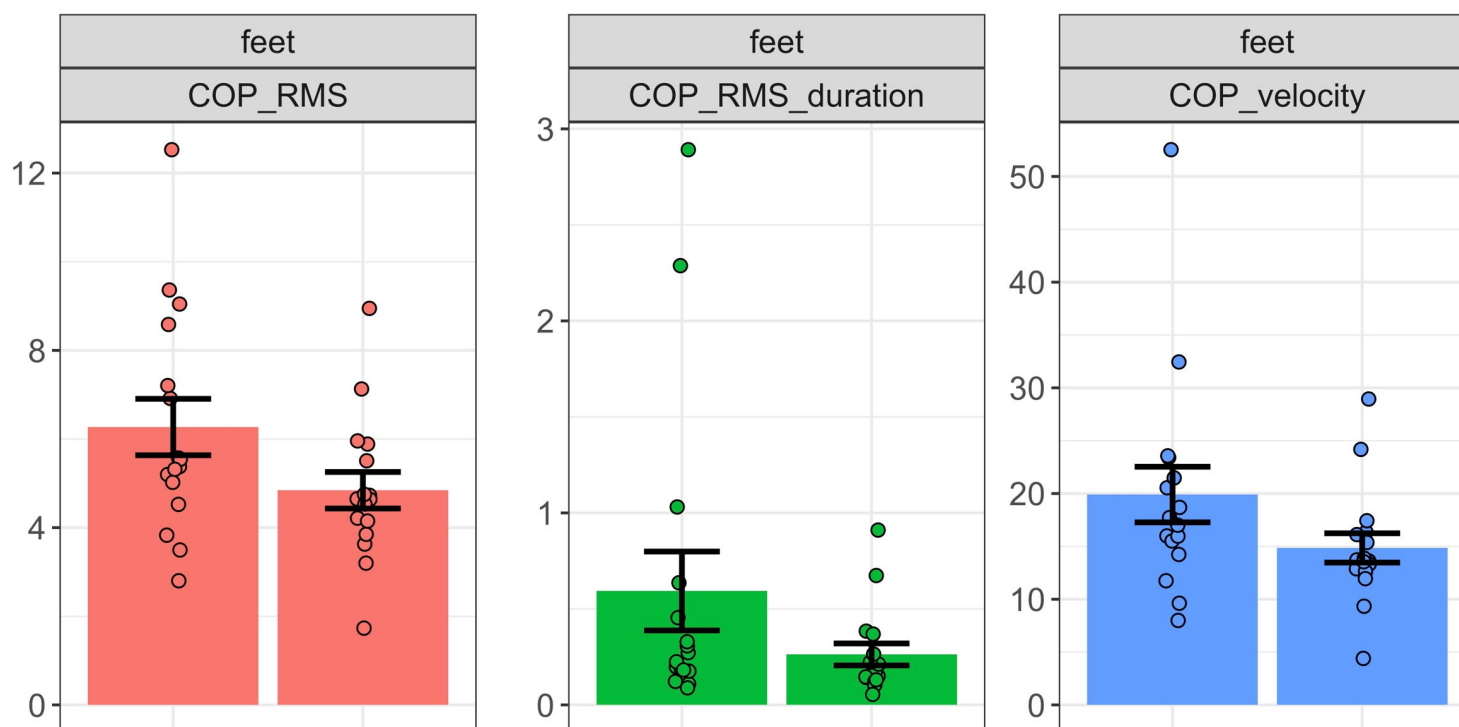


Time to Fall Approaches Normal



Audiol Neurotol. 2019

Postural Stability Improves





Single Sided Deafness in Children

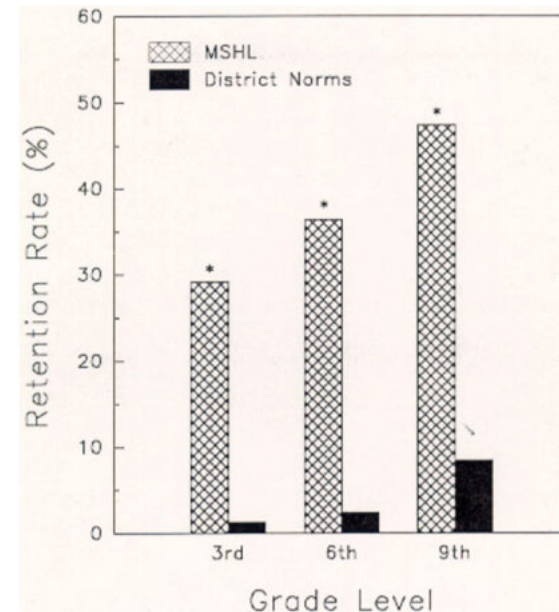


Single Sided Deafness in Children



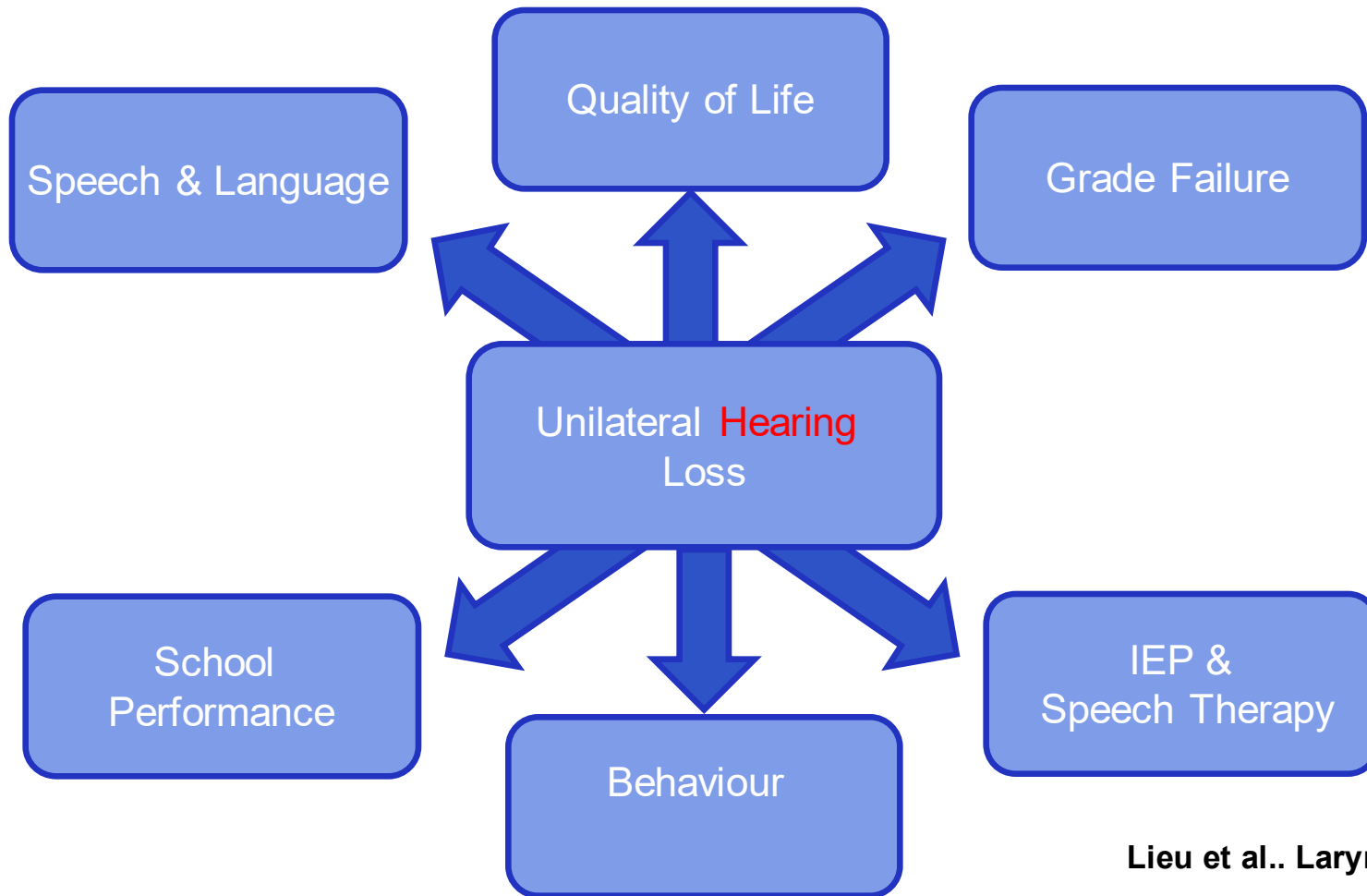
Unilateral Hearing Loss Has Impact

- 1/3 failed at least one grade
- 50% failed a grade or needed additional resources at school



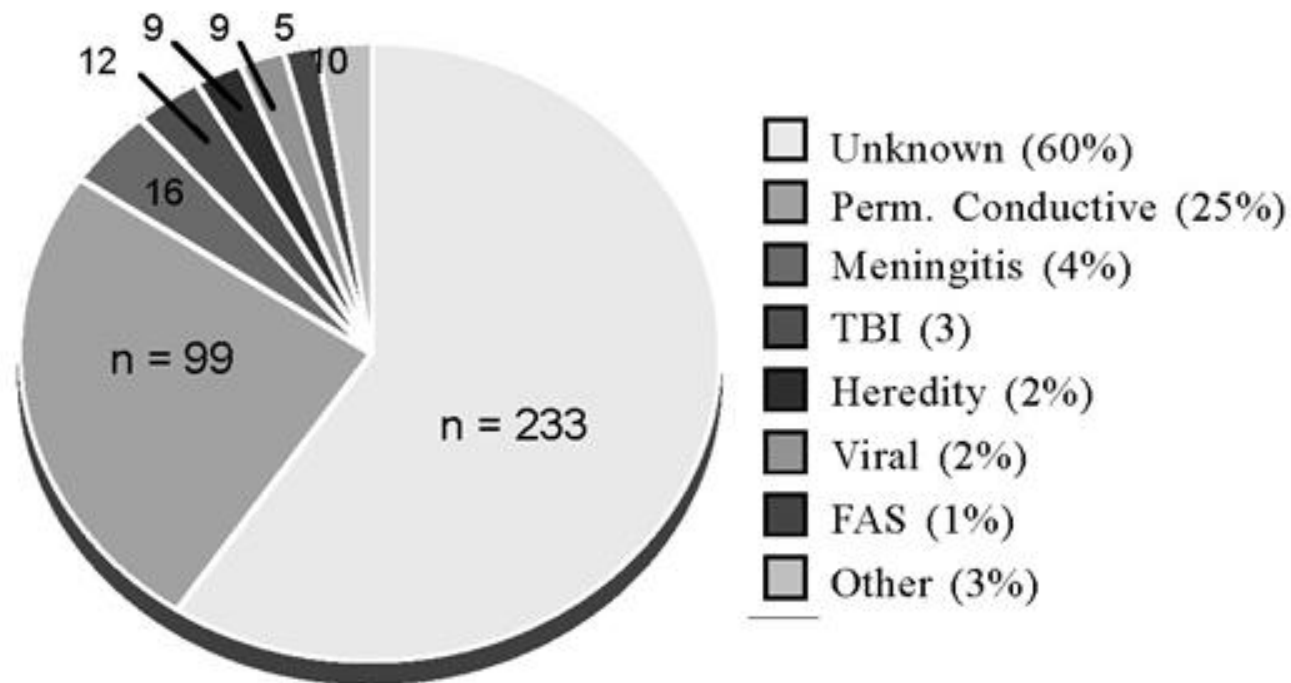
Bess and Tharpe, Int J Pediatr Otorhinolaryngol. 1991

Consequences of SSD in Children



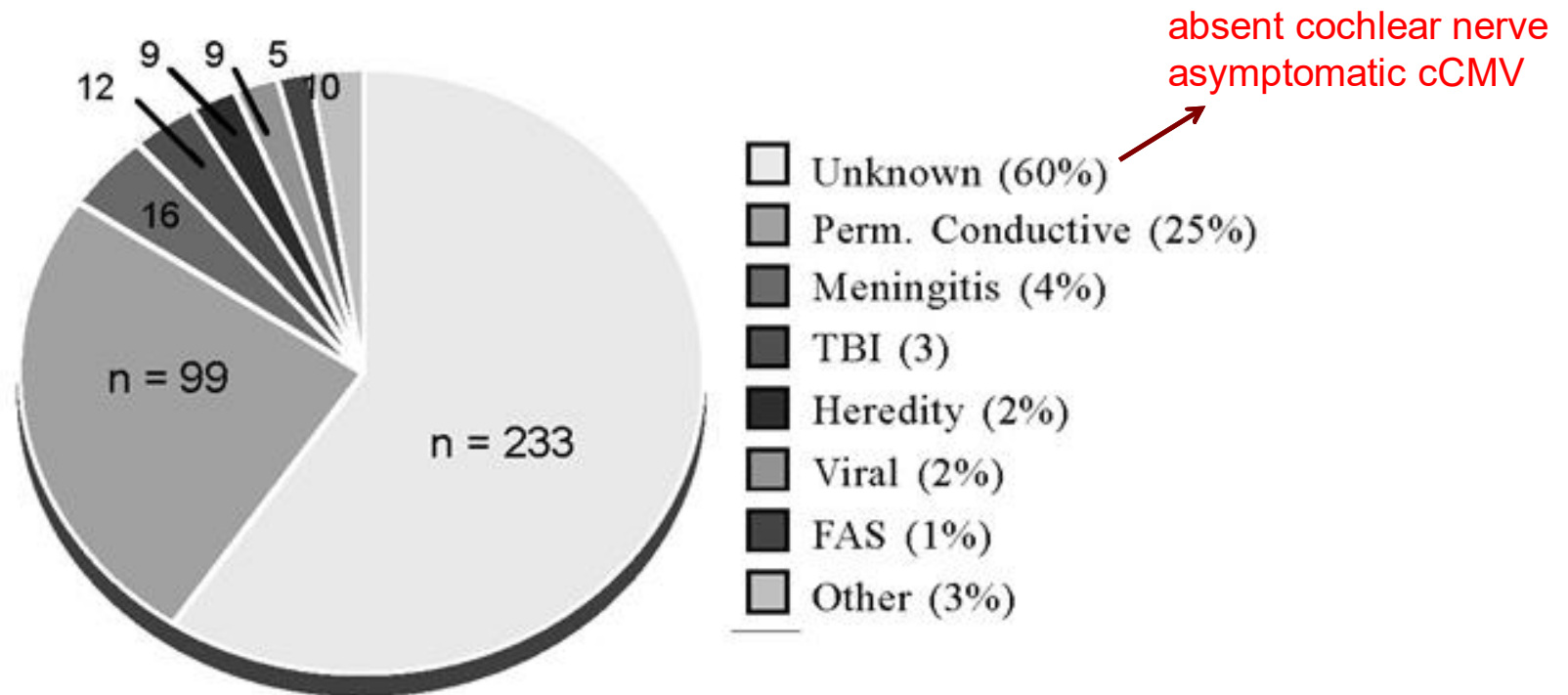
Lieu et al.. Laryngoscope. 2012

Etiology of SSD circa 1990



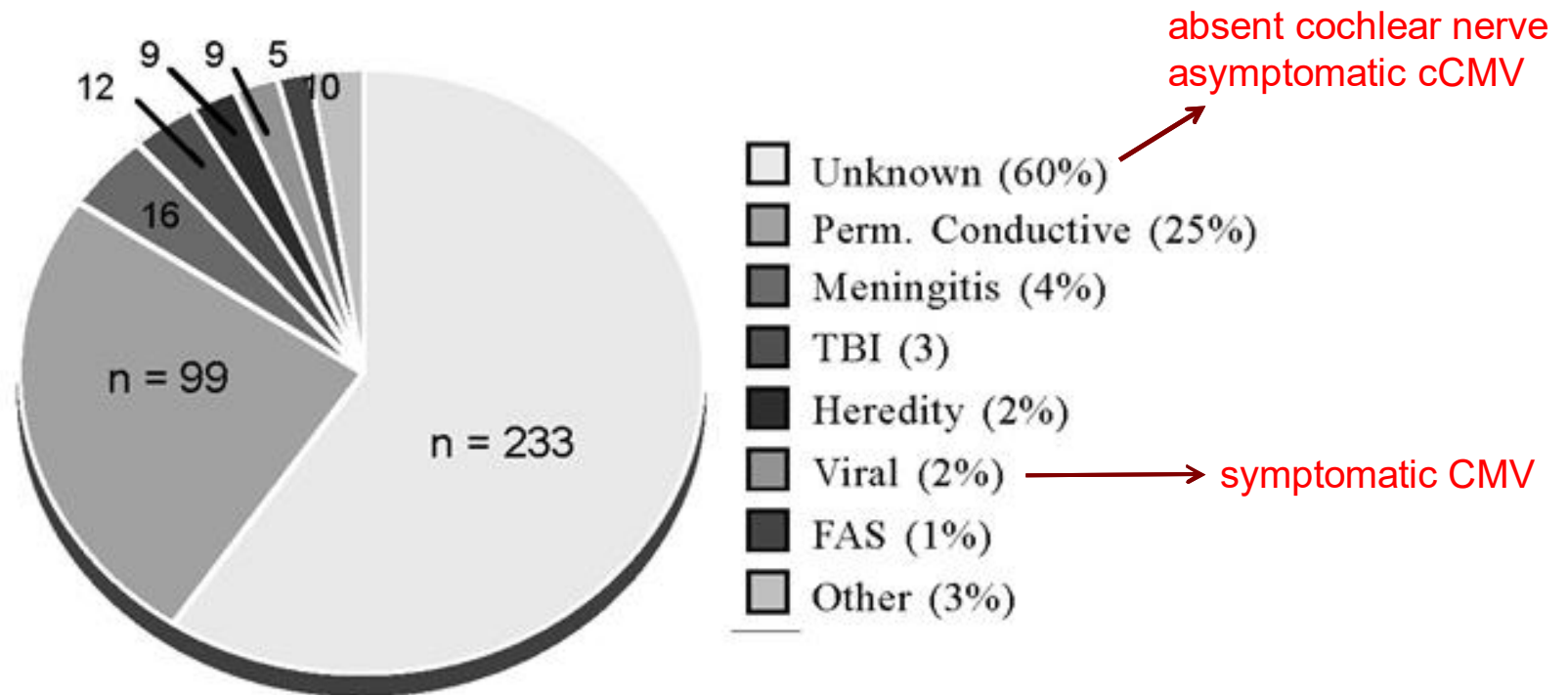
English et al, LSHSS 1999

Etiology of SSD circa 1990



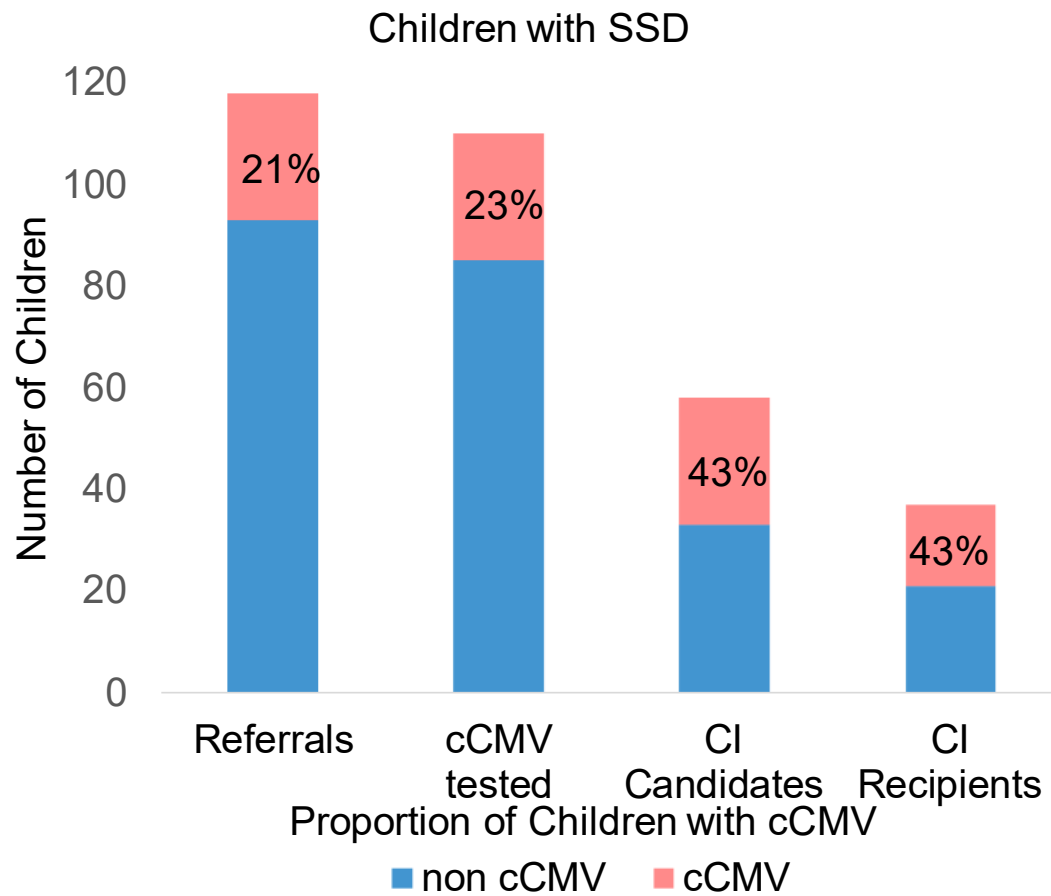
English et al, LSHSS 1999

Etiology of SSD circa 1990



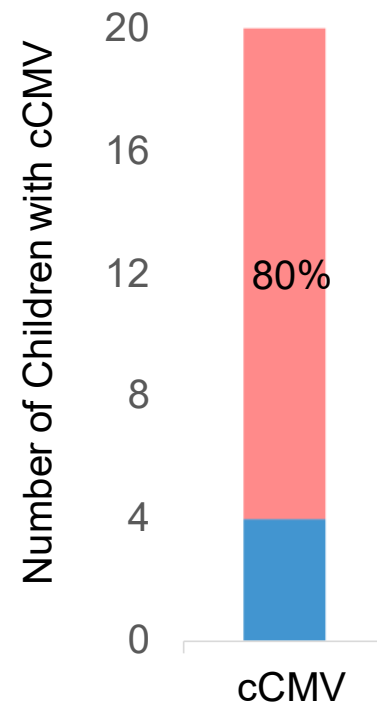
English et al, LSHSS 1999

cCMV is Common in SSD



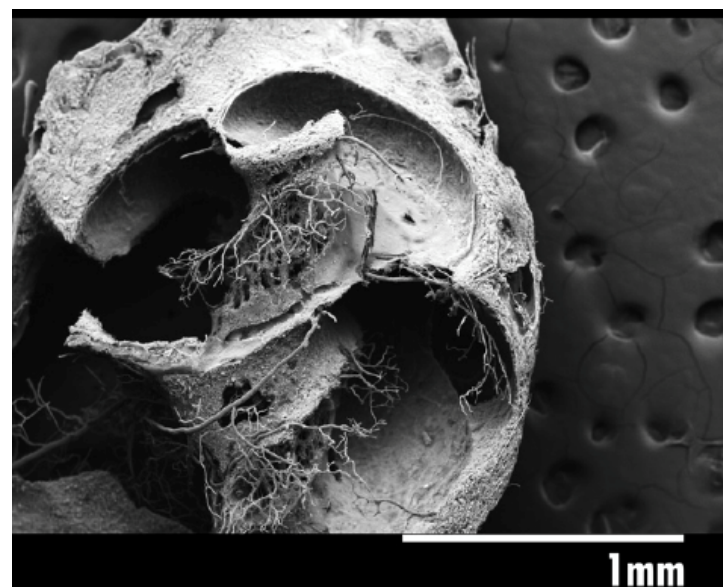
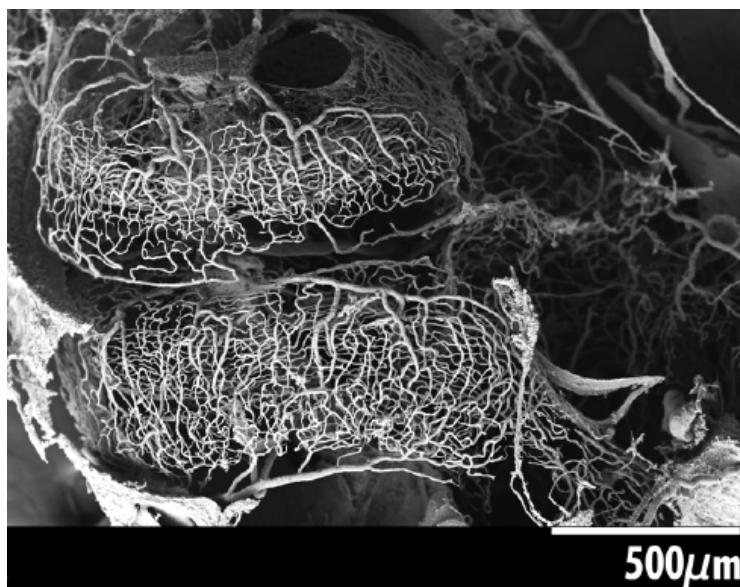
Children with cCMV Proceed to Implant

Children with SSD and cCMV



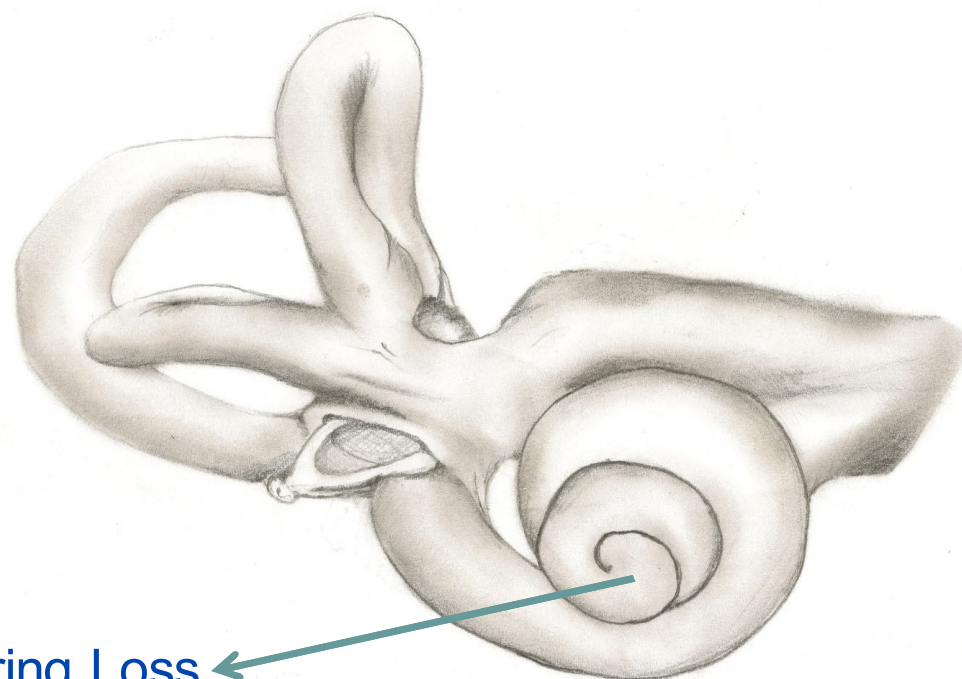
■ non-implanted ■ implanted

Congenital Cytomegalovirus



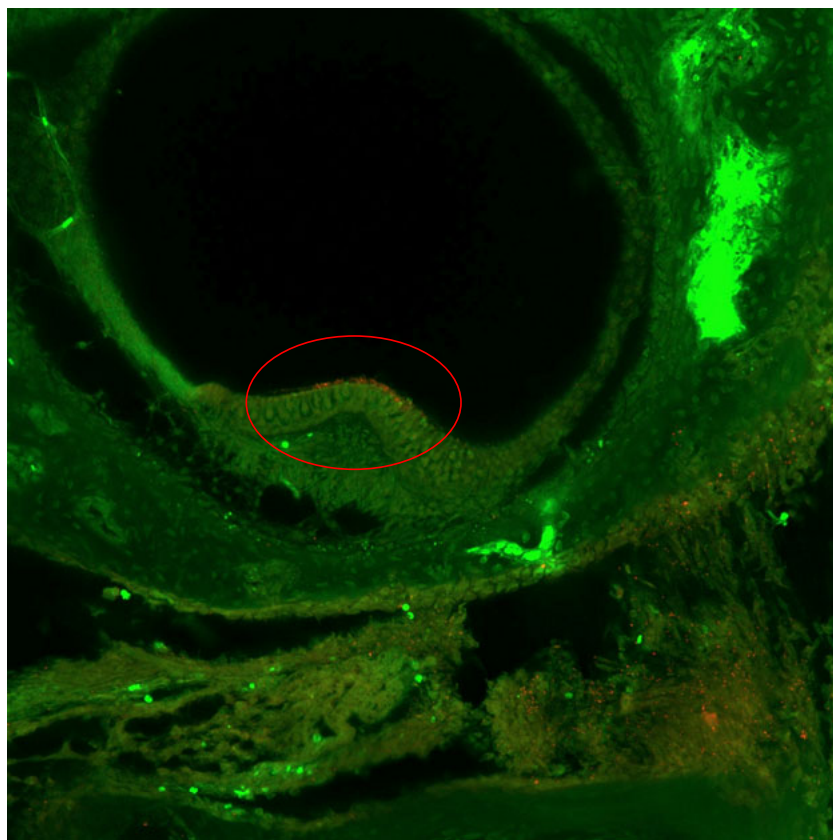
photos courtesy of Dr. Robert Harrison

Congenital Cytomegalovirus



20-50% Sensorineural Hearing Loss

Congenital Cytomegalovirus



photos courtesy of Dr. Robert Harrison

Congenital Cytomegalovirus

92% Vestibular end-organ Dysfunction

33% Complete Bilateral Loss

50% Progressive

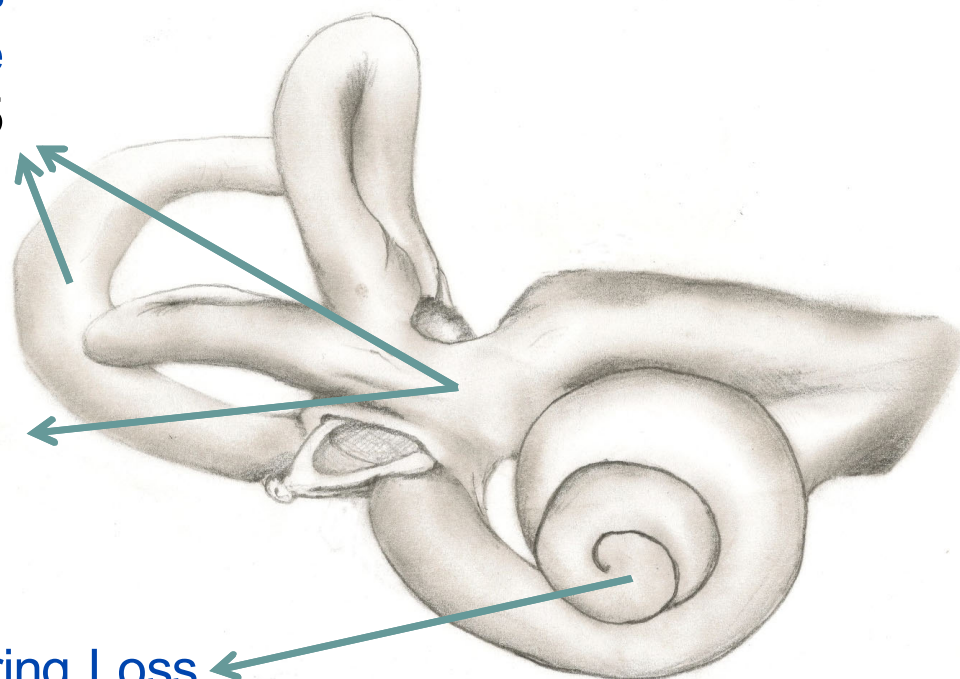
Bernard et al. Pediatrics. 2015

50% Otolithic Dysfunction

cVemp

Maes, Ear & Hearing. 2016

20-50% Sensorineural Hearing Loss

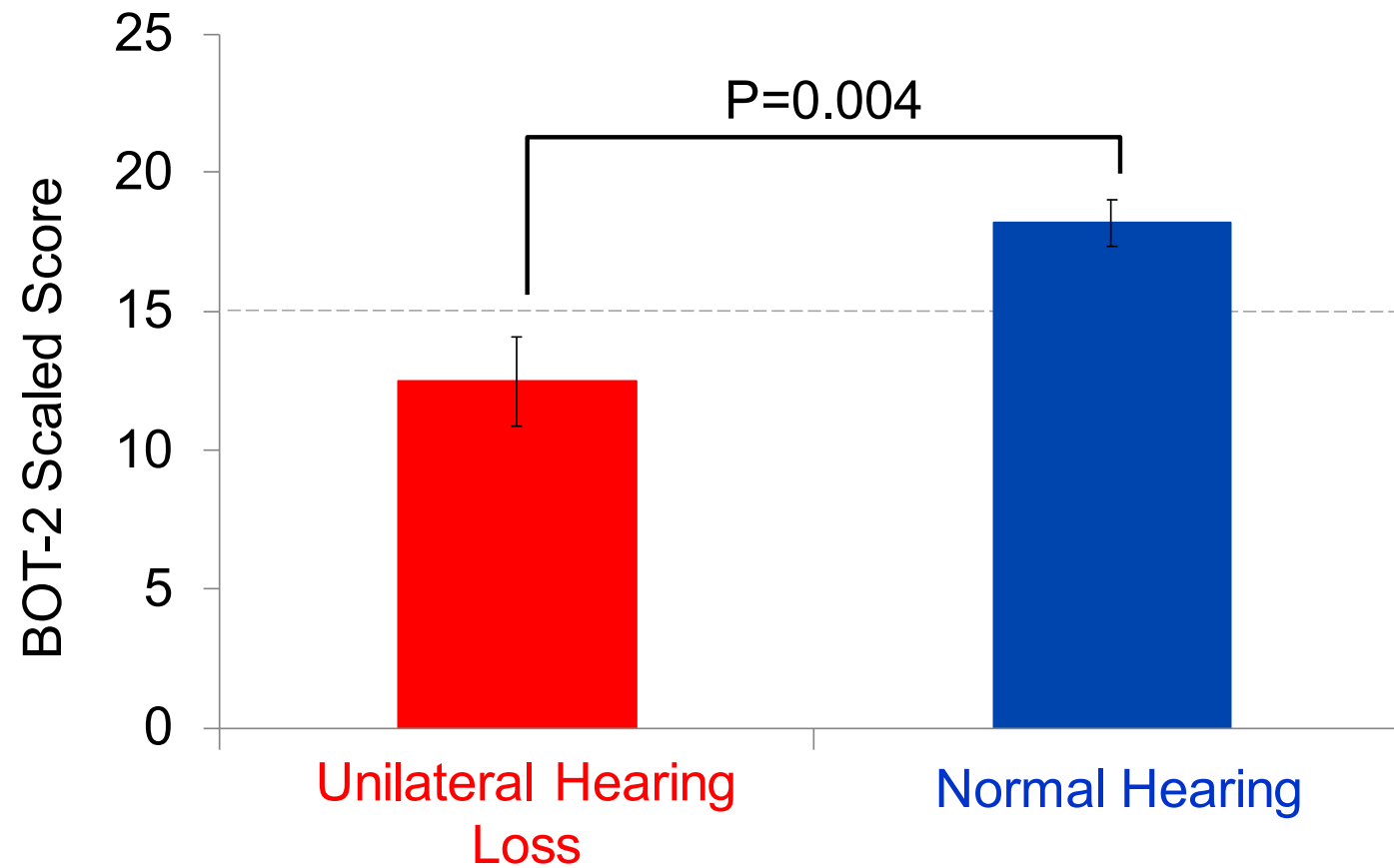


SickKids®

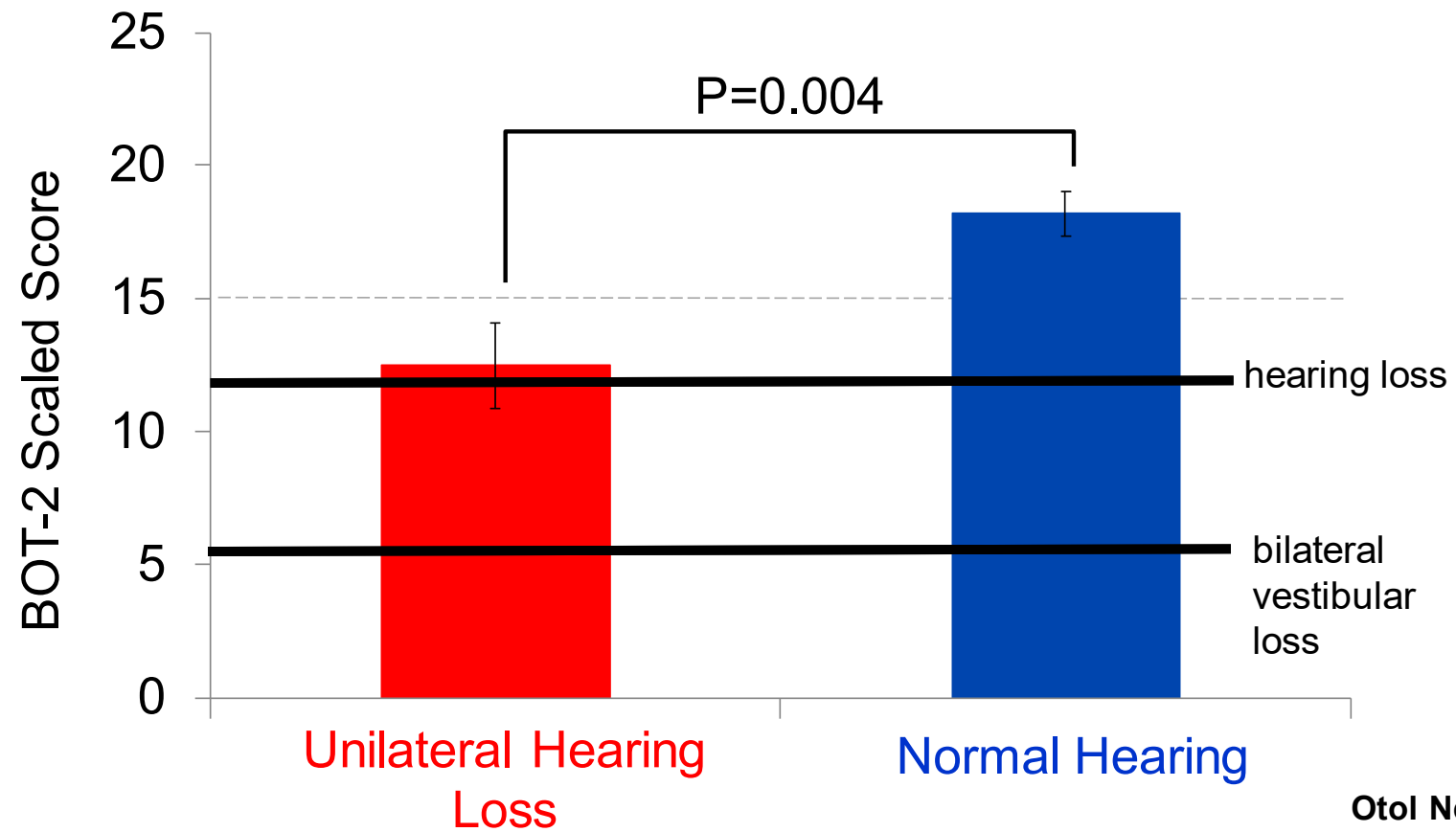
COCHLEAR
IMPLANT
PROGRAM



Balance is Impaired in SSD



Balance is Impaired in SSD



Vestibular Dysfunction in SSD

Horizontal Canal Dysfunction

48% Caloric Reduction

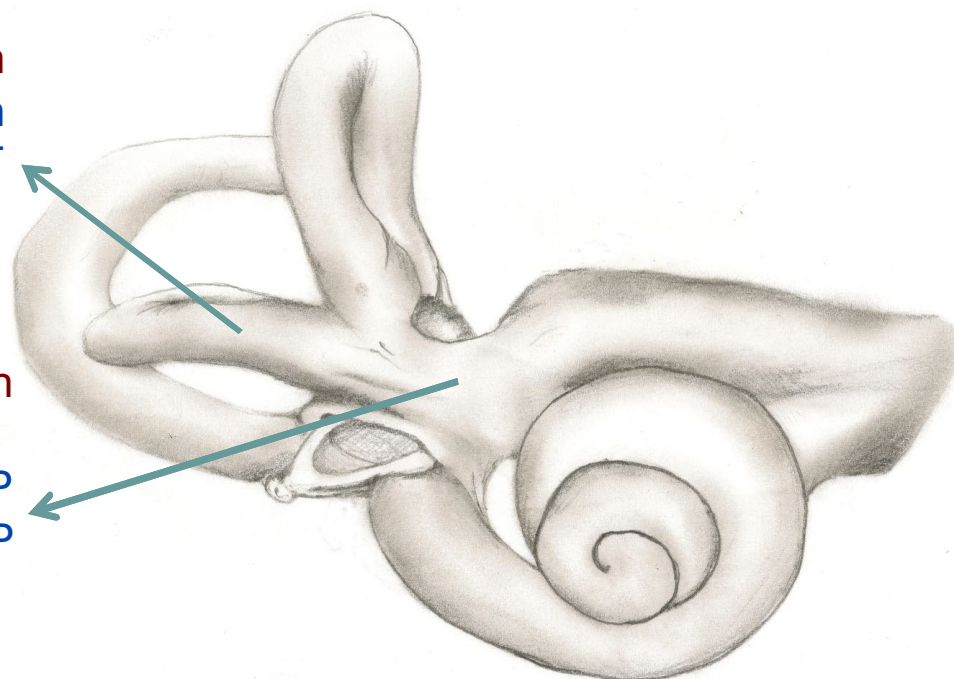
50% VOR loss vHIT

Otolithic Dysfunction

62.5%

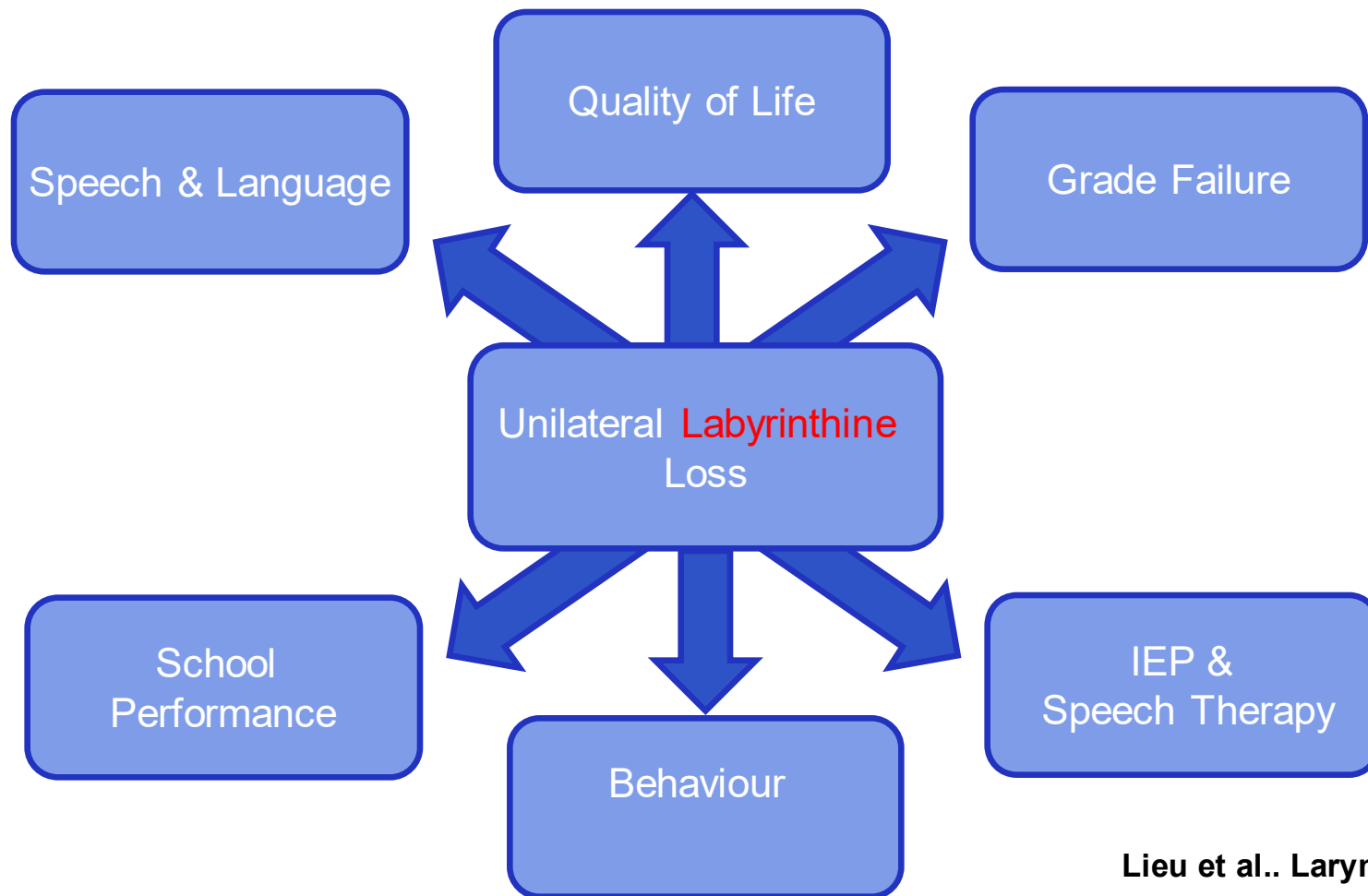
Utricular 42% oVEMP

Saccular 25% cVEMP



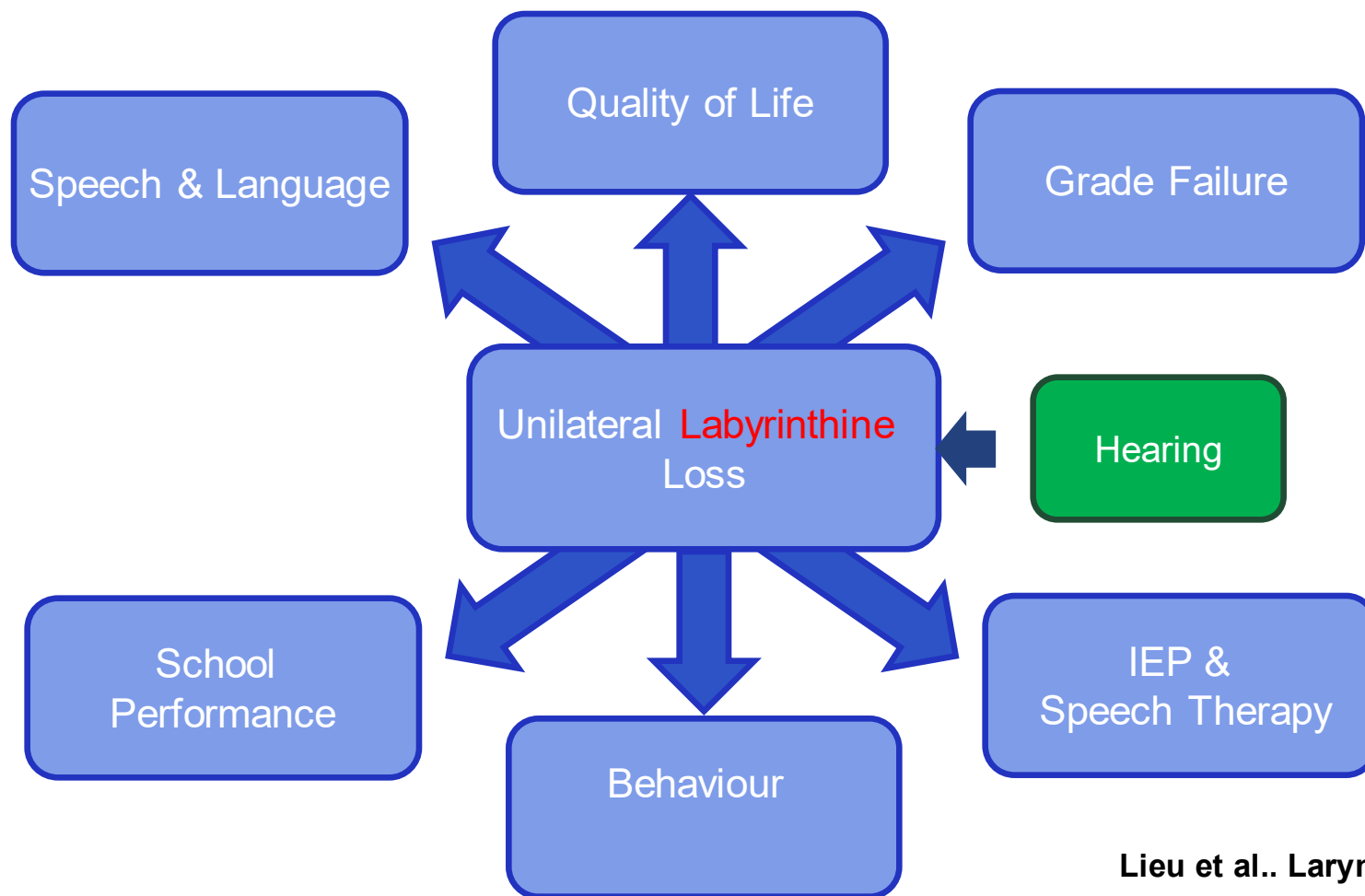
N = 20

Consequences of SSD in Children



Lieu et al.. Laryngoscope. 2012

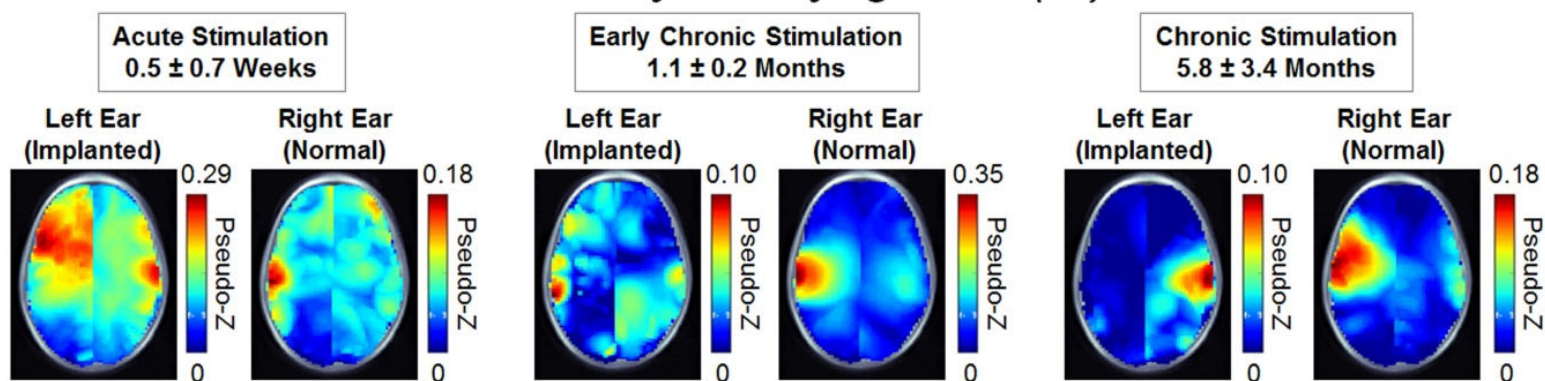
Consequences of SSD in Children



Lieu et al.. Laryngoscope. 2012

Aural Preference Restored

Source Activity Underlying Peak 1 (P1)



Duration of Stimulation

Summary

- sensory deficits lead to impairment
 - performance and development
- implants restore sensation
 - limits
 - compensation with effort
 - cross-modal?
- outcomes are complicated
 - extend beyond hearing & language
 - many gaps in knowledge exist

What Drives Plasticity in Children?



What Drives Plasticity in Children?



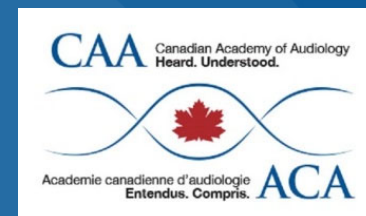
CAA Conference 2021

Delta Grand Marriott Hotel, Kelowna, B.C.
Save the date: October 13-16, 2021 in
Kelowna, B.C.



Upcoming and On Demand Webinars

canadianaudiology.ca/webinars/



- Therapeutic Techniques for Counseling Complex Patients with Hearing Challenges and Their Families with Michael Hoffman - Nov 12
- Measuring and Understanding Tinnitus - including patients views on the 'Meaning of Life' with Richard S.Tyler Nov 19th
- Adult amplification and Aural Rehabilitation with Gurjit Singh - Nov 24th
- Audibility-based Hearing aid Candidacy for Children with Ryan McCreery Nov 10th

Thank you