

Balance and Hearing: What Cochlear Implants Have Taught us about the Developing Vestibular System

SickKids®

COCHLEAR
IMPLANT
PROGRAM

Sharon L. Cushing



Disclosures

- Speaker's Bureau
 - Interacoustics
- 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

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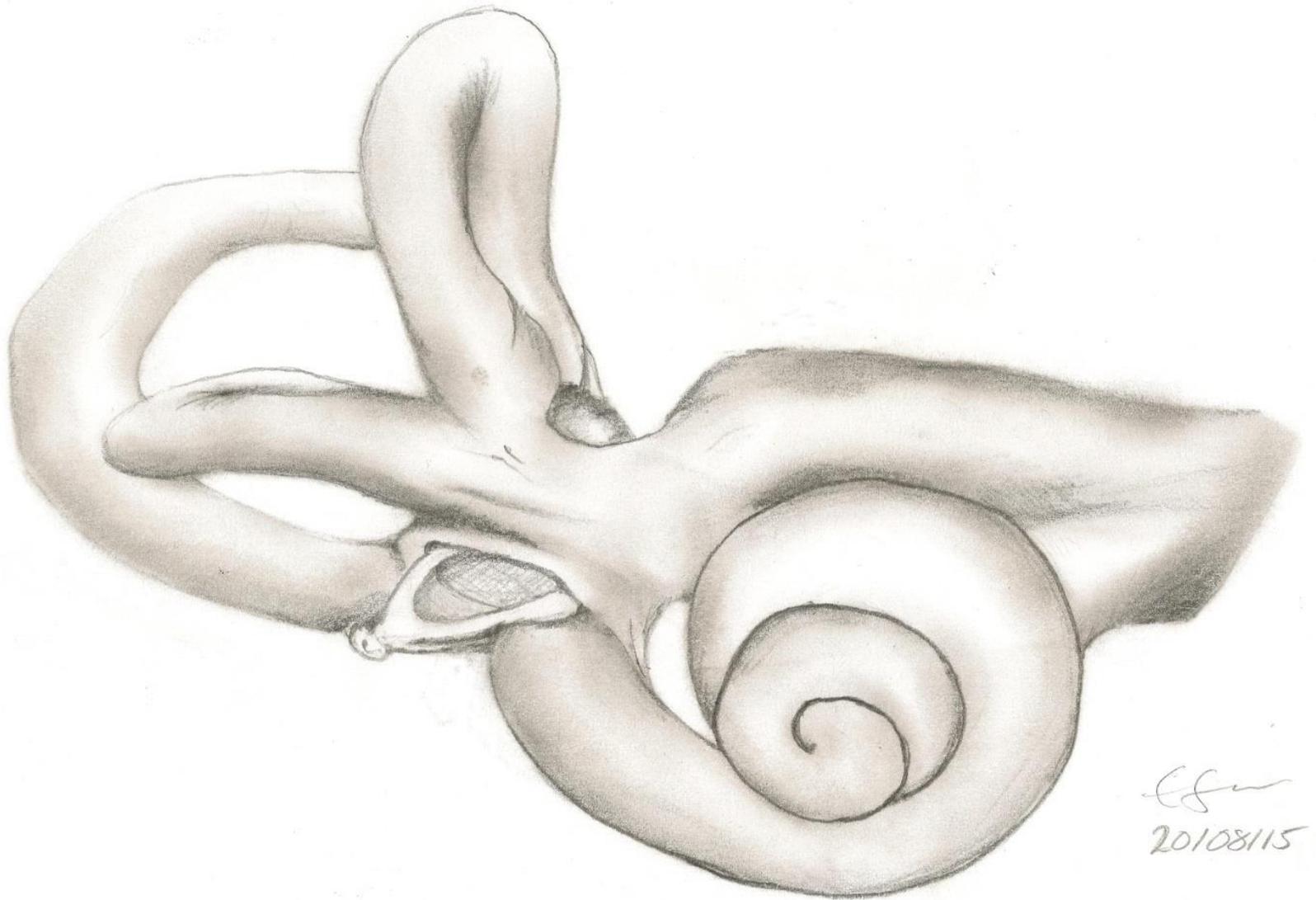


Unilateral



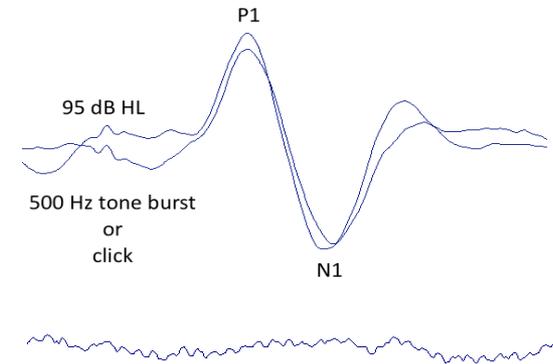
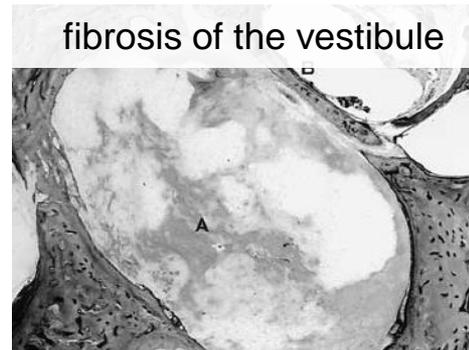
Bilateral



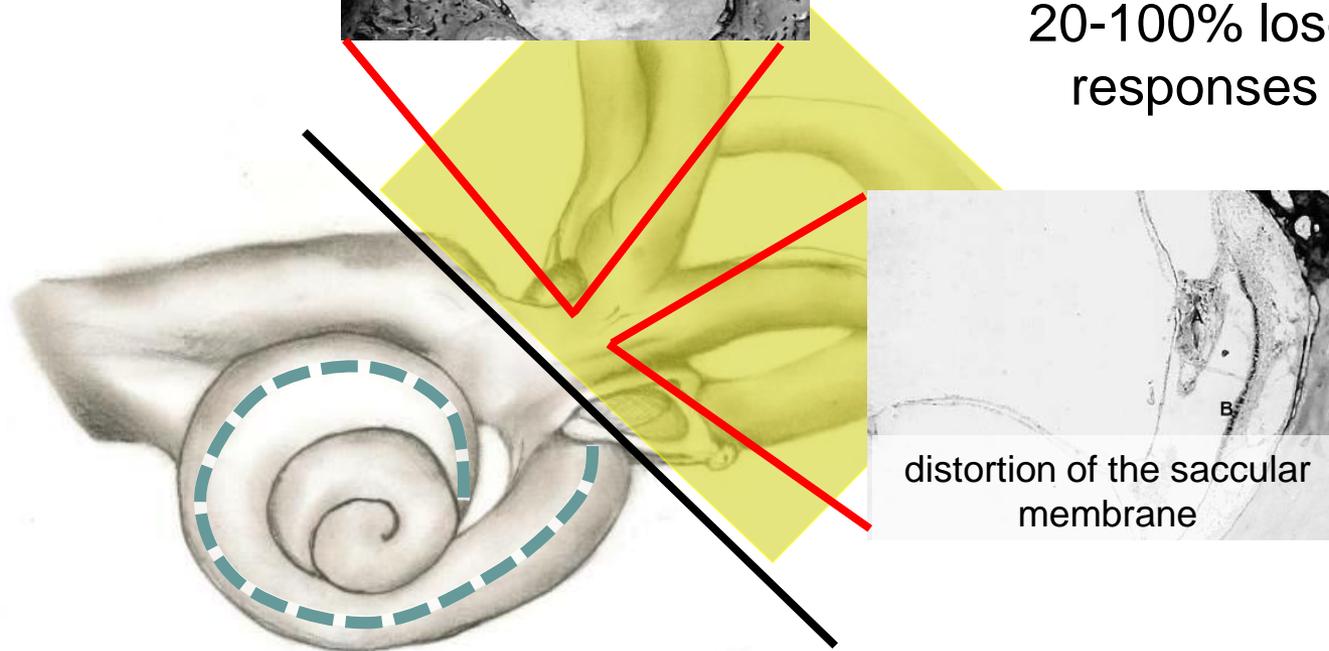


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Vestibular Effects of Cochlear Implantation (*negative*)



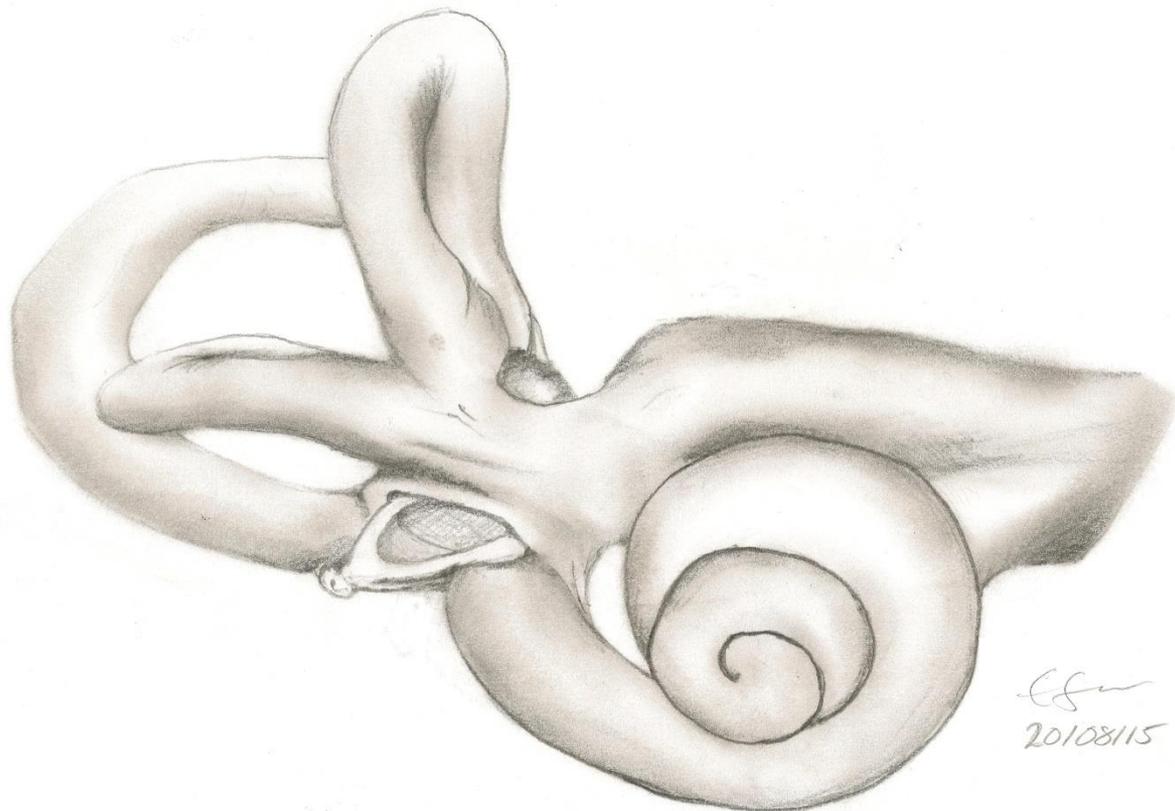
20-100% lose cVEMP responses after CI



Tien et al, 2002

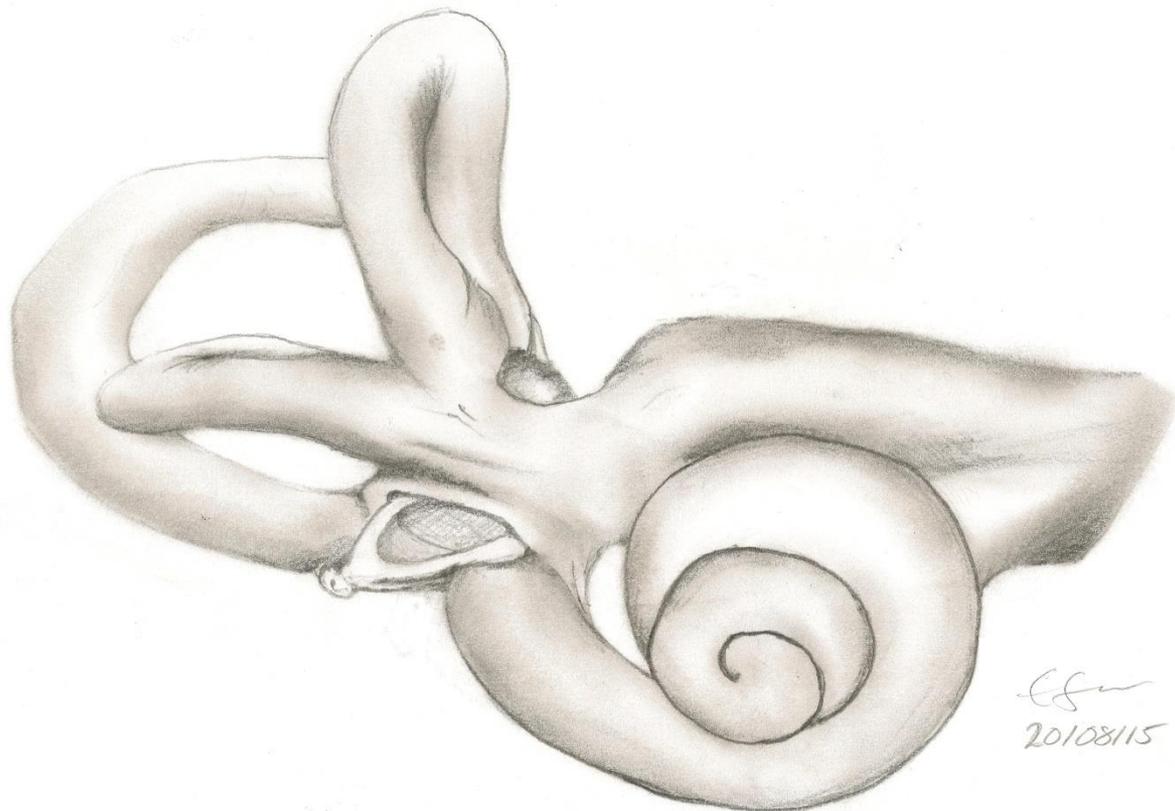
Vestibular Dysfunction is Common in Children with SNHL

70% dysfunction
35-40% complete
areflexia



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

Measuring Balance

Bruininks-Oseretsky (BOT2)

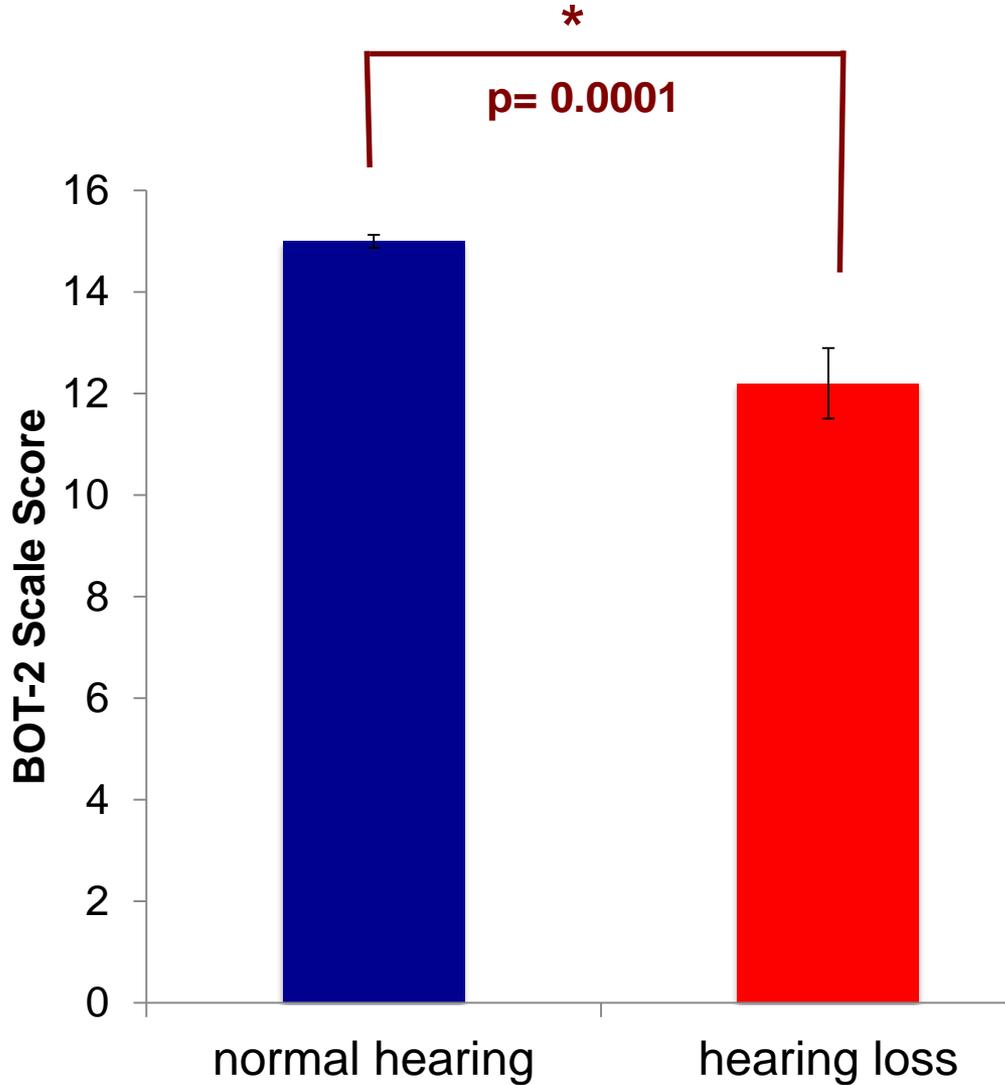
Balance Subtest

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.

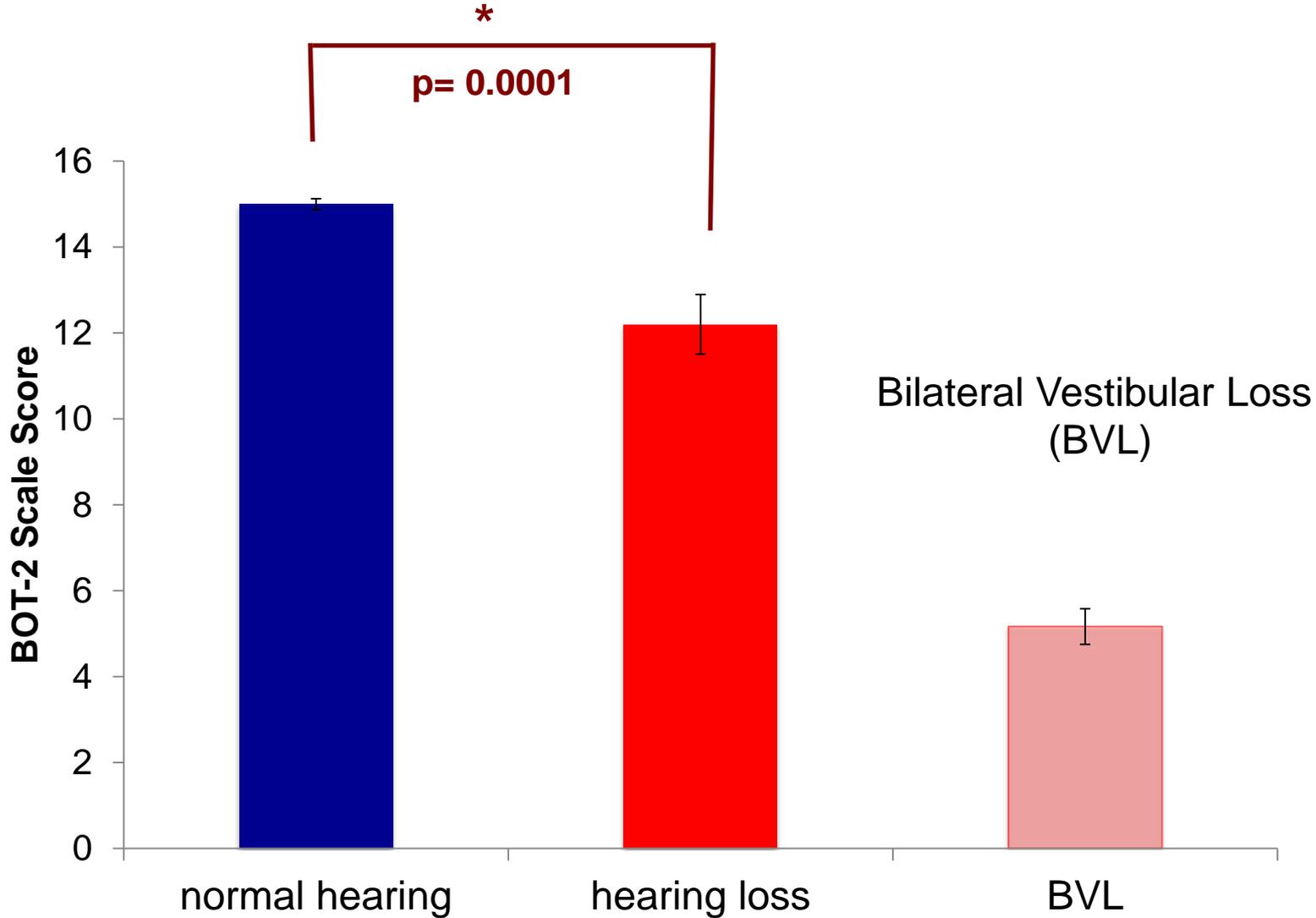
N= 190 children with CI tested



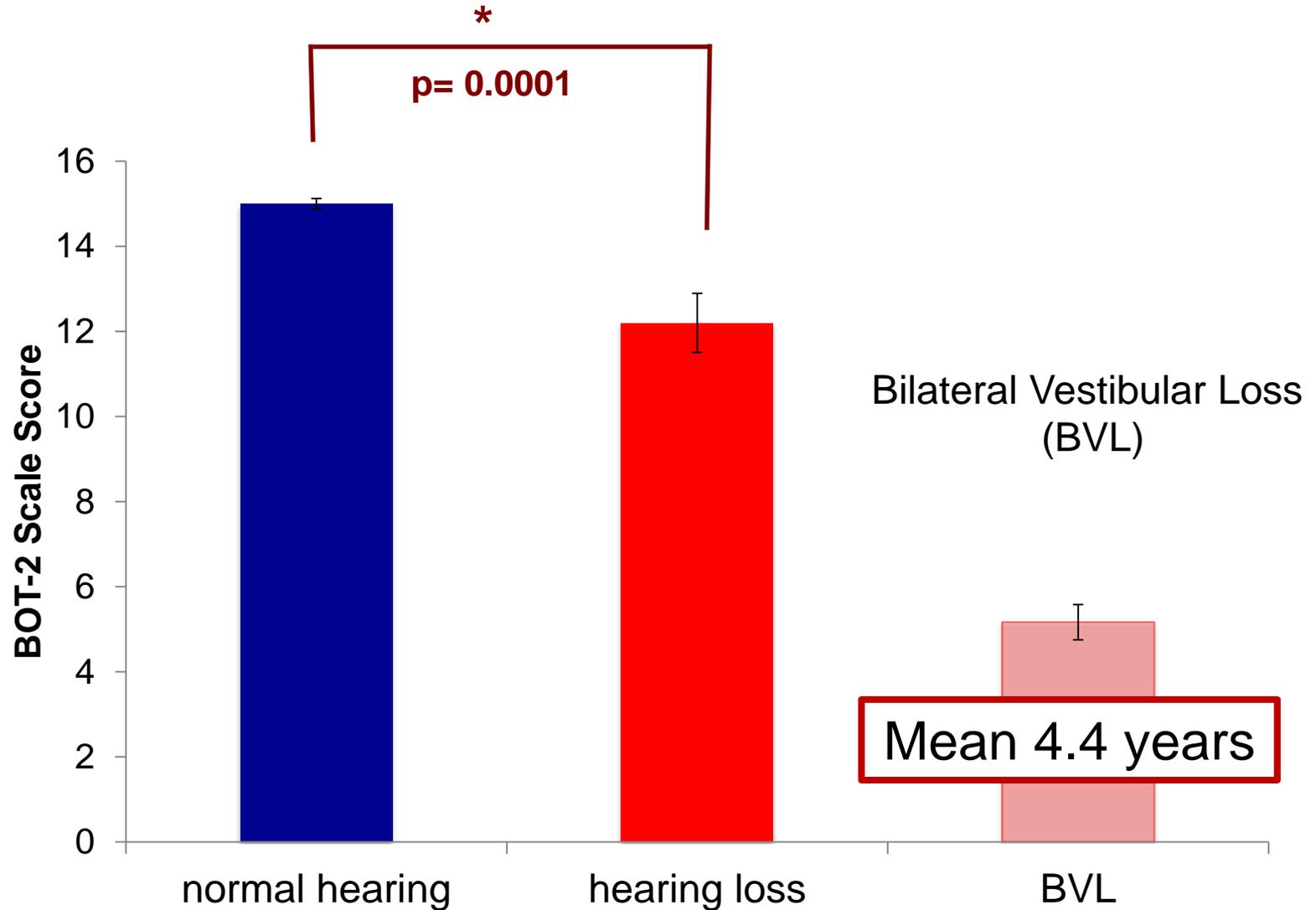
Vestibular Loss Impacts Balance



Vestibular Loss Impacts Balance



Vestibular Loss Impacts Balance



Vestibular Loss Impacts Balance

balance deficits are not always apparent

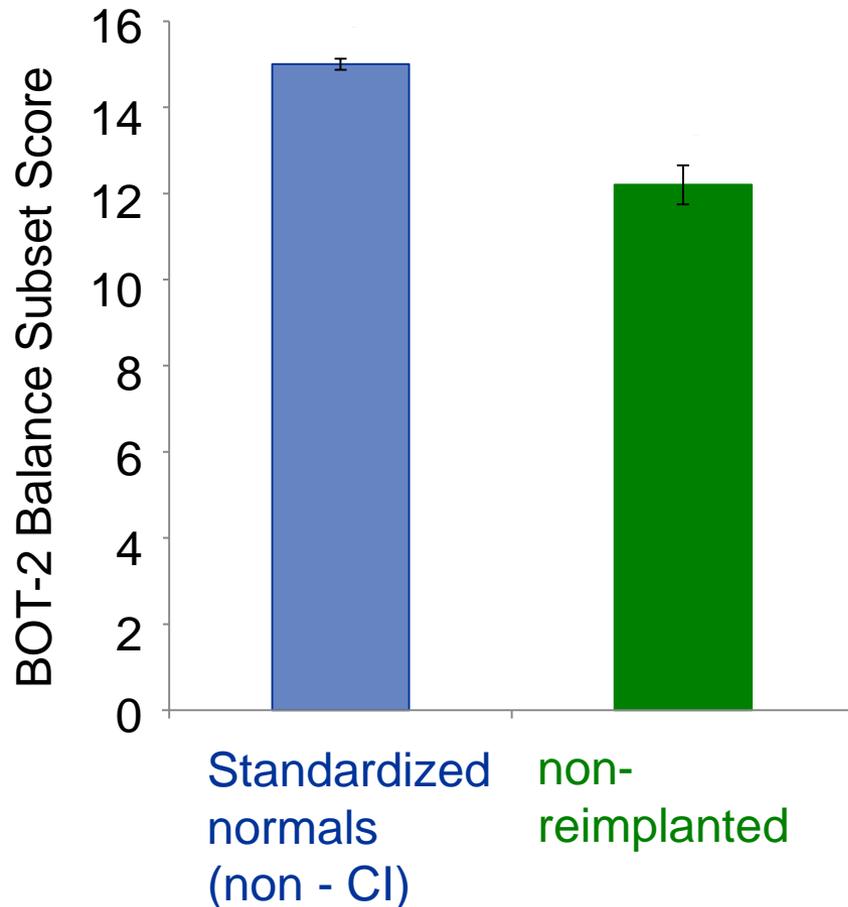




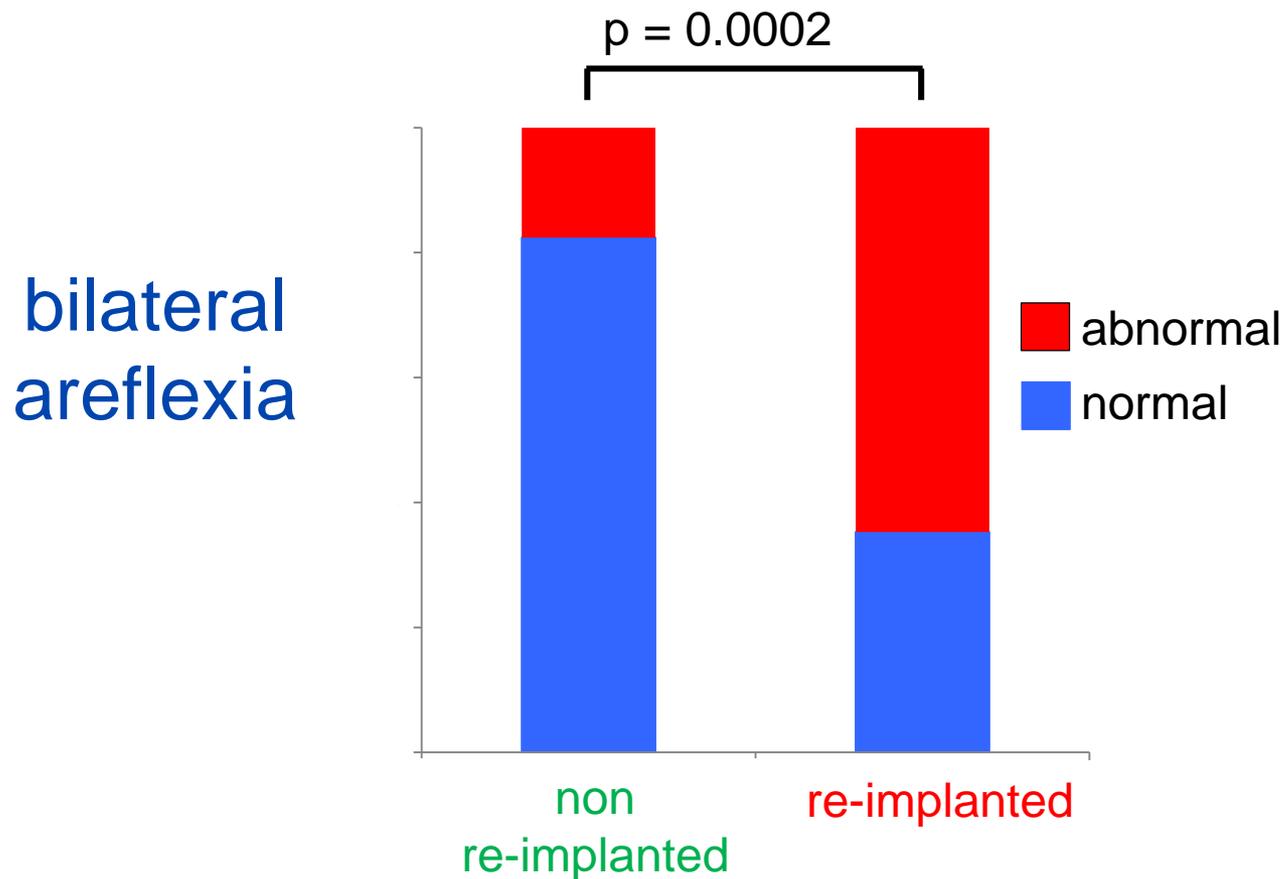


When Balance is Poor – Implants Fail

- 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

Screening Vestibular & Balance Function 4 Easy Steps



1. motor milestones
2. motor function
3. horizontal canal
4. otoliths

Motor Milestones – Upper Limits



- head control
 - 4 months
- sitting
 - 9 months
- walking
 - 18 months

Screening Vestibular & Balance Function 4 Easy Steps



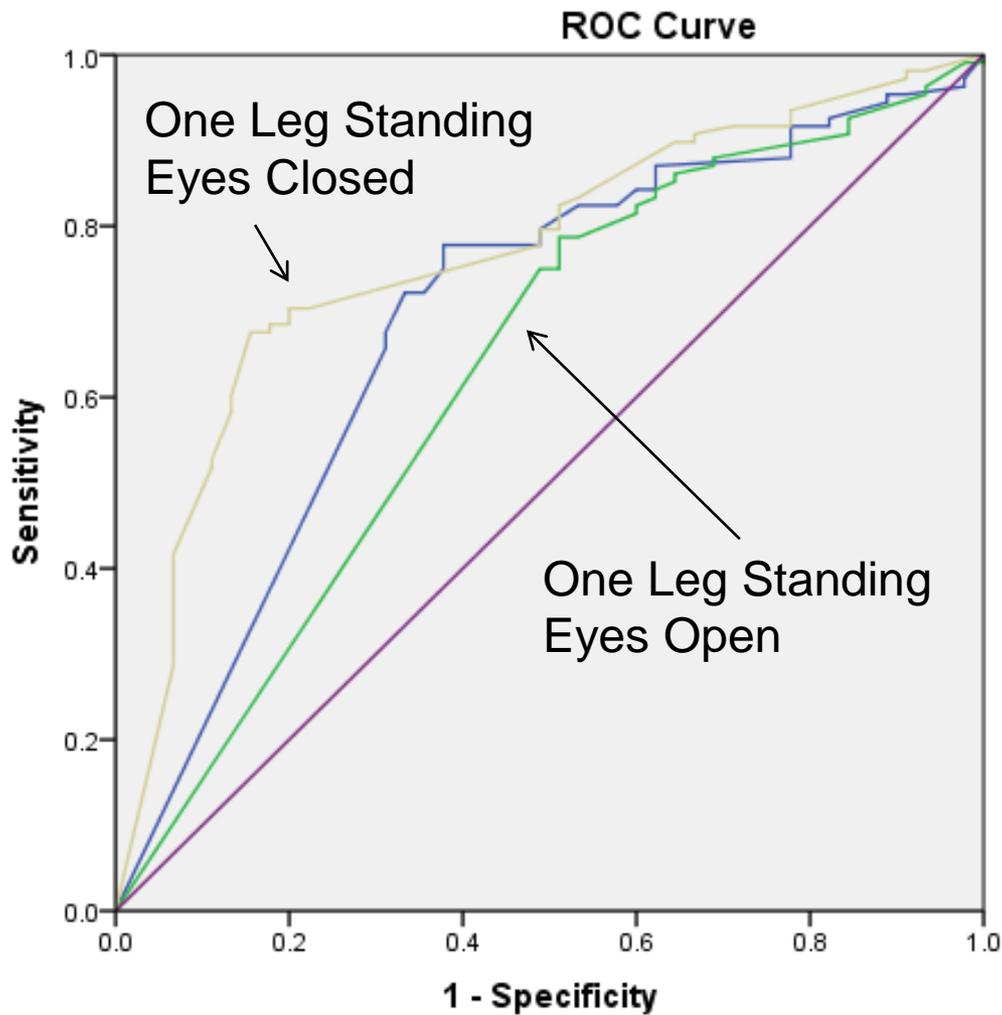
1. motor milestones
2. motor function
3. horizontal canal
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One Leg Standing

Age	Duration (sec) 1 foot standing
30 months	1 (briefly)
36 months	2
4 years	5
5 years	10

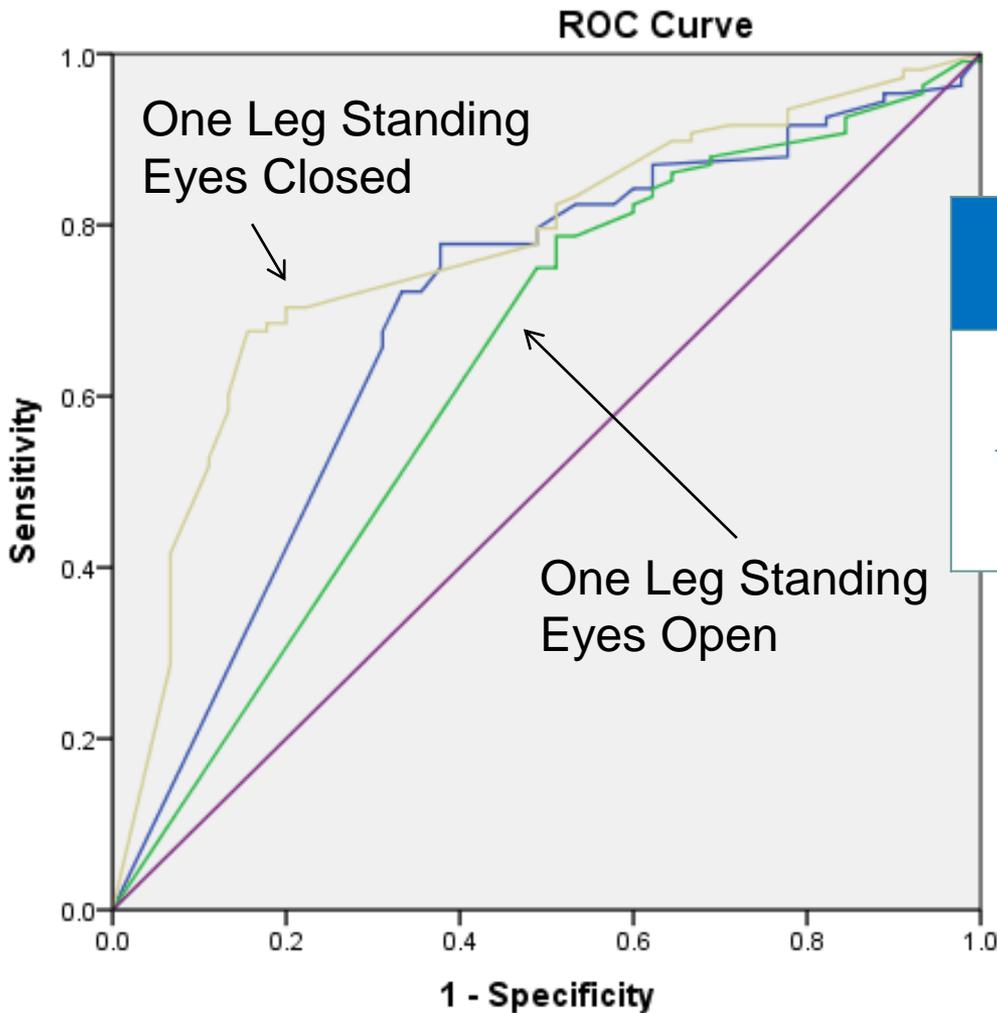


One Leg Standing



Diagonal segments are produced by ties.

One Leg Standing



Children age 5 and older

One Leg Standing	Sensitivity	Specificity
Eyes Open < 8 seconds	100%	49%
Eyes Closed < 4 seconds	90%	84%

Diagonal segments are produced by ties.

Screening Vestibular & Balance Function 4 Easy Steps



1. motor milestones
2. motor function
3. horizontal canal
4. otoliths

Assess Horizontal Canal Function



video Head Impulse Testing (vHIT) (EyeSeeCam, Interacoustics)



video Head Impulse Testing (vHIT) Synopsis



Infants < 6 months cannot visually suppress the VOR



Infants < 6 months cannot visually suppress the VOR

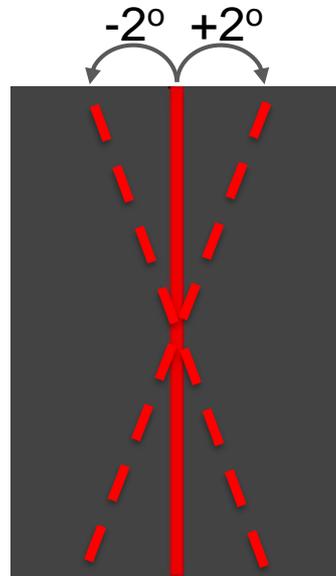


Screening Vestibular & Balance Function 4 Easy Steps



1. motor milestones
2. motor function
3. horizontal canal
4. otoliths

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)

Subjective Visual Vertical Chronos System, Interacoustics



Screening Vestibular & Balance Dysfunction

4 Easy Steps

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1. motor milestones
2. motor function
3. horizontal canal
4. otoliths

Vestibular and Balance Dysfunction Risk by Etiology

- Cochleovestibular anomalies
 - Phelps
 - Waardenburg
 - Klippel Feil
 - Pendred
- Ototoxicity
- Usher Syndrome
- Meningitis

- CMV
- ANSD

- Connexin
- Mitochondrial?
- Autoimmune?

Vestibular and Balance Dysfunction Risk by Etiology

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Ototoxicity in Cancer

cochleotoxicity \approx vestibulotoxicity

25-90%

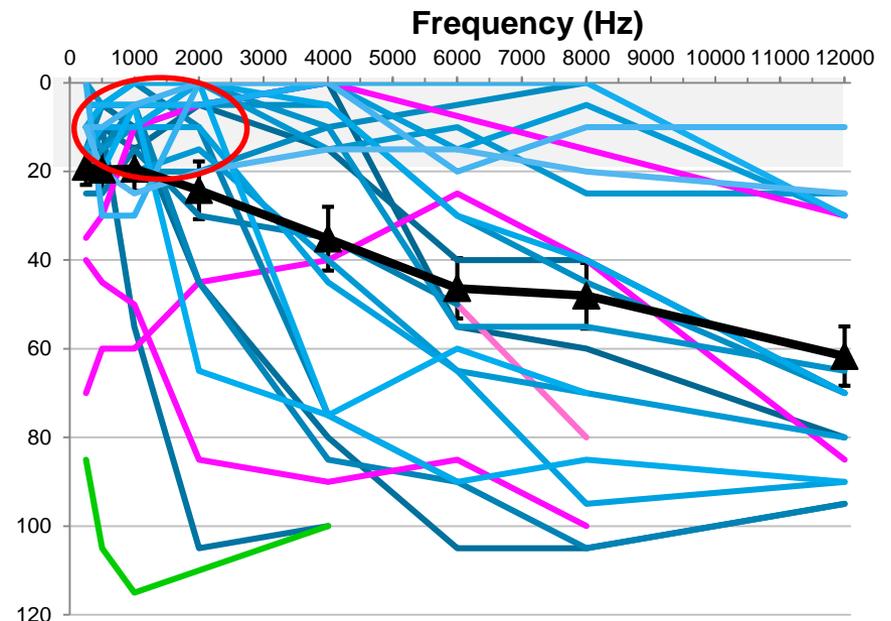
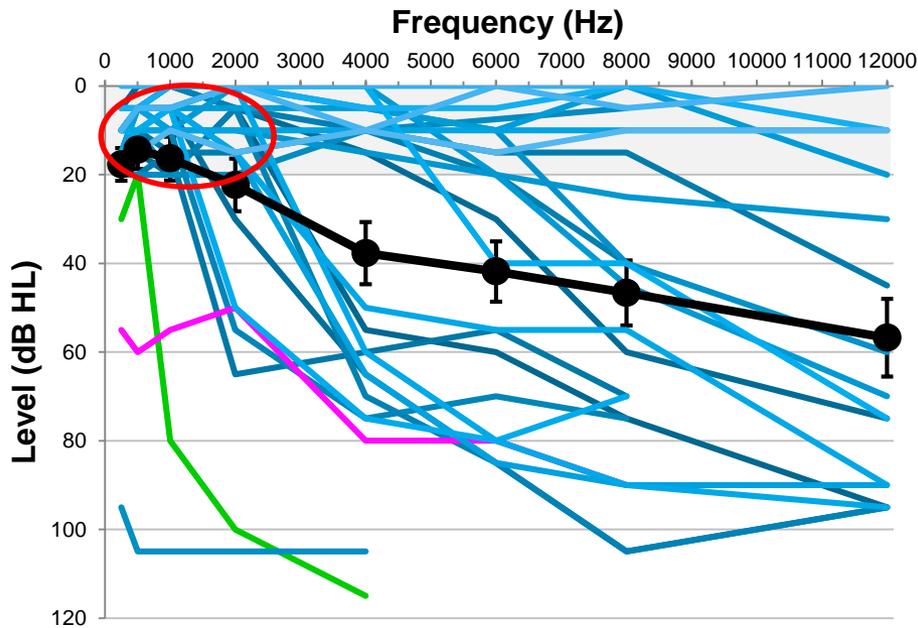
?

platinum based therapy
aminoglycosides
cranial radiation
surgical therapy

Ototoxicity Related Hearing Loss

Left Ear

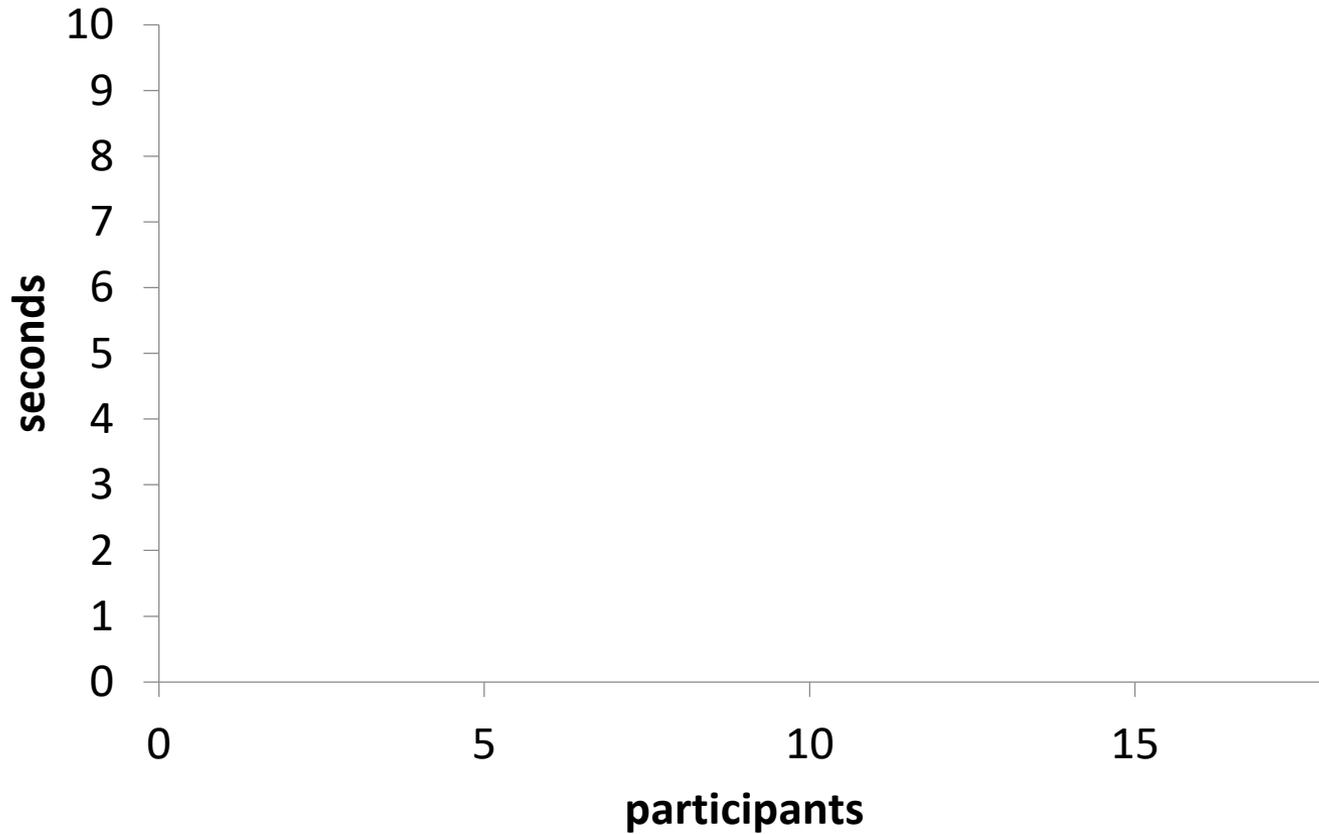
Right Ear



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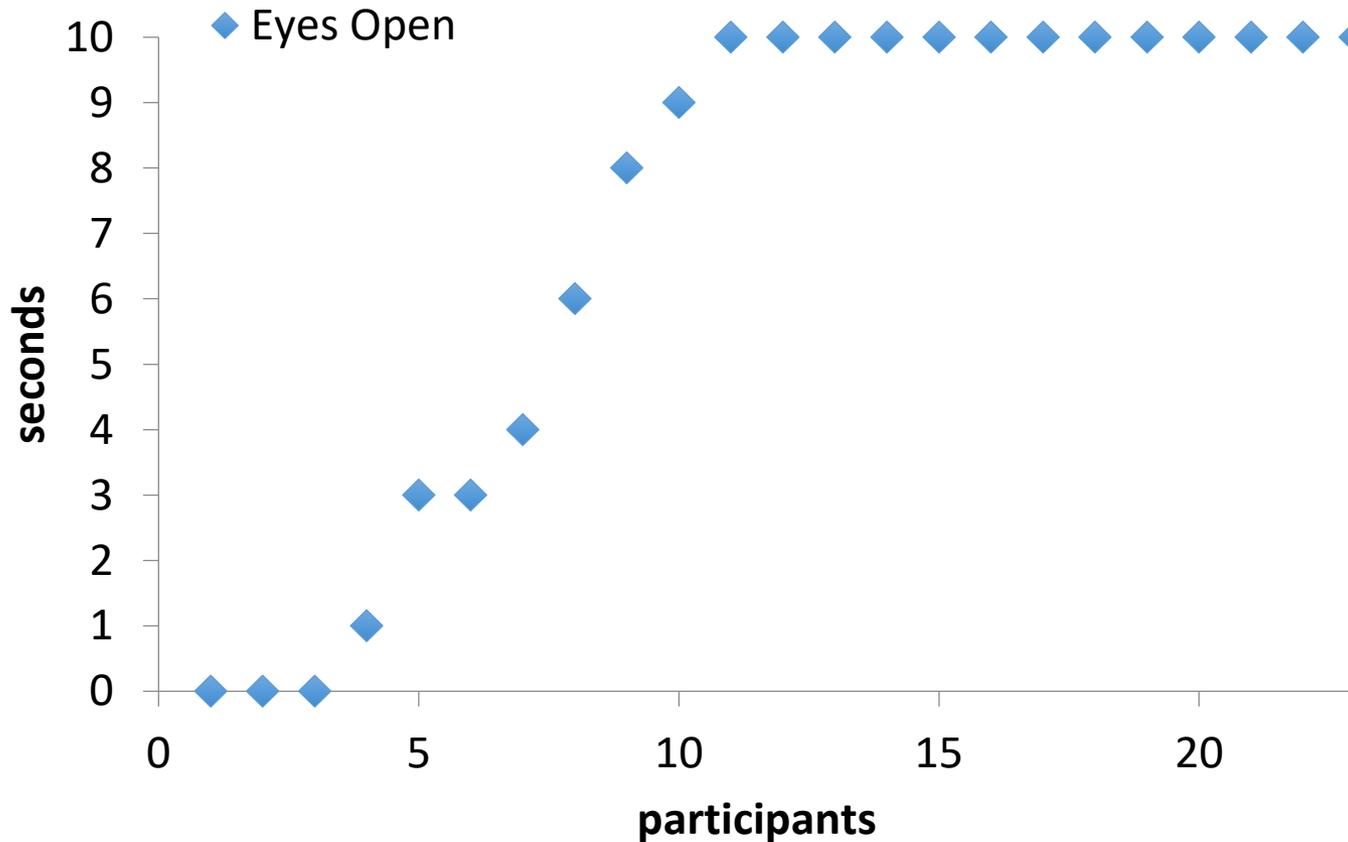
— Conductive hearing loss
— Cochlear implant

Ototoxicity Related Balance Impairment



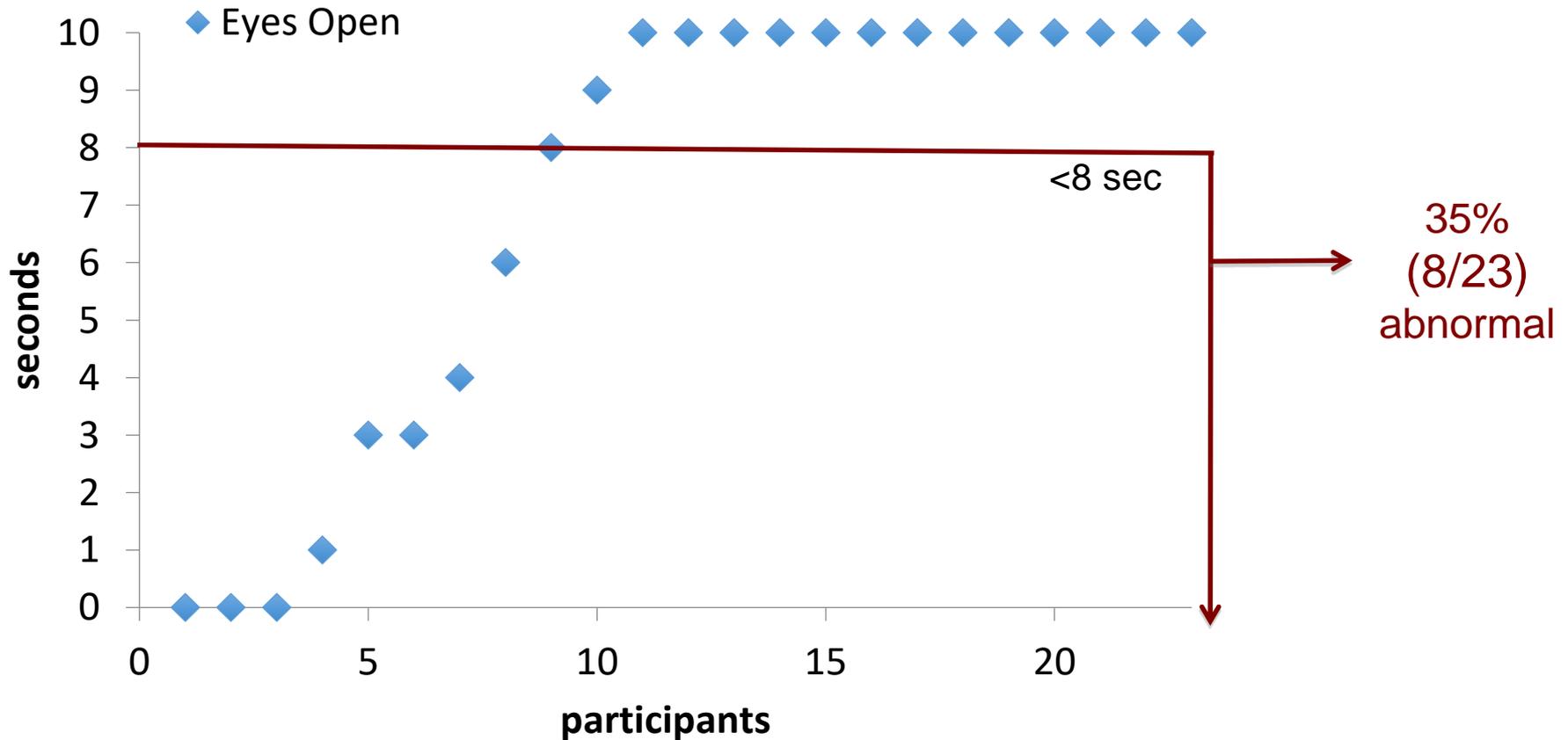
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Ototoxicity Related Balance Impairment



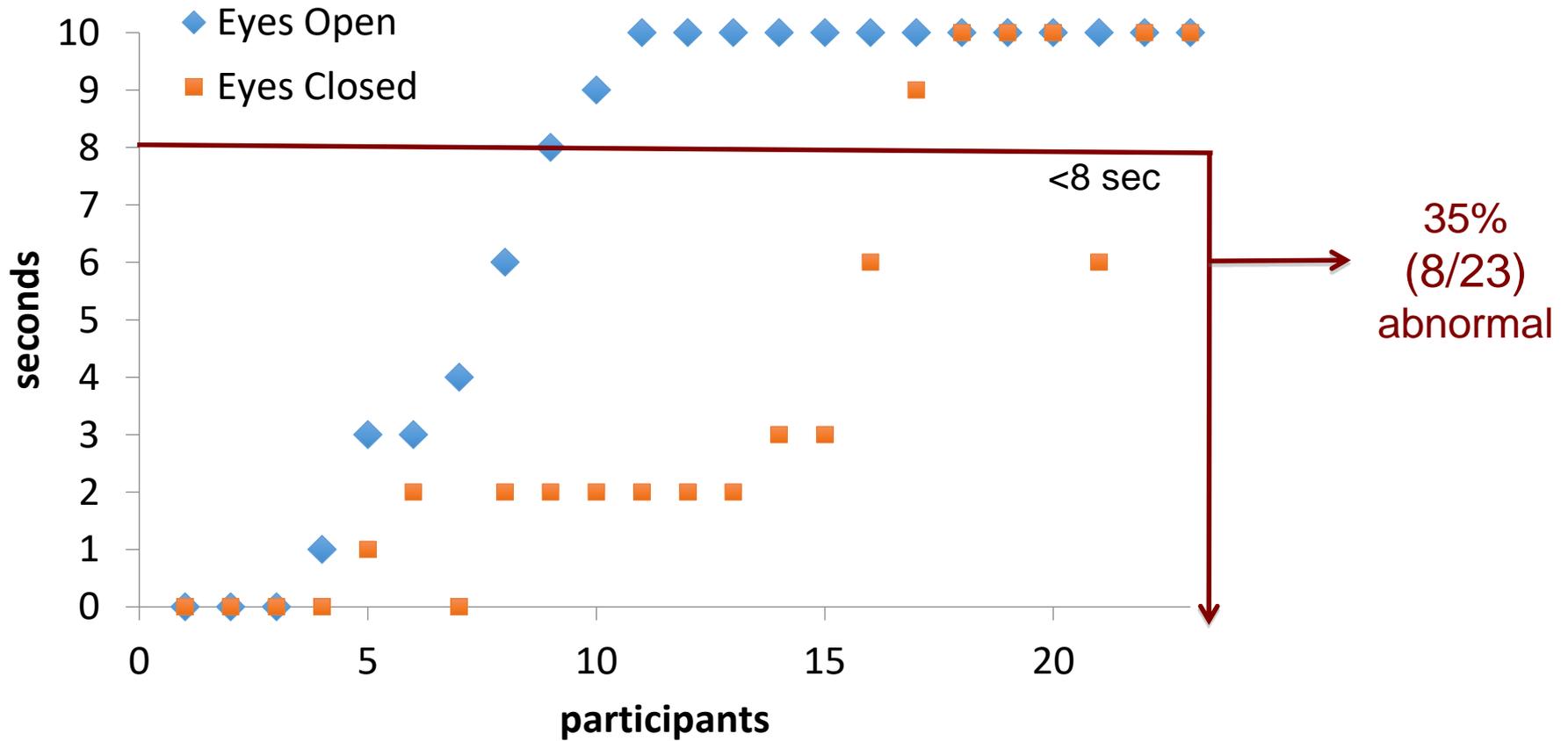
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Ototoxicity Related Balance Impairment



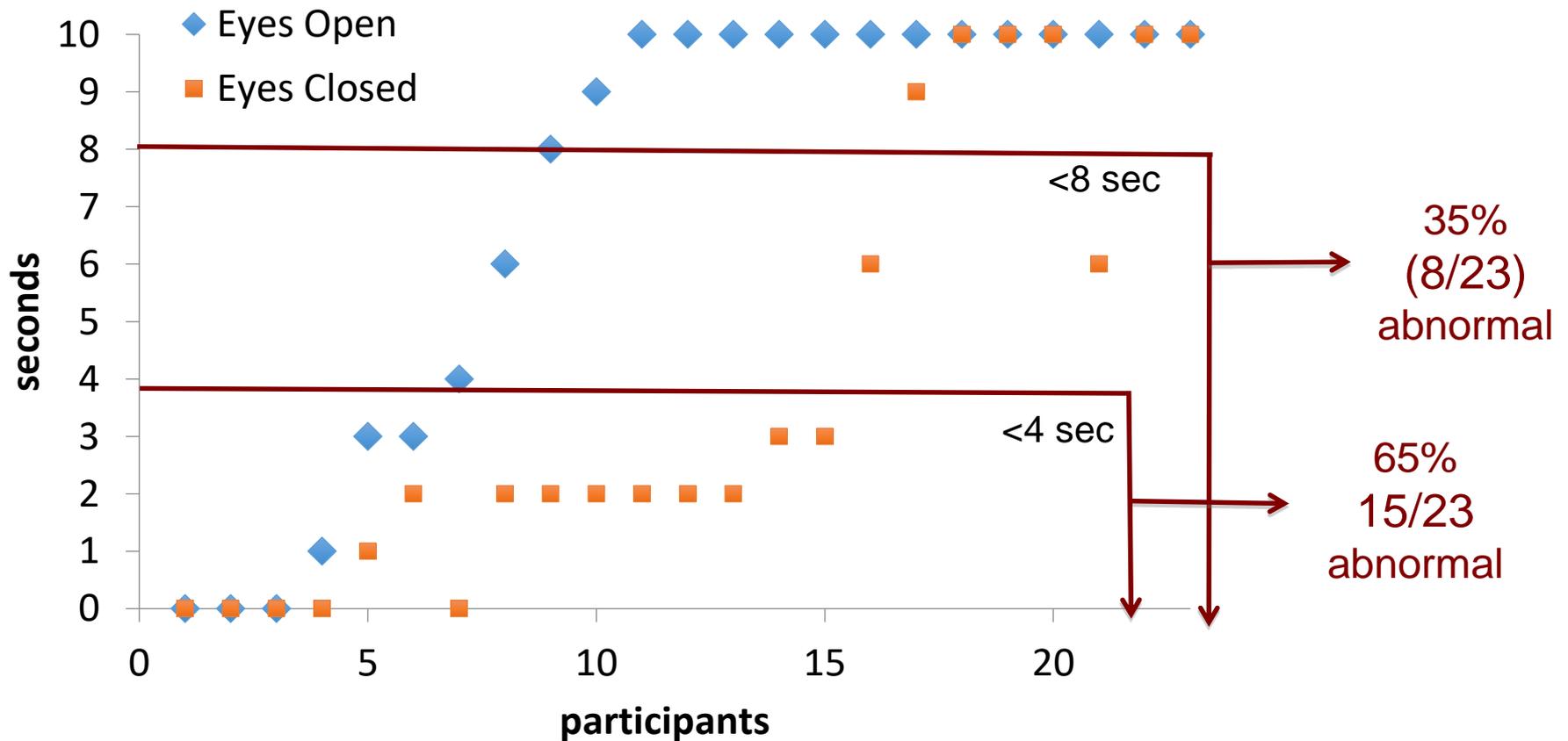
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Ototoxicity Related Balance Impairment



n=26

Ototoxicity Related Balance Impairment

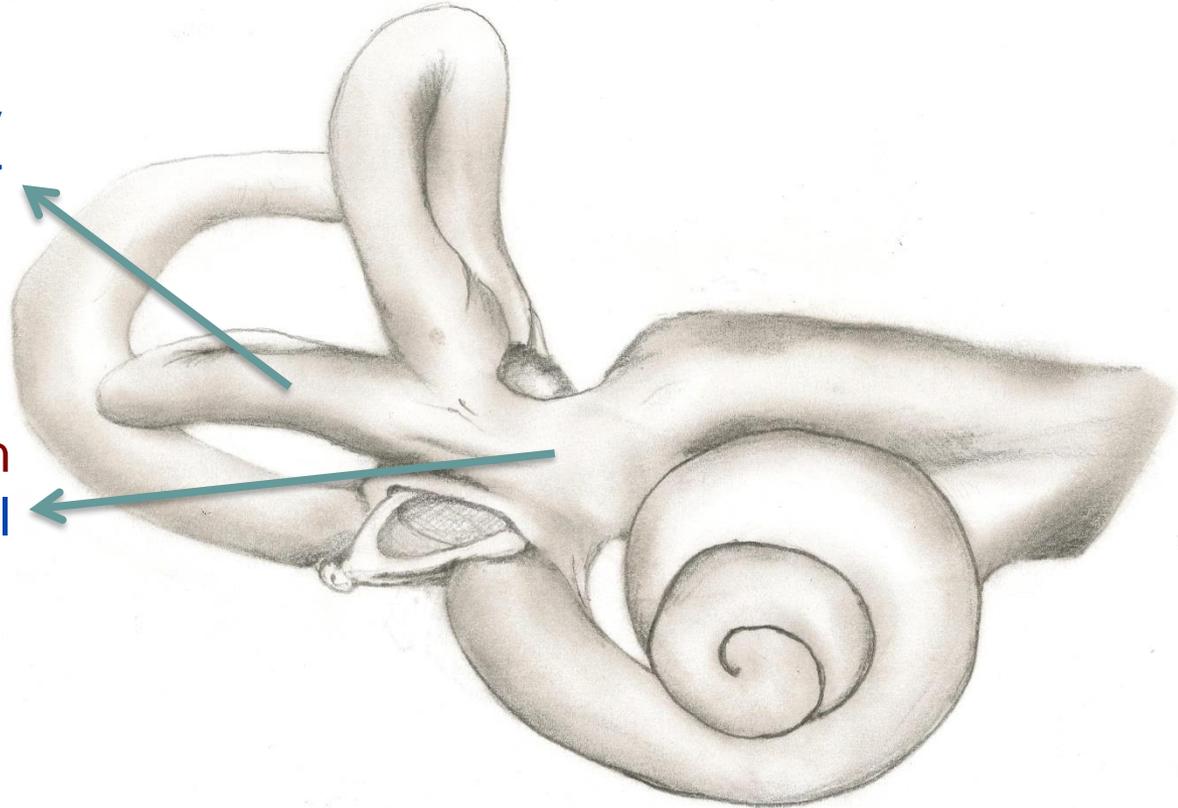


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Vestibular Impairment from Ototoxicity

Horizontal Canal Dysfunction
50% Dynamic Visual Acuity
52% VOR loss vHIT

42% Utricular Dysfunction
Subjective Visual Vertical



Ototoxicity in Cancer

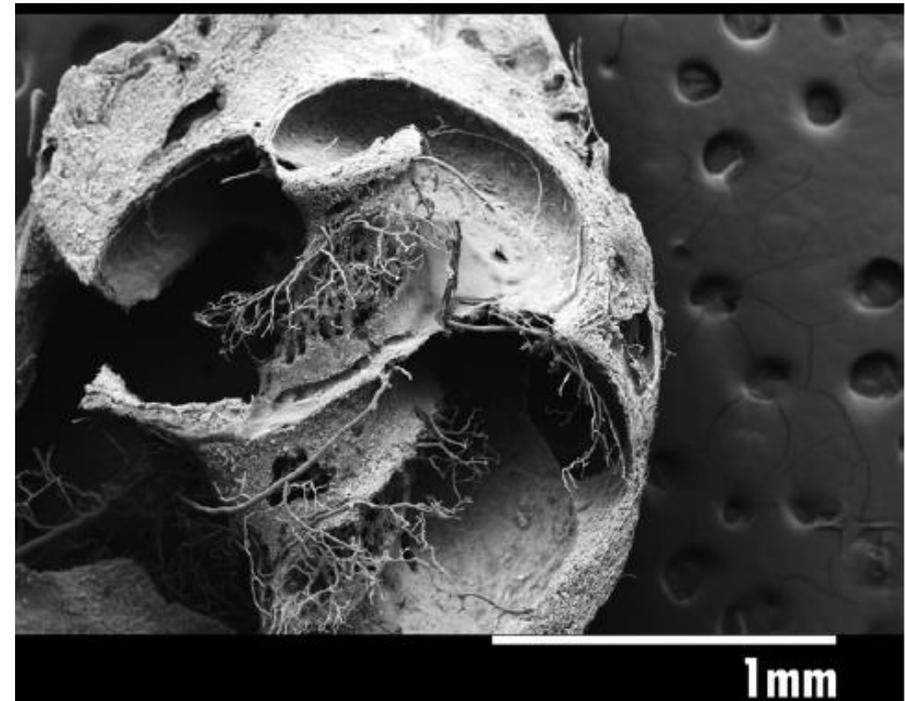
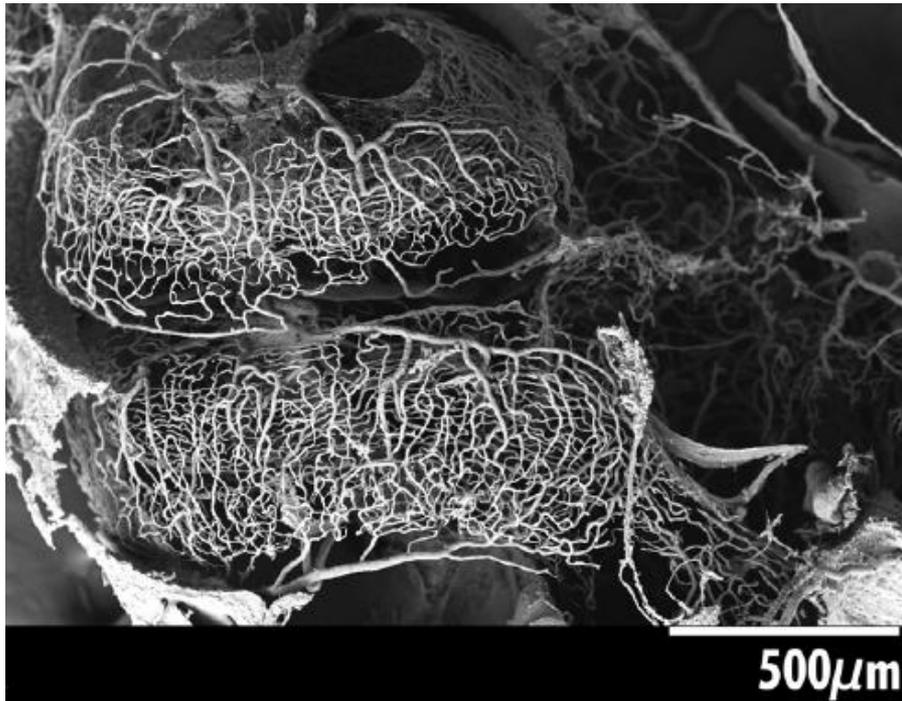
cochleotoxicity \approx vestibulotoxicity

25-90%

50%

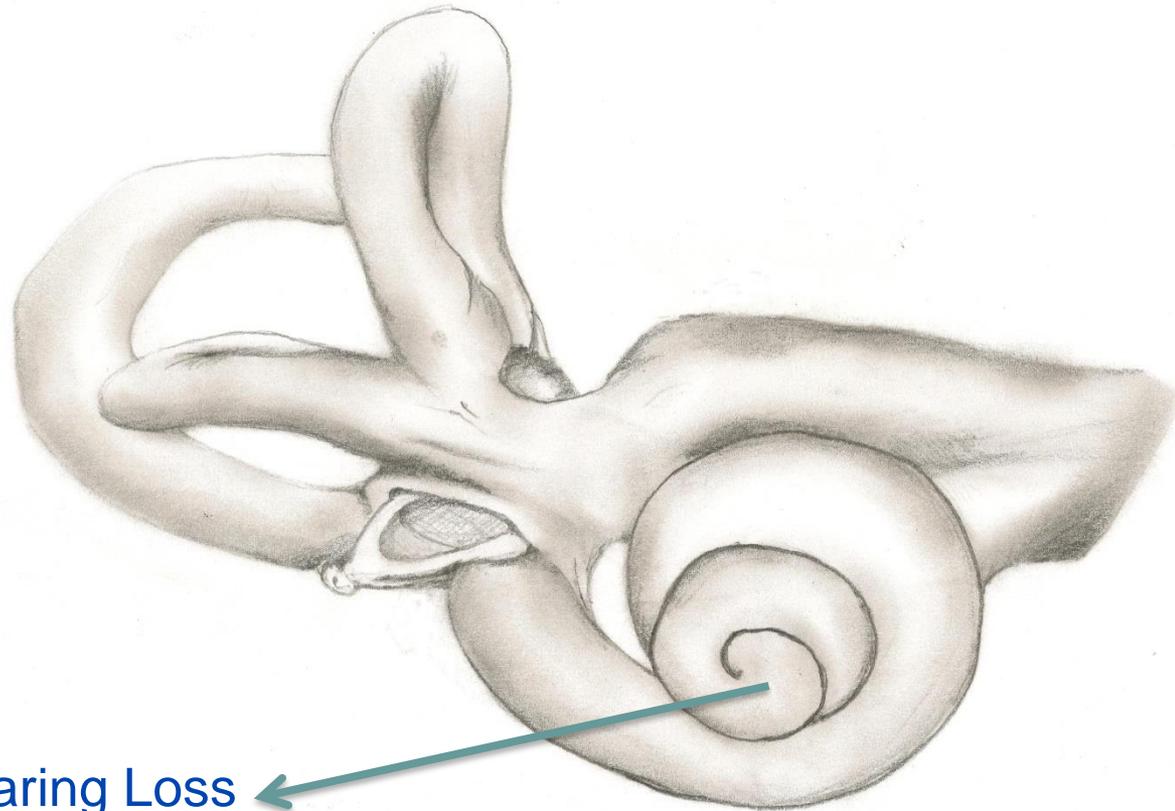
platinum based therapy
aminoglycosides
cranial radiation
surgical therapy

Congenital Cytomegalovirus



photos courtesy of Dr. Robert Harrison

Congenital Cytomegalovirus



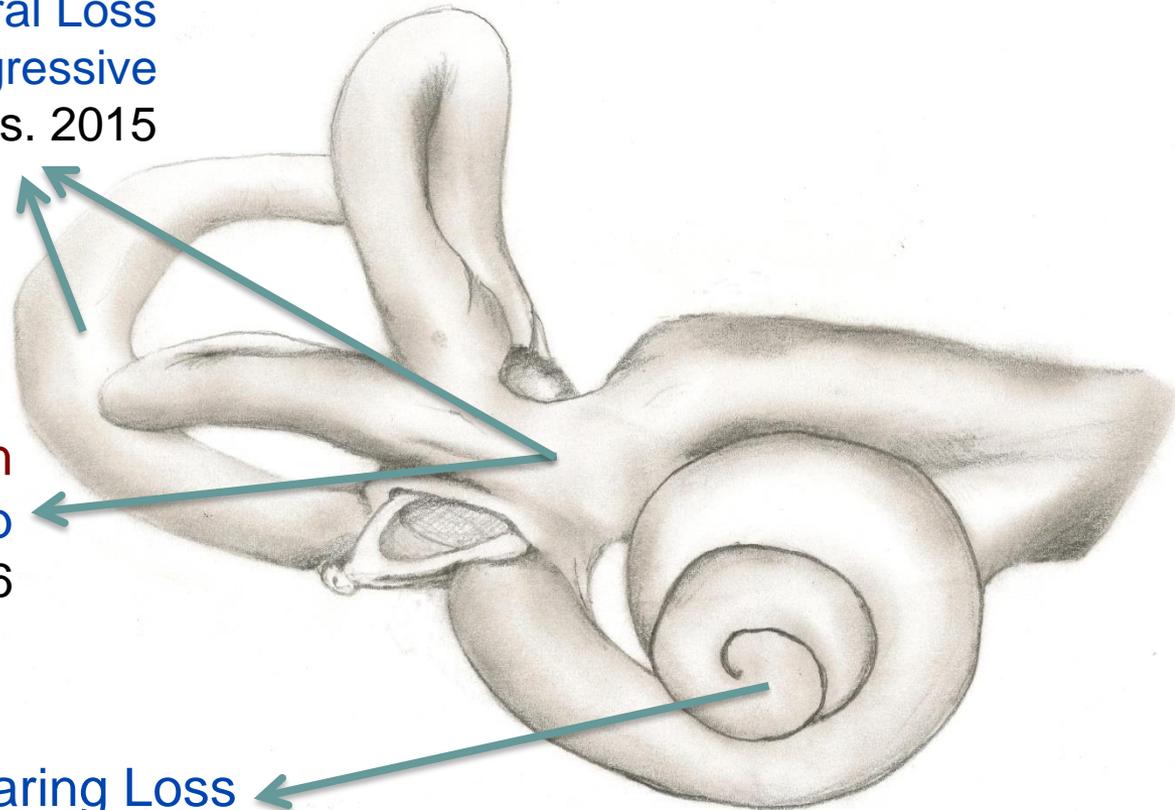
20-50% Sensorineural Hearing Loss

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



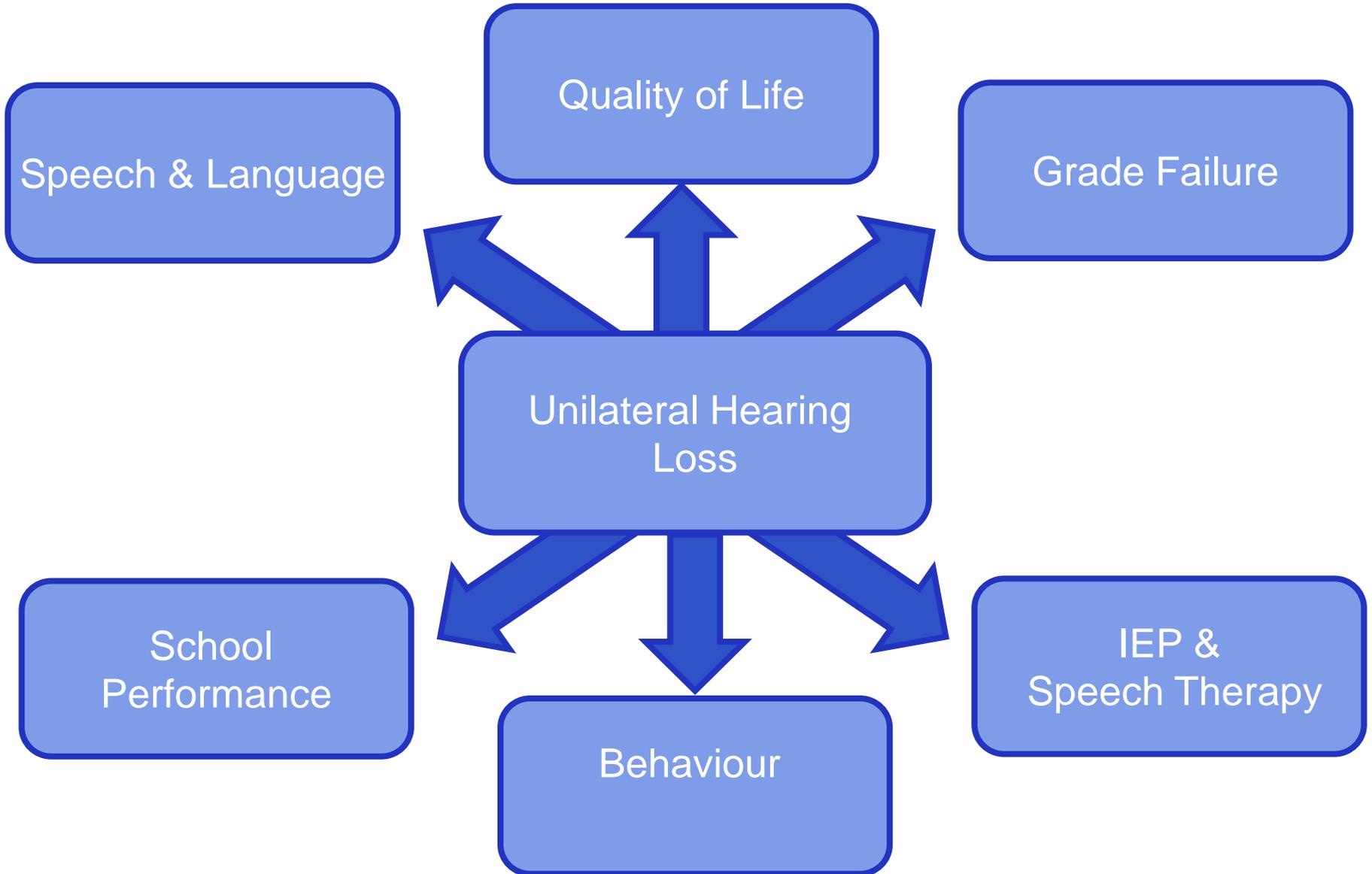
Single Sided Deafness in Children



Single Sided Deafness in Children

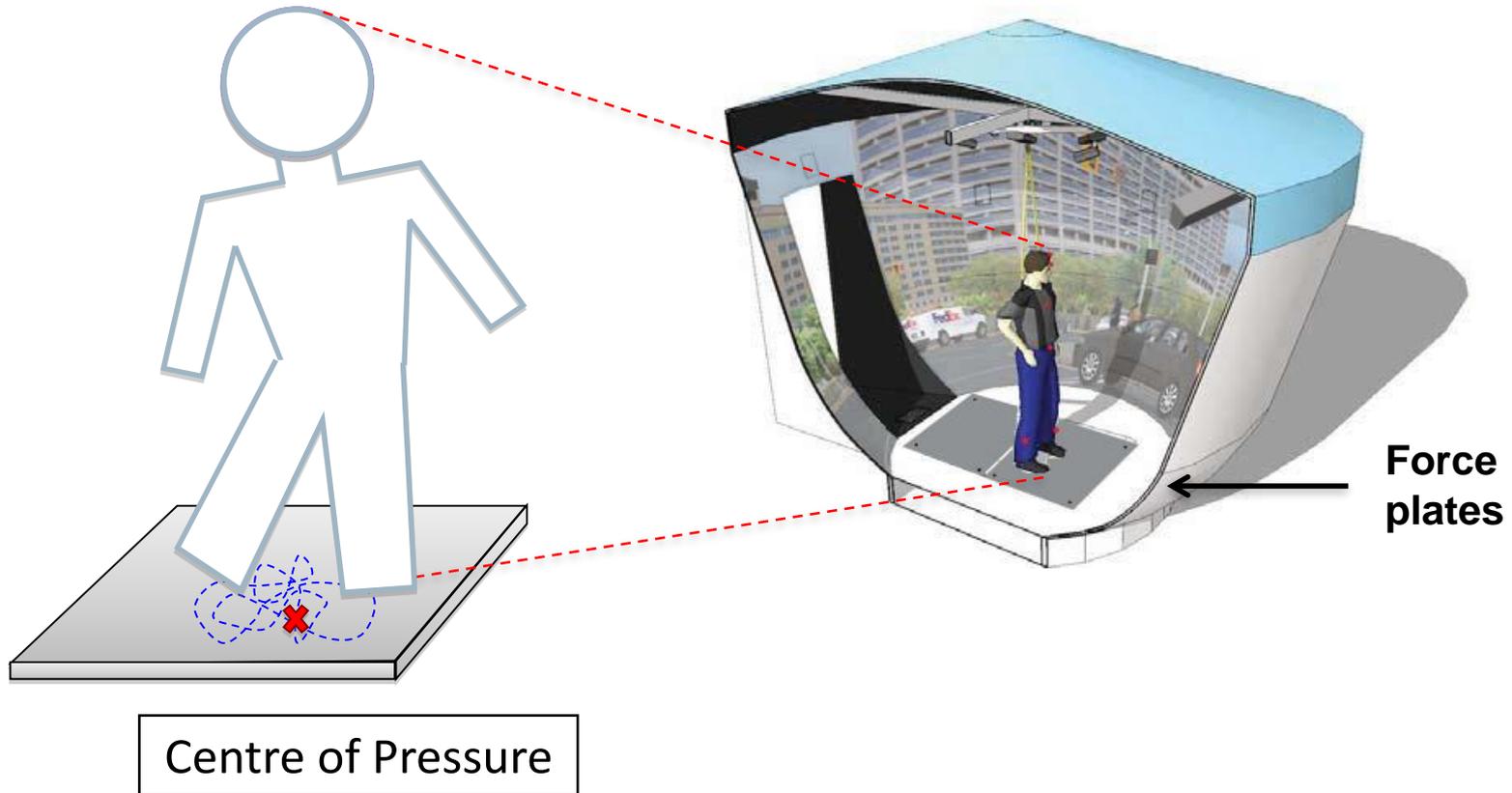


Consequences of SSD in Children

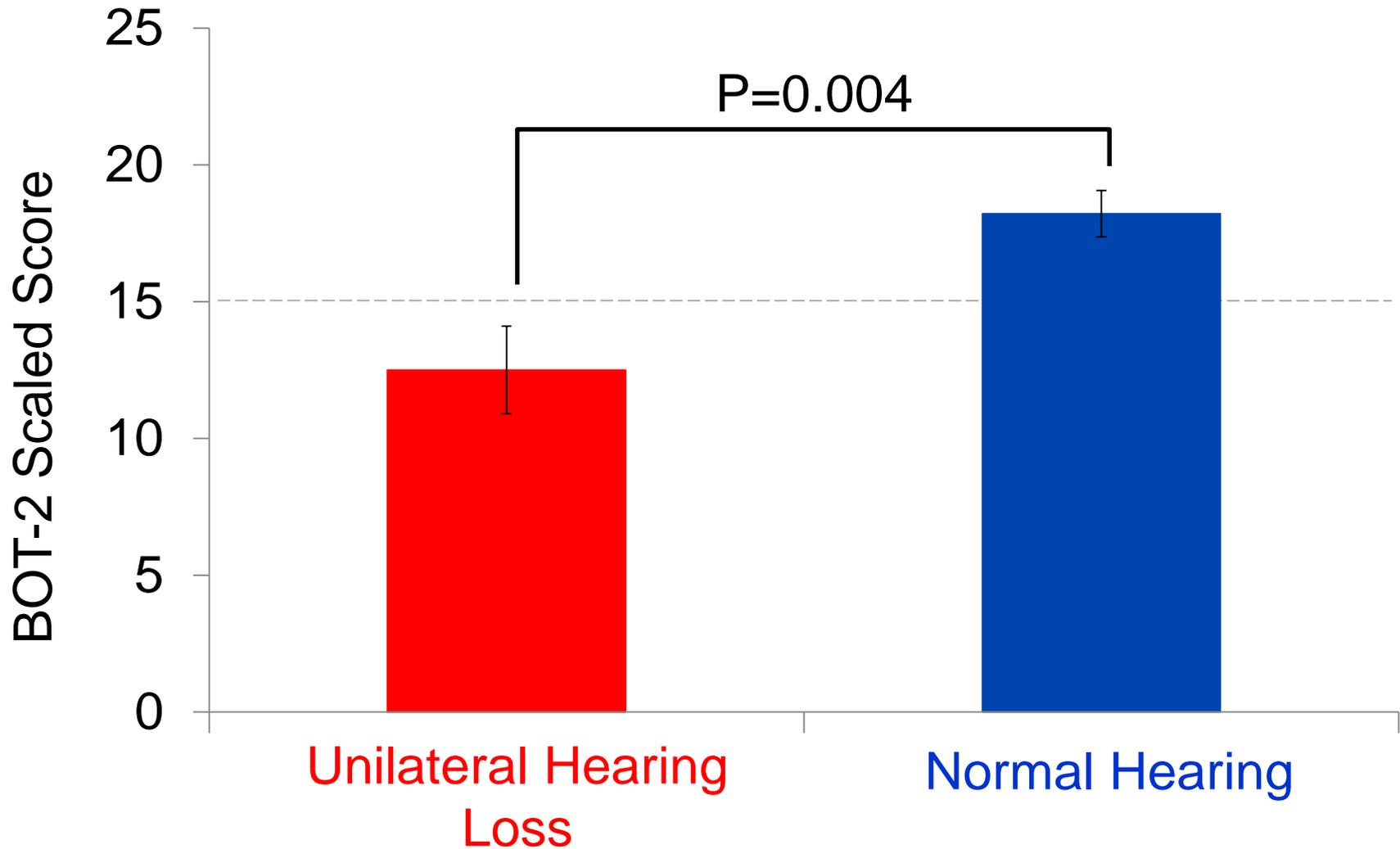


Measuring Balance Impairment

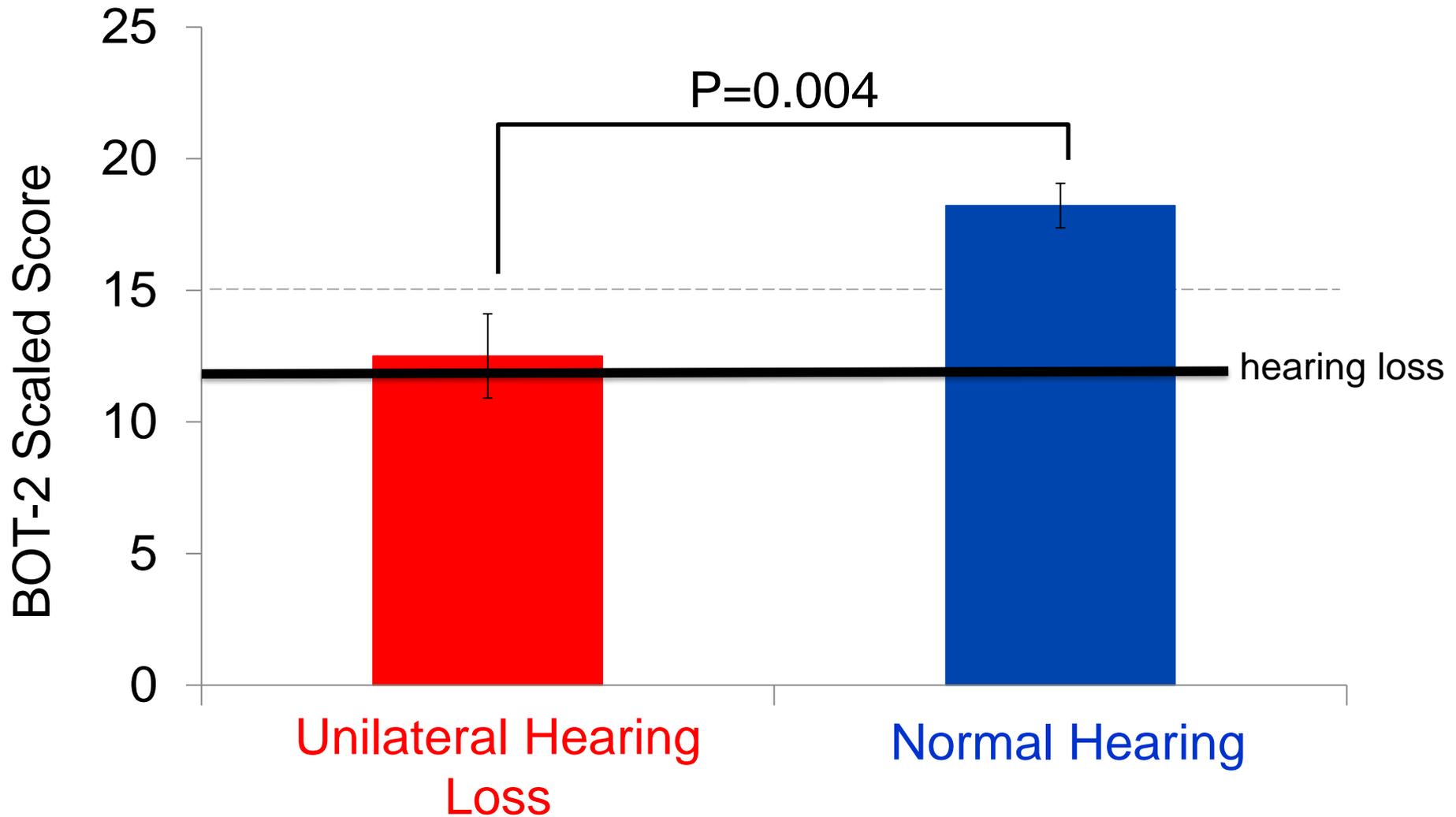
- quantitative measurement of postural control



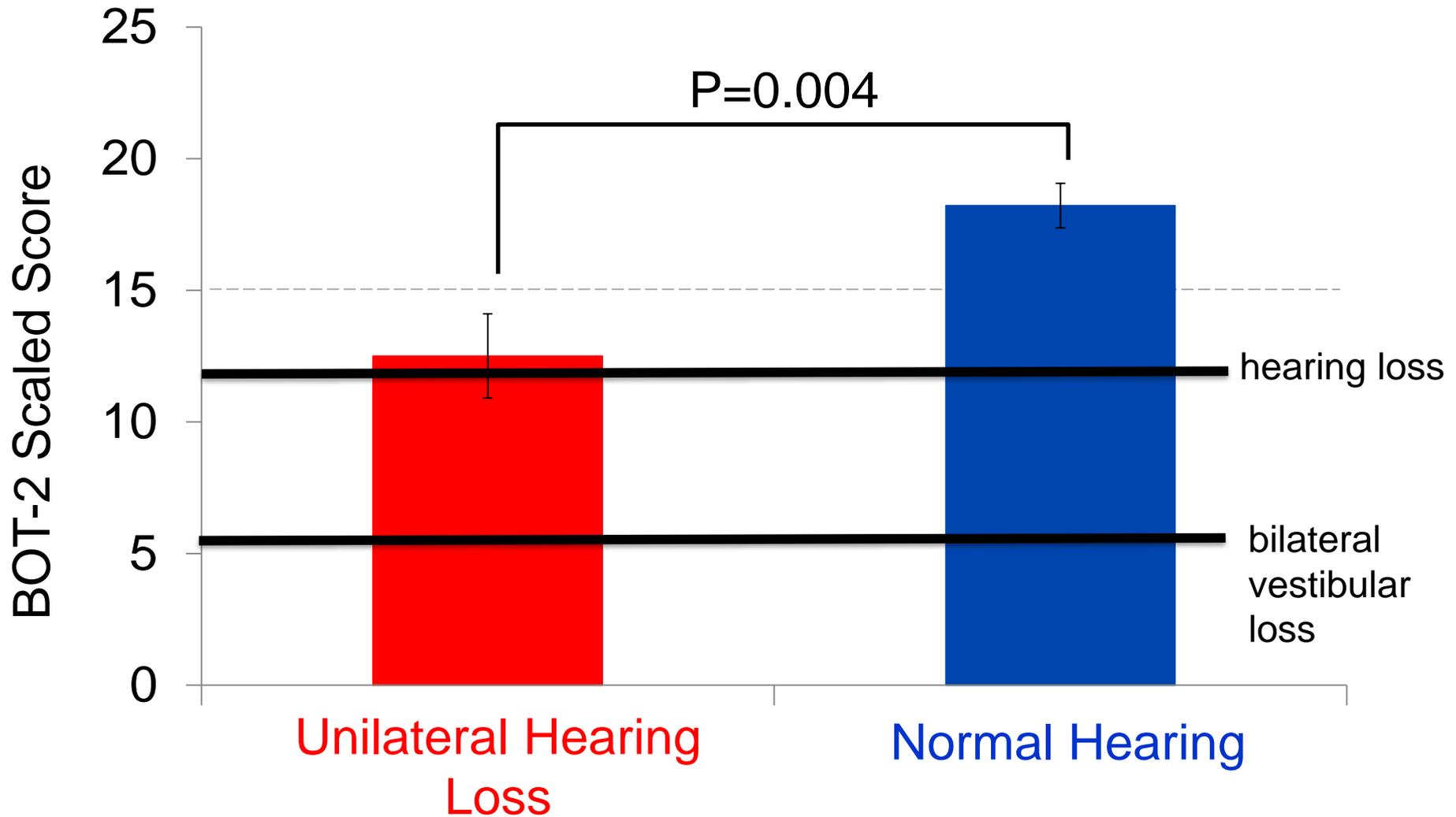
Balance is Impaired in SSD



Balance is Impaired in SSD



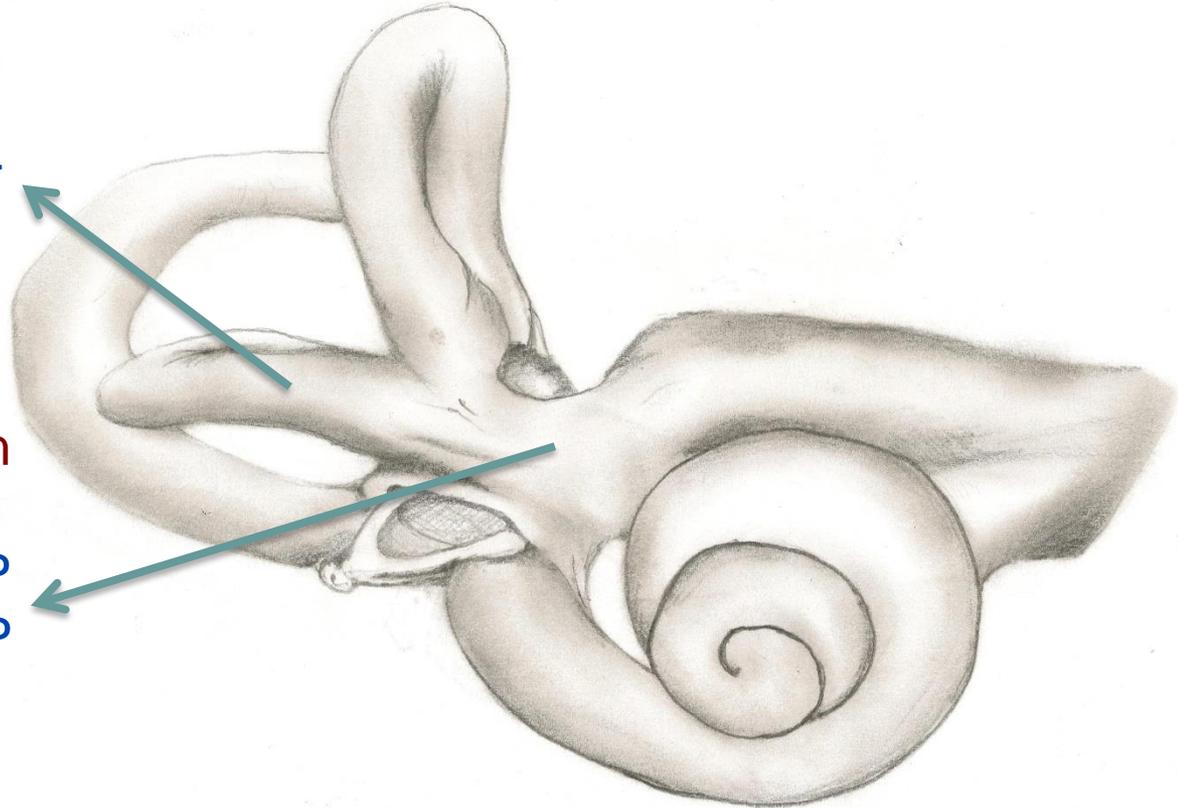
Balance is Impaired in SSD



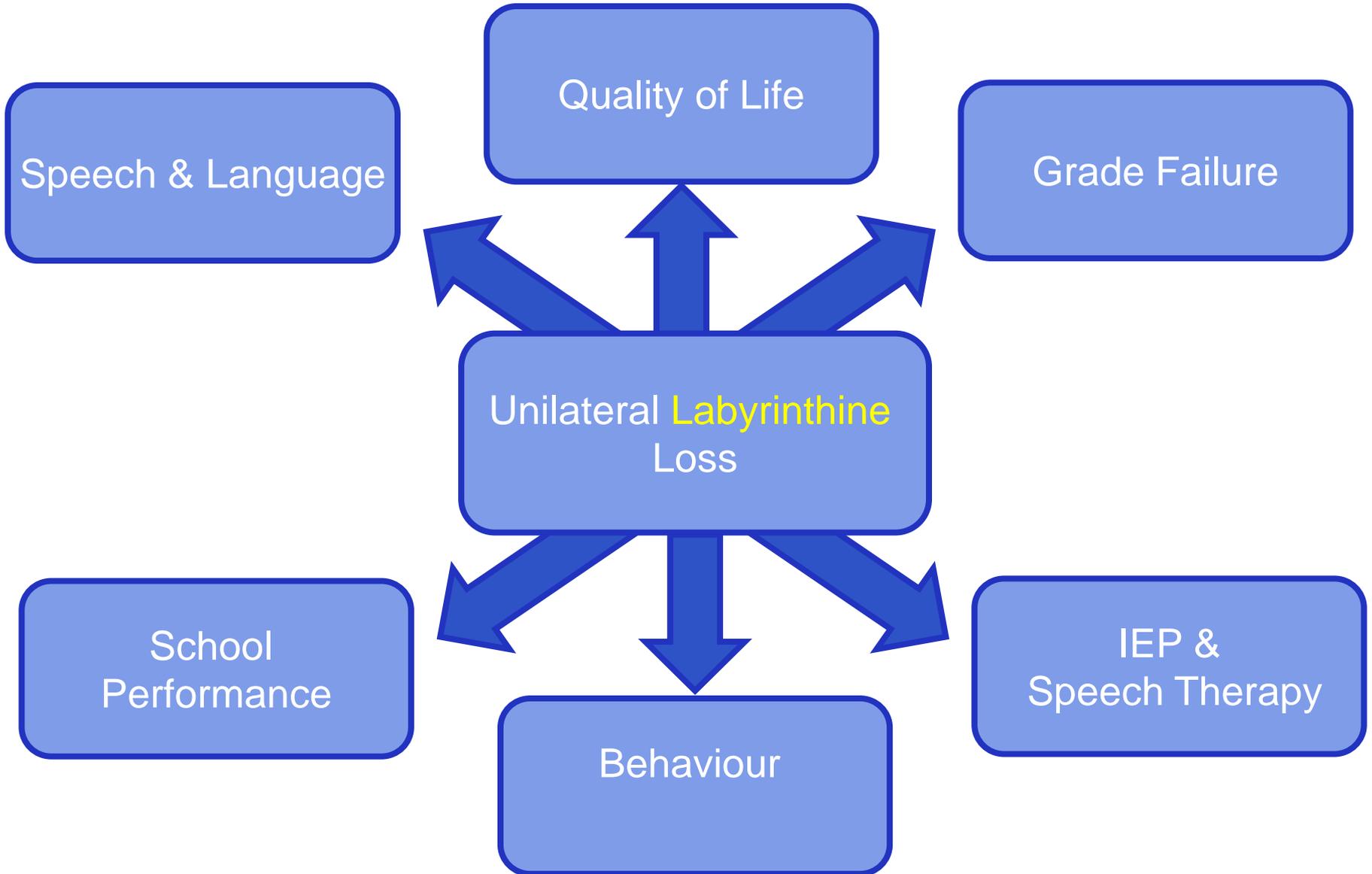
Vestibular Dysfunction in SSD

Horizontal Canal Dysfunction
48% Caloric Reduction
50% VOR loss vHIT

Otolithic Dysfunction
62.5%
Utricular 42% oVEMP
Saccular 25% cVEMP



Consequences of SSD in Children



Cognitive Decline

SickKids®

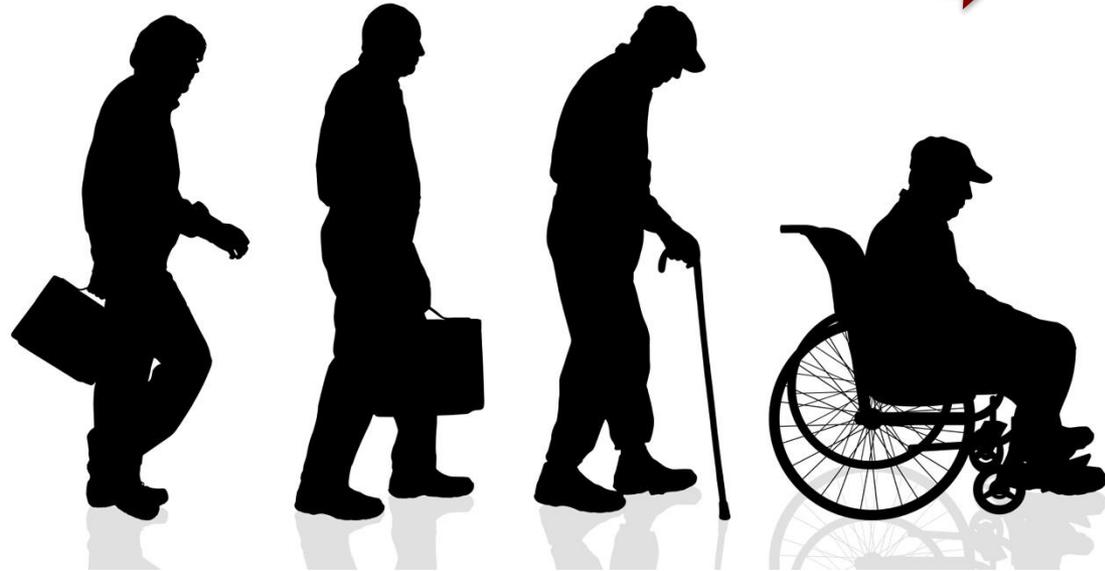
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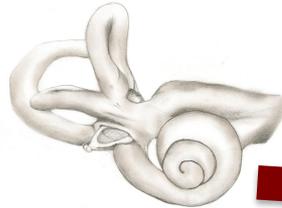
Cognitive Decline



Cognitive Decline

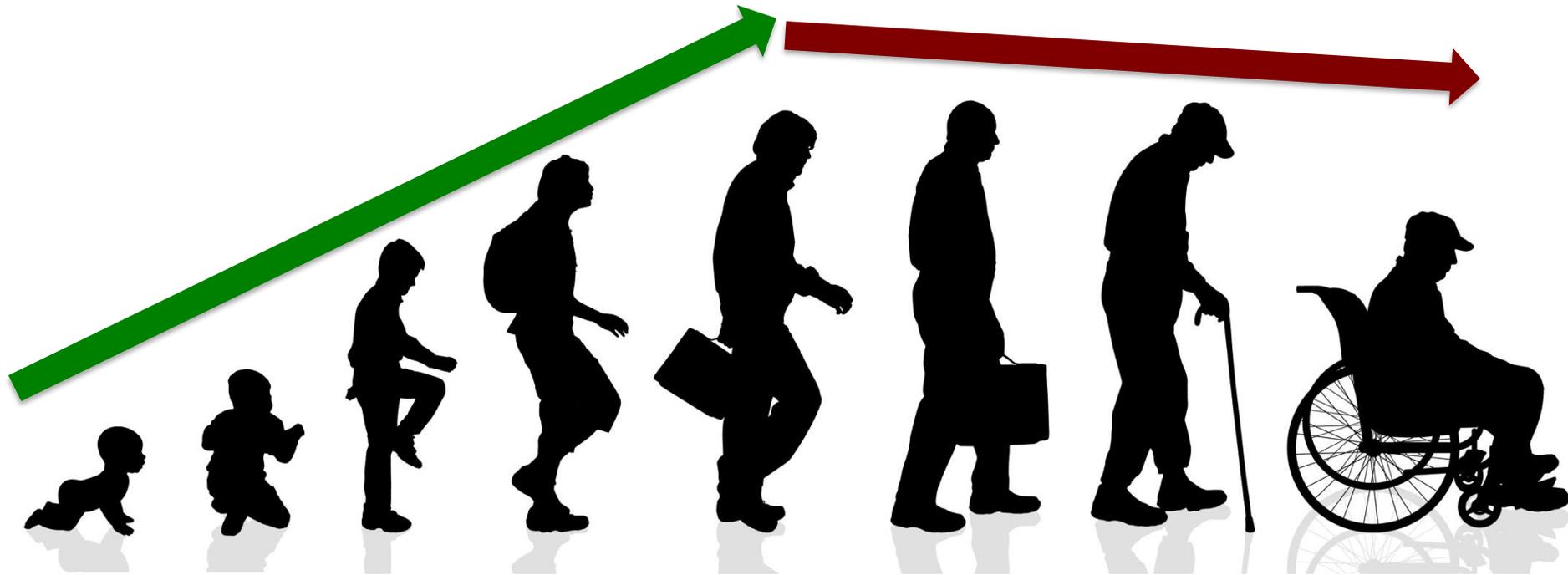


Cognitive Decline





Cognitive Incline



Disclosures (cont'd)

Everything I have learned about brain development
in children I have learned from 2 people



Dr. Karen Gordon



Xavier Sloan

Auditory Deprivation

Reduced activity



Duration of deafness

6.5 yr

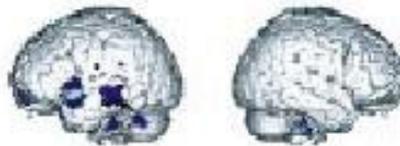


6.5 yr



11.2 yr

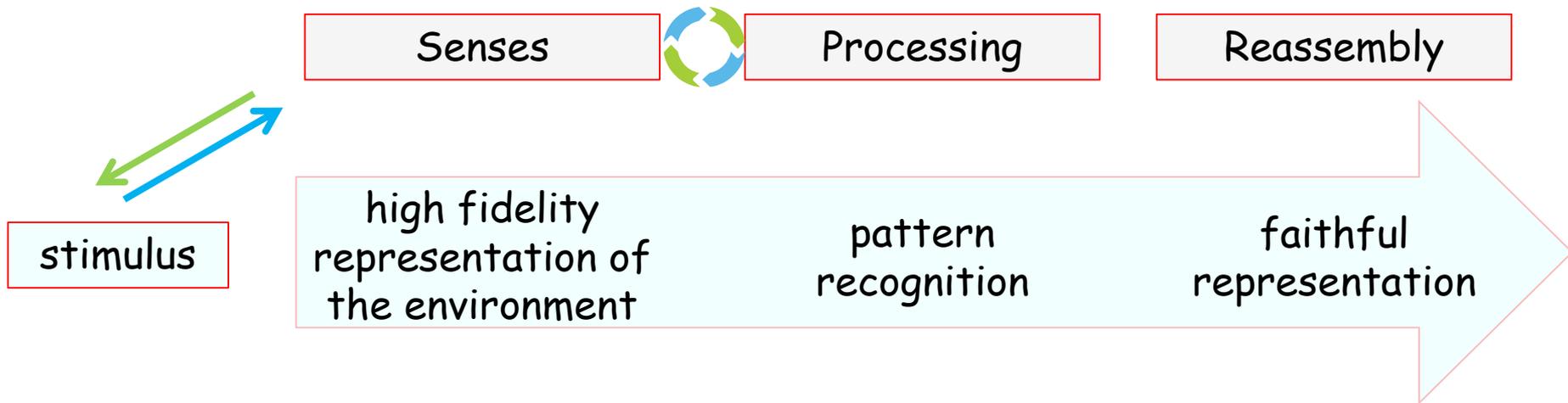
Activity no longer reduced



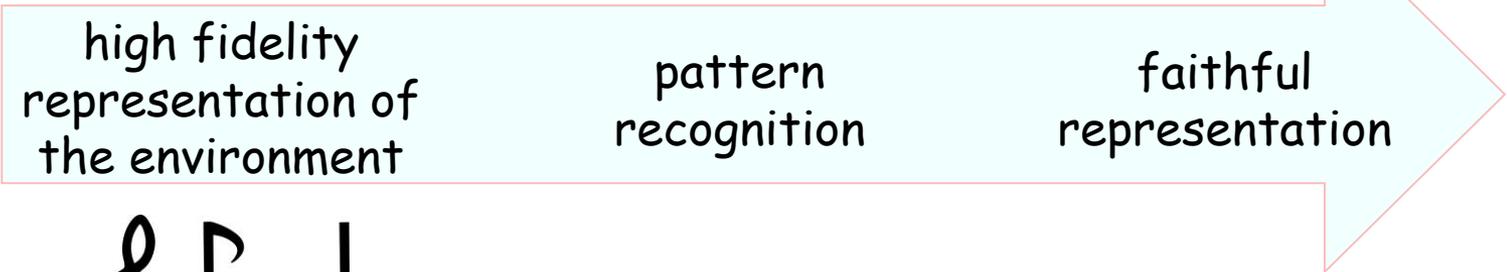
20.3 yr

Lee, et al., Nature, 2001

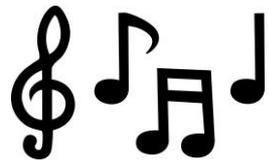
Sensory Assembly



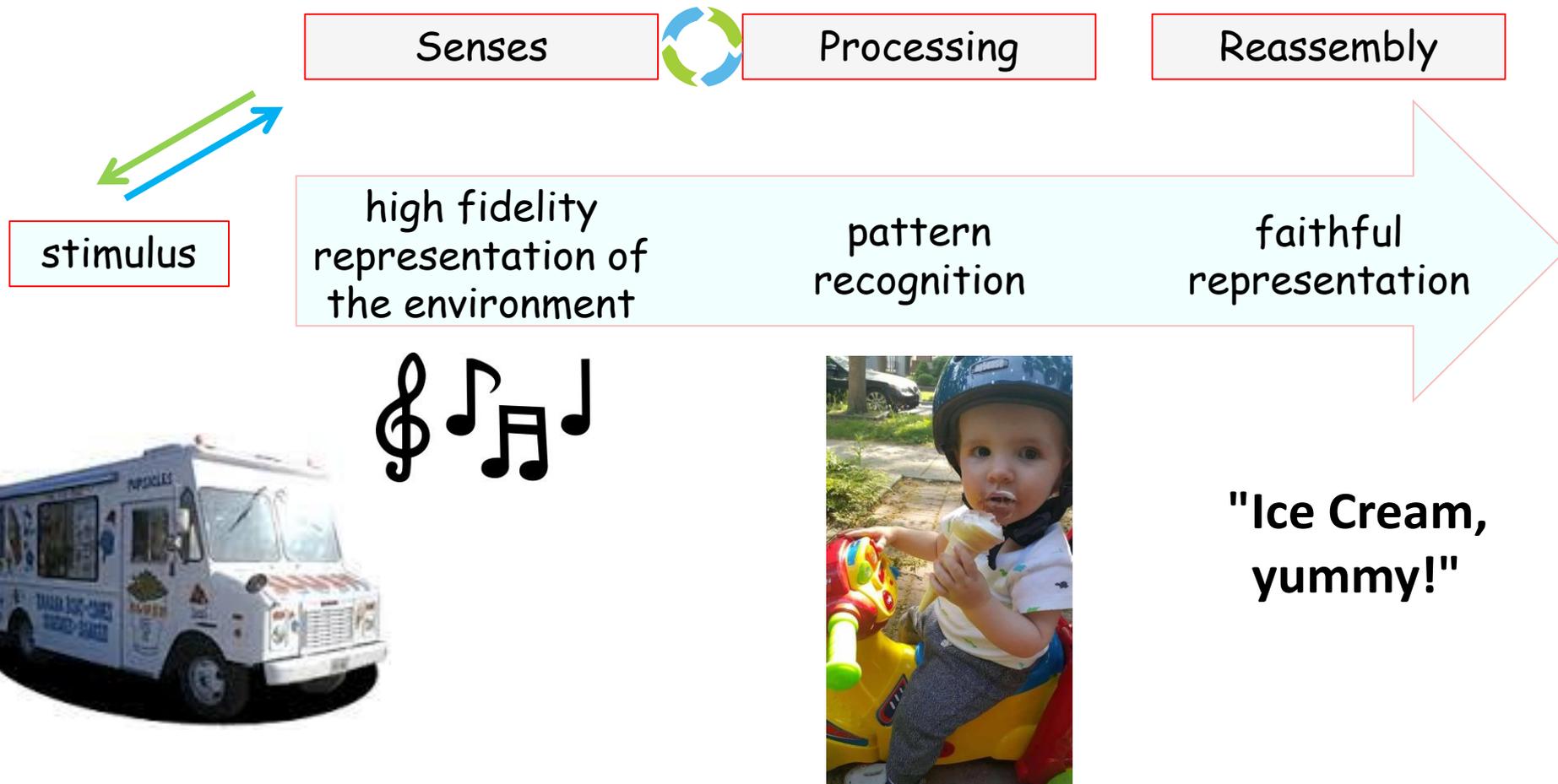
Sensory Assembly



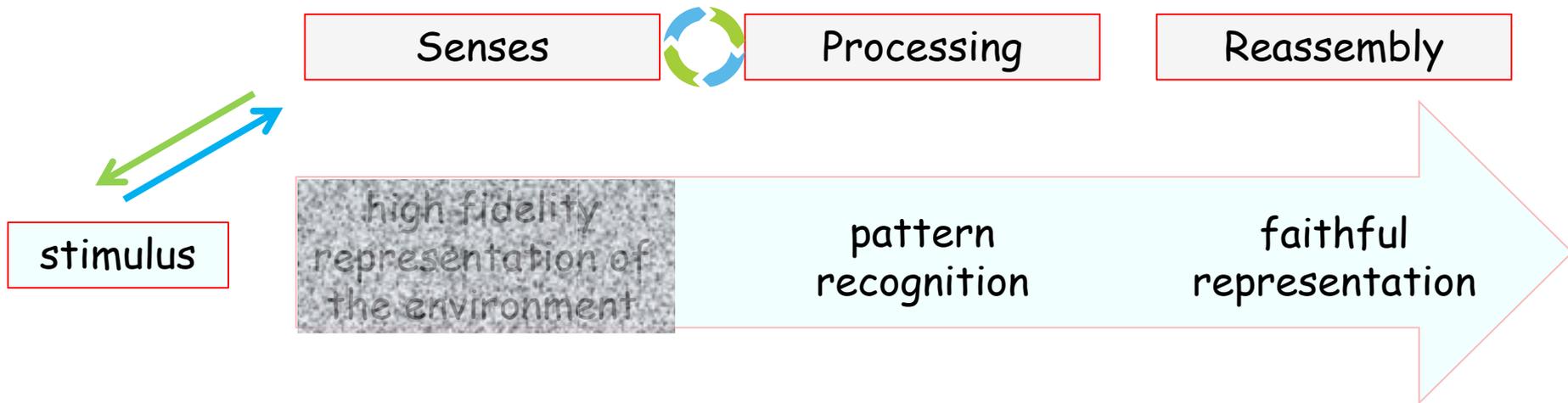
stimulus



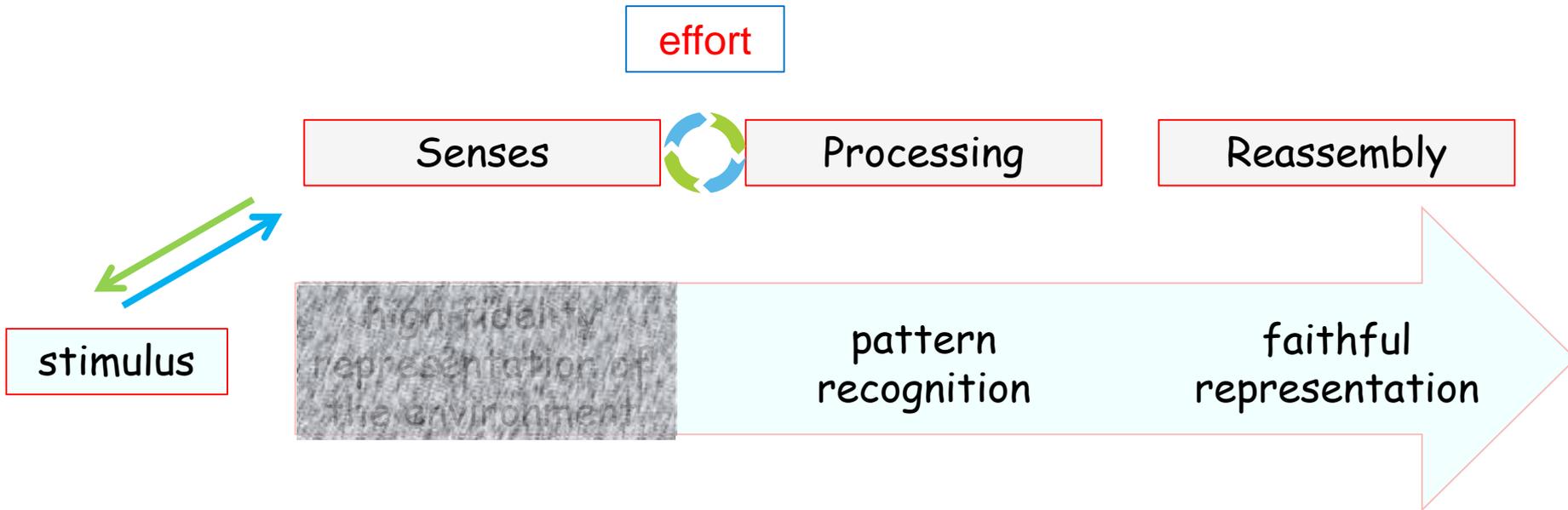
Sensory Assembly



Sensory Assembly - Cochlear Loss

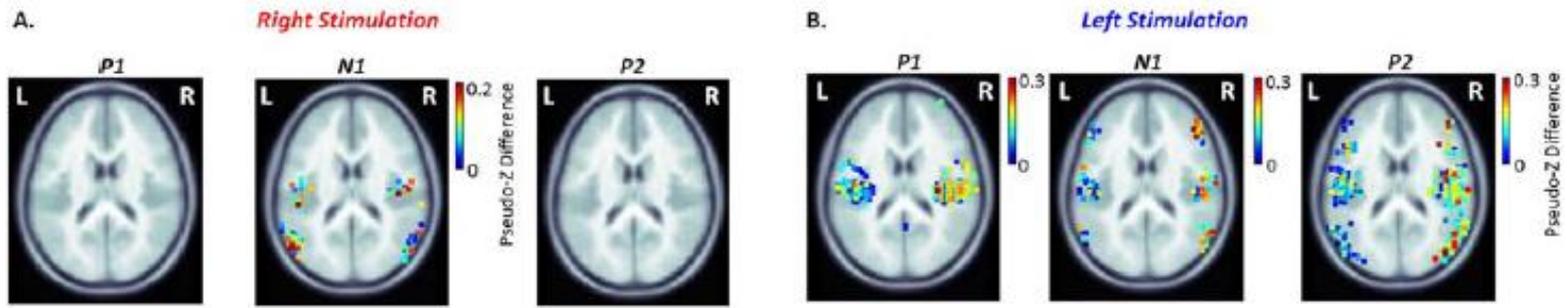


Sensory Assembly - Restored



How Do Humans Listen?

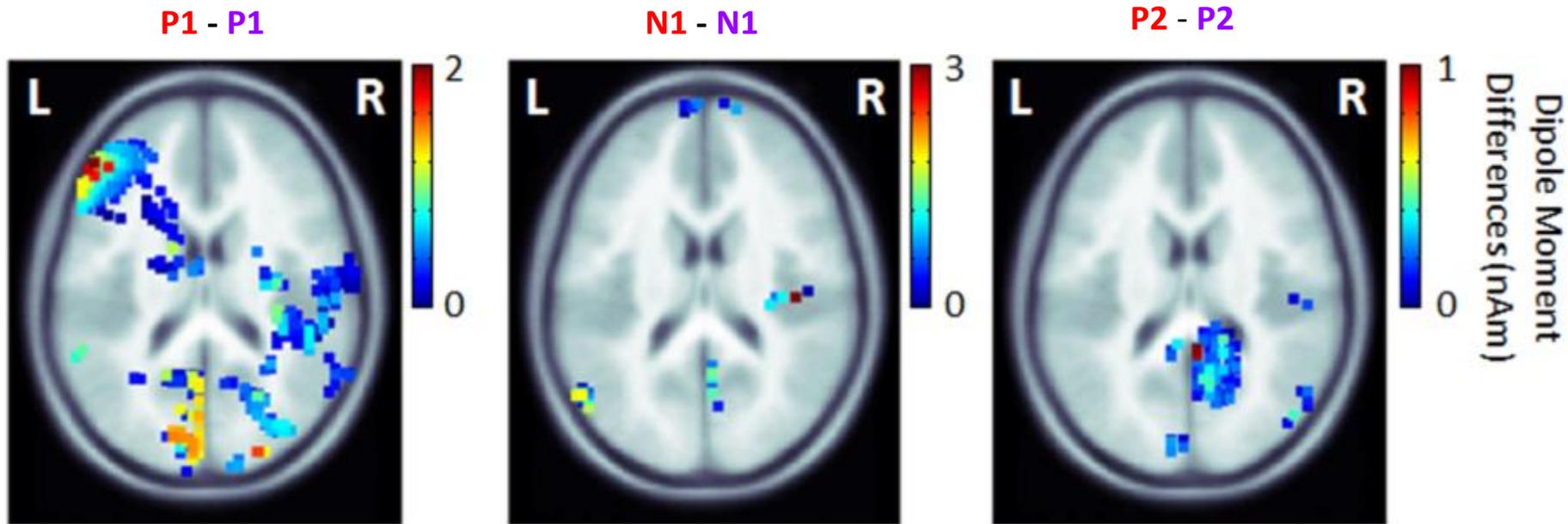
Normal Hearing Adolescents



How Do Humans Listen?

Right Stimulation

CI Experienced – Normal Hearing Dipole Differences

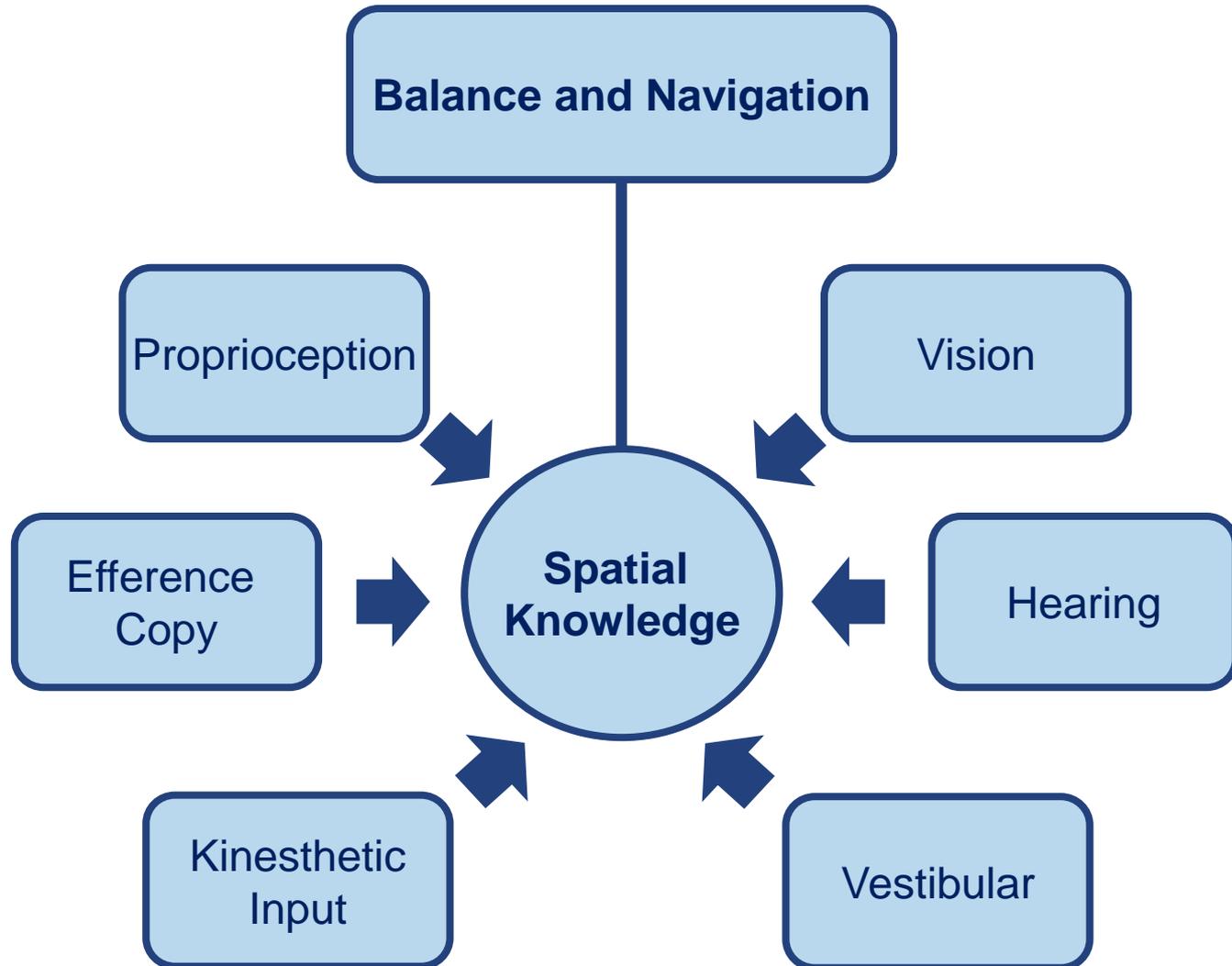


Jiwani et al.,
Submitted

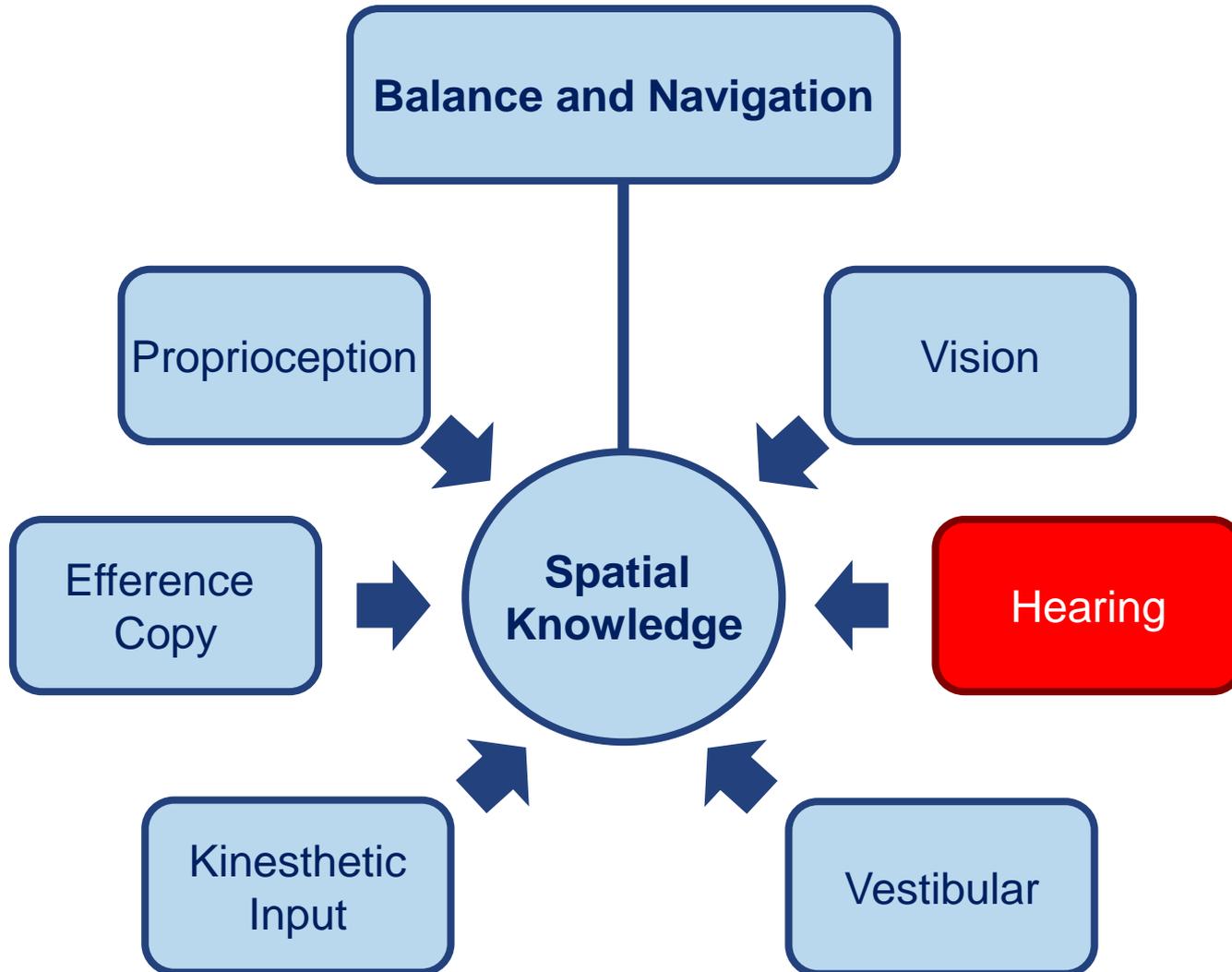
Cognitive Load



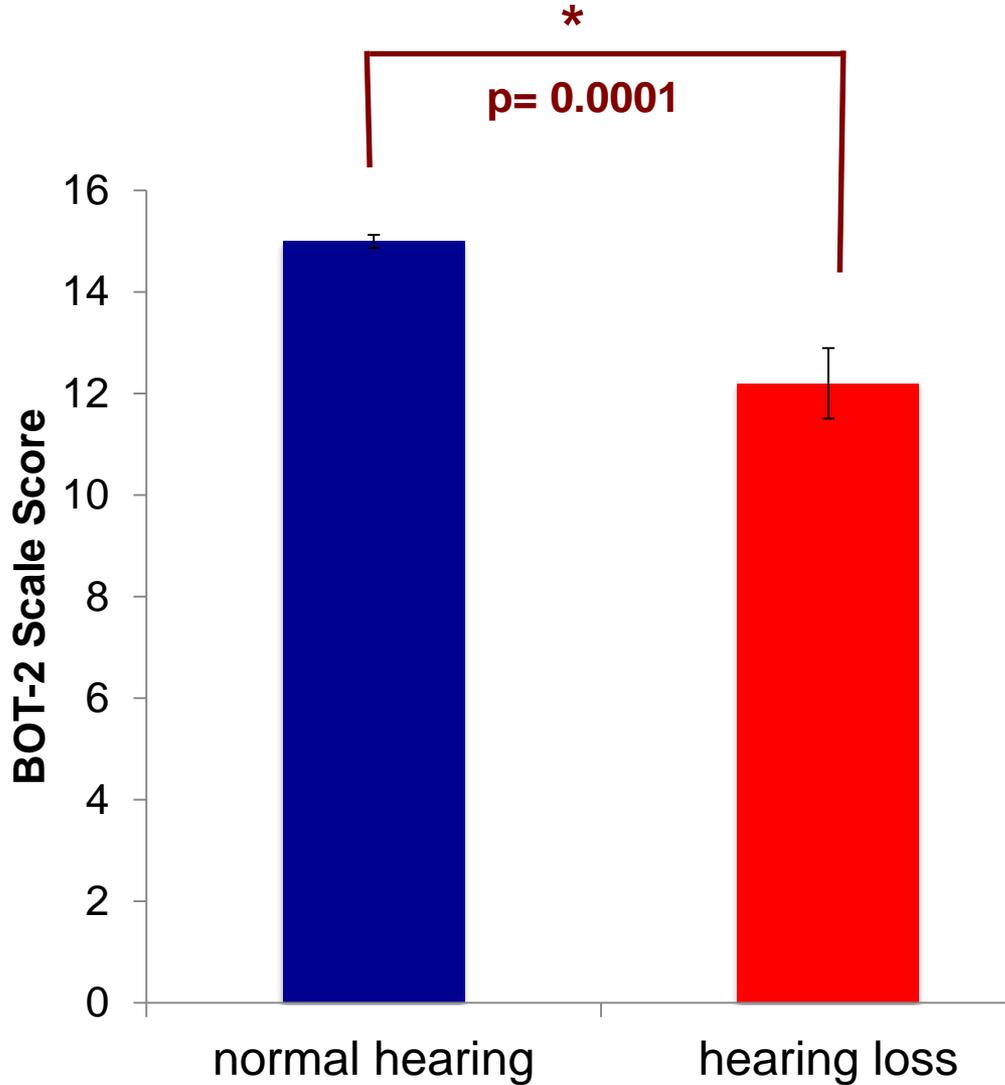
Cognitive Load



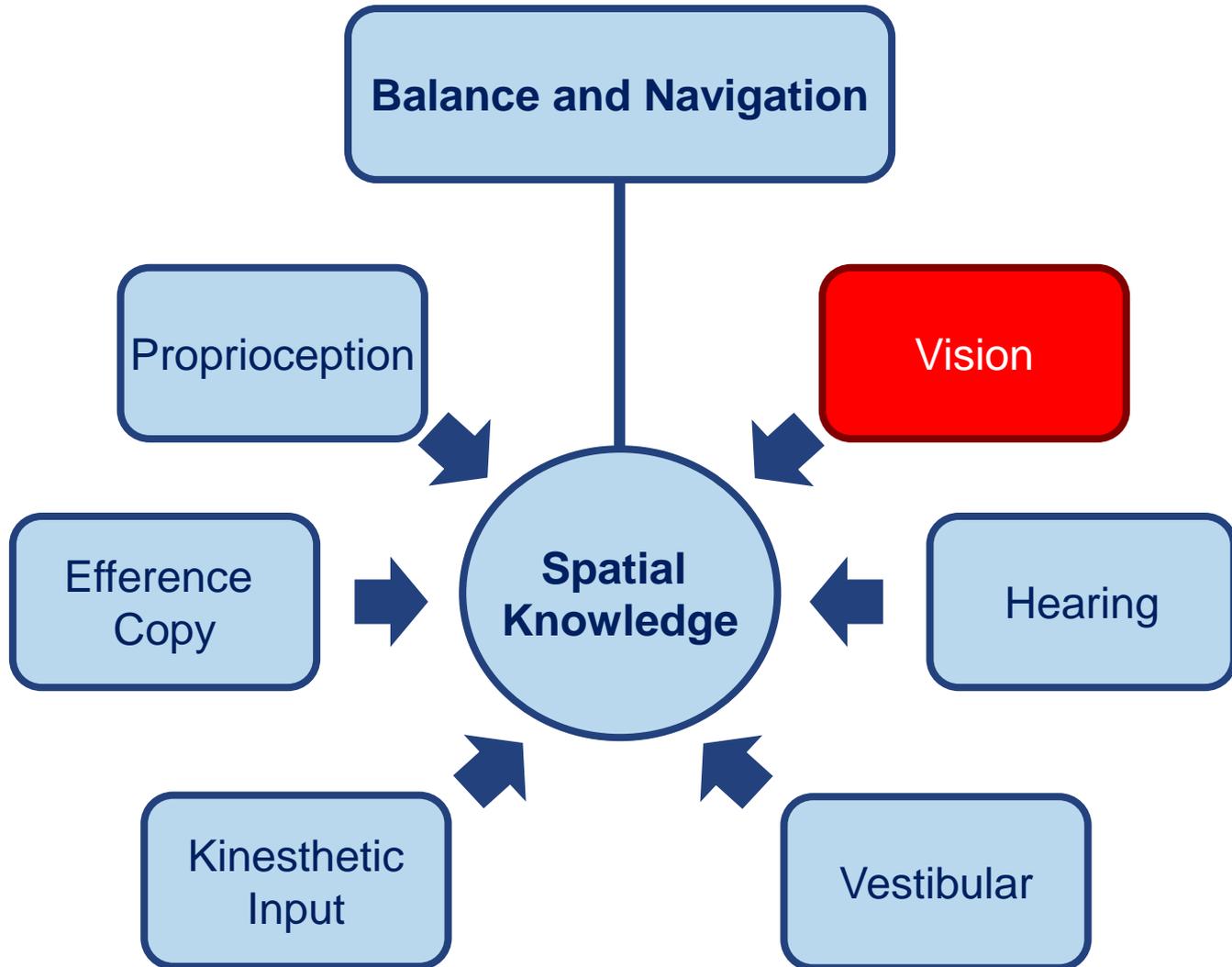
Cognitive Load



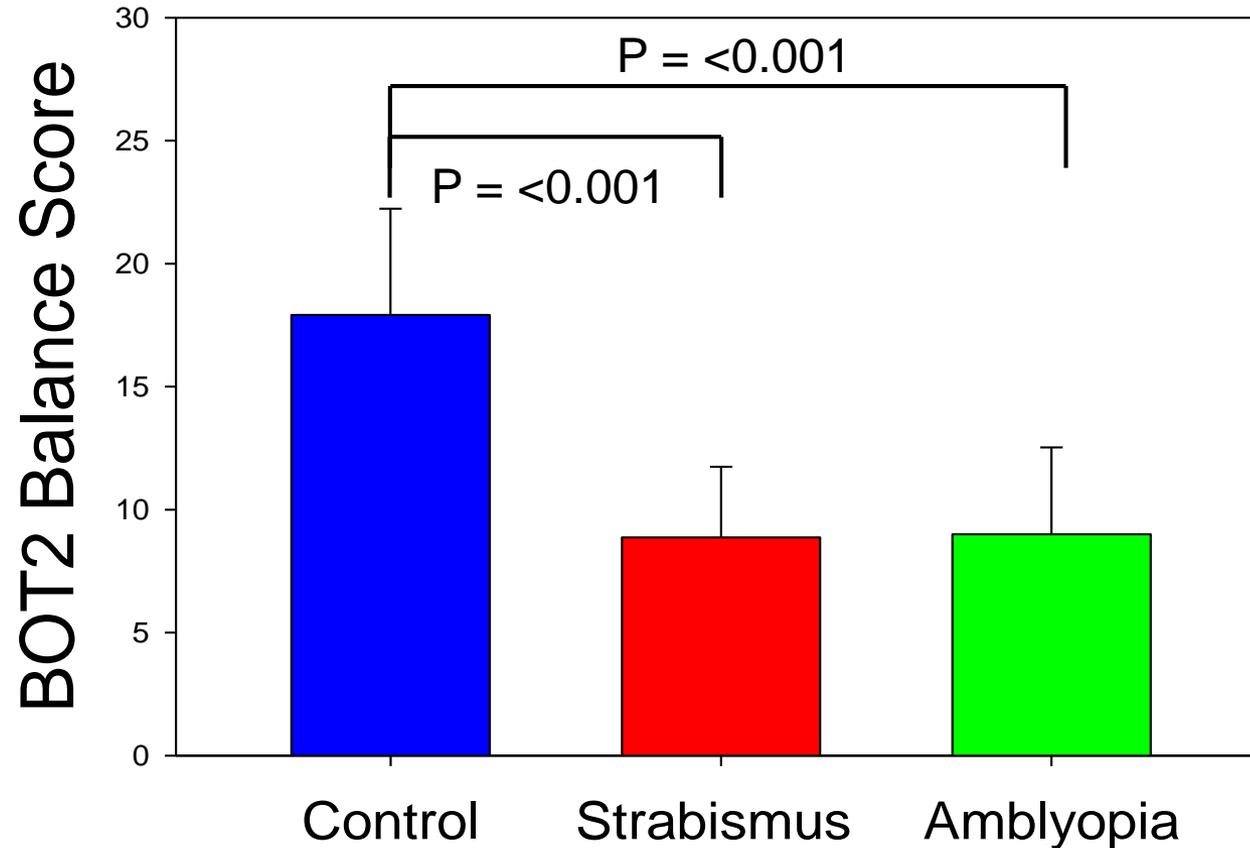
Vestibular Loss Impacts Balance



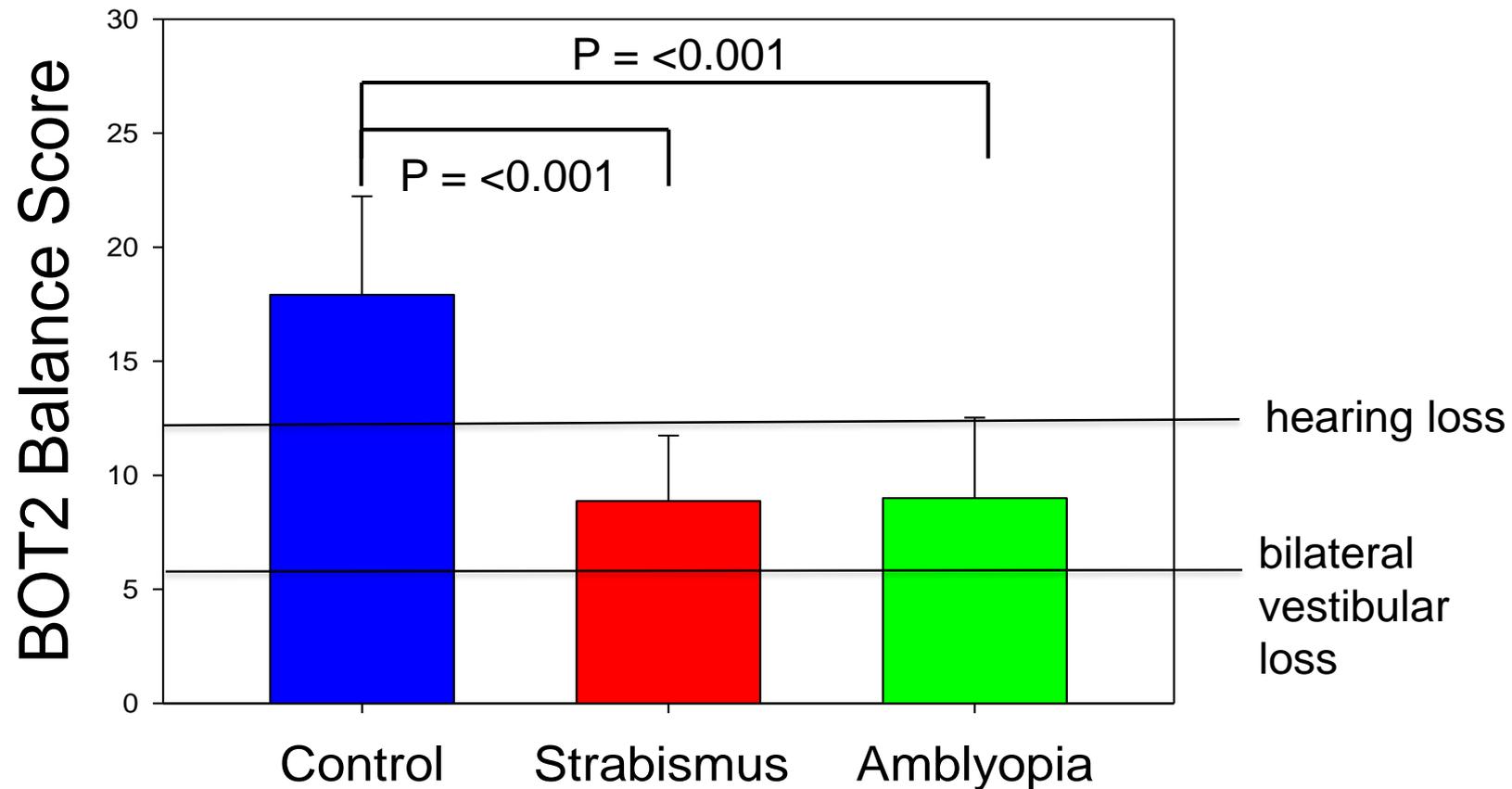
Cognitive Load



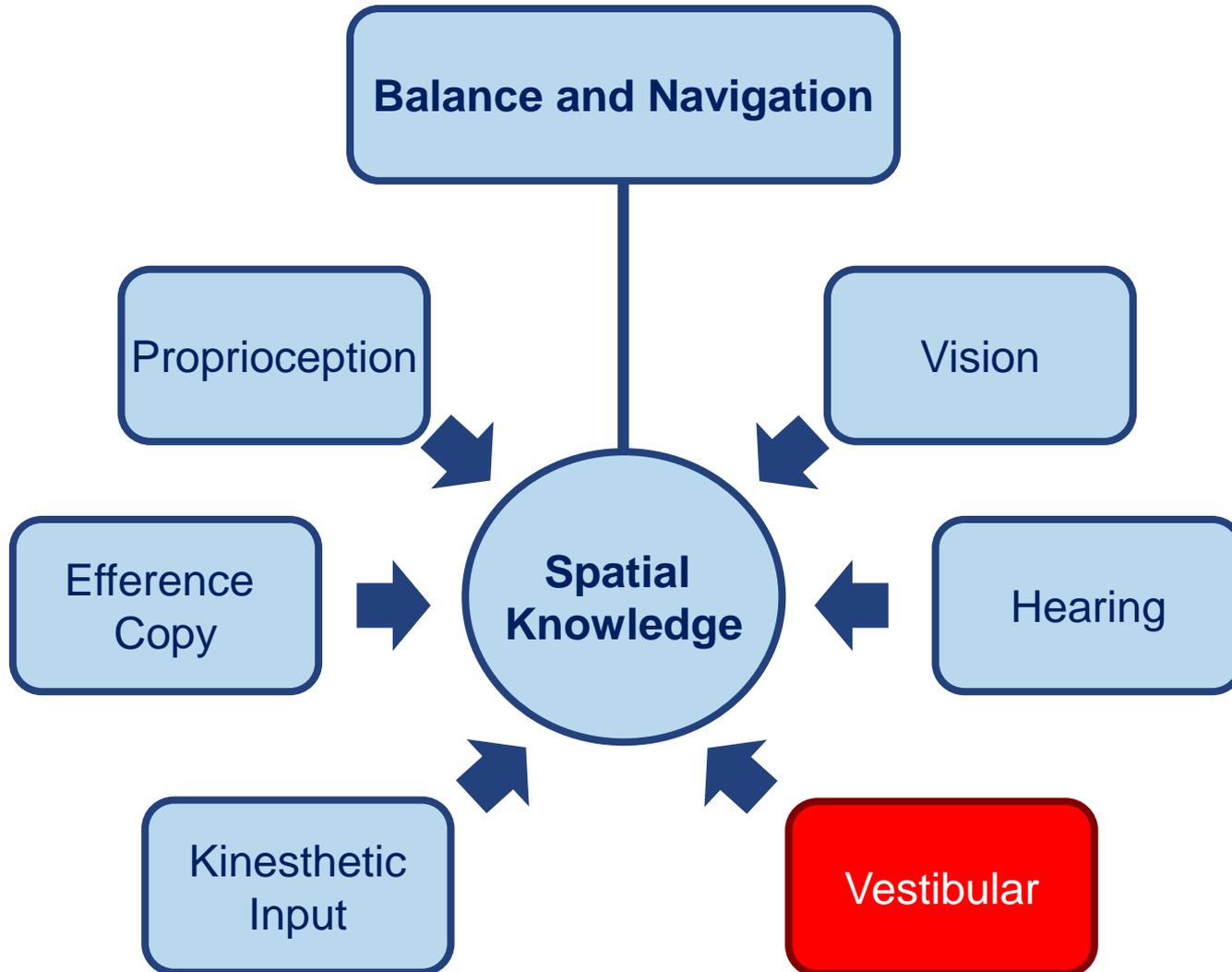
Balance is Impaired in Developmental Visual Anomalies



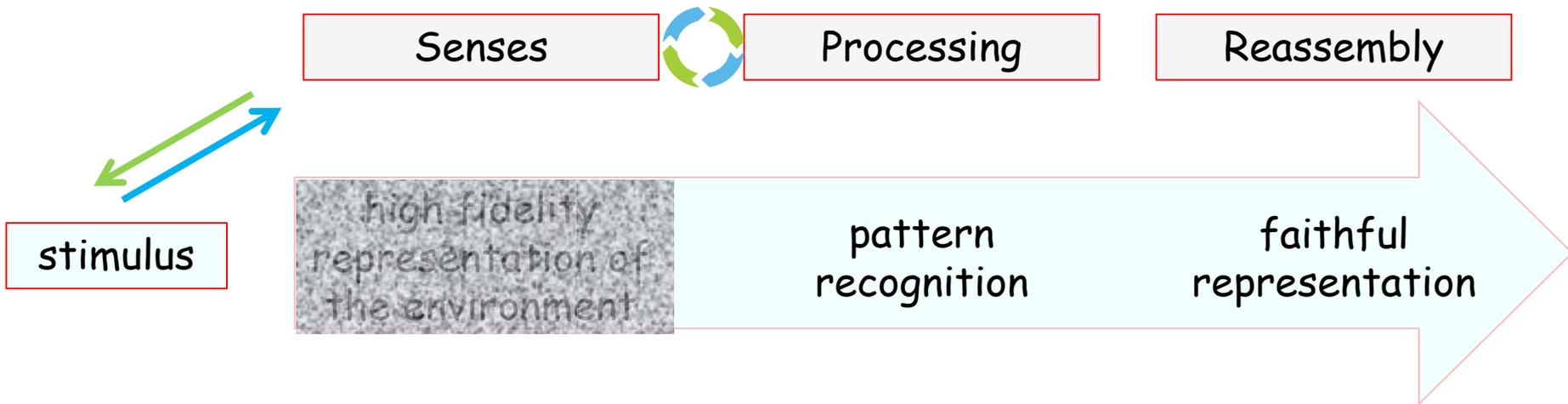
Balance is Impaired in Developmental Visual Anomalies



Cognitive Load

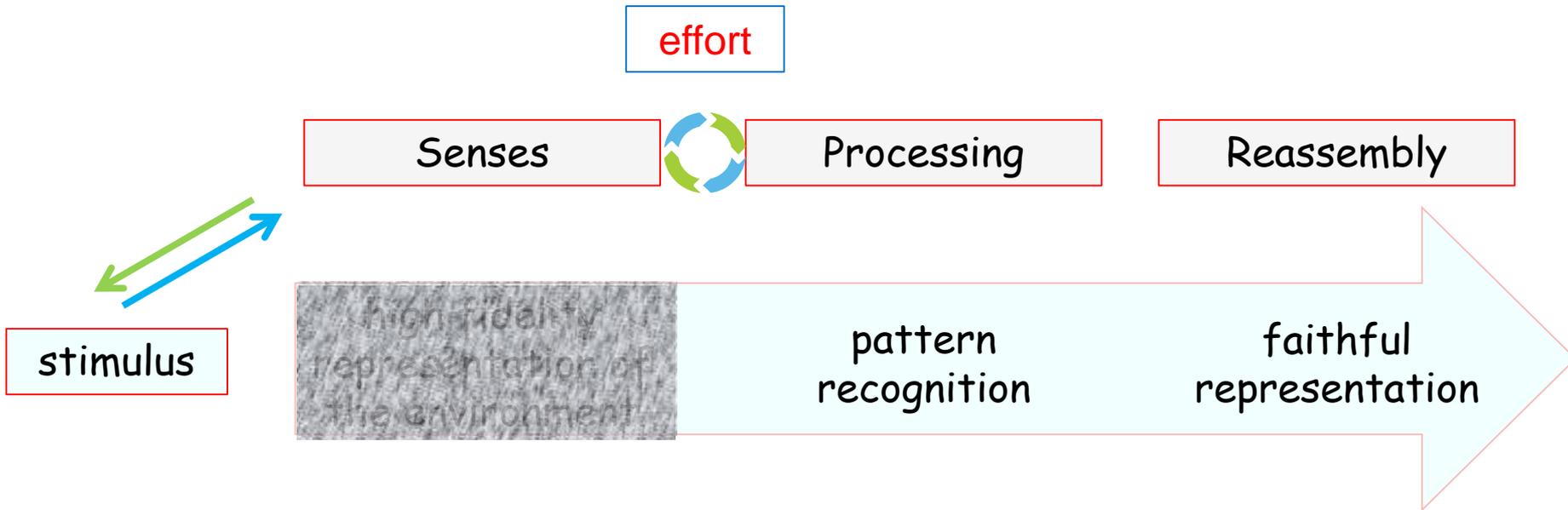


Sensory Assembly - Vestibular Loss



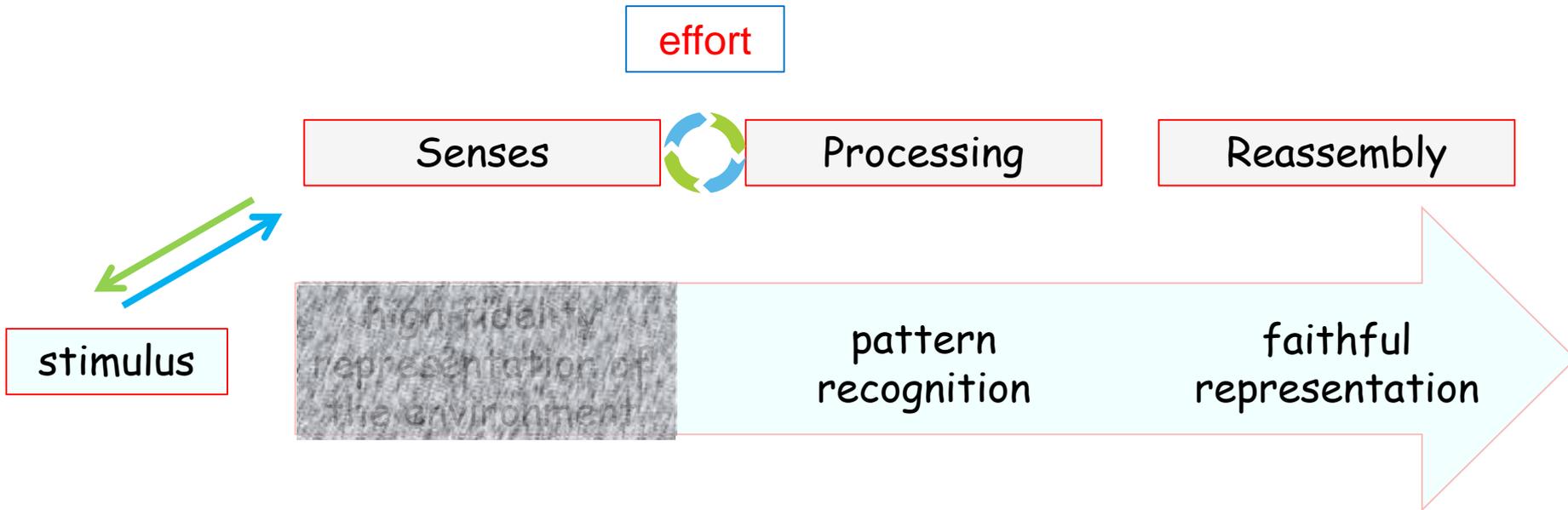


Sensory Assembly – Vestibular Loss

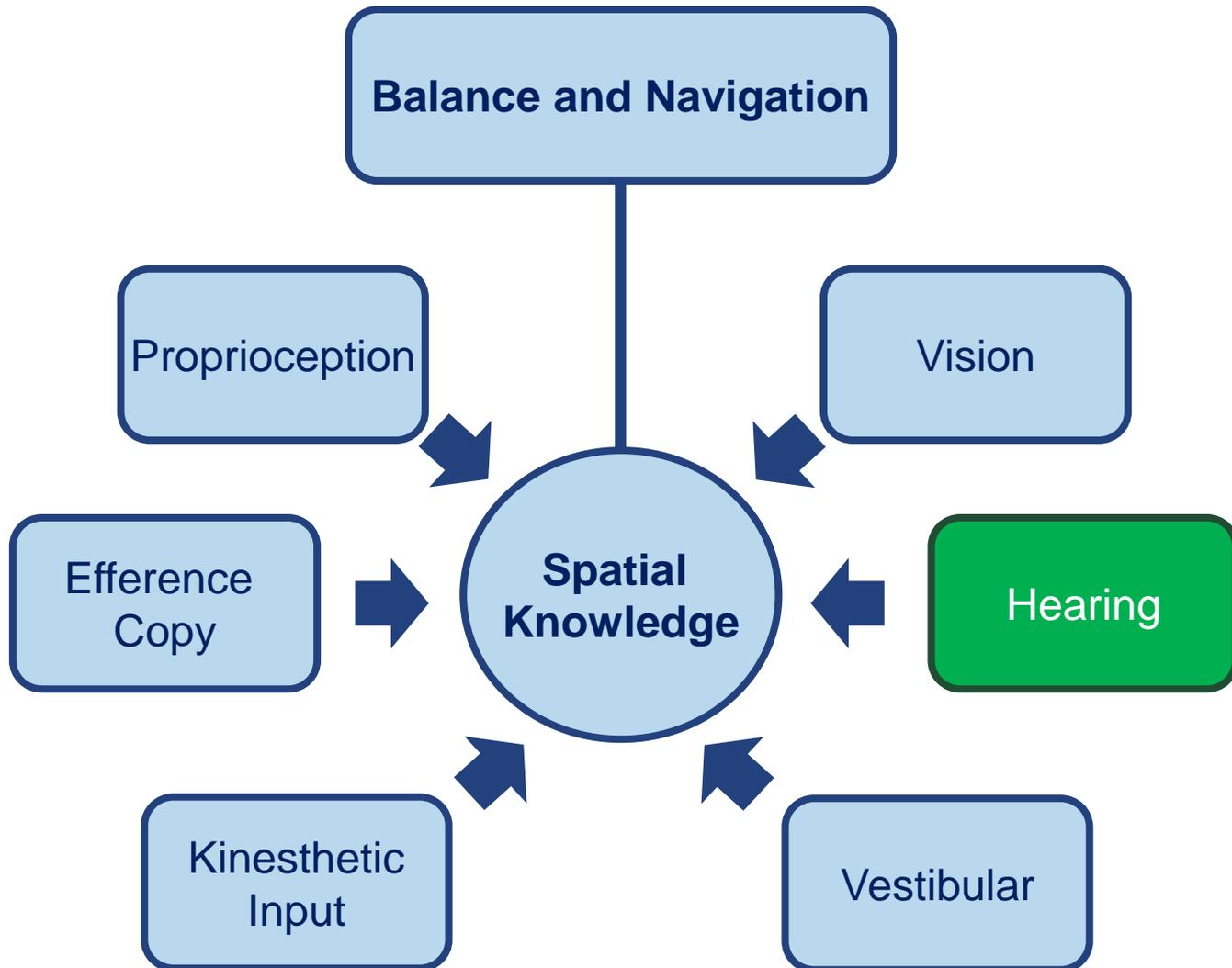




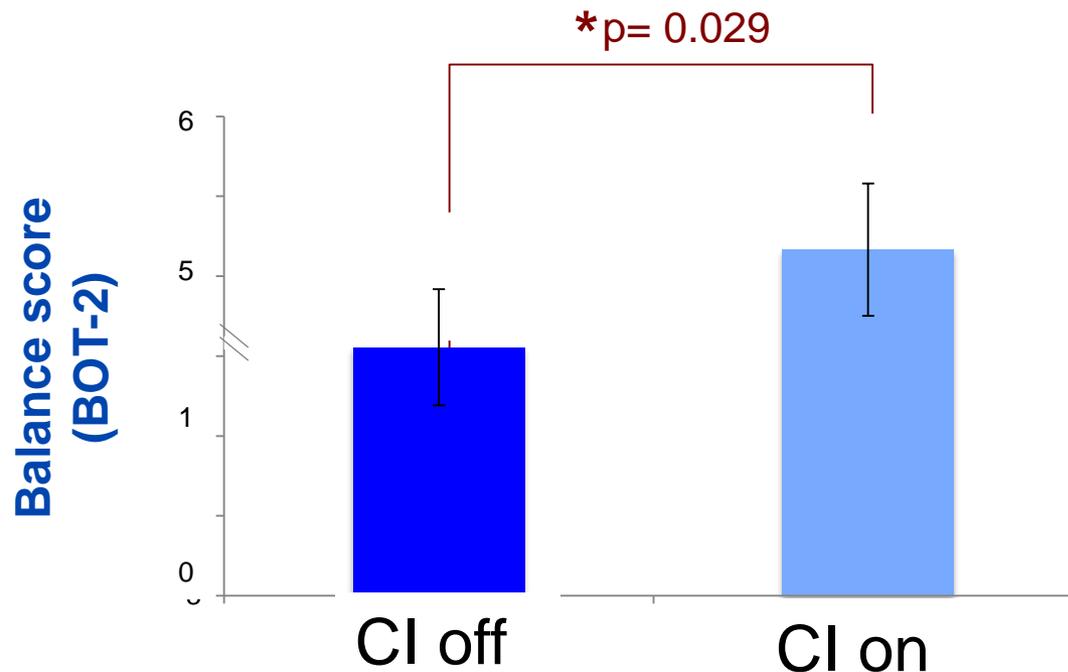
Sensory Assembly – Vestibular Loss



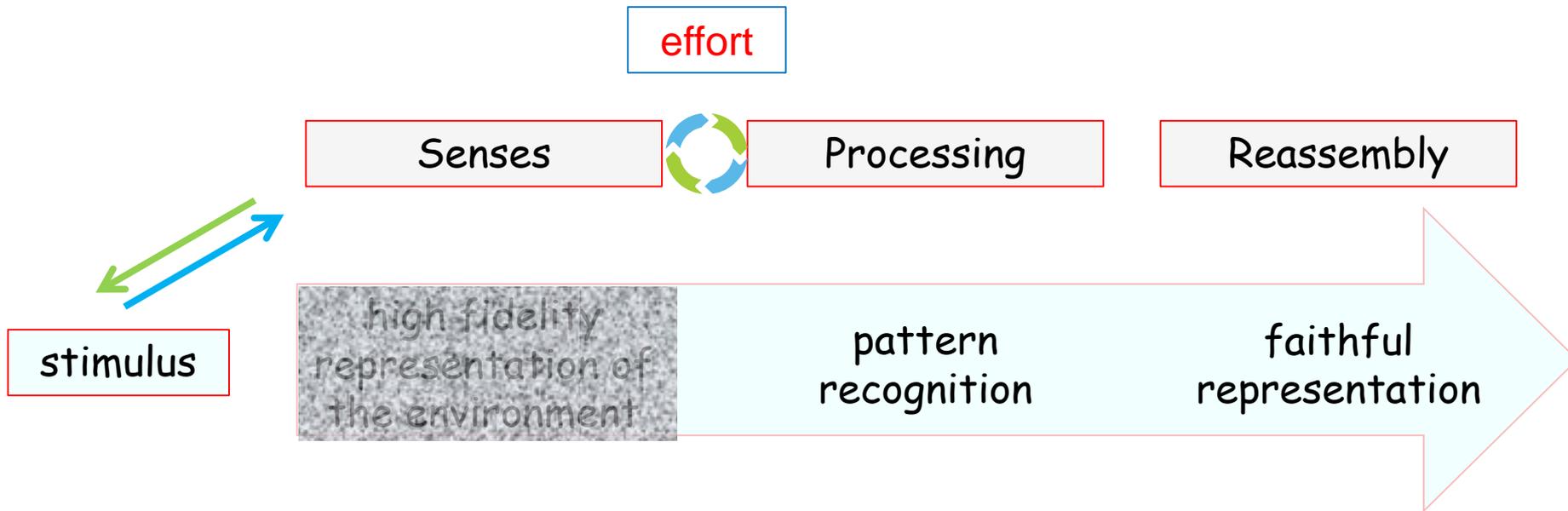
Cognitive Load



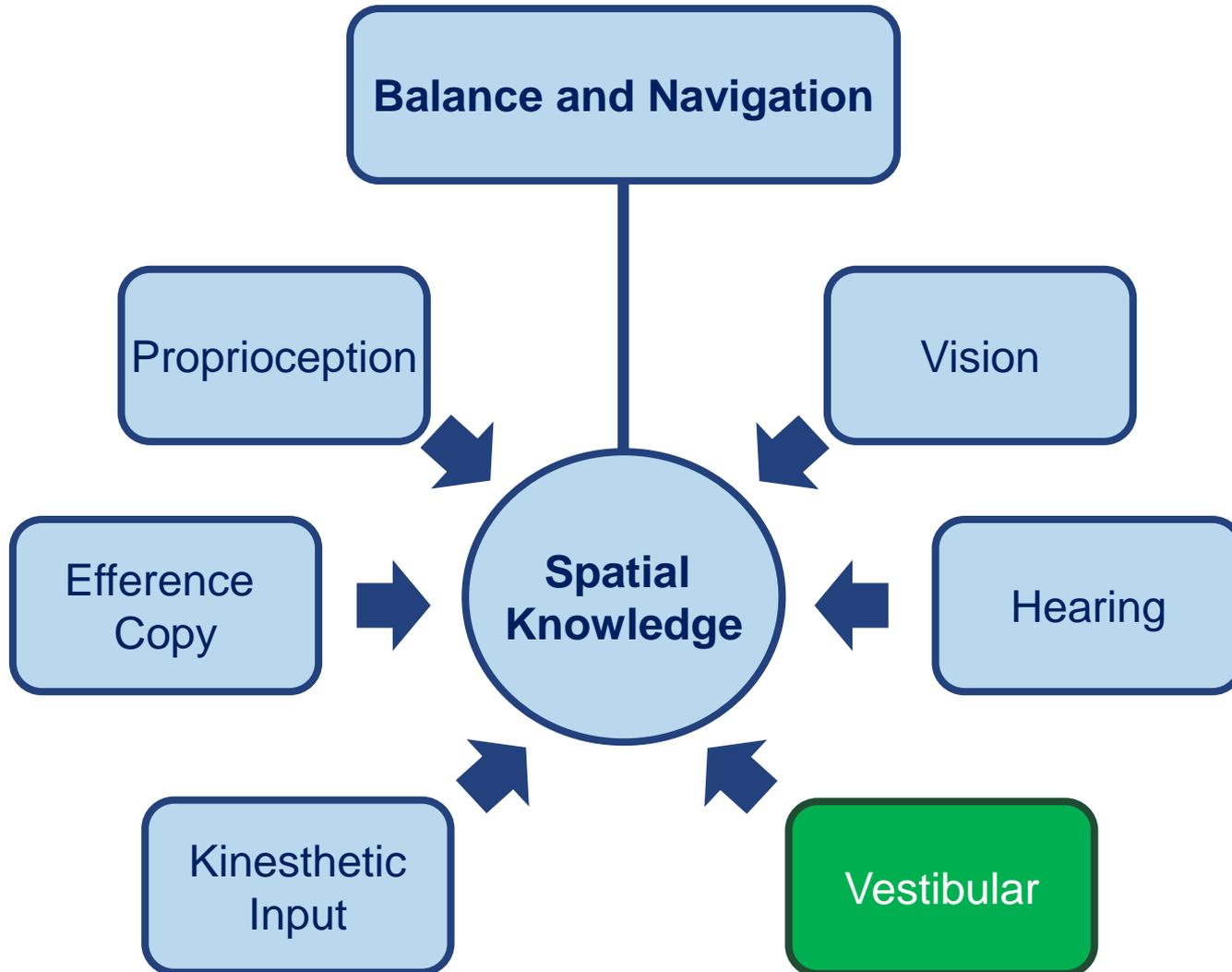
Vestibular Effects of Cochlear Implantation (*positive*)



Sensory Assembly – Restored?

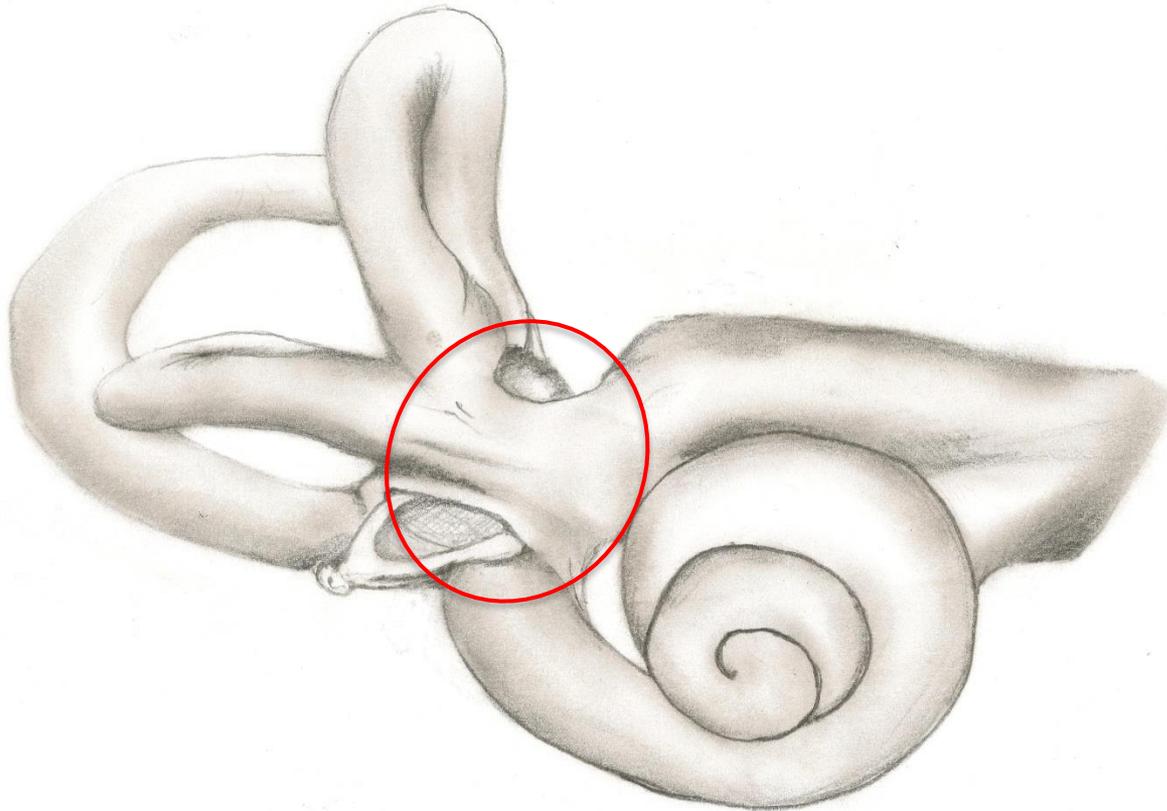


Cognitive Load



Hypothesis

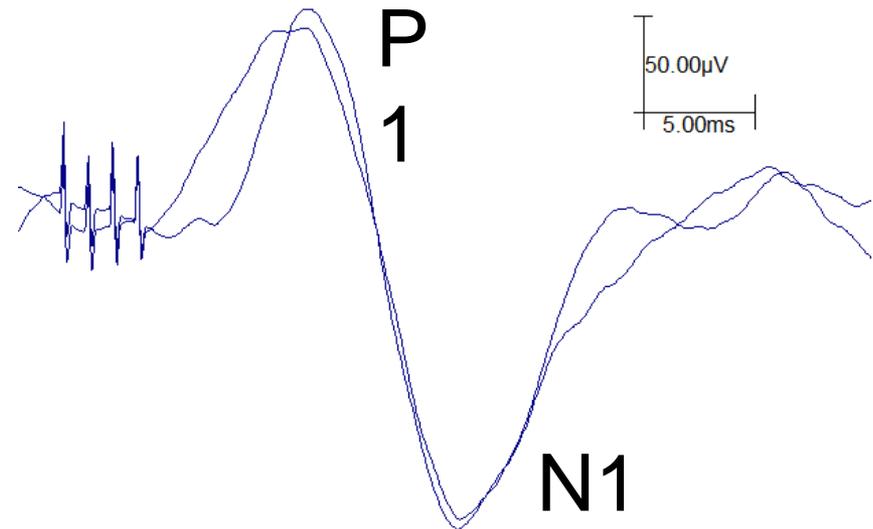
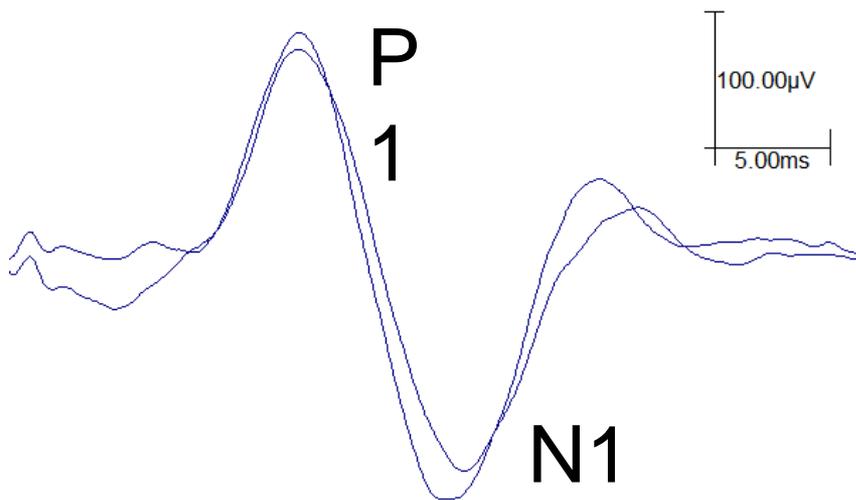
- Electrical current from a cochlear implant can spread to the vestibular system



VEMP Waveforms (same ear)

acoustic

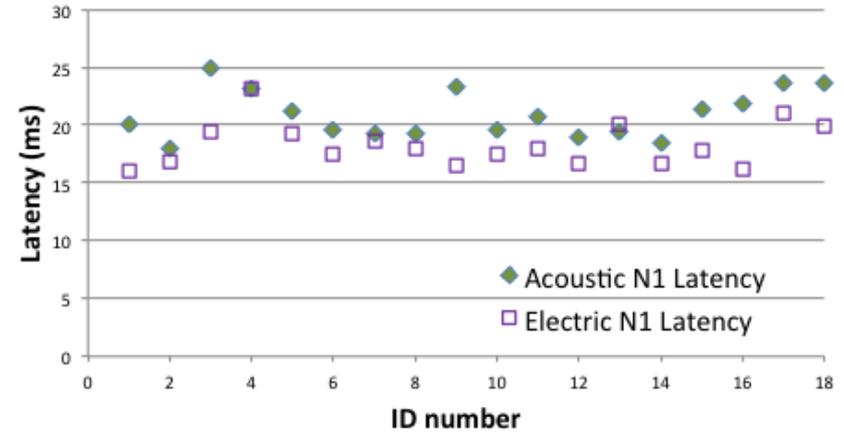
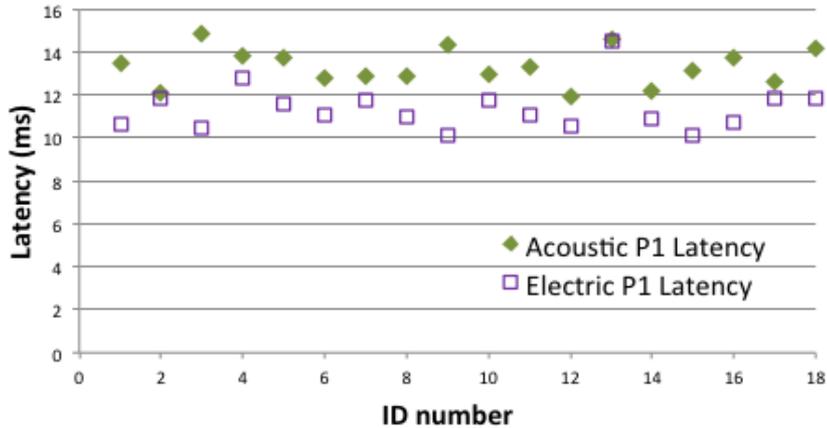
electric



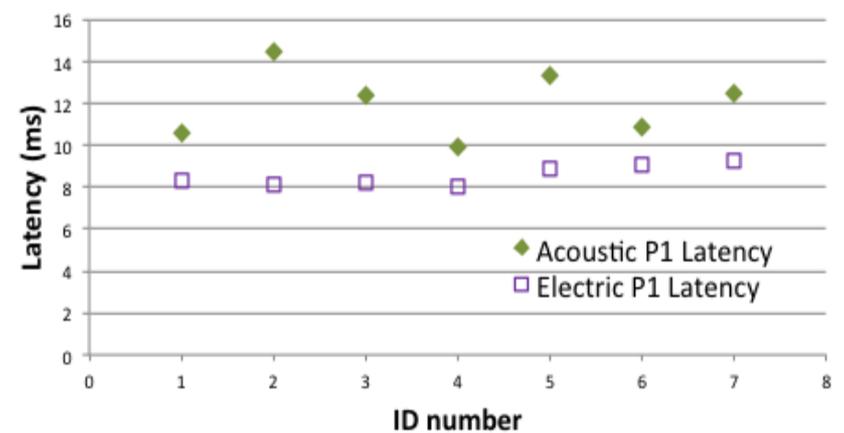
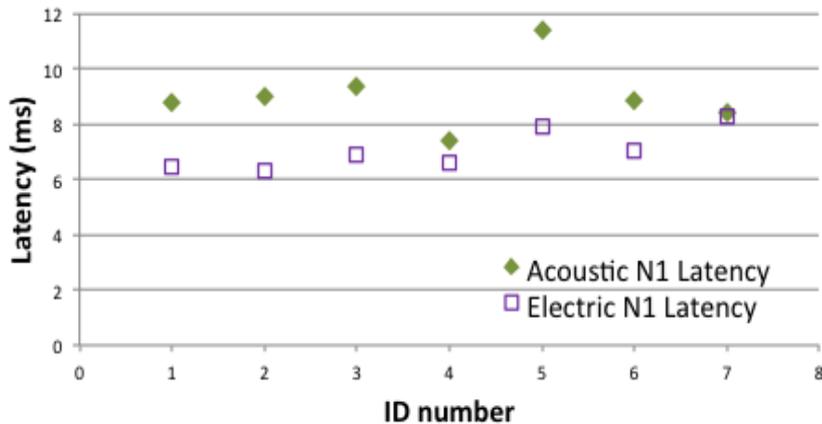
Laryngoscope. 2016

Electrically-Evoked VEMPs are Faster

P1 and N1 peak latencies of cervical VEMPs



N1 and P1 peak latencies of ocular VEMPs



p<0.01, paired t-test

Vestibular Abnormalities Cause Functional Deficits

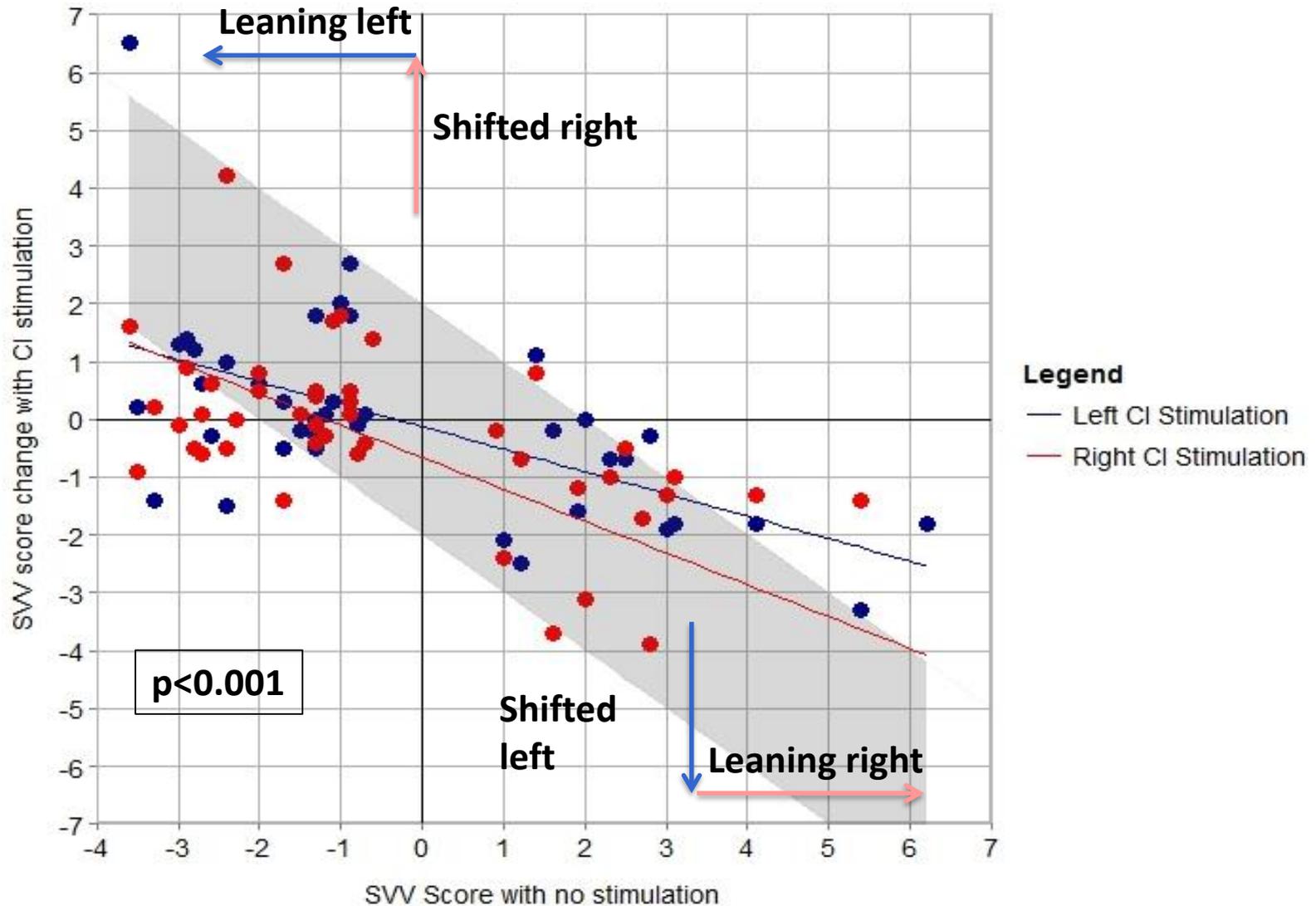
SickKids®

COCHLEAR
IMPLANT
PROGRAM

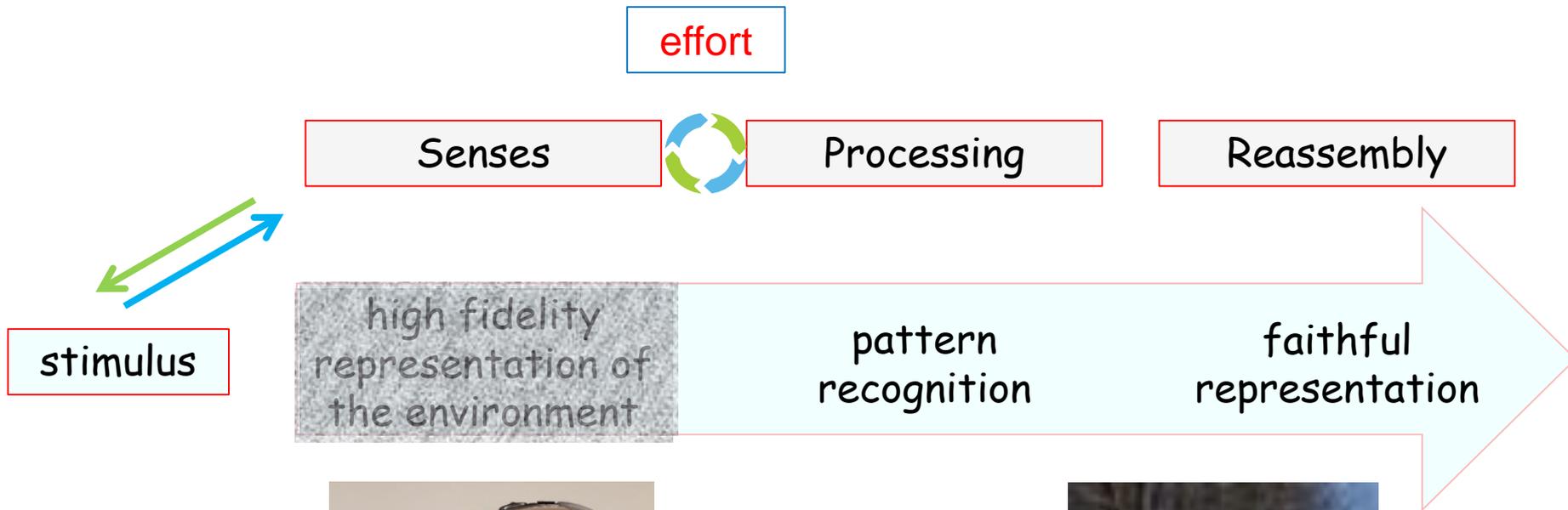


by Stephanie Jewell

Electrical Stimulation Shifts the Perception of Vertical Back to Center



Sensory Assembly – Restored?



TransCochlear Stabilization of Balance

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



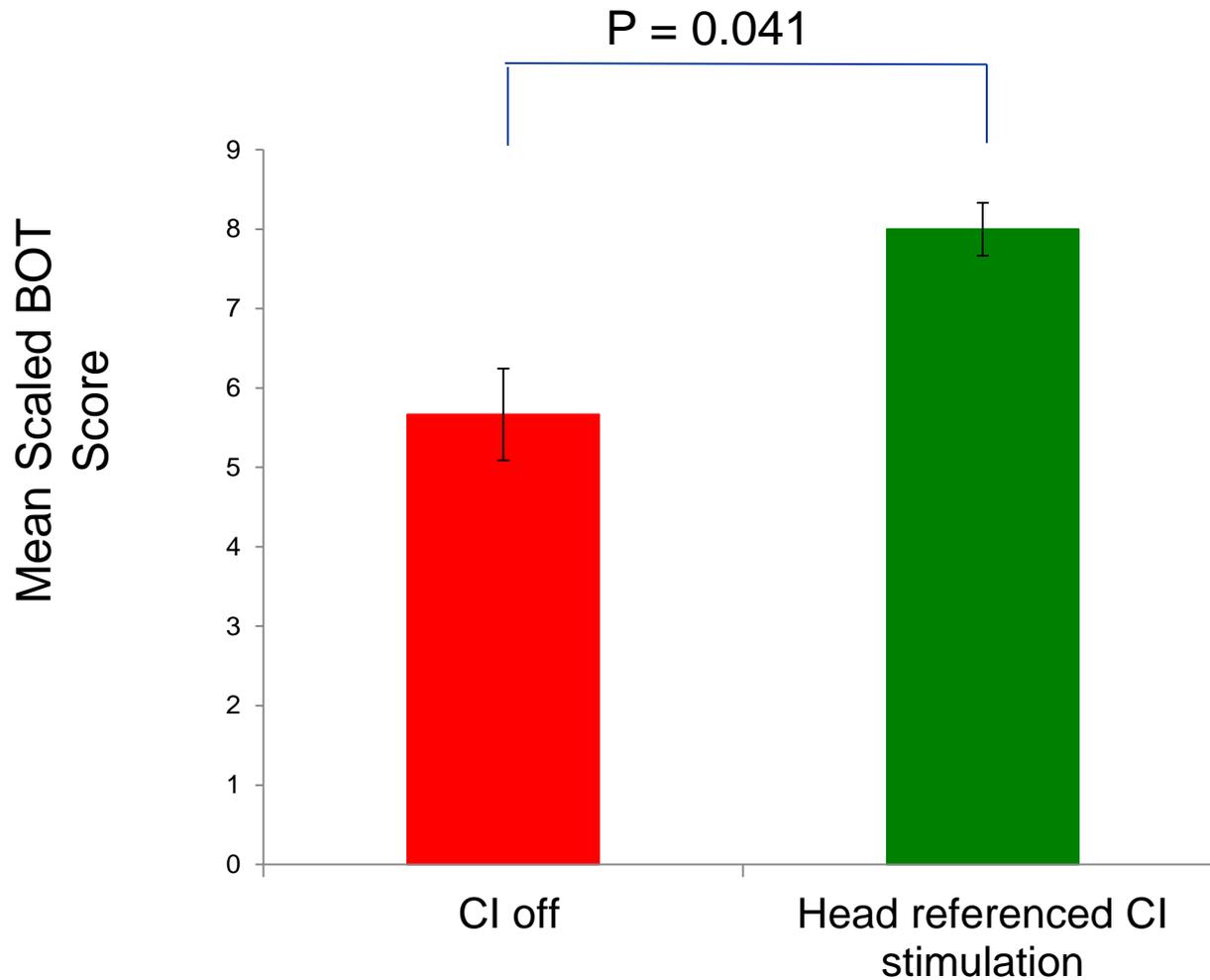
OFF



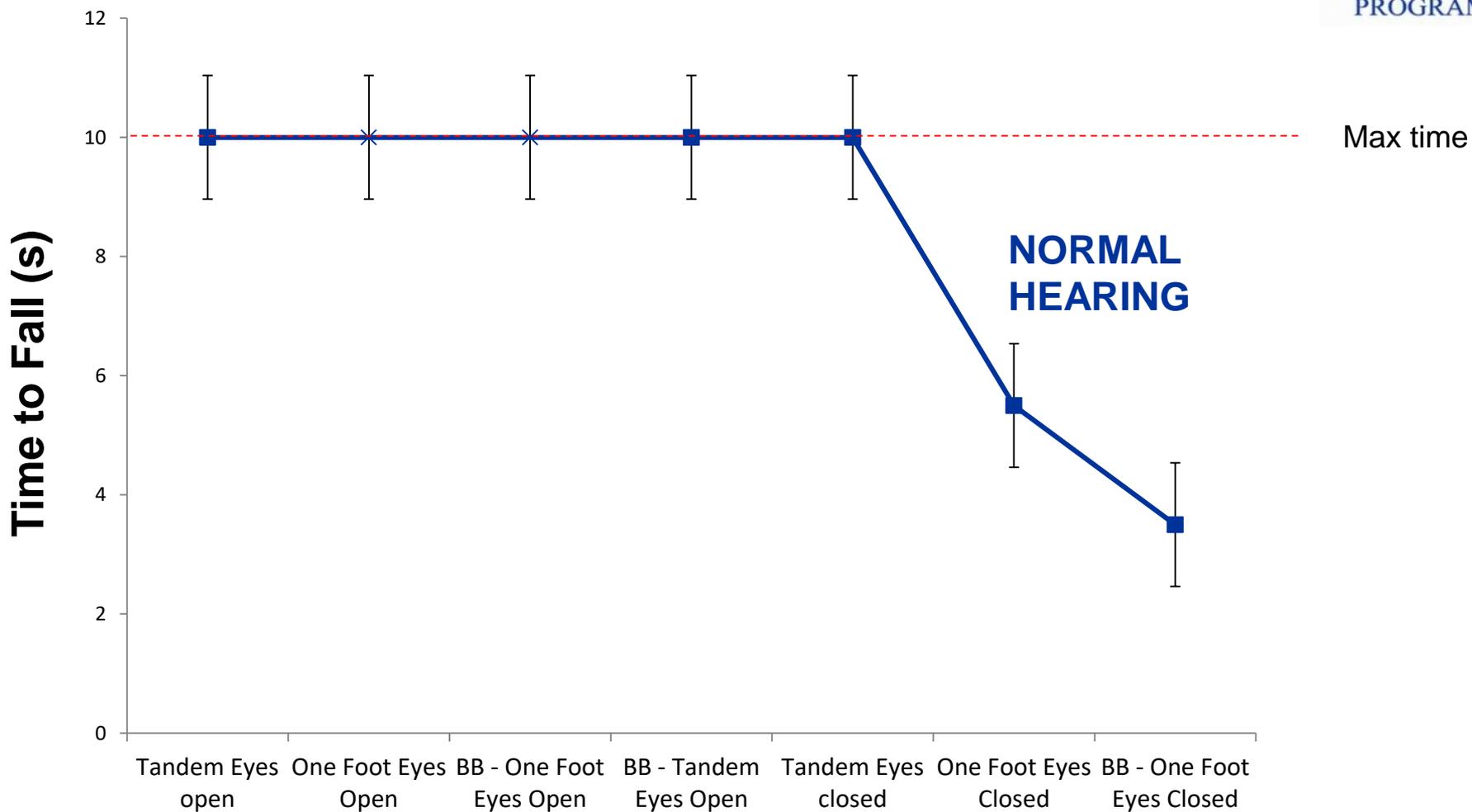
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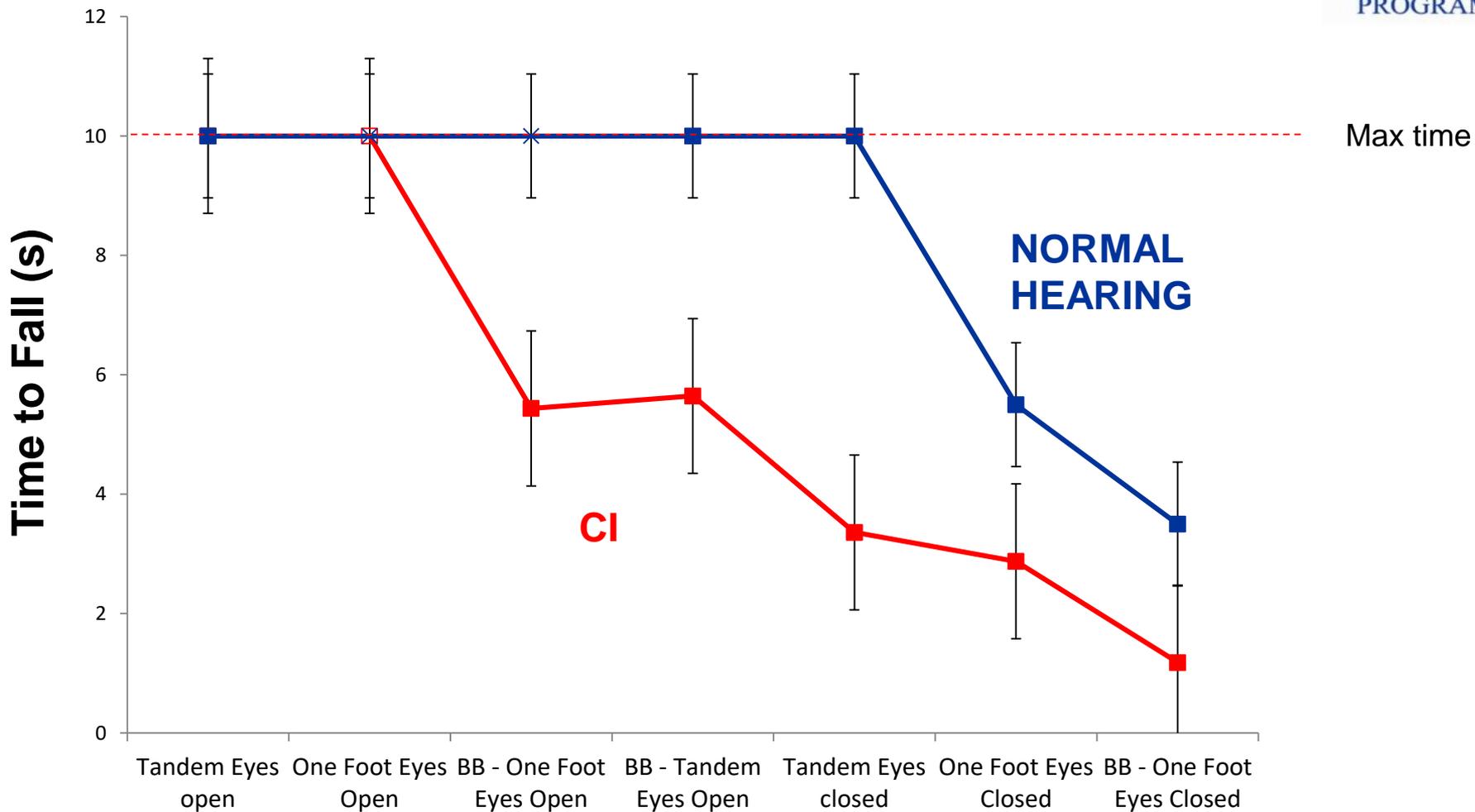
Hearing and Implants Help Balance



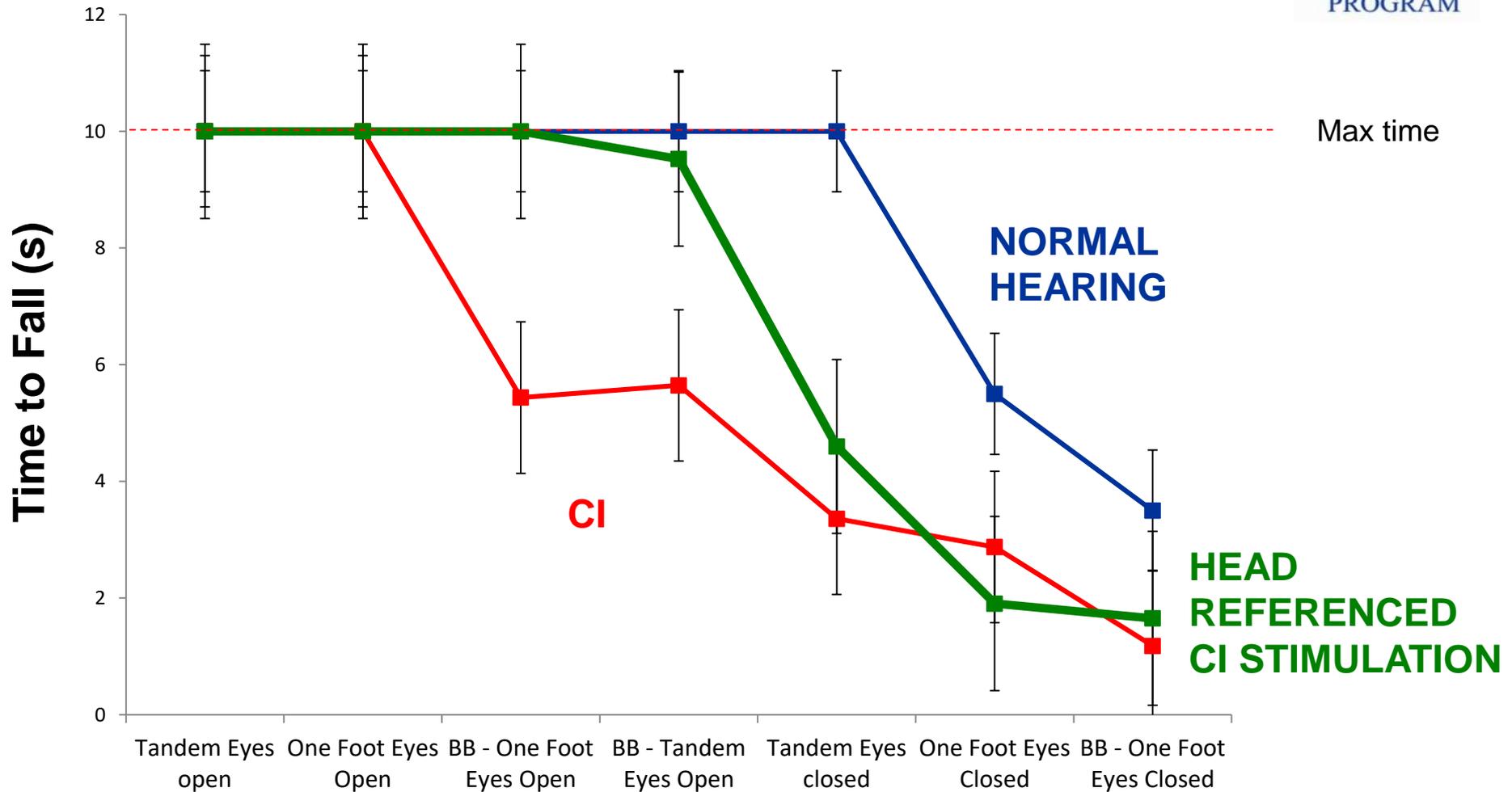
Time to Fall Approaches Normal

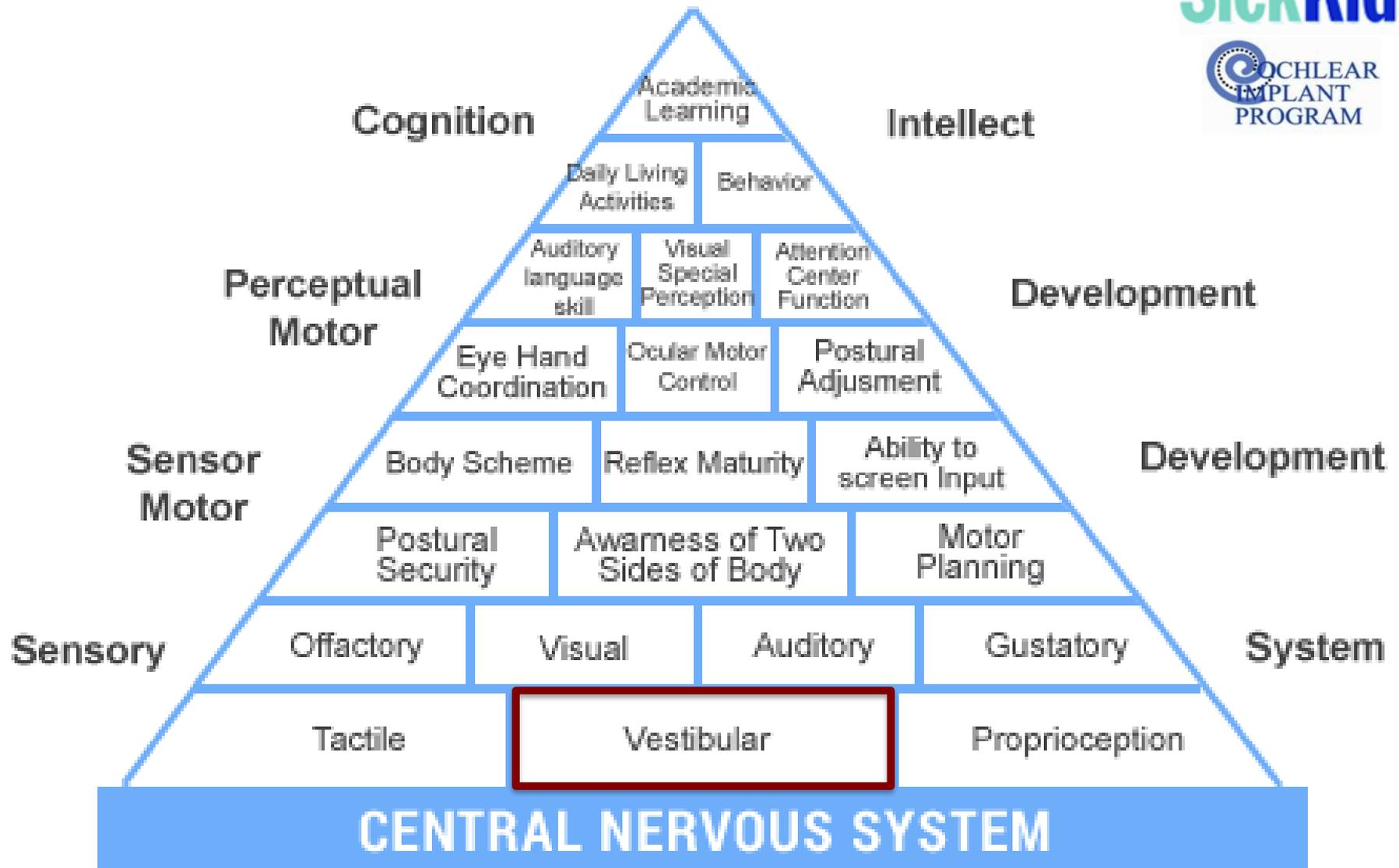


Time to Fall Approaches Normal



Time to Fall Approaches Normal





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

Summary

- vestibular & balance dysfunction
 - common
 - matters
- screening is feasible
- performance impaired by labyrinthine
- implants can stimulate an impaired vestibular system

What Drives Development in Children?

