Plasticity of the auditory cortex: Reorganization following hearing loss and intervention

CHRISTINE BRENNAN, PhD CCC-SLP

Abstract

A consequence of pediatric hearing loss is reorganization of the auditory cortex. These changes can result in delayed and/or abnormal maturation and functioning of the auditory cortex. This presentation will review previous and more current investigations of cross-modal re-organization that occurs following auditory deprivation. Specifically, we will discuss how this re-organization results in the recruitment of cortical resources of the deprived auditory modality by other, more intact sensory modalities (e.g. visual or somatosensory systems). Abnormalities in cortical maturation and cross-modal plasticity have been proposed as a source of variability underlying speech perception outcomes after cochlear implantation (CI) in pre-lingual and post-lingual bilateral deafness. More current investigations suggest that similar outcomes may be observed in single-sided deafness as well. This presentation will review the re-organization that occurs in both bilateral and single-sided deafness and the implications for early identification and intervention.

Recommended Readings


Learner outcomes

1. Understand and explain investigations that show cross-modal re-organization following auditory deprivation.
2. Explain how the related abnormalities in cortical maturation following auditory deprivation may impact underlying speech perception outcomes.
3. Discuss the implications of cortical re-organization related to early identification and intervention.

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Maturation of the Central Auditory Pathways in Children
The cortical auditory evoked potential (CAEP) response

During development, both intrinsic and extrinsic factors have a profound influence on cortical maturation of the auditory system.

The cortical auditory evoked potential (CAEP) response can be used as a biomarker to assess the development of the central auditory pathways.

In infants and young children, the CAEP response is dominated by a single obligatory, positive peak known as the P1 component.

In early childhood, the P1 CAEP response occurs around 200 and 300 milliseconds.

The source of the P1 component is the primary auditory cortex and the thalamus.
The cortical auditory evoked potential (CAEP) response

As a child's auditory system becomes refined by intrinsic and extrinsic inputs, the latency of the P1 CAEP response decreases rapidly early in childhood. Then gradually into adulthood, eventually reaching a peak latency of 50 to 70 milliseconds.

Synaptogenesis & Pruning

During the early developmental period, there is a period of synaptogenesis occurring in auditory cortex.

- Synaptogenesis — rapid increase in connections between neurons
- Pruning — reduction of unutilized connections

Peaks in Synaptogenesis
Synaptogenesis & Pruning of Auditory Cortex

After about 3.5-4 yrs...
Extrinsically driven factors
  - Including sensory stimulation and/or deprivation
Cause refinement and pruning of central auditory pathways

(Kral & Sharma, 2012; Kral et al., 2005)

Pre-adolescent changes in the CAEP

Later CAEP components, known as the N1 and P2, emerge

Pre-adolescent changes in the CAEP

The N1 and P2 components reflect higher order auditory processing with sources in primary and secondary auditory cortex

(Eggermont & Ponton, 2003; Kral & Sharma, 2012; Kral et al., 2003; Sharma, Dorman, & Spahr, 2002a; Sharma et al., 2007)

In typically developing, normal hearing children, the N1 component emerges as early as 3 yrs and can be reliably observed by age 7 yrs

(Gilley et al., 2005)
Sensitive period for cortical auditory development

Synaptogenesis of auditory cortex coincides with a critical or sensitive period for cortical auditory development in children. During this time, the central auditory pathways are maximally plastic.

(Sharma, Dorman, & Kral, 2005; Sharma et al., 2002; Sharma, Dorman, & Spahr, 2002a; Sharma & Dorman, 2006; Sharma, et al., 2005)

Sensitive period for cortical auditory development

During this sensitive period...

Extrinsic sensory stimulation has the ability to profoundly shape the development and organization of the auditory cortex.

(Sharma, Dorman, & Kral, 2005; Sharma et al., 2002; Sharma, Dorman, & Spahr, 2002a; Sharma & Dorman, 2006; Sharma, et al., 2005)

Increased vulnerability during the sensitive period
Single-Sided Deafness

Single-sided deafness
Single-sided deafness (SSD)
Normal hearing in one ear and severe-profound sensorineural hearing loss in the contralateral ear

Single-sided deafness - Prevalence
The prevalence of SSD is estimated to affect 3-6% of the population (Ross et al., 2010)
Incidence is higher among children and teenagers (2-5 per 1,000) (Hassepass et al., 2013; Tharpe & Sladen, 2008)
Single-sided deafness - Children
Common causes of pediatric SSD include:
- viral complications, meningitis, head trauma, enlarged vestibular aqueduct syndrome (EVA), and genetic disorders (Bess & Tharpe, 1984)
Congenital SSD occurs in about 1 in 3,700 newborns (Mehl & Thomson, 2002)

Single-sided deafness – Adults
Most common causes include:
- Meniere’s disease, viral infection, autoimmune disorders, acoustic neuroma or vestibular schwannoma, trauma and temporal bone fractures, unilateral nose exposure, and ototoxic drug exposure (Giardina, Formeister, & Adunka, 2014)
Sudden sensorineural hearing loss (SSNHL) accounts for the greatest proportion of patients with SSD

Problems w/ SSD
Adults & children w/ SSD demonstrate difficulties with:
- Sound localization
- Ability understanding speech in everyday listening situations (Cabral et al., 2016; Hassepass et al., 2013; Lieu et al., 2010; Wie et al., 2010)
- Social-emotional well-being
- Health-related quality of life (Kitterick, Lucas, & Smith, 2015; Wie et al., 2010)
Problems w/ SSD

Children with SSD are also at risk for:

- Behavioral problems
- Delays in
- Speech-language development
- Social-emotional development
- Educational progress

(Bess & Tharpe, 1984; Fischer & Lieu, 2014; Lieu et al., 2013; Martínez-Cruz, Poblano, & Conde-Reyes, 2016)

Current Tx options for SSD

Contralateral routing of signal (CROS) hearing aids

- Re-routes sounds arriving at the deaf ear to the good ear

Current Tx options for SSD

Bone conduction hearing devices (BCHD)

- Transmits sound arriving to the deaf side to the normal hearing cochlea via bone conduction
Current Tx options for SSD

Adults and children with SSD have not traditionally been considered cochlear implant (CI) candidates. This is due to excellent speech perception abilities in their normal hearing ear. Also, CIs are not currently approved by the Federal Drug Administration (FDA) in the United States for SSD.

But, there is evidence that this deprivation leads to reorganization and recruitment of the auditory pathway by other sensory modalities. So... maybe CIs should be considered for SSD.

Cochlear Implants (CI)

Consists of an external speech processor & a surgically implanted internal electrode array:
- Receives and transmits incoming sound to internal electrode array
- Provides direct stimulation to the auditory nerve
Cochlear Implants (CI)

Unlike other currently available rehabilitation options, CIs are...
- The only available intervention method that provides direct stimulation of the auditory pathways arising from the SSD ear
- The only device with potential to achieve true binaural hearing in SSD patients

SSD & CI

Several recent studies have demonstrated the benefits of CI in adult or pediatric SSD patients
- Including subjective reduction in tinnitus, measureable benefits hearing in noise, and improvements in sound localization

(Årvidt et al., 2011, 2015; Cabral et al., 2016; Cadieux et al., 2013; Dorman et al., 2015; Finet et al., 2012; Friedmann et al., 2016; Giardina et al., 2014; Hansen et al., 2013; Sharma et al., 2016; Vermiere & Van de Heyning, 2009)

Cross-Modal Re-organization in Bilaterally Deaf Children
Cross-modal plasticity
Can occur as a result of decreased or abnormal sensory input
- Cortical regions in the deprived modality become vulnerable to the recruitment by the remaining, intact sensory modalities

Intra-modal plasticity is another form of cortical plasticity
- Brain changes are induced within a particular cortical area as a result of increased or decreased input to that sensory system

Cross-modal re-organization by vision in animal models
In congenitally deaf cats there is enhanced peripheral localization abilities (vision) by the posterior auditory field (Lomber et al., 2010)
- The deaf cats also show enhanced peripheral visual abilities

Similarly, enhanced visual motion detection appears to be sub-served by dorsal auditory cortex (Lomber et al., 2010)

Cross-modal re-organization by vision in children w/ deafness
Increased visual motion detection in humans with prelingual hearing loss onset (Hauthal et al., 2013; Shiell, Champoux, & Zatorre, 2016)

Cortical visual evoked potentials (CVEPs) in bilaterally deaf CI children and normal hearing children (Campbell and Sharma, 2016)

The CI children had larger CVEP amplitudes with earlier latencies
The CI children demonstrated additional activation of cortical areas typically associated with auditory processing (e.g. IFG, STG) for the higher order N1 and P2 CVEP components
- Normal hearing children only activated visual regions in the cortex
Cross-modal re-organization by the somatosensory system

Cortical auditory evoked potentials (CAEPs)

Normal hearing and early-implanted children demonstrated expected activation of auditory cortex compared to the late-implanted children.

The late-implanted group had significant activation of post-central gyrus in somatosensory cortex.

This suggests abnormal cross-modal processing of auditory stimuli.

Cross-modal re-organization by the somatosensory system

Delayed or abnormal P1 CAEP responses are observed in congenitally deaf children fit with cochlear implants (CI) after age 3.5 years.

Immature development of primary auditory cortex if auditory deprivation extends beyond this age (Sharma et al., 2002a)

- Delayed or abnormal P1 responses in late-implanted children
- Absent N1 and P2 components in late-implanted children
- Suggests deficits in higher-level auditory processing (Eggermont & Ponton, 2003; Gilley et al., 2005; Sharma, Campbell, & Cardon, 2015)
Early-implantation
Children who are early-implanted under the age of 3 yrs show typical N1 patterns
71% of early-implanted children developed an N1 response between ages 6-9 yrs
100% of the children between ages 9-12 yrs
BUT... the late (post 12 yrs) implanted children rarely develop the N1/P1 components

(Gilley et al., 2005)

What happens when auditory input is introduced after a prolonged period of deafness in childhood?
There is a functional de-coupling between primary and secondary auditory cortices in late-implanted children
This leaves secondary auditory cortex vulnerable to repurposing by other sensory modalities (such as vision & somatosensation)
And an increase in neurons responding to non-auditory inputs sub-cortically

(Dodd et al., 2002; Niparko et al., 2003; Niparko et al., 2005)

Better outcomes given early implantation
Behavioral evidence in CI children
Children implanted early in life (best by age 1-2 years)
Have the potential to attain normal language outcomes
Whereas late-implanted children continue to show language delays even after implantation

(Gerr, 2004, 2006; Nozu et al., 2010; Svirsky et al., 2004; Tajudeen et al., 2010)
Bilateral sequential CI in deaf children

There are detrimental effects of unilateral auditory deprivation in bilateral sequential cochlear implantation in deaf children.

Unilateral CI use restricts bilateral development of the central auditory pathways in bilaterally deaf children.

- Resulting in permanent and irreversible strengthening of the pathways arising from the first implanted ear (Gordon et al., 2003)
- These changes remain evident even 3-4 years after sequential implantation of the second ear (Gordon, Wong, & Papsin, 2013)

Bilateral sequential CI in deaf children

In bilaterally deaf children:

- Speech perception benefit from the 2nd implant is significantly reduced...
- If there are very long periods between sequential implantation (Ilg et al., 2013)

Effects of unilateral auditory deprivation

Animal models of early-onset SSD

- Unilateral hearing experience in early childhood results in abnormal representation or dominance of the normal hearing ear
- This results in several consequences from deficits:
  - auditory spatial awareness
  - sound localization
  - problems with higher-order auditory processing (Kral, Hubka, & Tillein, 2015; Tillein, Hubka, & Kral, 2016)
Functional Significance of Cross-Modal Plasticity in Deaf Children

Cross-modal plasticity may serve as a source of variability in speech perception outcomes

- Pediatric and adult CI patients who exhibit higher levels of metabolic activity in dorsolateral pre-frontal cortex
  - Have higher speech perception scores
- CI patients w/ higher levels in ventral visual processing regions
  - Have lower speech perception scores (Lee et al., 2007; Giraud & Lee, 2007)
- There may be a dorsal/ventral dichotomy of cortical areas for good versus poor clinical outcomes (Giraud & Lee, 2007; Lee et al., 2007)
- Cross-modal re-organization by vision and somatosensation is negatively related to CI outcomes in children (Campbell & Sharma, 2016; Sharma, Campbell, & Cardon 2015)

The recruitment of auditory cortex by vision may be functionally correlated to increased reliance on visual cues as a result of auditory deprivation

Late-implanted pediatric CI recipients exhibited
- Higher levels of visual-only speech perception (lip-reading abilities)
- Higher auditory-visual gains (speech perception performance in auditory-visual task compared to performance in the auditory-only task)
- In comparison to early-implanted children who exhibited higher auditory-only speech perception scores

(Bergeson et al., 2005)
Recruitment of Frontal Cortical Networks for Sensory Processing in Hearing Loss

Other simultaneous changes in cortical resource allocation that occur as a result of auditory deprivation

Frontal cortices are activated during challenging listening situations in adults with hearing loss (Peelle et al., 2011; Wingfield & Peelle, 2015)

Frontal Cortex is associated with:
- Attention
- Working Memory
- Problem Solving
- Higher Level Thinking
- Organization

CAEPs were recorded in a group of adults with mild-moderate hearing loss and a group of normal hearing adults

The hearing loss group demonstrated decreased activation of temporal auditory cortical areas and simultaneous recruitment of frontal cortices
- But the group of normal hearing adults showed only activation of temporal cortex (Campbell & Sharma, 2013)

Similar evidence of frontal cortex activation has been observed in both normal hearing and hearing impaired adults under adverse listening situations (Campbell & Sharma, 2013; Peelle et al., 2011; Sharma et al., 2016; Wingfield et al., 2006; Wingfield & Peelle, 2015)

So... in addition to cross-modal re-organization by the visual and somatosensory modalities as a result of decreased input to auditory cortex
- Frontal cortices are also simultaneously recruited as a means to facilitate top-down processes
Developmental cortical neuroplasticity in a child with SSD before and after CI

DOCTORAL WORK BY HANNAH GLICK
AT UNIVERSITY OF COLORADO AT BOULDER

Single Subject Study
- Female child w/ a progressive idiopathic sensorineural
- HL in right ear identified at age 5 years
- HL progressed to severe-profound by age 9 years
- Hearing in the left ear was normal
- Trials with a CROS hearing aid and frequency modulated (FM) system (<6 months)
- Unaided speech awareness threshold was 80 dB HL in the right ear
- Speech recognition threshold was 5 dB HL in the left ear
- At the age of 9.86 years, the child received a CI in her right ear

Immature development of the pathways in response to stimulation of the SSD ear
Normal hearing in left ear recruits contralateral (right) STG, MTG, IFG

A gradual morphological change in the child's CAEP response for her SSD ear following CI implantation

Eventually results in an age-appropriate waveform morphology in the SSD ear

“Normalization” of auditory pathway

By 14 months post-CI we see robust contralateral activation of only auditory temporal areas in left STG & MTG

The decrease in frontal activation post-CI suggests a decrease in listening effort

The robust contralateral activation suggests more typical development of auditory pathways post implantation
Improved behavioral testing results following SSD-CI

Speech perception and sound localization tested at 33 months post-CI
Sentence-level speech perception in the context of surround-sound restaurant noise
- Target sentences presented via a loudspeaker to the child's SSD (right) ear, w/ noise coming from other speakers

There was a 25% improvement in performance from pre-CI status
The improved score fell in the range of scores achieved by adult SSD-CI subjects tested in the same environment

Summary & Conclusions

SSD results in changes in the auditory pathway, including making this region susceptible to recruitment by other sensory processes
CIs are not currently approved by the FDA for SSD, but newer research adds to the growing body of evidence supporting potential benefits of CI in SSD
Adults with SSD-CI show improved hearing and cortical reorganization
Single-subject study revealed “normalization” of the auditory pathway for a young child receiving a SSD-CI within 2-3 years of implantation
More research is needed, but an argument for early SSD-CI is emerging