Communication and Functional Skills in Children with cochlear implants and additional disabilities

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Objectives

- Identify red flags for additional disabilities in children with cochlear implants.
- Describe how cognitive abilities and language abilities impact functional communication in children who are deaf/hoh with implants.
- Identify strategies to improve interdisciplinary teaming for improved functional outcomes in children with cochlear implants and additional disabilities. Participants will be able to describe the impact of atypical learning on communication and function.

Why it matters

- Additional disabilities in children who are deaf/hoh are more common than you may realize.
- We want children to achieve to their maximal potential, but often the complication of hearing and an additional disability makes it hard to know how to approach intervention.
- Seeing a child meet a goal is very rewarding, even if the child is not keeping up with peers.

Hearing Loss and Additional Disabilities

<table>
<thead>
<tr>
<th>Type of Disability</th>
<th>Hearing Loss GRI data</th>
<th>General Population Varieties of sources</th>
</tr>
</thead>
<tbody>
<tr>
<td>No Additional Disability</td>
<td>60%</td>
<td></td>
</tr>
<tr>
<td>Cognitive (Intellectual Disability)</td>
<td>9.8%</td>
<td>2.5%</td>
</tr>
<tr>
<td>Cerebral Palsy</td>
<td>?</td>
<td>0.3%</td>
</tr>
<tr>
<td>Blindness</td>
<td>3.9%</td>
<td>0.03%</td>
</tr>
<tr>
<td>ADHD</td>
<td>6.6%</td>
<td>5-10%</td>
</tr>
<tr>
<td>Specific Learning Disability</td>
<td>10.7%</td>
<td>5-10%</td>
</tr>
<tr>
<td>Other</td>
<td>12%</td>
<td></td>
</tr>
<tr>
<td>Autism</td>
<td>1.4%</td>
<td>0.9%</td>
</tr>
</tbody>
</table>

From 2007 Gallaudet Research Institute

Risk factors for developmental delays

- Neonatal history (LBW, preemie, asphyxia, IVH)
- Congenital infections
- Meningitis
- Environmental exposures (Pb)
- Failure to thrive
- Iron Deficiency Anemia
- Maternal Substance Abuse
- Environmental deprivation
- Family history of learning difficulties, attentional problems

Risk factors for developmental delay in deaf/ hoh

- Neonatal factors (prematurity, intraventricular hemorrhage, NEC, prolonged ventilation)
- Symptomatic congenital CMV
- Bacterial meningitis
- Some syndromes
- Family history of learning difficulties, attentional problems
Red Flags for atypical learning

- Global delay/cognitive
- Gross motor
- Fine Motor
- Vision
- Personal-social
- Language

Problem Solving/Global Developmental Delay

- Problem solving difficulties tend to imply delay in many domains (with the general exception of gross motor skills)
- Rate of progress is slower than peers
- Self help skills are often also delayed
- May have splinter skills which can be used for strategies in learning

Gross Motor Problems

- Low muscle tone (hypotonia)
  - Have to develop strength to overcome low tone
  - Sometimes less awareness of where you are in space
- High muscle tone (spasticity, cerebral palsy)
  - Can make it hard to move easily, might have troubles bending to sit or walking if legs are tight and cross over, might tiptoe walk.
- Balance
  - Might affect walking balance, riding a bike, ice skating
  - Some children are in constant motion to counter the difficulties with balance (bicycle stays upright when moving, not when it’s still)

Fine Motor Concerns

- Handed-ness
  - Developing handedness under a year of age
  - Minimal hand preference at older age
- Immature grasp patterns
- Poor hand strength/endurance
- Difficulty crossing the mid-line
- Motor planning

Red Flags for Vision Concerns

- Poor visual regard
- Poor tracking (up or down)
- Wiggling eyes
- Wandering eyes
- Head tilt

Functional Vision Assessment

- A teacher of the visually impaired is essential in the provision of services to children with dual sensory impairment
- The eye exam/ophthalmology exam only gives limited information about vision
- Getting a sense of how a child uses their vision and the best approach to provide information is critical
**Functional Vision Assessment**

- May assist you in determining:
  - Best lighting (light focused on the item, backlighting with a light box, etc)
  - Best angle or presentation of information
  - Best font size/contrast needs
  - Best speed with which we can present information (visual tracking)
  - Most visually relevant information for the child
  - Tactile adaptation of materials

**Difficulties with Personal/Social Skills**

- Independence, self help skills
  - May be impacted by motor skills, problem solving skills, expectations of caregivers

- Interest in other children (language vs. autism)
  - Should be commensurate with language level (…theory of mind literature…)
  - Some children have no interest, others want to play and communicate, but have breakdowns in their attempts to communicate

- Pretend/imaginary play
  - Somewhat based on language levels as well as imitation of what children see in their environment

**Foundations to communication**

- Access to language in appropriate format
  - If auditory, amplification sufficient to access
  - If visual, can see sufficiently to access

- Joint attention
  - Responding to our attempts to communicate or get the child’s attention, response to pointing, use of eye contact

- Making choices
  - Between objects, pictures of choice, based on words or signs

**Foundations to communication**

- Understanding cause and effect
  - When I push the button, Ernie pops up
  - If you put your toys away, we can go to the park

- Turn-taking
  - Playing with a ball back and forth, taking turns with a board game
  - Imitation of activities back and forth

**“Red Flags”**

- Indications of slow language learning rate in spite of appropriate, extensive intervention efforts (Chronological Age/Language Age gap widens)

- Persistent behavioral difficulties and/or attention deficits when good communication system in place

- Over-reliance on nonlinguistic means to understand others (tries to get around language, uses environmental cues or visual cues to understand and respond)

**“Red Flags”**

- Limited eye contact for social purposes
- Stereotypical behaviors; repetitive movements
- Lack of playing with variety of toys conventionally
- Difficulties retaining and generalizing information
- Poor speech intelligibility in spite of strong residual hearing
**Communication/Language**

- Learning rate does not match “expectations”
  - Re: residual hearing or communication access history
- Uneven development
  - Atypical expressive - receptive gap
  - Atypical receptive – expressive gap
  - Strong auditory skills, limited language progress

**Impact of UNHS on language outcomes**

- Universal Newborn Hearing Screening has allowed the earlier identification of children with hearing loss
- This subsequently enhances age of intervention with data to support language outcomes in the average range for early identified children
- With this improved language trajectory, we should be striving for earlier identification of children with co-existing developmental or communication disorders, however diagnosis can be challenging

**The Gap**

Children who are deaf/hoh and have a co-existing developmental disability often are under-achieving as compared to their cognitive potential

**Language lags behind matched peers**

- Expected: Language in line with nonverbal cognitive abilities
- Language profoundly behind nonverbal cognitive abilities
- ~22 points behind \( p=0.001 \)
- ~25 points behind \( p=0.002 \)

**Nonverbal Cognitive Abilities Most Predictive of Outcomes**

Some disabilities that directly affect communication (e.g., autism) may need to be studied separately

**Accounting for Development**

- One of the most difficult aspects of studying outcomes among children with additional disabilities is the choice of an appropriate comparison (control) group
- When language is the goal, the language skill set in children should be **similar to their developmental level**
5 Possible Developmental Trajectories

<table>
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<tr>
<th>Score</th>
<th>5</th>
<th>10</th>
<th>20</th>
<th>30</th>
<th>40</th>
<th>50</th>
<th>60</th>
<th>70</th>
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</thead>
<tbody>
<tr>
<td>Age in Months</td>
<td>1</td>
<td>6</td>
<td>11</td>
<td>16</td>
<td>21</td>
<td>26</td>
<td>31</td>
<td>36</td>
</tr>
</tbody>
</table>

- Maintained functioning comparable to age peers
- Achieved functioning comparable to age peers
- Moved nearer functioning comparable to age peers
- Made progress; no change in trajectory
- Did not make progress

Hebbeler, 2006

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Functional Skills

- Aside from functional listening skills, day-to-day functioning has been excluded from existing literature on outcomes of children who are deaf/hard of hearing
- Functional skills tend to be the focus in the field of physical medicine and rehabilitation and special education programs (though impacted by requirement of grade-level competencies)

Pediatric Evaluation Disability Inventory

- Standardized measure of daily functional abilities across 3 domains for children ranging in abilities from 6 months to 7.5 years
  - Self-Care
  - Mobility
  - Social functioning
- Has 197 questions parents complete
- Skills are rated with
  - Scaled scores (max score of 100)
  - Standard scores (mean of 50)

Pediatric Evaluation Disability Inventory

- Useful in treatment planning and identifying specific areas where assistance is needed
- Sensitive to change over time

Mobility Domain

- Toilet Transfers
- Chair/Wheelchair Transfers
- Car Transfers
- Bed Mobility/Transfers
- Tub Transfers
- Indoor locomotion methods
- Indoor locomotion: distance/speed
- Indoor locomotion: pulls/carries objects
- Outdoor Locomotion: methods
- Outdoor Locomotion: distance/speed
- Outdoor locomotion: surfaces
- Upstairs
- Downstairs

Self Care Domain

- Food textures
- Use of Utensils
- Use of Drinking Containers
- Toothbrushing
- Hairbrushing
- Nose care
- Handwashing
- Washing Body and Face
- Pulver/Front-opening garments
- Fasteners
- Pants
- Shoes/Socks
- Toileting Tasks
- Management of Bladder
- Management of Bowel
**Social Function Domain**

- Comprehension Word Meanings
- Comprehension of Sentence Complexity
- Functional Use of Communication
- Complexity of Expressive Communication
- Problem-resolution
- Social Interactive Play (adults)
- Peer Interaction (child of similar age)
- Play with Objects
- Self-Information
- Time Orientation
- Household Chores
- Self-Protection
- Community Function

**Children with CI and additional disabilities**

- 1st CI cohort with control group (2007)
  - Hearing controls were matched according to chronologic age and nonverbal cognitive abilities
- 2nd CI prospective cohort (2008-2011)
  - Hearing controls were not included

**Study Design and Methods**

- Early development assessed with the Revised Gesell Developmental Schedules
  - A developmental evaluation for young children
  - Evaluates a child's problem solving and motor skills (gross and fine motor) as well as personal social, language skills (not used for study) prompting a functioning age
- All children evaluated pre-cochlear implant
- Controls evaluated with same tool (2-3 years of age)
- Developmental quotients were calculated

**Outcome Measures**

- **Preschool Language Scale - 4th edition**
  - Evaluates a child's understanding of language and expressive language abilities (receptive and expressive language skills)
  - Language quotients were used if the child obtained a standard score of 50 (lowest possible score calculated)
- **Pediatric Evaluation Disability Inventory**
  - Essential daily functions related to mobility, self-care, and social-communication
  - Useful in treatment planning and identifying specific areas where assistance is needed

**Characteristics**

<table>
<thead>
<tr>
<th></th>
<th>CI N=14</th>
<th>Controls N=7</th>
<th>p</th>
</tr>
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<tbody>
<tr>
<td>Age diagnosis (mos)</td>
<td>6 (0-58)</td>
<td>---</td>
<td></td>
</tr>
<tr>
<td>Age CI (max)</td>
<td>30 (14-134)</td>
<td>---</td>
<td></td>
</tr>
<tr>
<td>Duration CI (max)</td>
<td>28.5 (11-63)</td>
<td>---</td>
<td></td>
</tr>
<tr>
<td>Nonverbal CQ</td>
<td>59 (30-80)</td>
<td>50 (22-89)</td>
<td>0.5</td>
</tr>
<tr>
<td>Age study* (max)</td>
<td>64 (25-146)</td>
<td>78 (49-90)</td>
<td>0.2</td>
</tr>
<tr>
<td># therapies</td>
<td>3 (0-4)</td>
<td>2 (0-5)</td>
<td>0.2</td>
</tr>
<tr>
<td>Total therapy hrs</td>
<td>1.75 (0-8)</td>
<td>2 (0-4.5)</td>
<td>0.8</td>
</tr>
<tr>
<td>Male</td>
<td>8 (57%)</td>
<td>5 (71%)</td>
<td>0.7</td>
</tr>
</tbody>
</table>

*All last PEDID visit

**Developmental Issues**

- Cognitive
- Motor
- Oral/Motor
- Other 1 issue 2 issues 3 issues

- Variability in cognitive development among children with CI.
Functional Skill Standard Scores in children with CI and AD as compared to DD-matched hearing controls

Score of 50 is average or age appropriate


PEDI Domain Standard Scores

Presented at SENTAC, 2010; European Pediatric CI mtg (Athens, Greece)

Adjusted for cognitive abilities, parental income

Domain Scaled Scores

On scaled score, 100 is highest possible score (skills up to the age of 7)

Adjusted for cognitive abilities, parental income, age

Social Function Items: 6mos – 1 year

Awareness of others

Social Function Items: 1.5-2 yrs

Understands 10 words

1 Step commands

Requires immediate help

Initiates play

Interacts briefly
Social Functioning

After controlling for cognitive abilities, age at implant, time since implantation, age at PEDI not associated with scores

Peer Interactions

Receptive language is associated with these skills

Progress in PEDI skills over time

- Prospective group (n=6)
- Measured PEDI scores
  - at baseline (pre-implant)
  - 6 months post-implant
  - 12 months post-implant

PEDI Changes over time

Social Function Items: 6mos - 1 year

Social Function Items: 1.5-2 yrs

What have we learned?

- Children make measurable progress post-implant:
  - Increase in functional communication skills
  - Still behind relative to their age
- In terms of functional communication abilities, children with cochlear implants seem to have similar skills as their hearing “peers” based on scores, however may be different on specific questions
- Must consider (and STUDY) alternative outcomes

Case Example Discussion

Case

- HH didn’t pass his newborn hearing screen, however passed at follow-up
- His hearing loss was thought to be progressive and noticed around a year of age with profound hearing loss by 2 ½ years of age
- The cause of his hearing loss was enlarged vestibular aqueduct
- He received bilateral cochlear implants at just over 3 years of age
- He walked at 19 months of age

Case HH

- It had been difficult to get a good understanding of his CI thresholds as he shuts down in booth testing
- When he received a second opinion about mapping, there were concerns about settings outside usual parameters so he was re-mapped
- His new MAP seemed better with more responsiveness to sounds

Case HH

- Expressive Communication:
  - To get what he wants, he will usually take his parent’s hands to what he wants
  - They started using some pictures to help with communication and responded well to this (he went through many pages to find the toilet to indicate that he needed to go)
  - He also makes a specific sound to indicate he needs help or wants your attention.
Case HH

• He could say ma and ba and vowel sounds
• He would occasionally do a series of vowels as if saying I did it
• He was not yet jargoning with inflection and sounds more like singing than a jargon
• Apraxia has been raised as a possible concern, however his comprehension was also a notable problem

Case HH

• He had behaviors which were also concerning, including head-banging, sensory seeking and sensory avoidance (OT added to his services)
• At about 4 ½, they shifted his educational setting to provide a total communication and provided a lot more exposure to sign language
• In this setting, he expanded his sign fairly quickly with 10-15 signs, but did not quickly combine signs

Case HH

• At about 5 years of age, he completed cognitive and language testing
• His receptive and expressive language skills were at an 18 month level
• His non-verbal IQ on the Leiter was 100
• We tried to address issues around attention and sleep through medication with some improvements

Case HH

• At 6 years of age, his family had more questions about what is going on, are there broader problems (beyond apraxia and receptive language) such as an autism spectrum disorder
• Their interests were driven by wanting to everything possible for him to ensure he makes progress and achieves independence
• This prompted a school observation

Questions

• How does he seem to learn?
• What has the environment implemented to help his learning pattern?
• What could they do differently?

Team Building

• Strive towards common goals
• Listen actively
• Communicate effectively between/among team members
• Be confident in what you know and recognize when you don’t know something
• Learn from others/collaboration
• Be open to new ideas and strategies
• Think outside the box
• Consider co-treatment when appropriate
• Try something and tweak it when it doesn’t work
Goals/ Planning
Rules to guide instruction
• Focus on the donut, not the hole
• Celebrate successes great and small
• If a dead man can do it, it is not an appropriate objective
• Use meaningful contexts to make concepts explicit
  “I would tell them to be an advocate for your child and let the professionals know how your child is reacting.”
  Dr. T. Jones, Gallaudet University

Routine-Based Intervention
• Identify routines that are salient to family and child
• Determine sounds that may be relevant in the environment
• Work towards increasing hierarchy of communication skills
  Tools found at www.kidsdbci.org/links-and-resources.html
  “It’s a process and it may be difficult at times, but in the end, it was all worth it.”

Tools to Guide Intervention
• Tools found at www.kidsdbci.org/links-and-resources.html
• Routine Based work-sheet
• Home and Sound inventory
• Hierarchy of Communication Skills
• Gestural Assessment

Putting it all together
• A combination of
  – establishing realistic goals with family as the hub
  – using structured salient assessment/planning tools
  – trying creative and varied approaches
  – observing what happens and adapting as needed
• May be an effective tool-kit for complex children

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