Cochlear Implant Candidacy for children who are multiply involved: A developmental-pediatric perspective

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Some important premises

- There is a high rate of additional disabilities in children who are deaf/hard of hearing.
- Comparing these children to typically developing deaf/hard of hearing children when evaluating outcomes is inappropriate.
- Development is ever-changing due to on-going brain development (therefore, the earlier we implant children, the less accurate our predictions and surveillance is necessary).
- Most childhood development tends to follow specific patterns in early childhood (it’s about the brain).
- Family and child support and adaptations are critical.
<table>
<thead>
<tr>
<th>Type of Disability</th>
<th>Hearing Loss GRI data</th>
<th>General Population Variety of sources</th>
</tr>
</thead>
<tbody>
<tr>
<td>No Additional Disability</td>
<td>60%</td>
<td>2.5%</td>
</tr>
<tr>
<td>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 Spectrum Disorders</td>
<td>1-4%</td>
<td>0.9%</td>
</tr>
</tbody>
</table>

From 2007 Gallaudet Research Institute

CDC MMWR 2012
Risks and Etiologies

• Etiology can confer risk and may not protect from other risk factors for developmental problems
  – Risk factors for hearing loss often overlap with risk factors for developmental delay
  – Risk factors for developmental delay can co-occur in children with hearing loss unrelated to the cause of hearing loss

• Disability labels do not tend to provide an effective guide to our understanding of a child’s capacities
Specific Disability Label Not Very Predictive

a. scatter plot of nonverbal cognition and language

- Log Receptive Quotient
  - 1.5
  - 2.0
  - 2.5
  - 3.0
  - 3.5
  - 4.0
  - 4.5
  - 5.0

- Nonverbal cognitive quotient
  - 20
  - 30
  - 40
  - 50
  - 60
  - 70
  - 80
  - 90
  - 100
  - 110
  - 120

- $R^2 = 0.68$

b. scatter plot of disability diagnosis and language

- Disability Diagnosis
  - CP
  - CHARGE
  - GLOBAL
  - OTHER

- Log Receptive Quotient
  - 1.5
  - 2.0
  - 2.5
  - 3.0
  - 3.5
  - 4.0
  - 4.5

- $R^2 = 0.07$
Appropriate Comparison Group

- Children who are deaf/hoh with co-existing developmental disabilities learn differently than children who are deaf/hoh without a disability, making this comparison group inappropriate.

- Understanding the range of outcomes within disability categories and using these benchmarks to measure progress and guide expectations is more appropriate (non-verbal cognitive skills a large contribution to overall outcomes across disability categories).
Development is a Process

Center on the Developing Child
HARVARD UNIVERSITY

Human Brain Development
Neural Connections for Different Functions Develop Sequentially

Sensory Pathways (Vision, Hearing)
Language
Higher Cognitive Function

FIRST YEAR

Birth (Years) (Months)

And it’s about the brain

- There is rapid brain growth in the first two years of age
- Myelination of the brain continues into early adulthood
5 Possible Developmental Trajectories

- 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

Age in Months

Score

Hebbeler, 2006
Typical vs Atypical Patterns

• Motor Development
  – Head to toe
  – Proximal to distal
  – Primitive reflexes to protective responses
  – Balance

• Delay
  – Follows the usual trajectory, just at a later time
  – Rate of progress over time is important

• Atypical development
  – Atypical patterns of motor movements

• Take home point: you need a good reason to justify the motor delay in children who are deaf/hoh (inner ear malformation, vision impairment, syndrome associated with motor delay, brain-based process)
Typical vs Atypical Patterns

• Cognitive development
  – Infants and Toddlers learn through exploration of the environment and sensory input
  – Preschooler’s learn through language, spatial experiences, use concrete problem solving and have magical thinking, no abstraction
  – Early Childhood begin to have more logical thinking, but still concrete
  – Late Childhood/Adolescence use abstract and logical thinking

• Delay (non-verbal problem solving for deaf/hoh):
  – Follows the usual trajectory, just at a later time
  – Rate of progress over time is important, tends to plateau

• Atypical Development:
  – May have varying learning profiles, could be suggestive of a specific learning disability
Language: Considerations

• Language
  – What is hearing?
  – What is communication environment?
  – Are there unexplained (atypical) patterns of language development (processing, good understanding, poor speech)?
  – How is a child developing in their non-verbal, gestural, and pragmatic language?

• Why the gap matters?
  – When we don’t recognize cognitive potential, we miss children with high cognitive potential who have low average language levels and we have been satisfied with this
Estimated Functional Skills
Estimated Functional Skills
Adjusted mean Communication Scores

Communication Function Score

Range of nonverbal IQ

TOTAL  IQ >100  IQ 80-100  IQ <80

HIGH LANGUAGE  LOW LANGUAGE

Manuscript to be submitted to JDBP
Adjusted mean Communication Scores

![Graph showing communication function scores across different ranges of nonverbal IQ.](Image)

- **TOTAL IQ >100**
- **IQ 80-100**
- **IQ <80**
Behavior: Considerations

• Behavior
  – Understood in the context of communication needs
  – Understood in the context of the child’s overall developmental levels (i.e., if a child is functioning at a 2 year level, anticipate an attention span that is commensurate with most 2 year old’s)
  – Recognize when there may be risks for emotional difficulties (parent-child relationship, exposure to abuse, domestic violence)
  – Recognize when there are neurobiological factors contributing to behavior (attention, impulsivity, hyperactivity)
Integrating the Information

- Finding a Developmental Pediatrician with an understanding of typical development in children who are deaf/hoh
- Comprehensive History (risks for hearing, development)
- Physical Examination
- Laboratory, Genetic, and Imaging studies
- Broad based developmental assessment evaluating a number of domains
  - Gross Motor
  - Fine Motor
  - Cognitive
  - Language
  - Personal-Social
How the information is used?

• Is it about just determining candidacy?

• In our center, the pre-implant process is rarely to determine candidacy, but rather to recognize or identify all areas of need which will require support for overall child’s development.

• Information is used to guide conversations with families about anticipated rate of progress, adapting typical intervention strategies to support the individual child’s needs.
Family Perspective

• Deaf/hh Plus is meant to be a positive term, not in any way negative or insensitive to the child who has medical issues along with hearing loss. In fact, I see it as an “A+” or “B+,” meaning the child carries additional positive qualities. But it is a gift that needs to be carefully unwrapped. And it may not appear to be a gift when you first receive it. Time helps you appreciate, understand and unfold the possibilities. And the “Plus” most often means the child and family has added responsibilities and requires additional expertise."

• — Candace Lindow-Davies, MN Hands & Voices

http://www.cohandsandvoices.org/plus/index.html