Learner Outcomes

After attending this session, participants will be able to:
1. Define national trends in the diagnosis and treatment of children with hearing loss who have additional disabilities;
2. Recognize the range of co-occurring conditions that can happen with children with hearing loss; and
3. List accommodations and other strategies that can be employed during the diagnosis and treatment of children with hearing loss who have additional disabilities.

Multiple Disabilities Defined

- The combination of two or more disabling conditions
- These conditions may include deafness in association with blindness, mental retardation, autism, and/or physical impairment

“It is the reduction in possibilities for compensation, whether spontaneously or after intervention, that makes a child multiply disabled”

(Knoors and Vervloed, 2003)

ETIOLOGY OF HEARING LOSS AND MULTIPLE DISABILITIES

Causes of HL in Children

- Genetic (50%)
- Non-Genetic (25%)
- Unknown (25%)

Disclosure

- I currently receive royalties for three books published with Plural Publishing:
  - Telepractice in Speech-Language Pathology
  - Telepractice in Audiology
  - Assessing Listening and Spoken Language In Children with Hearing Loss
- I am a partner and co-founder of the 3C Digital Media Network, LLC.
Congenital Anomalies & In Utero Infections
- Charge Syndrome
- In Utero Infections:
  - CMV
  - Herpes
  - Rubella/German Measles
  - Syphilis
  - Toxoplasmosis

CHARGE Syndrome
- CHARGE Syndrome is an autosomal dominant disorder from mutations in the chromodomain helicase DNA-binding protein 7 gene (Vissers et al., 2004).
- The diagnosis of CHARGE is based on a combination of the following features:
  - Facial clefting
  - Genitourinary defects
  - Tracheoesophageal fistula
  - Short stature
  - Developmental delay
- Hearing loss and cognitive and developmental sequelae are common features of CHARGE.
- Studies have shown the CI outcomes in children with CHARGE are often:
  - Improvement on either closed-set or modified open-set word and sentence recognition
  - Identification of syllable patterns and Ling sounds
  - Outcomes will vary

Congenital Cytomegalovirus (CMV)
- CMV infection is the most common intrauterine viral infection
- Sensorineural hearing loss (SNHL) is the most common sequela affecting 22 to 65% of affected children (Dahle, et al., 2000; Fowler & Boppa, 2006)
- CMV can result in several other developmental disabilities, including blindness, cognitive impairments, and motor impairments.
- Results of research performed with CI children with CMV:
  - Scores on measures of speech intelligibility and closed set sentences were lower than non-CMV children (Rameriz Inscoe and Nikolopoulos, 2004)
  - There was a wide range of performance for both CMV and non-CMV children (Rameriz Inscoe and Nikolopoulos, 2004)
  - Outcomes in Speech Perception Categories for CMV children were highly variable, with some children making minimal progress and others making substantial gains (Lee et al., 2005)

Postnatal/Acquired Conditions
- Low birth weight / Hypoxia
- Ototoxicity
- Hyperbilirubinemia
- Meningitis

Disabilities That Occur with Deafness (%)

<table>
<thead>
<tr>
<th>Disability</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Deafness with No Other Disabilities</td>
<td>60.1</td>
</tr>
<tr>
<td>Learning Disability</td>
<td>10.7</td>
</tr>
<tr>
<td>Intellectual Disability</td>
<td>9.8</td>
</tr>
<tr>
<td>Attention Deficit Disorder (ADD/ADHD)</td>
<td>6.6</td>
</tr>
<tr>
<td>Blindness and Low Vision</td>
<td>3.9</td>
</tr>
<tr>
<td>Cerebral Palsy</td>
<td>3.4</td>
</tr>
<tr>
<td>Emotional Disturbance</td>
<td>1.7</td>
</tr>
<tr>
<td>Other conditions</td>
<td>12.1</td>
</tr>
</tbody>
</table>

Multiple studies have found that 50-75% of deaf/HOH children with multiple disabilities have more than one additional disability.

Gallaudet Research Institute
Intellectual Disability (10%)

- Largest Group: Down Syndrome
- Most children with intellectual disabilities have HL in one or both ears
  - Some estimates approach 80%
- Intellectual disabilities affect language, learning, socialization
- Intellectual disabilities are exacerbated by hearing loss

Learning Disabilities (11%)

- The DSM definition
  - IQ-achievement discrepancy
- Four types of learning disorders:
  - Reading, written expression, mathematics, and learning disorder “not otherwise specified”
- Domain-based
  - Focus on cognitive abilities,
  - Phonological awareness, listening comprehension, word retrieval

Learning Disabilities (cont.)

- Described for over 100 years but remarkably little research
  - Each child is unique
  - Unable to use traditional research methods
- Educational Challenges
  - Programs that provide structured intervention
  - Specific, clearly stated objectives, focusing on the individual strengths and needs of each child.

Attention Disorders (7%)

- ADD/ADHD (5% in general population)
  - Inattention
  - Distractibility
  - Impulsivity
  - Hyperactivity
- Can interfere with all aspects of development
  - Educational achievement
  - Socialization
- Parents and teachers may over-identify
- Need for Comprehensive Medical Evaluation
  - Medication for AD

Visual Impairment (4%)

- Genetic Syndromes
  - Usher (Types I and II)
  - Goldenhar
- Congenital / Perinatal
  - CHARGE, CMV, meningitis

Deaf-Blindness

- Child cannot be accommodated in special education programs designed for children with deafness or blindness.
- Approx 11,000 deaf-blind children in U.S.
- Most have severe developmental disabilities (CI)
Cerebral Palsy (3.4%)

- Three Main Forms
  - Spastic CP: stiff, difficult movement
  - Athetoid CP: involuntary and uncontrolled movements
  - Ataxic CP: disturbed sense of balance, position in space, and overall lack of coordination

- Classification made according to the age of diagnosis
  - 70% of CP occurs prior to birth (prenatal)
  - 20% occurs during the birthing period (perinatal)
  - 10% occurs during the first two years of life (postnatal)

- Remarkably little research on HL in CP

**IMPLICATIONS FOR AUDIOLOGICAL MANAGEMENT**

### Audiologic Assessment

- Many of same components
  - Test battery
  - Coordination of multiple disciplines
- Delays can be prevented by competent, proactive management

### Physiologic Measures (ABR, OAE, etc.)

- To define nature/degree of HL
  - Sedation
    - More likely to be needed
  - Communication with medical staff is critical
    - O.R. Evaluation
      - When there are respiratory risks
      - More expensive but “worth it” to achieve timely follow-up
      - Communication with medical staff enables multiple procedures (e.g. earmolds)

### Behavioral Assessment

- Begins with VRA, developmental age of ~six months
- Multiple disabilities should not preclude attempts to obtain behavioral assessment
  - You don’t know until you try; results can be surprising
- Modifications often needed
  - Infant seat; head support; careful observation
- Begin with sound field but individual ear data needed as soon as possible
  - Consider use of child’s earmolds w/ insert receivers

### Goal of Audiologic Assessment

- To obtain the info needed to proceed with hearing aid selection and fitting
  - Approximation of HL degree and configuration
  - Need accurate estimate of low and high frequencies
- Children with multiple disabilities tend to be later diagnosed and fitted
- Need to identify centers with the experience and institutional support to manage these children well
Considerations in HA Selection

- Flexibility in hearing aid circuitry
- Volume control covers
- FM may be useful for young children on ventilators
- Remote controls for children with poor head control
- Multiple care providers necessitate easy operation/maintenance of devices

Close Follow-up is Essential

- 1 month
- Every 3-4 months
- On-going diagnostic assessment
- On-going electroacoustic analysis
- On-going monitoring for OME
  - Health implications
  - Hearing aid implications

Assessment of Aided Performance

- Formal and Informal
- Referral for Cochlear Implant
  - At one time, children with MD not considered CI candidates
  - Many centers will – if no medical contraindications and reasonable chance of benefit
  - Success may need to be defined differently

PARENT / FAMILY PERSPECTIVES

Guiding Principles (Ewing and Jones, 2003)

1) Every child is capable of learning even when disabilities are multiple and severe
2) Peer acceptance and social relations are essential for all children including those with multiple disabilities
3) Families and communities are critical to success
4) Service delivery based on a transdisciplinary model results in better coordination and sharing of knowledge and resources

Communicating with Families

- Children with multiple disabilities pose many challenges for professionals
- Focus on areas of expertise and specialization is needed, but need to consider implications for family
- Families are profoundly affected by a child with special needs, especially when the history includes developmental disabilities and serious illness.
Family Perspectives

“Having a child with disabilities affects the entire family. It is something we live with every minute of every day. The simplest things that happen every day remind us of the fact that we’re different.”

“The best thing professionals can do is admit they don’t have all the answers, and that no one else does either.”

“Give power to families by reminding them that they are the experts on their child, because they are with the child every day and most professionals only see them in the clinic or school.”

“Even other families with children who have disabilities cannot know what it’s like for another family. Many professionals recommend involvement in family support programs, but it’s important they realize this doesn’t solve all the problems.”

“It took me a long time to seek support outside my own immediate family. I was still grieving the loss of my “perfect” child, and I didn’t want to share that heartbreak with strangers.

“Everyone is different, and everyone needs to seek support in their own time. Professionals can help by reminding families that support is available if they need it.”

“Your patience and understanding is even more valuable than anything you can fix.”

COCHLEAR IMPLANTATION & CHILDREN WITH MULTIPLE DISABILITIES

More Children with Additional Needs are Receiving Cochlear Implants

■ Based on surveys, approximately 25% - 50% of children followed by some cochlear implant programs have multiple disabilities
■ The number has increased over the years due testing and programming techniques that do not require full participation on part of the child
Rehabilitation/Therapy

- Goals
  - Improve child's auditory/listening skills
  - Establish communication system

Specific Strategies

- Create a listening environment
- Create a communicative environment
- Hand Cue
- Acoustic Highlight
- Auditory Sandwich
- Parent participation
- Dialogue
- Developmental Order
- Follow the Child's Lead
- Flexibility is the key!!

Move Along a "Continuum of Cueing"

- Least to Most – Verbal Cueing

Move Along a "Continuum of Cueing"

- Least to Most – Verbal to Hand-over-hand

Important Considerations

- Child's Developmental Level
- Child's Motor Limitations
- Child's Sensory Needs

Establishing Language/Communication Mode(s)

- Do not limit yourself to one method
- Children with hearing loss and additional special needs may need alternative communication modes
- After assessment and consideration, consider picture systems or other alternative communication devices
Establishing Language/Communication Mode(s)

- Strategies may change over time for a child
- Just because you start with one “plan” does not mean you are “stuck” with it. However, be clear to caregivers that you are changing your recommendation, your prognosis, or your approach.

Remember...

Monitoring progress is essential to determine the need for change and/or future interventions.

Every session is “diagnostic.”

Success Is Individual

- Defining success may be difficult in children with additional disabilities
- Seeing change may take months and years of tuning
- Help the family identify success
- The success is a result of the family’s hard work and dedication
- The feeling of success encourages the family to continue on this journey

Collaboration & Interprofessional Service Delivery Is Critical

- All professionals working with the child must work together
- Different professionals bring different areas of expertise to the discussion
- Management discussions amongst team members may initiate ideas of alternative and creative test and therapy techniques to meet the needs of the child

Case 1: 18 mon. old.

- Congenital CMV
- DNP Newborn Hearing Screen
- ABR at 1 month
- Cortical Blindness
- Swallowing disorder
- Seizure Disorder requiring medication
- Parent had appropriate expectations at time of candidacy evaluation
- Received a CI at 12 months of age
- Child is wearing the device consistently, becomes upset when device does not work, alerts to some sounds (not consistent)
Case 2

- Hearing loss diagnosed at 5 months of age
- Fit with hearing aids at 6 months of age
- Enrolled in PT, OT, and AV Therapy
- Received cochlear implant at 2 years 8 months of age
- Diagnosis – Autism
- Currently, child is in elementary and while her spoken language is excellent, she continues to require support related to pragmatics, socialization

Case 3

- Identified with bilateral profound sensorineural hearing loss at birth; received hearing aids by 3 months of age
- Diagnosed with Pierre Robin Syndrome
- Cleft of the palate, nares - now repaired
- Eating and swallowing issues; continued to use a feeding tube
- Received a cochlear implant on right side at age 3 years; wears a hearing aid in left ear; bilateral implantation not possible due to cochlea deformity on left side
- Began using sign at home and uses some sign at school
- Vocalizations are hoarse due to multiple surgeries for cleft
- Now, enrolled in public school using iPad and other AAC devices; he vocalizes and uses occasional signs to communicate his wants and needs

Adaptive Intervention for Children with Hearing Loss & Additional Needs

- Level of vision, hearing, and time in sound must be considered in all communication. CI must be worn!
- All therapy approaches need to be individualized - in consideration of both the child AND the caretaker
- Every interaction needs to be in spoken words, regardless of what the targeted communication mode is for the child
- Auditory-verbal therapy needs to be adapted when implemented with children who experience deaf-blindness and have received a cochlear implant

Adaptations for Children with Hearing Loss & Additional Needs

- Utilize routines to teach communication skills
- Augment communicative input:
  - Touch cues: systematic touches, made on the child's body, to convey meaning (e.g., touch corner of mouth to elicit vocalization)
  - Object cues: objects / partial objects utilized to convey meaning (bubble wrap handed to child, to indicate, "Time to take a break")
**Intervention: Follow The Child’s Lead**

- **Phase 1 - Intervention**
- **Strategy:** Follow the Child's Lead - “Responsiveness”
- Interpret eye gaze (Verbalize what you think she wants)
- Interpret facial expressions (Verbalize what you think she is telling you)
- Respond to ALL forms of the child's potential communication:  
  - body movement, reaching, gestures, sounds, words
- Imitate the sounds the child makes
- Imitate and expand her vocalizations (i.e., child says "mrmr," interpret as 'more' or 'wa' as 'water')
- Respond directly to the intent of the child's initiated communication (honor intent, if appropriate)

**Intervention: Narrative Description**

- Describe a motor action that is occurring: of your child, yourself, others (i.e., "Suzie walking, walking - pushing, good pushing," "Mommy opening cereal," etc.)
- Make a verbal comment, related to (i.e., "Suzie's so happy today," "Daddy's home!")
- Describe an action / activity, pairing it with an object (i.e., While putting on shoe, get child's attention, bring shoe to your face and say, "Shoe, shoe, shoe, put on shoe.")

**Parent/Caregiver vs. Child Behaviors**

**Parent**
- Provides opportunities to participate
- Follows child lead in behavior, vocalizations, nonverbals
- Descriptive talk regarding objects and actions
- Gives word to non-verbal communication

**Child**
- Learns to participate, even to a small degree, in activities
- Child learns to initiate, make choices
- Learns to listen and notice what is being described in activity
- Learns basis for communicative intent

**Intervention: Cue Responses**

- Cue Response to Environmental Sound
- Implement Auditory Sandwich (to facilitate child's COMPREHENSION)
- Incorporate Critical Elements in Speech (specific words)

**Intervention: Response to Environmental Sounds**

- Point out sounds that naturally occur in the environment (e.g., fan on heating unit, humming sound of computer / refrigerator)
- Call attention to "episodic" sounds that take place within the child's earshot (i.e., "I hear Daddy's car," "noisy dog," "telephone is ringing!")

**Parent/Caregiver vs. Child Behaviors**

**Parent**
- Gives directions (Pauses, provides assistance, if needed)
- Auditory sandwich - Auditory-visual-auditory for objects / actions
- Models gestures / pointing, provides choices with assistance

**Child**
- Responds to directions with / without support; increases turn-taking
- Increases receptive language and vocabulary
- Increases assisted gestures, toward intentional communication
Intervention: Opportunities for Imitation

- Provide models and opportunities for the child to communicate by imitation.
- Shapes child's communication forms to the next level of complexity (i.e., reaching to pointing, vowel to CV, one word to two).
- Child is expected to listen, to respond, to communicate - via body movement; contact, distal, or representational gesture; single or repetitive vocalization; word approximation; total communication; speech.

Intervention: Providing Choices

- Shift from giving directions or automatically providing objects to offering choices (i.e., "Which one do you want?)
- Offer choice through visual, tactile, and / or kinesthetic modes
- Begin with choice of preferred, with "nothing" alternative
- Move to providing choice between preferred and non-preferred
- Present choice among two or more preferred items / objects / foods / activities

Intervention: Auditory Sandwich - Expression

- Make direct request for child response, utilizing the "auditory sandwich"
- Augment verbal input with movement, touch or object cues, gesture, and / or manual sign
- Provide opportunity for child to use a variety of communicative functions: reject, protest, request (action, object, attention, continuation), greet, comment, offer, ask / answer a question

Parent/Caregiver vs. Child Behaviors

Parent

- Uses concentrated model of vowel & CV's (for imitation)
- Provides more gestures to be imitated
- Provide opportunities for word imitation
- Uses "Up the ante" in cueing strategies

Child

- Begins to imitate more sounds
- More gestures to request objects / actions, protest; use of gestures more consistently
- Approximates more simple words: "wa/water," "ou/out"
- Changes body movement to gestures, word approximation

Strategies

- Increase in the number of opportunities provided for the child to communicate
- Provision of narrative description related to child, self, another aspect of the present context (e.g., another person, family pet)
- Incorporation of critical elements in spoken language (e.g., child's name, partner's name, preparatory cues prior to touching / interacting with child)
- Overall implementation of the auditory sandwich technique

Strategies

- Demonstrating RESPONSIVENESS to a wider variety of the child's behaviors
  - Imitating the child's vocalizations
  - Responding to the child's facial expression
  - Recognizing and responding to the child's body movements
  - Assigning meaning to the child's gestures
  - Noticing and following the child's apparent communicative intent
- Expansion of the critical elements in the child's spoken communication
Unilateral losses

- Children with unilateral hearing loss are 10 times more likely to fail a grade by age 10.

- Almost 50% of children with at least a 30dB hearing loss in one ear, have failed one or more grades...or are receiving support services.

Potential Impact of Hearing Loss

<table>
<thead>
<tr>
<th>Age</th>
<th>Vocabulary</th>
<th>Age</th>
<th>Vocabulary</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Normal Hearing</td>
<td>Profound Hearing Loss</td>
<td></td>
</tr>
<tr>
<td>4/5 yrs.</td>
<td>2,000</td>
<td>4/5 yrs.</td>
<td>&gt;500</td>
</tr>
<tr>
<td>7/8 yrs.</td>
<td>22,000</td>
<td>7/8 yrs</td>
<td>nouns, some verbs &amp; adj.</td>
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<tr>
<td>17 yrs.</td>
<td>80,000</td>
<td>17 yrs.</td>
<td>&lt; 3rd grade</td>
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</tbody>
</table>

Potential Language Delays Based on Levels of Hearing Loss

- 15-26 dB loss
  - 1.2 year delay

- 27- 40 dB loss
  - 2.0 year delay

- 41-55 dB loss
  - 2.9 year delay

- 56-70 dB loss
  - 3.5 + year delay

Language is NOT taught; it is acquired.

Comments made by Teachers...

- “I don’t think she needs the speakers, she hears me fine.”
- “I have a loud voice...I don’t need the microphone.”
- “She just has selective hearing and hears when she wants to.”
- “He hears me all the time when I call his name and I don’t have a microphone on.”
Classroom noise

In a typical classroom...the noise levels can reduce the student’s ability to understand 60% or as low as 27% without appropriate acoustics (ex. carpet, etc.)

Why isn’t speech/lipreading enough?

- 30% to 35% of English sounds are visible
- 13 - 15 speech movement per seconds, eyes can detect only 8 – 9 movements
- Language required
- 23% of hard of hearing people become effective lip readers—these are children
- Fatigue factor—concentration!

The Why’s

Why can’t I just talk louder?

- It seems like talking louder would help...however, studies show that talking louder only increases vowel energy and not consonant energy.
- Ironically, yelling increases audibility but not intelligibility.
- They need speech 10 times louder than the background noise.

Social & Emotional Issues that can occur with Children with Hearing Loss

Students with hearing loss in the regular classroom may exhibit one or more of the following traits.

1. Hesitant to speak.
2. Pretends to understand when he/she does not understand.
3. Does not ask questions to help his/her understanding.
4. Needs to watch other students to understand instructions.
5. Is not aware of what is happening or how to respond in social situations.
6. Does not understand why he/she is being disciplined.
7. Does not pick up social courtesies, graces or tact immediately.
8. Takes the fact that he/she has a hearing loss.
9. Hasn’t worn his/her hearing aid.
10. Is withdrawn.
11. Is overly aggressive.
12. Has fewer or younger friends.
13. Has family problems.

Understanding the Student with Hearing Loss

1. Because speech reading is fatiguing, students with hearing loss may have difficulty holding their attention for long periods.
2. Students, including deaf and hard of hearing, can learn to look attentive and appear to understand when they do not. Ask direct questions.
3. It is better for understanding, for other persons to be in close proximity to the deaf and hard of hearing student.
4. Speak to the student and call his/her name for attention. Touching or tapping the arm should not be any more necessary than with other students.
5. Remind hearing students to speak in complete sentences, to enunciate clearly and to face in the direction of the student with hearing loss.
6. Students with hearing loss need to learn any “in” expressions or words used by hearing students.

Final Thoughts...

- Children with hearing loss who have additional disabilities have their own needs but can learn to communicate and experience success academically.
- Parents and professional may have to adjust their expectations while maintaining high standards for learning.
- Children who are deaf will succeed on their own terms.
- Interprofessional collaboration and teaming is essential to meet the various needs of most children who are deaf.
- We seem to be seeing an increase in children who are deaf+; we need to be prepared to meet the needs of the family and the child.
Thank You for Listening!

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