Special Populations:
An Audiologic Approach to Managing Children with Craniofacial Anomalies

Brittney Sprouse, Au.D.
Pediatric Audiologist

2016 Early Hearing Detection & Intervention Conference
Course Objectives:

• Discuss the importance of early audiologic monitoring of children with craniofacial anomalies.

• Evaluate audiologic protocols and recommended guidelines for the management of children with craniofacial anomalies.

• Examine audiologic data collected from children diagnosed with Pierre Robin Sequence (site study).
Cleft Craniofacial Anomalies:

- **Cleft Lip and Cleft Palate**
  - Can occur simultaneously or separately
  - Unilateral or bilateral
  - Occurs in approximately 1 in every 600 births
    - CDC recently estimated:
      - 2,650 born with cleft palate
      - 4,440 born with a cleft lip with or without a cleft palate
  - Audiologic Implication:
    - Susceptible to recurrent hearing loss due to middle ear fluid
Non-Cleft Craniofacial Anomalies:

- Occurs in approximately one in every 1600 births

Microtia:

- Abnormal pinna (small/malformed external ear)
- Four types
  - Ranges from Type 1 (mild) to Type 4 (severe)
- Unilateral (most common) or bilateral
- Occurs in approximately 1–5 in every 10,000 births
- Audiologic Implication:
  - Conductive hearing loss
Non-Cleft Craniofacial Anomalies:

- **Atresia:**
  - Absence or closure of the external ear canal
    - Almost always accompanied by abnormalities of the middle ear bones
  - Unilateral (most common) or bilateral
  - Occurs in approximately 1 in every 10,000–20,000 births
  - Audiologic Implication:
    - Conductive hearing loss
Non-Cleft Craniofacial Anomalies:

- **Ear tags/pits:**
  - Located in front of the ear
  - Unilateral or bilateral / multiple or solitary
  - Occurs in approximately 5-10 in every 1000 births
  - Audiologic Implication:
    - Can be indicative of a syndrome or structural abnormality associated with hearing loss
Non-Cleft Craniofacial Anomalies:

- Malformed Eyes:
  - Hypertelorism: Widely spaced eyes
  - Hypotelorism: Closely spaced eyes
  - Coloboma: Gap in the structure of the eye; can affect the eyelid, iris, retina, or optic nerve
  - Microphthalmia: Small eye globe
  - Anophthalmia: Absence of the eye globe
  - Audiologic Implication: Does not impact hearing; can be indicative of a disease or syndrome associated with hearing loss
Non-Cleft Craniofacial Anomalies:

• Hemifacial Microsomia:
  – The lower half of one side of the face is underdeveloped
  – Varies in severity
    • Unilateral or bilateral
    • Always includes maldevelopment of the ear and the mandible
  – 2nd most common facial anomaly after cleft lip/cleft palate
  – Occurs in approximately 1 in every 3,500–4,500 births
  – Audiologic Implication:
    • Degree of hearing loss depends on the structures of the ear involved
Non-Cleft Craniofacial Anomalies:

- **Craniosynostosis:**
  - Premature fusion of the sutures of the skull bones
  - Can affect one or more of the joints in the skull
  - Often occurs in isolation
    - Associated with a syndrome in 15–40% of patients
  - Occurs in approximately in 1 in every 2,000 births
  - Audiologic Implication:
    - May result in middle and/or inner ear abnormalities
    - Often the syndrome it’s associated with involves hearing loss
Early Hearing Detection and Intervention (EHDI):

• EHDI Components:
  - Newborn Hearing Screening
  - Early Childhood Hearing Screening
  - Diagnostic Audiology
  - Early Intervention
  - Family Support and Partnership
  - Medical Home
  - Data Management
  - Financing & Reimbursements
  - Program Evaluation

• Specialized Populations and EHDI Components:
  - Key component in a diagnostic audiological evaluation of infants and children
  - Risk indicators associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood
    - #5: Craniofacial anomalies including those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
Early Hearing Detection and Intervention (EHDI): 

• **1-3-6 Model**
  - Hearing screening on all infants by **1 month of age**
  - Diagnosis by **3 months of age**
  - Medical, educational, and audiologic intervention **by 6 months of age** to maximize developmental, educational, and communication outcomes

• **Key Program Goal**
  - Provide support and education for families about the importance of detection and treatment of newborn hearing loss
Early Hearing Detection and Intervention (EHDI):

• Craniofacial Anomalies and the 1-3-6 Model
  – How is the 1-3-6 model impacted by a diagnosis of a craniofacial anomaly?
  – How is early audiologic monitoring and management affected?
  – What is the role of the audiologist in this model?
  – How do we implement the components of EHDI?
• Craniofacial Clinic Multidisciplinary Team
  – Plastic and Reconstructive Surgery
  – Neurosurgery *
  – Genetics
  – Otolaryngologist (ENT)
  – Neuroradiology *
  – Ophthalmology *
  – Orthodontics
  – Audiology
  – Speech
  – Social Work *
  – Nurse
  – Child Life *
  – Team Coordinator
University of Chicago Medicine Comer Children’s

- Audiologic Protocol
  - What role does audiology play on the craniofacial clinic team?
  - What was being implemented?
  - What areas could be improved upon?

Searching for Best Practice
Guidelines And Recommendations:

• Audiologic Guidelines:
  - What are the current audiologic guidelines and recommendations?
The American Cleft Palate-Craniofacial Association:

- ACPA Audiologic Guidelines:
  - Recommend individual ear hearing results by 3 months of age
BC Children’s Hospital (2012):

• Current Programs:
  • Three Cleft Palate programs in British Columbia (BC) Canada

• Audiologic guidelines:
  • Wide variability of recommended follow-up
  • No distinct Audiologic protocol
  • ACPA guideline was not being met across BC

• Outcome:
  • Developed the Audiology Clinical Practice Guideline: Cleft Palate/Craniofacial and Syndromic Patients (March 2012)
Audiology Clinical Practice Guideline: Cleft Palate (CP) / Craniofacial (CF) and Syndromic Patients

- Includes recommended care paths specific to the hearing loss risk for each group of children.
- Specific recommendations include:
  - All CP/CF and Syndromic infants in BC will receive a full diagnostic auditory brainstem response testing (ABR) prior to 3 months of age, regardless of their screening outcome.
  - Close periodic follow-up is required in groups at risk for recurrent middle ear disorders.
  - Close periodic follow-up for infants with a permanent hearing loss.
  - Use of high frequency tympanometry for infants under 6 months of age.
  - Assessment of ipsilateral acoustic reflexes using broadband noise stimuli.
Audiology Clinical Practice Guidelines:

- Service Description and Clinical Outcome:
  - Newborn hearing screening
  - Diagnostic ABR obtained prior to 3 months of age
    - Ear specific and frequency specific
    - Air Conduction (500, 2000, 4000 Hz) & Bone Conduction (500 and 1000 Hz)
  - Audiologic assessment at 9 months and annually from 2-6 years of age
    - Tympanometry
    - Ipsilateral acoustic reflexes
    - Otoacoustic emissions
    - Behavioral thresholds (500-6000 Hz bilaterally); age appropriate speech testing
    - If hearing remains unknown after 2 attempts, consider sedated ABR
    - Link with needed community resources
  - Hearing Loss (HL)
    - If a conductive HL is observed at any time, refer to PCP or ENT
    - Audiologic assessments following myringotomy and/or ventilating tubes (within 2-3 months)
    - Permanent HL: audiologic assessments every 6 months for at least one year until stable, then at the discretion of the audiologist for ongoing monitoring
• Audiologic Protocol
  
  — What role does audiology currently play on the craniofacial clinic team?
    • Increased visibility on the team
  
  — What is currently being implemented?
    • Protocol changes (consistent follow-up, further specific diagnostic testing)
    • Co-treatment of patients among other disciplines
    • Opportunities to enhance EHDI at our facility
  
  — What additional areas could be improved upon?
    • Review of outcomes since making changes in protocol
    • Continue to improve the implementation of the 1-3-6 model
    • Continue to reduce lost to follow-up
Craniofacial Syndrome—Pierre Robin Sequence:

- Pierre Robin Syndrome now known as a Sequence
- A set of abnormalities affecting the head and face consisting of:
  - Micrognathia (Small lower jaw/mandible)
  - Glossoptosis (Tongue placed farther back than normal)
  - Cleft palate
  - Respiratory obstruction
- The most common anatomic deviation associated with a non-isolated cleft
- Occurs in approximately in 1 in every 8,500–14,000 births (higher occurrence in twins)
- Can occur in conjunction with other syndromes
  - Stickler syndrome (10–30 %)
  - Velocardiofacial syndrome (10%)
Pierre Robin Sequence Study:

- **Overview of the study:**
  - Retrospective study
  - Multidisciplinary study
  - 34 patients (PRS and non-cleft patients)

- **Audiology:**
  - UNHS screening results
  - Follow-up diagnostic test results
  - Treatment and intervention
Pierre Robin Sequence Study:

**PRS STUDY**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>34</td>
</tr>
<tr>
<td>PRS</td>
<td>19</td>
</tr>
<tr>
<td>Other syndrome</td>
<td>5</td>
</tr>
<tr>
<td>Passed UNHS</td>
<td>8</td>
</tr>
<tr>
<td>SNHL</td>
<td>4</td>
</tr>
<tr>
<td>Mixed HL</td>
<td>2</td>
</tr>
<tr>
<td>Permanent CHL</td>
<td>2</td>
</tr>
<tr>
<td>Hx of OM/tubes</td>
<td>15</td>
</tr>
</tbody>
</table>
THANK YOU!

Contact Information:

brittney.sprouse@uchospitals.edu