Defining UHL/SSD

- One ear with normal hearing (15 dB PTA) and one ear with a severe to profound loss OR any degree of permanent hearing loss with word recognition scores ≤ 60%

UHL - Prevalence

- Unilateral hearing loss, single sided deafness
- 0.1 – 5 % of school aged children suffer from UHL / SSD
- 0.04 – 3.4 % of infants at birth
  - Approximately 1/3 of children identified during NHS have a UHL
- Large variability due to different methods applied in the different studies
- Open question: how many patients with UHL / SSD use any form of hearing aids?
  - Probably very few...

Why we should know about UHL

- 25% - Hearing loss progresses to bilateral (Colorado Pilot, 2002)
- Increased learning difficulties
  - 13-54% receive special education services (Bess, Klee & Culbertson, 1986; Oyler, Oyler, & Matkin, 1988; English & Church, 1999)
  - 36% on IEPs (Colorado Department of Education, 2002)
- Provide monitoring and/or intervention to address educational problems
Consequences of UHL: Psycho-social consequences

Qualitative Study: Focus groups
- Children: focus groups with children with UHL and their parents
- Children felt annoyed, uncomfortable and difficult when addressed on the poorer ear side
  - Pretended to understand conversations especially in noisy places to not be embarrassed
- Parents concerned about children not understanding teachers well
  - Learning spelling words
  - Classroom instructions
  - During conversations

Quantitative Study: PedsQL
- 85 children and their parents, out of which 24 normal hearing children, 32 UHL, 29 BHL
  - Children with UHL reported poorer QoL than NH children although on most scales but no statistically significant difference was found
- Specific questionnaire addressing the needs of UHL was developed and tested with 35 children with normal hearing, 35 with UHL and 45 with BHL
  - No difference in rating between children with BHL and UHL!
  - There is a need to provide a solution for these children

Opportunity

1. Examine the literature and answer the following questions:
   a) How do children with UHL/SSD compare to their normal-hearing peers?
   b) What are the effects of UHL/SSD?
   c) Does amplification improve auditory access for patients with UHL/SSD?
   d) Does amplification benefit patients with UHL/SSD?
   e) Can we define those who are at greater risk?
2. Establish a specialty team
3. Develop a testing protocol utilizing functional outcome measures
4. Design a pilot study to evaluate outcomes and contribute to the evidence
5. Generate an evidence-based clinical pathway for the treatment of childhood UHL
Development of SSD Specialty Team

- Team of 5 audiologists:
  - Evaluations and treatment

- Team meets bi-monthly to discuss:
  - Discuss study progress and barriers
  - Issues related to devices: hearing aid retention, verification, programming, updates, education and concerns; new developments
  - Managing unique populations including infants, toddlers and patients with developmental delays
  - Feedback from families
  - On-going education for audiology, ENT, speech pathology, educators, etc.

Evaluation Protocol

Data Collection:
- History
- Hearing test
- BKB-SIN
- Functional outcome measures

Discussion:
- Amplification Options
- Parent Questions
- Parent motivation and expectations

Referrals:
- Speech and AR

1. LittleEars (Med-El) – infants
2. Peach (Ching and Hill) – until~6 years
3. HEAR-QL (Judith Lieu) – ≥ 7 years
4. PBQ (modified-Eby) – (Perceived Benefits questionnaire)
**Evaluation Protocol**

**Data Collection:**
- History
- Hearing test
- BKB-SIN
- Functional outcome measures

**Discussion:**
- Amplification Options
- Parent Questions
- Parent motivation and expectations

**Referrals:**
- Speech and AR

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**Additional Considerations**

- Is the family interested in surgical options?
- Any hearing loss in better ear?
- Is hearing loss etiology known to be progressive?
- Does your child have any developmental challenges?
- Does your child have chronic ear infections or drainage?
- Do they participate in activities – what type?
- Size of ear canals – occlusion with CROS?
- Does your child have any sensory issues?
- How does your child handle loud environments?

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**Opportunity**

1. Examine the literature and answer the following questions:
   1. How do children with UHL/SSD compare to their normal-hearing peers?
   2. What are the effects of UHL/SSD?
   3. Does amplification improve auditory access for patients with UHL/SSD?
   4. Does amplification benefit patients with UHL/SSD?
   5. Can we define those who are at greater risk?
2. Establish a specialty team
3. Develop a testing protocol utilizing functional outcome measures
4. Design a pilot study to evaluate outcomes and contribute to the evidence
5. Generate an evidence-based clinical pathway for the treatment of childhood UHL
Pilot Study:

**Subjects** – school-aged children 7-17 years of age with one ear with normal hearing and one ear with a permanent profound hearing loss.

**Definitions:**
- Normal hearing - ≤ 20 dB HL 500Hz-8kHz
- Severe to Profound hearing loss - ≥ 80 dB HL and/or poorer than 60% word recognition scores

**Methods** – Follow SSD protocol and obtain appropriate consent forms from interested participants.

Flowchart 1 – Pre Treatment

**Referral Source**

- **New Private Patient**
  - Diagnosis of SSD
  - Counsel family
  - Refer to ENT for sensorineural work-up
  - Give family UHL tip sheets
  - Recommend a SSD evaluation

- **New ENT Patient**
  - Diagnosis of SSD
  - Counsel family
  - Sensorineural work-up
  - Give family UHL tip sheets
  - Recommend a SSD Evaluation

- **Established patient with SSD**
  - Recommend a SSD evaluation

Flowchart 2

**Test Battery:**

1. Hearing Test (optional at 1 month)
2. Amplification verification – when appropriate* (CROS aids)
3. Speech in noise test (BKB-SIN)
4. Aided testing with speech and tonal stimuli using insert plug and ear muff on good ear
5. PBQ-modified and HEAR-QL

Flowchart 3

**No Treatment follow up at 6 months and 12 months**

**No Treatment Group**

**Test Battery:**

1. Hearing Test
2. Speech in noise testing (BKB-SIN)
3. PBQ and HEAR QL
4. Discussion of current issues and concerns
5. Discuss and consider any new treatment options
### Perceived Benefits Questionnaire - modified

<table>
<thead>
<tr>
<th>Question</th>
<th>Never</th>
<th>Almost Never</th>
<th>Sometimes</th>
<th>Almost Always</th>
<th>Always</th>
<th>Improved</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. My child is frustrated when communicating (e.g., with friends, family, or teachers)</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>2</td>
<td>1</td>
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<tr>
<td>2. My child is easily distracted while reading or writing</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>2</td>
<td>1</td>
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<tr>
<td>3. My child has difficulty understanding written material</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>2</td>
<td>1</td>
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<td>4. My child has difficulty keeping up with their schoolwork</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>2</td>
<td>1</td>
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<tr>
<td>5. My child has difficulty understanding written material</td>
<td>5</td>
<td>4</td>
<td>3</td>
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<tr>
<td>6. My child has difficulty understanding spoken material</td>
<td>5</td>
<td>4</td>
<td>3</td>
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<tr>
<td>7. My child has difficulty understanding spoken material</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>2</td>
<td>1</td>
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<tr>
<td>8. My child has difficulty maintaining eye contact when reading</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>2</td>
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<tr>
<td>9. My child has difficulty maintaining eye contact when writing</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>2</td>
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<tr>
<td>10. My child has difficulty maintaining eye contact when speaking</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>2</td>
<td>1</td>
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</tbody>
</table>

### PBQ – Preliminary results

### Hearing Environments And Reflection on Quality of Life – Measurement for Children

<table>
<thead>
<tr>
<th>Environments</th>
<th>Never</th>
<th>Almost Never</th>
<th>Sometimes</th>
<th>Often</th>
<th>Almost Always</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Is it hard to hear your friends when you are playing?</td>
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<tr>
<td>2. Do you have a hard time hearing your friends at recess?</td>
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<td>3. Is it hard to hear in gym class (Physical Education, PE)?</td>
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<td>4. Do you think you have a harder time hearing than your friends in noisy places (restaurants, ball games, field trips, etc.)?</td>
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<td>5. Is it hard to hear at restaurants?</td>
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<td>6. Is it hard to hear in your classroom?</td>
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<td>7. Is it hard to hear in the cafeteria (lunch room)?</td>
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<td>8. Is it hard to hear your friends when playing outside?</td>
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<td>9. Is it hard to hear your family (mom, dad, brother, sister)?</td>
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<td>10. Is it hard to hear when someone whispers to you?</td>
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<td>11. Is it hard to hear when someone dials the phone?</td>
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<td>12. Is it hard to hear movies or TV because of your hearing?</td>
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</tbody>
</table>

### HEAR-QL Cont.

<table>
<thead>
<tr>
<th>Activities</th>
<th>Never</th>
<th>Almost Never</th>
<th>Sometimes</th>
<th>Often</th>
<th>Almost Always</th>
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</thead>
<tbody>
<tr>
<td>14. Do you play with fewer people because of your hearing?</td>
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<td>15. Do you not play with certain people outside of school because of your hearing?</td>
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<td>16. Do you play with friends or relatives less often than you want to because of your hearing?</td>
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<td>17. Do you go to parties less because of your hearing?</td>
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<tr>
<td>18. Do you play fewer sports or participate in fewer activities than your friends because of your hearing?</td>
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<tr>
<td>19. Do your parents not let you do certain things because of your hearing?</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Feelings</th>
<th>Never</th>
<th>Almost Never</th>
<th>Sometimes</th>
<th>Often</th>
<th>Almost Always</th>
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</thead>
<tbody>
<tr>
<td>20. Do you feel shy when meeting new people because of your hearing?</td>
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<td>21. Does your hearing loss make you feel different from everyone else?</td>
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<td>22. Does your hearing cause you to be nervous?</td>
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<tr>
<td>23. Does your hearing make you angry?</td>
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<td>24. Do you worry about your hearing loss getting worse?</td>
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<tr>
<td>25. If you can’t hear someone, do you have a hard time asking them to speak louder or repeat what they said?</td>
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<tr>
<td>26. Do you feel different from others because of your hearing?</td>
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</table>
HearQL – Preliminary results

Conclusions

- Children with UHL exhibit a range of perceived handicap
- For many, families and children notice challenges
- Technological solutions can very successfully mitigate those challenges in children
- No one size fits all
- It helps to have a protocol to collect and provide information so that families can make informed decisions about what solutions are best for them

Take Home :

- Take a chance on your patients!
- For many patients – something is better than nothing!
- Learning about the options available for today increases the opportunity for improved options in the future.
- Never trust that you know the right answer – keep questioning and exploring!

Upcoming Research on SSD at CCHMC

Study by Andrew Dimitrijevic
Assistant Professor, Communication Sciences Research Center
Study Purpose:
- The use of EEG to investigate hemispheric differences in cortical activation for patients with SSD
Hypothesis:
- It has been shown that adults demonstrate neural plasticity through bilateral activation in spite of SSD
- if kids with SSD who do not seem to perform well in the real world are unable to demonstrate the same contralateral activation
Methods:
- Look at left and right auditory cortical activations
Based on the following article:
Thank you for listening