SSD – indication and results of cochlear implantation, bone conduction devices and CROS hearing aids in children

S. Arndt, F. Hassepass, R. Laszig, R. Beck, A. Aschendorff, T. Wesarg

Department of Oto-Rhino-Laryngology and Implant Center Medical Center - University of Freiburg, Germany
2 Cases - both Single Sided Deaf
(6 months of age)

Diagnosis with NHS

Case 1:
- Unilateral sensorineural deafness
- Morphology of the pinna unremarkable

Case 2:
- Unilateral atresia of external ear canal
- Malformation of the pinna
- Conductive hearing loss in objective evaluation

different treatment methods
Common complaints of patients with unilateral deafness

- **70-93%**: high degree of difficulties in hearing in background noise
  (Coletti et al. 1988; Ruscetta et al. 2005; Priwin et al. 2007; Wie et al. 2010)

- **Difficulties in localization**
  (Wie et al. 2010, Hol et al. 2010, Flynn et al. 2010)

- **54-84%**: tinnitus
  (Quaranta et al. 2004; Priwin et al. 2007; Wie et al. 2010)

- **Frequent headaches, stress, fatigue**
  (Borton et al. 2010, Wie et al. 2010)

- **Social isolation, psychological problems**
  (Borton et al. 2010, Wie et al. 2010)
Special problems in children with unilateral deafness

- 12-41%: need of additional educational assistance
  (Bess & Tharpe 1986; Bovo et al. 1988)

- Increased need for speech therapy in comparison to their normal-hearing peers
  (Lieu et al., 2010, 2012)

- 22-35%: increased rate of grade failures
  (Bess and Tharpe 1988; Brockhauser et al. 1991; Cho Lieu et al. 2004)

- Low self-esteem and increased fatigue; negative affect on academic success of children
  (Kuppler 2013)
Treatment options
Rehabilitation in patients with SSD/AHL 1

Treatment options

- **Contralateral Routing of Signals ((Bi)CROS)-Hearing aids**
  
  A Rehabilitative Approach to the Problem of Unilateral Hearing Impairment: The Contralateral Routing of Signals (CROS)
  

  advantage: non invasive, no artefacts in MRI examination
  disadvantage: hearing aids on both ears, no binaural hearing, no prevention of unilateral auditory deprivation

- **Bone Conduction hearing Implants (BCI) percutaneous/ soft band**
  
  [The monaural pseudo-stereophonic hearing aid (BAHA) in unilateral total deafness: a study of 29 patients].
  

  advantage: no/ minimally invasive, minimal artefacts in MRI
  disadvantage: wound infection, no binaural hearing, no prevention of unilateral auditory deprivation
Rehabilitation in patients with SSD/AHL 2

Treatment options

- **FM systems**
  - advantage: non invasive, provide an increased SNR in educational settings, no artefacts in MRI examination
  - disadvantage: modus operandi is a big obstacle for application in everyday life, no binaural hearing, no prevention of unilateral auditory deprivation
Rehabilitation in patients with SSD/AHL 3

Treatment options

- **Cochlear Implant**

<table>
<thead>
<tr>
<th>Advantage</th>
<th>Disadvantage</th>
</tr>
</thead>
<tbody>
<tr>
<td>binaural hearing possible, intact skin (transcutaneous), no (further) deprivation of auditory pathway</td>
<td>invasive, rehabilitation, excessive artefact in MRI</td>
</tr>
</tbody>
</table>

Incapacitating unilateral tinnitus in single-sided deafness treated by cochlear implantation.

Comparison of pseudobinaural hearing to real binaural hearing rehabilitation after cochlear implantation in patients with unilateral deafness and tinnitus.
Results of different treatment options in adults
Unaided vs. CROS-test vs. BCI-test vs. CI (12 months)

37 adult SSD patients with acquired hearing loss
• with CROS and BCI significant superior results (blue brackets) vs. unaided in only one speech in noise condition
• CROS and BCI reduce SNR in one condition, no improvement in localization
Unaided vs. CROS-test vs. BCI-test vs. CI (12 months)

37 adult SSD patients with acquired hearing loss

- with CROS and BCI significant superior results (blue brackets) vs. unaided in only one speech in noise condition
- CROS and BCI reduce SNR in one condition, no improvement in localization
- with CI significant superior results (red brackets) in all SR conditions in noise compared to all treatment options and localization
- CI is the only option to increase the SNR in all conditions due to (partial) restitution of binaural hearing
Binaural benefits with conventional CROS HA?

n=7 adults; 12 months device experience; HSM sentence test, localization

- significant improvement of speech recognition in only one configuration representing the head shadow effect
- deterioration of speech SR when noise presented to poorer ear (n.s.)

Treatment option / CROS-HA
Binaural benefits with percutaneous bone conduction implants?

n=17 adults; 12 months device experience; HSM sentence test, localization

- head shadow effect: p<0.005
- summation effect: n.s.
- squelch effect: n.s.
- localization: p<0.05

- Significant improvement of speech recognition in one configuration representing the head shadow effect
- Improvement in localization

Treatment option / Bone conduction Implant
Results of different treatment options in children

CROS hearing: decrease or no improvement of speech comprehension in most conditions, difficult for the child during speech development

indicated in school aged children after speech development
Cochlear implantation in SSD-children
Freiburg study of CI treatment in SSD/AHL Children

Pre-examination in SSD

- Subjective (paed)audiometry comprising thresholds, SR of monosyllables, sentences in quiet and noise (if measurable in respect to age)
- Objective audiometry including ABR thresholds, ABR latencies, electrocochleography
- HR-CT and MRI
- Neuropediatric examination, if other developmental disorders are known or suspected

Extensive counselling with regard to

- Ramifications for daily life (localization, SR in noise)
- Availability of BCI /(BI)-CROS and other assistive technology
- Necessity for participation in a mandatory rehabilitation phase
Freiburg study of CI treatment in SSD/AHL Children

Audiometric tests

• Tests according to the children’s age difficult to compare the results
• Speech tests in children (Mainz -, Göttingen speech test, Oldenburg sentence test for children: OlKiSa)
• If possible: OlSa for adults in 3 test conditions with background noise
• Categories of auditory performance (CAP; Archbold et al. 1998)
• CI-usage time indicates potential benefit in children.

Subjective evaluation

• For children younger than 9 years: Child self-report and a parent report using the adapted SSQ-version for children and parents (Galvin 2007)
• For children older than 9 years: adult version of the SSQ originally developed by Gatehouse and Noble
Cause of deafness of all SSD/AHL pre-Evaluations
328 patients: 269 adults; 59 children

- **Adults**
  - labyrinthitis; 27
  - sudden hearing loss; n = 103
  - fracture; 13
  - ear surgery; 21
  - Menière; 10
  - childhood; 29

- **Children with acquired SSD/AHL n=28**
  - EVA; n=6
  - unknown; 1
  - Labyrinthitis; 4
  - premature; 2
  - mumps; 3
  - meningitis; 2
  - fracture; 3

- **Children with congenital SSD/AHL n=32**
  - **cochlear nerve hypo-/aplasia; n = 18**
  - EVA; 1
  - unknown; 7
  - CMV-infection; 4

---

- Treatment options / CI in children

Arndt et al. 2015
## Therapeutic decisions after preevaluation

<table>
<thead>
<tr>
<th></th>
<th>congenital (n=32)</th>
<th>acquired (n=28)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cochlear Implant (CI feasible)</td>
<td>11 (12)</td>
<td>24 (28)</td>
</tr>
<tr>
<td>CROS-HA</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Bonebridge</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Baha</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>iSense (FM receiver)</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>nothing</td>
<td>14</td>
<td></td>
</tr>
</tbody>
</table>
### Cochlear implant indications in SSD/AHL-children

<table>
<thead>
<tr>
<th></th>
<th>Congenital SSD/AHL (PE: n=32)</th>
<th>Acquired SSD/AHL (PE: n=28)</th>
</tr>
</thead>
<tbody>
<tr>
<td>number of indicated CI's</td>
<td>11</td>
<td>28</td>
</tr>
<tr>
<td>number of CI surgeries</td>
<td>10</td>
<td>24</td>
</tr>
<tr>
<td>age at surgery/yrs</td>
<td>5.7 (1.5 – 13.8)</td>
<td>11.1 (5.7-17.9)</td>
</tr>
<tr>
<td>duration of deafness/yrs</td>
<td>5.7 (1.5 – 13.8)</td>
<td>2.4 (0.3-10)</td>
</tr>
</tbody>
</table>

- Treatment options / CI in children

Arndt et al. 2015
Results: **acquired SSD/AHL in children**

CI-OP: 18 children; results 12 months post-activation: OLSa (n=10); OlKiSa (n=4)

- results comparable to adult CI patients with acquired SSD
Results: **acquired SSD/AHL in children**

SSQ results: adapted version for children and parents (Galvin 2007): 12 months post-activation (n=10)

- significant improvement in all subscores of child and parent version of SSQ
- **Use of CI all day long in all children!!**
Different situation in congenital SSD children

- The development of auditory pathways is determined by neural plasticity and needs auditory input
- Asymmetric hearing influences brainstem development and poses hurdles for the realization of true binaural processing

It is still unclear in congenital SSD
- Whether the normal hearing ear sustains the formation of pathways at least partially
- Or if the extensive asymmetry accelerates lateral inhibition and reduces the window for successful intervention

Assumption
- Age for cochlear implantation in children with asymmetric hearing loss would not differ much from those with bilateral hearing loss
# Overview: congenital SSD in children

<table>
<thead>
<tr>
<th>Child</th>
<th>age @ diagnosis (year; month)</th>
<th>age @ surgery (year; month)</th>
<th>Etiology</th>
<th>BERA threshold (SSD ear, dB)</th>
<th>BERA threshold (better ear, dB)</th>
<th>education</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>NHS 1;10</td>
<td>1;10</td>
<td>CMV</td>
<td>&gt;90</td>
<td>20</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>NHS 1;9</td>
<td>1;9</td>
<td>unknown</td>
<td>&gt;90</td>
<td>20</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>NHS 3;0</td>
<td>3;0</td>
<td>hypoxia</td>
<td>&gt;90</td>
<td>30</td>
<td>kindergarten</td>
</tr>
<tr>
<td>4</td>
<td>2;6</td>
<td>3;2</td>
<td>CMV</td>
<td>&gt;90</td>
<td>20</td>
<td>kindergarten</td>
</tr>
<tr>
<td>5</td>
<td>NHS 4;0</td>
<td>4;0</td>
<td>unknown</td>
<td>&gt;90</td>
<td>20</td>
<td>kindergarten</td>
</tr>
<tr>
<td>6</td>
<td>NHS 4;4</td>
<td>4;4</td>
<td>CMV</td>
<td>&gt;90</td>
<td>20</td>
<td>kindergarten</td>
</tr>
<tr>
<td>7</td>
<td>NHS 4;8</td>
<td>4;8</td>
<td>EVA</td>
<td>&gt;90</td>
<td>20</td>
<td>kindergarten</td>
</tr>
<tr>
<td>8</td>
<td>NHS 5;2</td>
<td>5;2</td>
<td>unknown</td>
<td>&gt;90</td>
<td>20</td>
<td>kindergarten</td>
</tr>
<tr>
<td>9</td>
<td>4;5</td>
<td>6;8</td>
<td>ototoxic</td>
<td>&gt;90</td>
<td>20</td>
<td>school</td>
</tr>
<tr>
<td>10</td>
<td>0;4</td>
<td>13;10</td>
<td>CMV</td>
<td>&gt;90</td>
<td>20</td>
<td>school</td>
</tr>
</tbody>
</table>
## Results

### CI-usage; speech comprehension; subjective evaluation

<table>
<thead>
<tr>
<th>Child</th>
<th>Age (y;m)</th>
<th>Time after 1(^{th}) fitting (y;m)</th>
<th>CI-usage</th>
<th>CAP</th>
<th>Speech test @ 65 dB</th>
<th>Speech comprehension with CI only</th>
<th>Subjective evaluation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2;1</td>
<td>1;0</td>
<td>all day</td>
<td>-</td>
<td>too young</td>
<td>-</td>
<td>Delayed general and motor development</td>
</tr>
<tr>
<td>2</td>
<td>2;7</td>
<td>1,0</td>
<td>all day</td>
<td>3</td>
<td>Mainzer I</td>
<td>100%</td>
<td>Repeats words and imitates sound heard by CI only</td>
</tr>
<tr>
<td>3</td>
<td>3;4</td>
<td>0;6</td>
<td>all day</td>
<td>-</td>
<td>Mainzer I</td>
<td>60%</td>
<td>Repositions transmission coil, shows localization</td>
</tr>
<tr>
<td>4</td>
<td>5;1</td>
<td>2;0</td>
<td>all day</td>
<td>5</td>
<td>Göttinger II</td>
<td>80%</td>
<td>Describes hearing with CI similar to unimpeded ear</td>
</tr>
<tr>
<td>5</td>
<td>5;0</td>
<td>1;0</td>
<td>all day</td>
<td>5</td>
<td>Göttinger II</td>
<td>70%</td>
<td>Asks for speech processor, immediate feedback, when batteries are empty</td>
</tr>
<tr>
<td>6</td>
<td>7;8</td>
<td>3;6</td>
<td>school</td>
<td>3</td>
<td>Göttinger II</td>
<td>0%</td>
<td>No speech discrimination with CI only, tendency towards non-use</td>
</tr>
<tr>
<td>7</td>
<td>5;2</td>
<td>0;6</td>
<td>all day</td>
<td>5</td>
<td>Göttinger II</td>
<td>0%</td>
<td>Improved speech discrimination, localization improved</td>
</tr>
<tr>
<td>8</td>
<td>5;9</td>
<td>0;6</td>
<td>all day</td>
<td>5</td>
<td>Göttinger II</td>
<td>0%</td>
<td>Mainstream school planned</td>
</tr>
<tr>
<td>9</td>
<td>7;5</td>
<td>0;6</td>
<td>all day</td>
<td>5</td>
<td>Freiburg monosyll.</td>
<td>20%</td>
<td>Checks status, changes batteries</td>
</tr>
<tr>
<td>10</td>
<td>15;2</td>
<td>2;0</td>
<td>school</td>
<td>5</td>
<td>Freiburger monosyll.</td>
<td>0%</td>
<td>Focus on contralateral side, tendency towards non-use</td>
</tr>
</tbody>
</table>
Results – children > 4 years at implantation

SSQ scale for children and parents (Galvin 2007)

Tendency towards non-use

Speech

Spatial

Quality

<table>
<thead>
<tr>
<th></th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kind 6</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eltern</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th></th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kind 6</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eltern</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th></th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
<th>präop</th>
<th>postop</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kind 10</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eltern</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Summary: Treatment of SSD/AHL in Children

Acquired SSD/AHL
• Comparable results to adult CI patients with acquired SSD/AHL

CI: Favorable treatment option for this special group of SSD children, as it is the only opportunity to restore binaural hearing abilities

Congenital SSD/AHL
• Anatomy should be evaluated with caution: app. 60% aplasia of the auditory nerve
• Limited results in two children, most likely due to late implantation or as a result of their CMV infection
• Promising results in children with implantation before age of 4 yrs
• Outcomes of auditory habilitation with CI vary significantly

Intensive counseling of parents and patients is indispensable!

• CROS hearing: decrease of SNR in most conditions, difficult for the child during speech development- maybe wise later FM in Kindergarten or preschool?