Cochlear Implantation in Pediatric Single Sided Deafness

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“OFF-LABEL”

Single Sided Deafness is an FDA “OFF-LABEL” indication for cochlear implantation
Effects of Single Sided Deafness (SSD)

Keller & Bundy (1980)
*Child Care Health Dev*

*Pediatrics*

Bess, Culbertson & Davis (1984)
*Ear Hearing*

Culbertson & Gilbert (1986)
*Ear Hearing*

**Average Results**

Math = 30th percentile
Language = 25th percentile
Social = 32nd percentile

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<tr>
<th>Subject</th>
<th>Normal Hearing</th>
<th>Unilateral Hearing Loss</th>
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<tbody>
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<td>Math</td>
<td>30th percentile</td>
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Methods

• Single Side Deafness (SSD)
  • Better Ear – PTA < 30 dB
  • Affected Ear – PTA > 85 dB

• Squelch Effect
  • LNT – speech signal in front, noise signal behind

• Binaural Summation
  • AzBio – speech and noise from same speaker in front

• Presentation
  • 50 dB
  • Recorded voice and multitalker babble and/or speech noise

• Condition Evaluated
  • Normal ear only
  • Binaural condition = normal ear + cochlear implant
Who are the patients?

**Patient 1**
- Congenital SSD
- Normal MRI
- Speech delay at 18 mo – no improvement after therapy
- Implant 29 mo/o

**Patient 2**
- Chronic middle ear disease & tympanoplasty at 6 y/o on SSD side
- Delayed Kindergarten: Once enrolled school recommended to repeat
- ADHD, disruptive behavior disorder & expressive language disorder
- Diagnosed with SSD at 7y/o & a mild CHL in the contralateral ear.
- Hearing aid and FM at school
- MRI revealed bilateral EVA
- October parents counseled that he would repeat 1st grade
- Cochlear Implant in Dec. of 1st grade at 7 y/o

**Patient 3**
- Adopted from Russia at 4y/o – prior history unknown
- Diagnosed with SSD at 6y/o
- Language testing was below the 3rd percentile in all areas. Only had spoken English for 2 years.
- Normal MRI
- FM used at school
- Behavior & attention problems
- Cochlear Implant 7 y/o

**Patient 4**
- CHARGE Syndrome
- Failed NBHS: Unilateral moderate SNHL – fit with hearing aid
- Speech regression 4 y/o
- 4y/o diagnosed with SSD & contralateral mild CHL
- Abnormal MRI consistent with CHARGE Syndrome
- Cochlear Implant 4 y/o – inserted through remnant of vestibule

**Patient 5**
- Passed NBHS
- Failed hearing screen – diagnosed with SSD at 4y/o
- Juvenile rheumatoid arthritis at 5y/o
- Kindergarten to present – Mixed language disorder-receptive and expressive
- Academic problems
- Social problems in noisy environments
- Normal MRI
- Cochlear Implant 12 y/o

**Patient 6**
- Congenital SSD
- Normal MRI
- Congenital hypothyroidism
- Diagnosed with articulation disorder at 25 mo/o & started speech.
- Abnormal MRI (bilateral cochlear dysplasia and areas of gliosis raising concern about possible congenital CMV)
- Cochlear Implant 34 mo/o

**Patient 7**
- Congenital SSD
- Normal MRI
- Maternal aunt with SSD who struggled academically and at work due to hearing loss.
- Presented at 4 y/o – mom wanted to restore hearing
- Cochlear implant 5 y/o
- At the time of initial activation – she repeatedly asked to have the implant turned up to the level of her normal hearing ear
Patient 1
Patient 2
Patient 3
Patient 4
Patient 5
Patient 6
Patient 7
AzBio
**Patient 1**
- Congenital SSD
- Implant 29 mo/o
- 3 mo implant use – language normal
- Currently 4 y/o – recent language testing

**Patient 2**
- Chronic middle ear disease & tympanoplasty at 6 y/o on SSD side
- Delayed Kindergarten: Once enrolled school recommended to repeat
- ADHD, disruptive behavior disorder & expressive language disorder
- October parents counseled that he would repeat 1st grade
- After implant successfully completed 1st grade – has done well in school since

**Patient 3**
- Adopted from Russia at 4y/o – prior history unknown
- Language testing was below the 3rd percentile in all areas. Only had spoken English for 2 years.
- Behavior & attention problems
- Parents and teachers report improvement in behaviors & school performance

**Patient 4**
- CHARGE Syndrome
- Speech regression 4 y/o
- WIPI
  - Binaural = 96%
  - Normal Ear = 92%
- Subjective speech improvement

**Patient 5**
- Passed NBHS
- Kindergarten to present – Mixed language disorder-receptive and expressive
- Academic problems
- Social problems in noisy environments
- Parent and teachers report improvement in academic performance

**Patient 6**
- Congenital SSD
- Diagnosed with articulation disorder at 25 mo/o & started speech.
- Abnormal MRI (bilateral cochlear dysplasia and areas of gliosis raising concern about possible congenital CMV)
- Objective speech perception measures improved
- Continued speech and language problems
- Progression of hearing loss in contralateral ear

**Patient 7**
- Congenital SSD
- Maternal aunt with SSD who struggled academically and at work due to hearing loss.
- Mom wanted to restore hearing
- At the time of initial activation – she repeatedly asked to have the implant turned up to the level of her normal hearing ear
- Star
Conclusion

• Children with SSD receive binaural benefit from cochlear implantation
  • Binaural squelch affect seen
  • Binaural summation seen
Single Sided Deafness is an FDA “OFF-LABEL” indication for cochlear implantation.