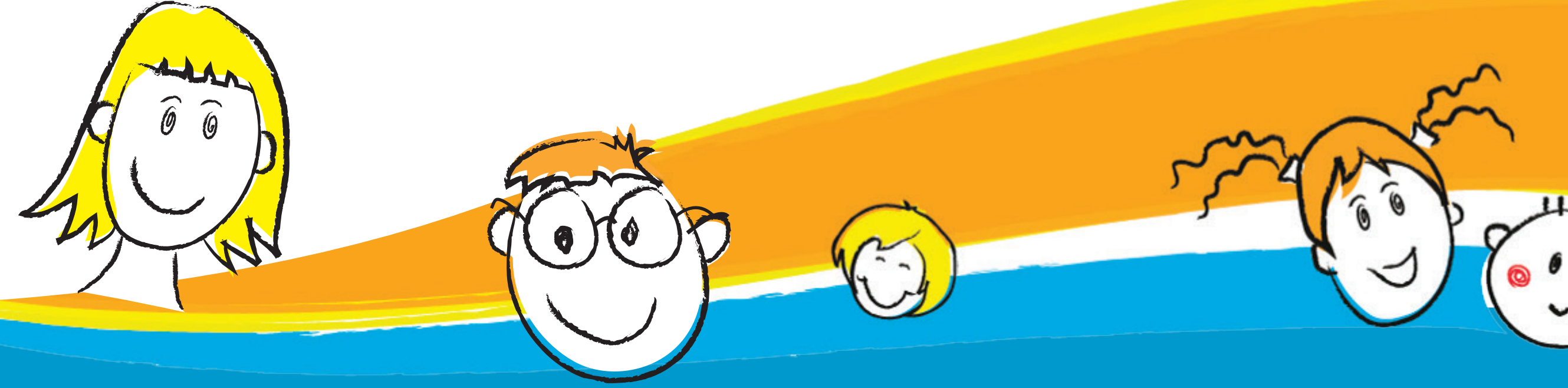


Managing Unilateral Hearing Loss (UHL) in Infants: Why one ear is not enough

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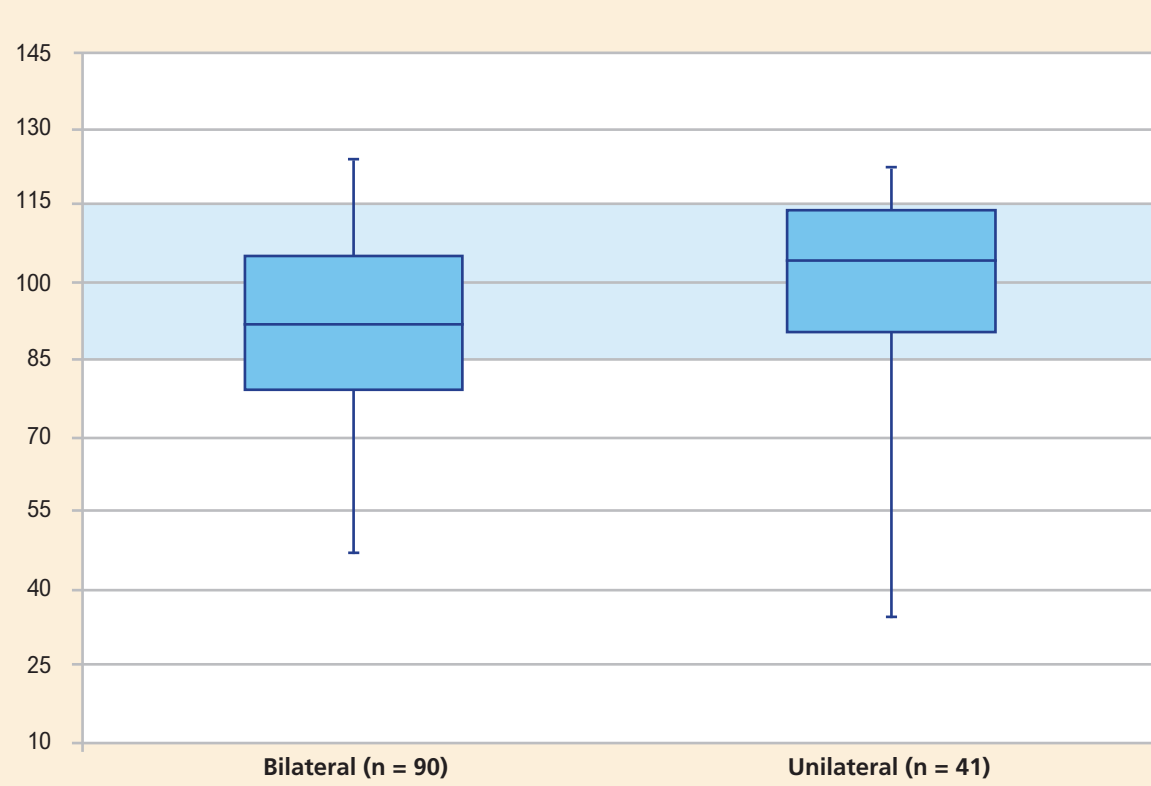
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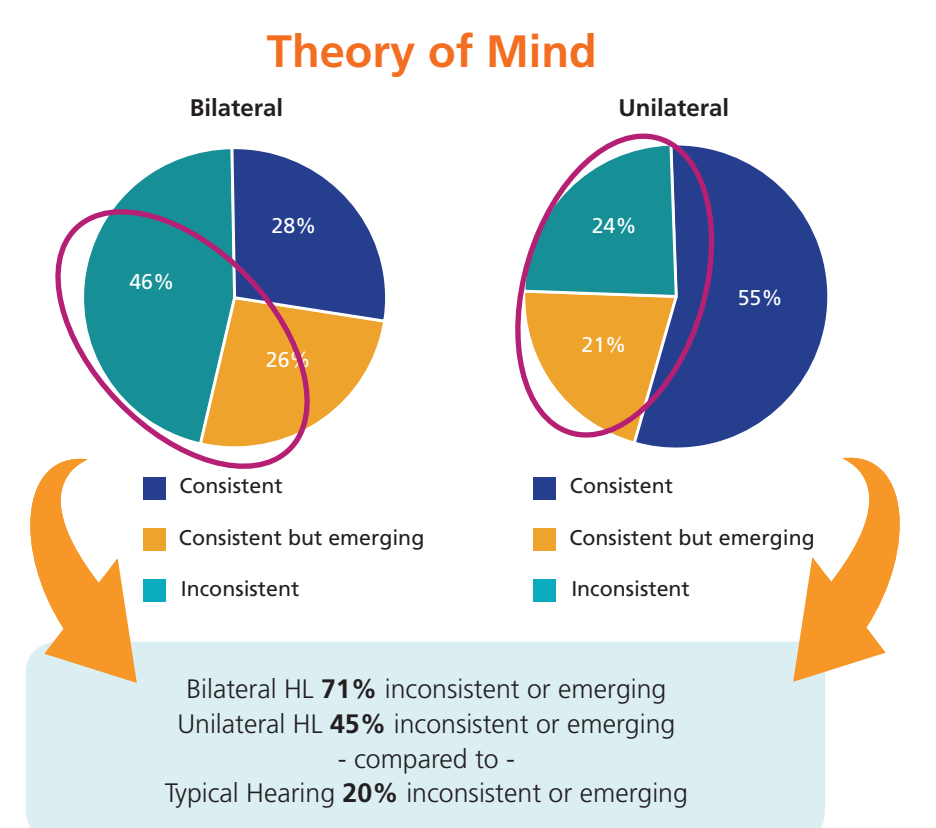
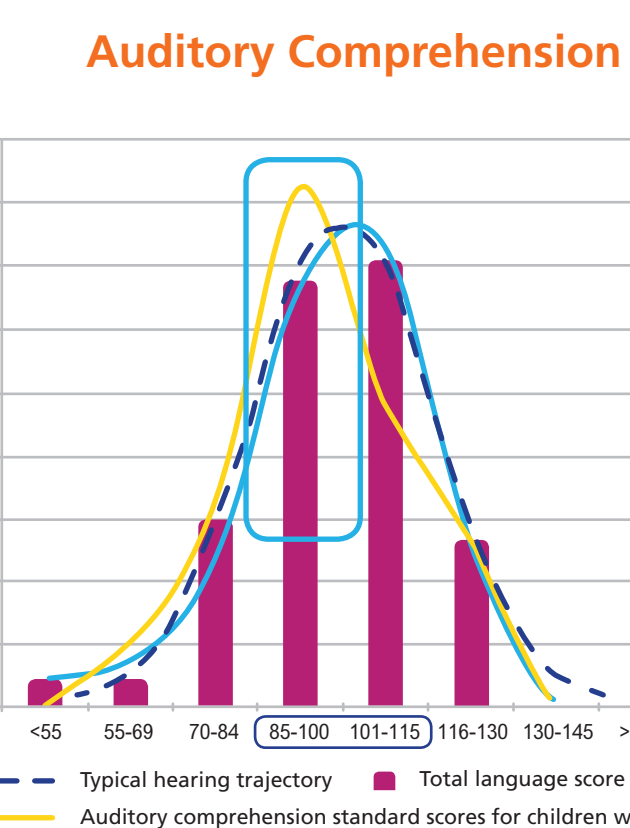
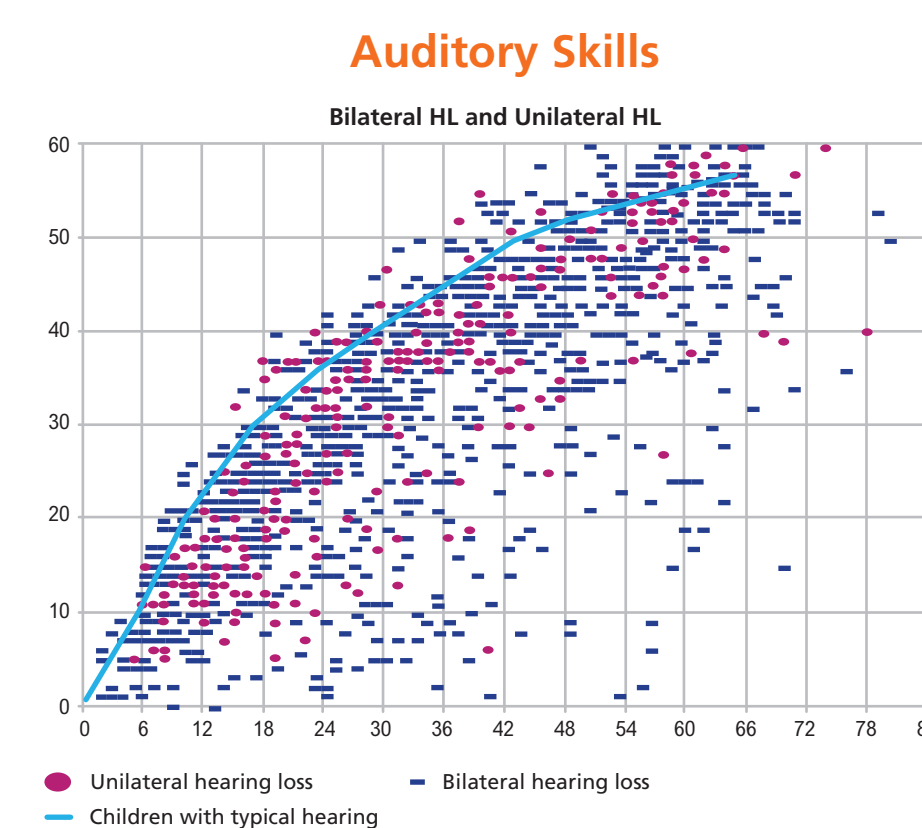
The Problem



Graduated language assessment results at 5 years indicate:
 - The median language results at 5 years indicate many children with UHL achieve language within the normal range (as with Bilateral children in the TSC program), HOWEVER
 - The lower quartile range for children with UHL is greater indicating the range of poor outcomes is worse than with bilateral hearing loss.

Parents are faced with a range of choices for their child with unilateral hearing loss.
 - The challenge for health professionals is knowing what advice to provide, and what is evidence based best practice recommendations for each child

Outcomes

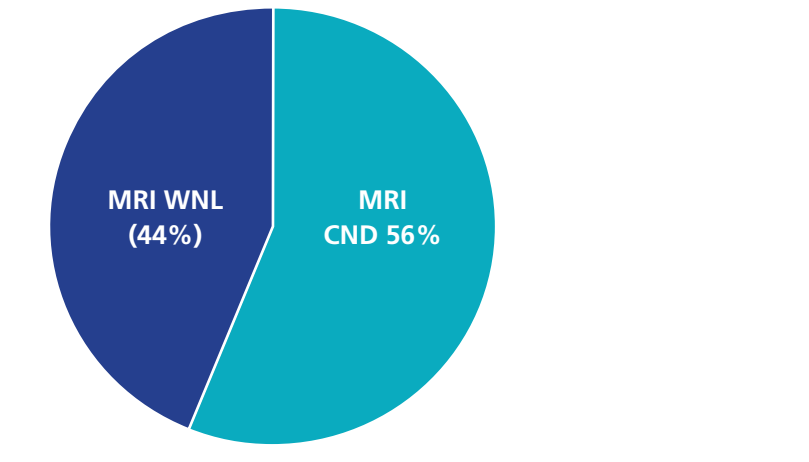
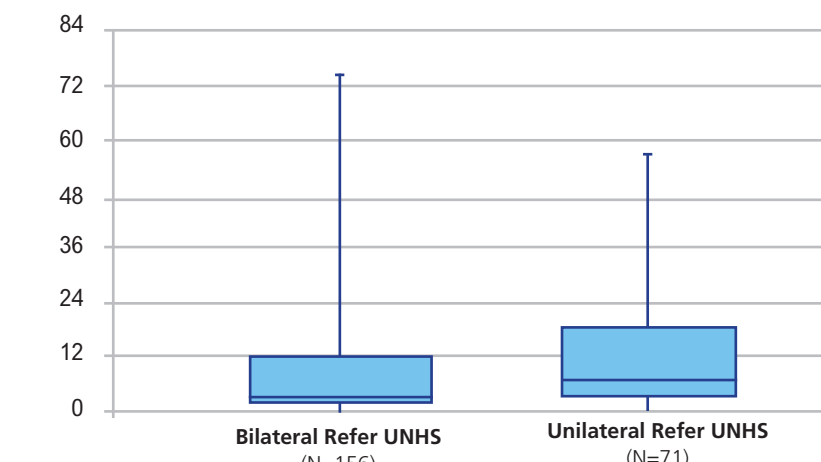
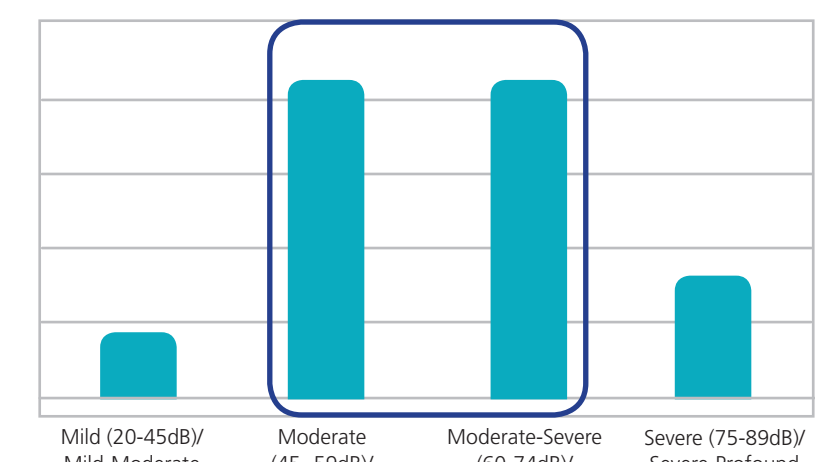
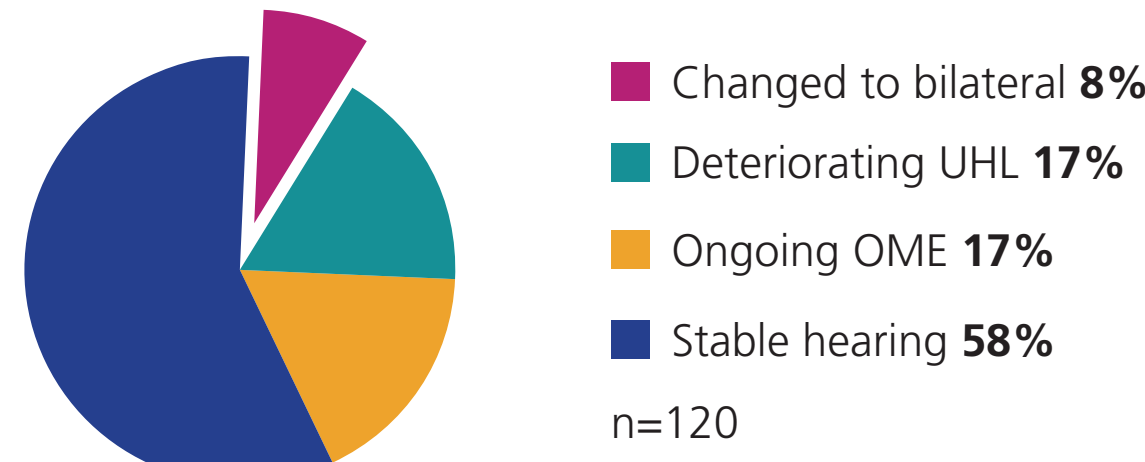
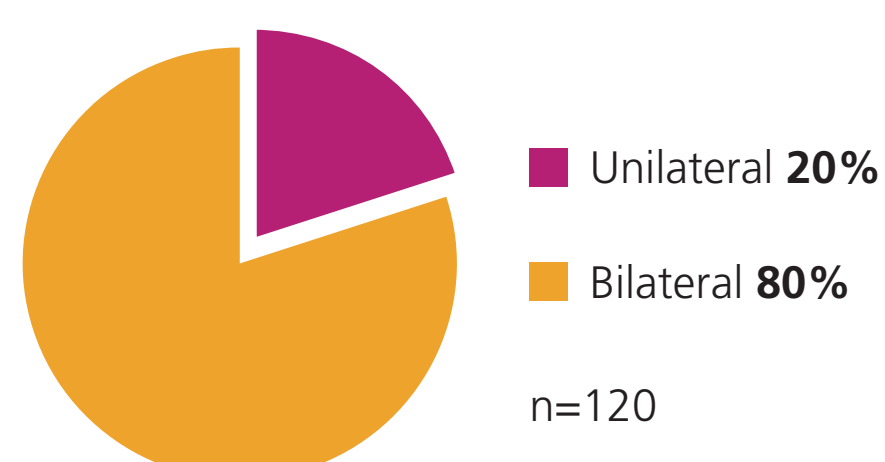


• Even though standardised language assessments may be normal, many children with UHL have poorer listening skills than children with typical hearing.
 • Listening with one ear may not be enough for these children to achieve their communicative potential

• Potential discrepancies become apparent once the standard score is broken into details
 • Skew of Auditory Comprehension scores for children with UHL lower than that of typical hearing

• Statistically significant amount of children with UHL have inconsistent or delayed patterns of theory of mind development than reported of typical hearing children

Demographics



• 20% children with UHL in TSC EI program (Incidence reported at 0.6:1,000 (WHO, 2009))
 • 92% referred for diagnosis at birth,
 • 10-22% will have a progressive bilateral loss

• 42% children with UHL had changing hearing levels
 • In line with reported figures of 37-58% deterioration of hearing threshold in that ear

• Majority of children fitted with hearing aid fall into moderate, mod-sev, mod-profound hearing loss levels

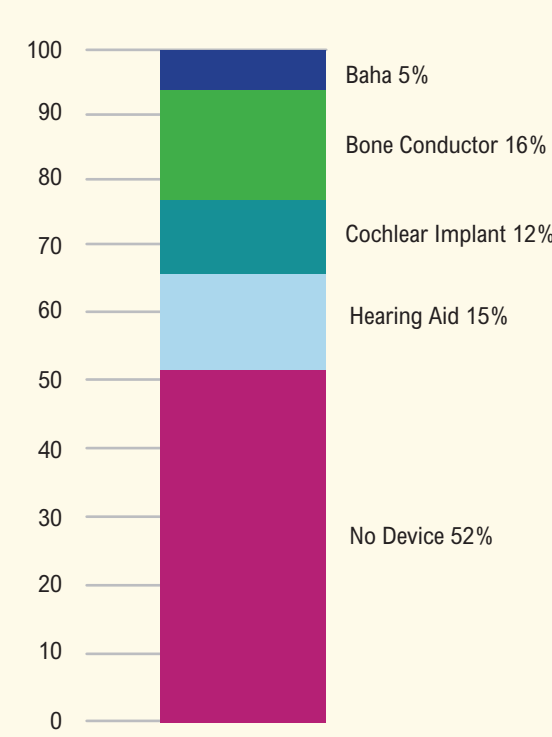
• Average age of entry to EI closer to 12mths, compared to 6mths for children with bilateral hearing loss

• 36/55 in group with MRI results
 • 20 of which showed to have Cochlear Nerve Deficiency (CND), including 5 (25% with ANSD)
 • 88% MRI WNL proceeded to Cochlear Implantation

The Challenge



Device options



HL in the worse ear



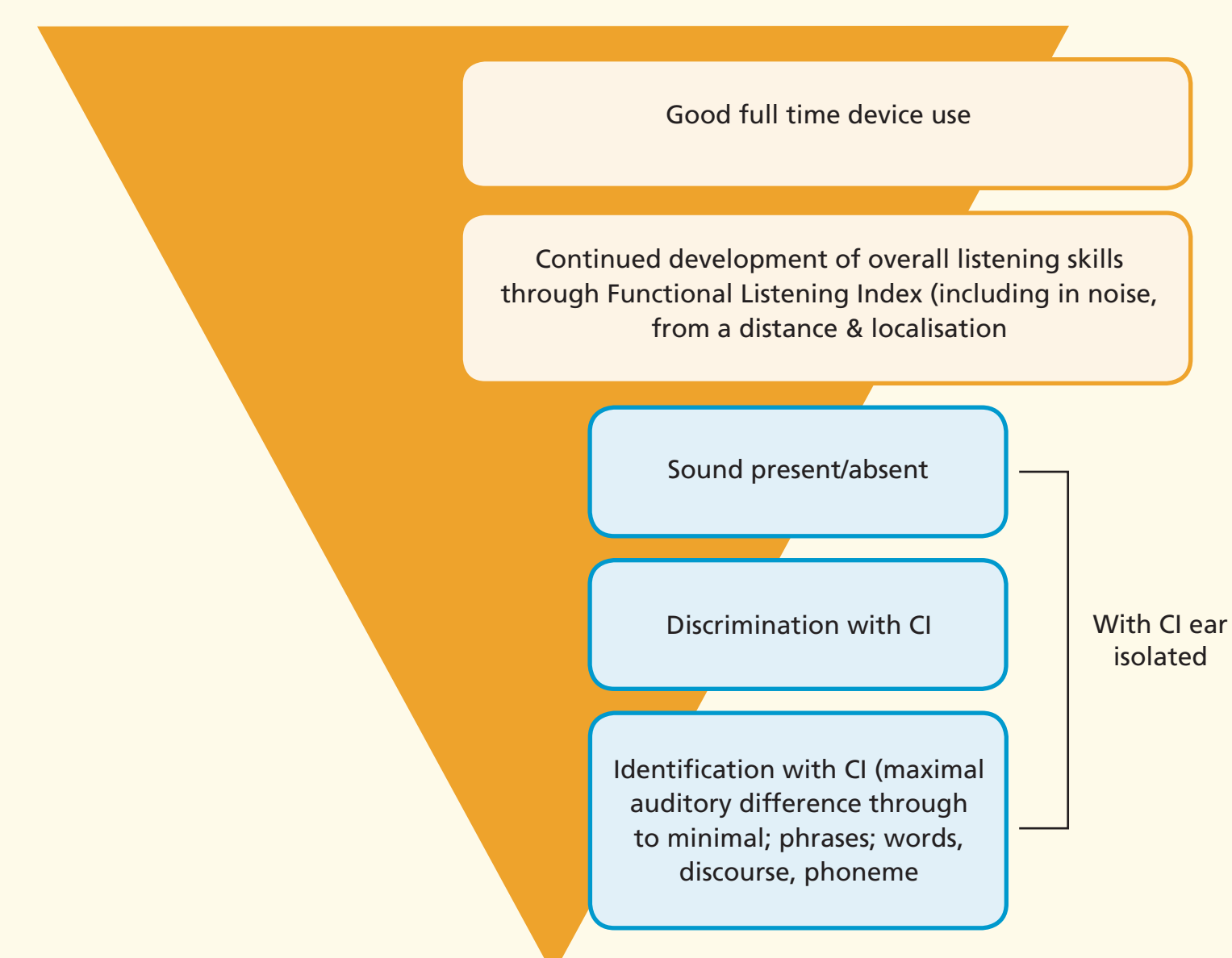
• Highest percentage age in central cohort (mod/mod-sev)
 • 55/89 (63%) hearing levels within traditional CI range

"I've been told my child has enough hearing to develop typical speech and language"
 "I can't get funding as the level of my child's hearing loss isn't significant enough"
 "To us it's simple. If our baby only had one arm, we'd do anything to make sure she had another one. It's the same with her ears"

Audition goals: UHL & CI

1. Measurable Improvement in access to sound
2. Changes in listening ease and confidence

• For children with bilateral HL, CI development focusses on building individual listening and auditory skills.
 • In children with UHL, this is reversed, with most work on integrated development of listening skills with only a small focus on individual CI work.



Cochlear Implant Experience with UHL

| Gender | Age at CI | Age at diagnosis (months) | CI ear | CI ear | Non-CI ear | HA/Baha | CI type | SP | Duration of CI (yrs) | Data logging | Con/Acq | MRI |
|--------|-----------|---------------------------|--------|--------|------------|---------|-----------|------------|----------------------|--------------|------------|-------------------|
| M | 2.5 | 0 | L | 80 | 20 | no | 0522 | CP910 | 0.1 | 5.0 | Congenital | WNL |
| M | 3.3 | 1 | L | 89 | 12 | no | 0522 | CP920 | 0.9 | 8.7 | Congenital | WNL |
| F | 3.8 | 0 | L | 81 | 20 | yes | 0522 | CP920 | 0.0 | na | Congenital | WNL |
| M | 3.9 | 35 | L | 90 | 13 | no | 0248E(CA) | CP910 | 1.6 | 7.6 | Congenital | WNL |
| M | 4.0 | 0 | L | 88 | 20 | no | 0422 | CP920 | 1.6 | 7.6 | Congenital | WNL |
| F | 4.7 | 44 | R | 99 | 21 | no | 0422 | CP910 | 2.8 | na | Congenital | WNL |
| M | 5.2 | 0 | R | 101 | 10 | no | 0248E(CA) | CP920 | 1.1 | 6.7 | Congenital | WNL |
| M | 5.6 | 21 | R | 100 | 9 | no | 0422 | CP920 | 2.0 | 10.8 | Acquired | Fused/Common VON |
| F | 6.0 | 51 | R | 62 | 13 | yes | 0422 | CP920 | 1.6 | 7.0 | Congenital | WNL |
| M | 6.4 | 1 | L | 120 | 13 | yes | 0422 | CP920 | 2.8 | 7.0 | Congenital | WNL |
| M | 6.7 | 0 | R | 120 | 15 | no | 0422 | CP920 | 1.7 | 4.6 | Congenital | WNL |
| M | 6.9 | 79 | R | 120 | 15 | no | 0248E(CA) | CP920 | 2.2 | 2.8 | Acquired | Cochlear fracture |
| F | 7.1 | 59 | L | 73 | 15 | no | 0522 | Not fitted | Non-used | na | Acquired | Ossification |
| M | 10.0 | 88 | R | 113 | 5 | no | 0248E(CA) | CP920 | 1.6 | 2.7 | Congenital | WNL |
| F | 10.4 | 36 | R | 120 | 30 | yes | 0512 | CP920 | 0.4 | 7.0 | Acquired | WNL |
| F | 10.7 | 48 | R | 98 | 20 | yes | 0512 | CP920 | 5.0 | na | Congenital | EMAS |
| F | 11.2 | 1 | L | 112 | 12.4 | no | 0248E(CA) | CP910 | 1.0 | na | Congenital | WNL |
| F | 43.9 | Adult (43 years) | L | 95 | 19 | no | 0422 | CP910 | 1.8 | 12.4 | Acquired | WNL |

| CHILDREN CURRENTLY UNDER EVALUATION | | | | | | | | | | | | |
|-------------------------------------|------|----|---|------|------|-----|--|--|--|--|------------|-----|
| M | 2.0 | 12 | R | >120 | 5 | yes | | | | | Congenital | WNL |
| F | 16.1 | 48 | L | 89.0 | 14.0 | no | | | | | Acquired | WNL |
| M | 0.8 | 0 | L | >120 | <20 | no | | | | | Congenital | WNL |
| F | 4.2 | 0 | L | 86.0 | 14.0 | no | | | | | Congenital | TBC |

| Gender | Median Age at CI (years) | Median Age at Diagnosis (months) | Median HA/BA (dB HL) | Median CI Age (months) | Median CI Age (years) | Median Duration of CI (years) | Median Data Logging (hrs) | Median Con/Acq (%) | Median MRI (%) |
|--------|--------------------------|----------------------------------|----------------------|------------------------|-----------------------|-------------------------------|---------------------------|--------------------|---------------------------|
| N 55% | 3.5 | 12 | 99dBHL (60-120dBHL) | 80 | 1.6 | 1.6 | 1.6 | 73% | 55-60% with WNL-BSDHC CND |
| F 45% | 3.5 | 12 | 99dBHL (60-120dBHL) | 80 | 1.6 | 1.6 | 1.6 | 73% | 55-60% with WNL-BSDHC CND |

References: Screening guidelines for principles and practice, Force & Force (2008); Joint Committee on Infant Hearing of the American Academy of Pediatrics, et al., (2013); Ross et al., (2008); WHO, (2009); Fitzpatrick, Whittingham, & Durieux-Smith (2014); Lieu, Tye-Murray, & Fu (2012); McKay, Gravel, & Tharpe (2008); Martinez-Cruz, Poblano & Conde-Reyes (2009); Fischer & Lieu (2014); Fitzpatrick et al., (2014); Holstrom, Gaffney, Gravel, Oyler, & Ross, (2008); Tharpe, (2008); Ead, Hale, DeAlwis & Lieu, (2013); Abel & Lam (2008); Johnstone, Nabelek, & Robertson (2010); Tavora-Vieira & Rajan (2015); Goberis et al., (2012); Firszt, Reeder, Dwyer, Burton, & Holden (2015); Johnstone et al., (2010); Van Deun et al., (2010); Yoshinaga-Itano, (2013); Lieu, Tye-Murray et al. (2015); Propst et al (2010); Tibbetts et al (2011); Sharma (2014); Heggdal et al. (2016)