Chapter 27. The Importance of Early Intervention for Infants and Children with Hearing Loss

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Chapter Outline

Learning objectives

The reader will be able to describe recent evidence and current findings from the Longitudinal Outcomes of Children with Hearing Impairment (LOCHI) study on 1) the efficacy of early intervention for improving language outcomes of infants and children with hearing loss, (2) the factors influencing child outcomes, and (3) how the evidence can be used to guide clinical management of children diagnosed with hearing loss to optimise outcomes.

Key Points

1. The literature on the effectiveness of intervention for improving outcomes of children with hearing loss at a population level has equivocal findings.
2. The LOCHI study addresses the evidence gap by prospectively evaluating the outcomes of a population cohort of early- and later-identified children who receive the same consistent post-diagnostic hearing services.
3. Early hearing aid fitting or cochlear implantation was effective in improving spoken language outcomes of children.
4. Factors associated with better outcomes also included better nonverbal cognitive ability, less severe hearing loss, absence of additional disabilities, higher maternal educational level, and the use of an oral communication mode (speech) during therapeutic intervention.
5. The presence of auditory neuropathy spectrum disorder (ANSD) was not a significant factor influencing outcomes.
6. Hearing aids prescribed according to the NAL or DSL procedures and verified using real-ear measurements provide audibility that supports language development.

7. The effectiveness of hearing aids for each individual should be evaluated so that the devices can be optimised. This might include the use of objective cortical assessments and parent reports of functional performance in real life such as the PEACH.
8. The evaluations could be used to monitor progress, or trigger referral for cochlear implant candidacy evaluation in children who demonstrate a lack of progress despite optimisation of hearing aids. This would ensure that children who need cochlear implants receive them early so that they can derive maximal benefits.

INTRODUCTION

About 4/1000 children are fitted with hearing aids or received cochlear implants by school entry for a permanent childhood hearing loss (PCHL).[1] The presence of PCHL has major adverse developmental and health impacts on children's lives[2] – including speech and language,[3,4] literacy,[2,5] mental health,[6] social and cognitive functioning,[7] educational achievement,[8] employment and socio-economic opportunity.[9] The associated lifetime cost of all care and lost productivity has been estimated to be USD117 million per birth cohort of 80,000 children.[10] The high incidence of PCHL, the advent of simple screening tests of high sensitivity and specificity, together with studies in the US[11,12] showing that children who received intervention before 6 months of age achieved better language outcomes by 3 years of age than those who received later intervention have provided a driving force for widespread implementation of universal newborn hearing screening (UNHS) programs. The ultimate goal of UNHS is to improve long-term outcomes of children with PCHL through early detection and early intervention, thereby reducing the cost of hearing loss to the individual and to society. However, high-quality evidence on the efficacy of UNHS and early intervention in achieving its goal, *at a population level*, was lacking.[13]

Does early intervention improve outcomes of children with hearing loss at a population level? What factors influence outcomes? What can professionals/clinicians do to ensure that children diagnosed with hearing loss are managed effectively? We address these questions by drawing on research evidence, including recent findings from a population-based study, the Longitudinal Outcomes of children with Hearing Impairment (LOCHI) study.

DOES EARLY INTERVENTION IMPROVE POPULATION OUTCOMES?

In this chapter, intervention refers to the post-diagnostic audiological management (i.e., hearing assessment, amplification fitting) as well as therapeutic intervention for speech, language and socio-emotional development. As the presence of PCHL compromises a child's auditory access to information from the sounds in their environment, treatment of the condition typically commences with fitting of hearing aids or cochlear implants to restore audibility. With widespread implementation of UNHS, it is now possible to treat the condition soon after birth, shortly after diagnosis.

What does the literature say?

In 2001 and 2008, the US Preventive Services Task Force commissioned systematic reviews on the efficacy of UNHS and early intervention for improving language outcomes. These reviews have revealed epidemiological and methodological flaws in previous studies,[14,15] including but not limited to sampling bias, lack of control for confounding variables, and reliance on parent reports. The Task Force called for a prospective, population-based study that directly compares the outcomes of early- and later-identified children.

The early studies also raised several uncertainties. First, they showed that intervention prior to 6 months of age led to improved early language function in hearing-impaired children measured at 3 years.[11,12] However, they also revealed no difference in outcomes between children who first received amplification at 7 to 12 months of age compared to those who first received amplification between 1 and 3 years old, which seems counter-intuitive. Second, there is no evidence as to whether outcomes are affected by when amplification is provided within the first 6 months of life. Third, the evidence does not indicate any interaction between degree of loss and the effects of intervention timing on outcomes. There must however be some interaction, as in the extreme case, no intervention is needed for the average child with normal hearing to achieve normal outcomes.

Table 27.1 summarises recent studies, including three population-based studies. Although observational studies that included convenience samples drawn from specific intervention programs revealed positive effects of early intervention, results from population-based studies were equivocal.

<Table 27.1 >

Kennedy et al[18] studied 120 children at a mean age of 7.9 years (range 6 -10 years), 61 of whom had access to UNHS, 57 of whom had hearing loss confirmed before 9 months of age. On average, children who were exposed to UNHS or confirmation of hearing loss before 9 months had higher receptive language scores than those who received later diagnosis (effect size 0.56, 95% CI 0.03 to 1.08, p=0.04), but remained around 1.9 standard deviation below population norms. Neither expressive language nor speech production benefited from early detection of hearing loss. In a subsequent evaluation of 76 of the children at a mean age of 17 years (range 13-19 years) by Pimperton et al,[19] there was an advantage of early intervention for reading comprehension (effect size 1.17, 95% CI 0.36 to 1.97, p<0.01), but not for oral word reading.

Korver et al[20] compared developmental outcomes at 4-5 years of age of 80 children born in regions with UNHS to those of 70 children in regions where distraction hearing screening was in place. The mean age at hearing aid fitting was 15.7 months for the former group, and 29.2 months for the latter. On average, there were positive effects of UNHS for gross motor skills (effect size 9.1, 95% CI 1.1 to 17.1, p<0.05) and social development (effect size 8.6, 95% CI 0.8 to 16.7, p<0.05), but not for language comprehension or expressive language.

More recently, Wake et al[21] compared UNHS and risk factor screening outcomes of 5-6 year-olds with outcomes of a cohort exposed to opportunistic detection a decade earlier and assessed at 7-8 years. The mean age at hearing aid fitting was 13.5 months, 17.9 months, and 24 months for the respective groups. In children without intellectual disability, benefits of UNHS were associated with expressive language (effect size: 8.2, 95% CI 0.5 to 15.9, p=0.04) and vocabulary (effect size: 8.1, 95% CI 0.8 to 15.4, p=0.03), but not with receptive language, behaviour, or health-related quality of life.

From a developmental perspective, the real benefit of earlier identification lies in the timeliness and quality of intervention following diagnosis. It is noteworthy that the population-based studies have reported delays in diagnosis and intervention following screening. Kennedy et al[18] found that 67% of their screened group of children had hearing loss confirmed by 9 months of age, compared with 27% of their unscreened group. Of children whose hearing loss was confirmed by 9 months, the mean age at intervention was 13 months. In a similar vein, Korver et al[20] indicated that there were delays in diagnosis of those born in UNHS regions, and that there was no guarantee that timely intervention occurred after confirmation. Further, there were variations in the quality of audiological intervention in regions with or without UNHS. Variations in services and technology are likely to influence outcomes. For instance, the trial reported by Kennedy et al[18] occurred before the implementation of the hearing aid modernisation program in the UK, and during a period when effective post-screening audiology and other services for those screening positive for PCHL in the newborn period were largely absent.

A Cochrane review on the efficacy of UNHS[22] found no randomised controlled trials comparing UNHS with either at-risk screening or opportunistic detection of hearing impairment.

Does the effect of early intervention vary with age of assessment?

It has been thought that one of the reasons for the equivocal findings in previous studies may be related to the age at which outcomes of children were evaluated. Fitzpatrick et al,[23] for instance, found no significant effect of age at intervention on 3-year outcomes of children who attended three therapeutic intervention centres. The authors suggested that the effect of early intervention on language development may not have manifested itself at this early age. Nevertheless, Tomblin et al[24] observed a positive effect of early age at fitting on language outcomes in a study using an accelerated longitudinal design in which multiple age cohorts entered the study between 6 months and 7 years of age. For children with mild to severe hearing loss (hearing loss averaged between 0.5 and 4 kHz [4FA HL] in the better ear to be between 25 and 75 dB HL) evaluated at 2 years of age, those who were fitted with hearing aids before 6 months of age had, on average, better levels of language development than those fitted after 18 months. The authors attributed this advantage to the longer duration of hearing aid experience in the early-fitted group at the time of evaluation. For children who were evaluated at 6 years of age, however, there were no significant differences in language scores between the early-fitted and the later-fitted groups. The authors concluded that "later fitting has only temporary effects on language development and may not have lifelong consequences." [24]

In contrast, studies that reported longitudinal follow-through of single cohorts showed a persistent effect of age at intervention on language and literacy development. Pimperton et al[19] found that confirmation of hearing loss by 9 months of age accounted for significant unique variance in reading comprehension in a population cohort of children evaluated at 13 to 19 years of age. They estimated that the effect size of the benefit of early confirmation of PCHL had increased from moderate to large between assessments of the same cohort at ages 8 and 17 years. In a similar vein, Yoshinaga-Itano et al[25] examined the longitudinal outcomes of children with hearing loss exceeding 56 dB HL in the better ear, who were using either cochlear implants or hearing aids. The authors found that gaps in development that were present when the children were 4 years of age persisted into the early school years. Geers and Nicholas[26] reported that earlier age at cochlear implantation was significantly associated with better language in 60 children who received a cochlear implant before 3 years of age. The positive effect of early implantation was observed not only when the cohort was evaluated at 4.5 years, but also when the children were evaluated again at 10.5 years. Indeed, the advantage observed in testing at the younger age was maintained in all aspects of spoken language; and the effect of duration of device experience no longer predicted language outcomes at 10.4 years than when it did at 4.5 years.

The question remains as to whether early intervention is effective in improving long-term outcomes of children with PCHL, at a population level. Any effectiveness of early identification has to be due not only to the earlier diagnosis but to the interventions that result. Therefore, the desired outcomes can be influenced by many factors including, but not limited to, age at intervention. Such factors may relate to the characteristics of the child, the family, and also the intervention. It is in this context that the effectiveness of early intervention for improving outcomes needs to be investigated. Early identification through UNHS incurs heavy financial costs. Even though its acceptance is widespread, its continual existence in an environment of intense competition for healthcare resources may be at risk if its effectiveness remains in doubt.

To address the evidence gap, we commenced a prospective, population-based study in Australia to directly compare outcomes of early- and late-identified children – the Longitudinal Outcomes of Children with Hearing Impairment (LOCHI) study.[27] Across the three most populous states of Australia, New South Wales, Victoria and Queensland, we invited all families with a child diagnosed with PCHL that presented for hearing services under 3 years of age to participate in the study. Depending on the stage of implementation of UNHS in the respective states during a narrow time window, the hearing loss of the children was identified via either UNHS or standard care. Nevertheless, all children received uniform post-diagnostic expert audiological intervention from Australian Hearing (AH), the government-funded organisation for hearing service provision nationally, at no cost to families. This means that the results of the children can be fairly compared, whenever and wherever their hearing loss was discovered. By measuring the outcomes of the cohort prospectively, this longitudinal study provides definitive evidence on the impact of congenital permanent hearing loss and efficacy of early intervention on population outcomes.

What have we learnt from the LOCHI study?

The LOCHI study included about 460 children with a range of characteristics that might impact on their development. The children were from culturally and linguistically diverse backgrounds. As described in Crowe et al,[28] about 80% of the children used spoken English only, with the remaining using a language other than English at home with or without English. For communication at home or in early educational environment, about 76% used speech, with the remaining children using sign with or without speech. About 37% of children have been diagnosed with disabilities in addition to hearing loss. The averaged hearing loss in the better ear ranged from mild (19%), moderate (33%), severe (18%) to profound degrees (32%). Of the children who were diagnosed via UNHS, the median age at hearing aid fitting was 3.3 months (interquartile range: 2.2 to 6.5). About 53% of children received hearing aids before 6 months of age.

At 3 years of age, speech pathologists evaluated children directly using standardised tests of receptive and expressive language as well as speech production. Parents also completed questionnaires to provide information on their children's psychosocial skills, auditory behaviours and functional performance in real-world environments. Ching et al[29] reported that after allowing for the effects of a range of demographic characteristics, there was a trend for 3-year language outcomes to decrease with later age at fitting of hearing aids for children using hearing aids (from 2.4 to 11 months, effect size: -2.4, 95% CI: -5.8, 1.1), although it did not reach the predetermined significance level. Nevertheless, later age at cochlear implantation was significantly associated with a decrement in language skills. Deferring the age at cochlear implant activation from 9.8 months to 23.5 months was associated with a degradation of 8.1 points or 0.54 SD in language outcomes (95% CI -14.5 to -1.8, $p<0.05$).

When children in the LOCHI study were assessed at 5 years of age, the results clearly attested to a significant positive effect of early intervention on children's language outcomes.[30] On average, those who received hearing aids earlier had significantly better receptive and expressive language than those who received hearing aids later. In a similar manner, there was a significant advantage of earlier cochlear implant activation on language outcomes. Importantly, the positive effect of early intervention was observed for children with or without disabilities in addition to hearing loss.

By assessing the same cohort at 3 and 5 years of age, the LOCHI study not only shows an advantage of early intervention, but also demonstrates that the effect increases over time, at least up to 5-6 years. This is especially evident for children with mild to severe hearing loss using hearing aids, unlike the findings based on multiple age cohorts reported by Tomblin et al.[24] Our findings are consistent with those reported for longitudinal cohorts in previous studies,[19,25,26] and reinforces the importance of timely intervention after diagnosis.

<Box 27.1>

FACTORS INFLUENCING OUTCOMES

Without question, there are numerous factors in addition to age at intervention that may account for diversity of outcomes within the population of children with PCHL. These factors may include those relating to the characteristics of the children such as their non-verbal cognitive ability (e.g., Geers et al[31]), severity of hearing loss (e.g. Wake et al[32]), and presence of additional disabilities (e.g. Dammeyer[33]); characteristics of the family such as communication mode at home (e.g. Percy-Smith et al[34]), maternal education[23], socio-economic status (e.g. Tobey et al[35]); as well as characteristics relating to therapeutic intervention, such as the communication mode during intervention (e.g. Meristo et al[36]; Geers & Sedey[37]), family involvement (e.g. Moeller[16]); and the quality of audiological intervention (e.g. Tomblin et al[24]). Although it is well recognised that an investigation of the effect of age at intervention needs to be examined together with the influence of multiple factors in the same regression model,[38] methodological constraints may have precluded this consideration in previous studies. By design, many studies excluded children with additional disabilities and children from diverse cultural and linguistic backgrounds (e.g. Korver et al[20]; Wake et al[32]). In the population studies that examined the effect of age at intervention, Wake et al[32] adjusted scores for severity of hearing loss and nonverbal cognitive ability, Kennedy et al[18] allowed for the effects of these two factors and maternal education, and Korver et al[20] adjusted for maternal education and chronological age at evaluation. The influence of other factors on outcomes remained uncertain.

Factors influencing outcomes in the LOCHI study

The LOCHI study provided the first systematic examination of the effects of a range of child-, family-, and intervention-related variables on outcomes of a population-based cohort of children with PCHL. At 3 years of age, female gender, absence of additional disabilities, less severe hearing loss, higher maternal educational level, and earlier age at cochlear implant activation were significantly associated with better outcomes. The significant effects of gender and maternal education level are not surprising, as they are compatible with those reported for children with normal hearing.[39] The effects of other predictors were also in the expected direction.

At 5 years of age, better spoken language outcomes were associated with earlier hearing aid fitting or cochlear implantation, less severe hearing loss, higher cognitive ability, no additional disabilities, higher maternal educational level, and the use of an oral mode of communication (speech) during early intervention.[30] The advantage associated with the use of speech should not be interpreted as a causal effect, as the study did not randomly assign children to 'speech' versus 'speech plus sign' groups. Parental decisions on communication mode appeared to be influenced by information from professionals, family and friends; considerations of practicalities of communication within the family; children's individual characteristics; and parents' own aspirations about their children's future opportunities.[40,41]

Furthermore, the longitudinal design of the LOCHI study allowed an investigation of whether early outcomes predicted later performance. As part of the test battery, the PEACH questionnaire was administered at 6 and 12 months after initial fitting of hearing devices. The PEACH comprised of 11 items that described a range of situations in real life[42] (the scale is freely downloadable from (http://outcomes.nal.gov.au/Assesments_Resources/PEACH%20ratings%20with%20coverpage%20260509.pdf). Parents were asked to rate their children's auditory behaviour and auditory/oral functional performance in real-world environments, based on their observations. This early measure accounted for unique variance of about 20% in language scores at 3 and 5 years of age.[30,43] The finding suggests that early monitoring of performance can assist with identifying children who may be at risk of language development. This early identification makes it possible for tailoring strategies in intervention to meet the needs of individual children.

<Box 27.2>

Factors that were found to have insignificant effects on outcomes

Some of the factors that have been posited in previous studies on convenience samples to influence outcomes were found to have insignificant effects in the population-based cohort of the LOCHI study. The factors included the presence of auditory neuropathy spectrum disorder (ANSD); and audibility-related factors comprising the proximity of hearing aid characteristics to prescriptive targets, choice of hearing aid prescription, and processing schemes in hearing aids.

Presence of ANSD

In the systematic review on audiological management of children with ANSD conducted by the American Speech and Hearing Association,[44] it was concluded that current literature had methodological limitations and there was insufficient evidence to guide clinical practice. The review called for "a prospective longitudinal study" to "address the efficacy of acoustic amplification and cochlear implantation in children with ANSD and the impact of this disorder on developmental outcomes".[44] The LOCHI study addresses this question by drawing on the prospective evaluations of the population-based cohort that included 47 children diagnosed with ANSD soon after birth. Of the 39 children with ANSD evaluated at 3 years of age, 22 were using hearing aids and 17 were using cochlear implants. They received early intervention provided by AH. The presence of ANSD *per se* was not a significant predictor of language outcomes of children at 3 years of age. The mean effect size of the presence of ANSD was close to zero, too small to be of statistical or clinical significance. On average, there was no significant difference in language outcomes between children with and without ANSD, regardless of whether they used hearing aids or cochlear implants.[45] Similar findings were obtained for evaluations conducted at 5 years of age (see **Fig. 27.1**). Instead of delaying intervention until a child with ANSD could provide reliable results in behavioural assessments of hearing, these findings from the LOCHI study support early amplification or cochlear implantation for children with ANSD, as for children without ANSD.

<**Fig. 27.1** >

<Box 27.3>

Audibility and hearing aid characteristics

A second factor relates to audibility provided by amplification. A recent multi-center study has examined aided audibility in 195 children whose hearing loss in the better ear (4FA HL) ranged between 25 and 75 dB HL.[46] The data showed that 55% of children had fitting that deviated by more than 5 dB root-mean-square (rms) error from prescriptive gain targets in both ears. On average, the proximity of hearing aid fittings to prescriptive gain targets was 6.6 dB in the frequency range between 0.5 and 4 kHz. For a certain degree of hearing loss, hearing aids that deviated more from prescriptive targets had lower gain, resulting in lower aided audibility. This has been associated with poorer language outcomes,[24] even when controlling for the influence of degree of hearing loss. In contrast, the LOCHI study found that hearing aid gain did not have a significant effect on child outcomes at 5 years of age, after allowing for the effects of age at fitting, severity of hearing loss, maternal education level, communication mode, nonverbal cognitive ability and the presence of additional disabilities. This finding must be interpreted in light of the consistent proximity of the fitting to prescriptive targets, which was 3 dB rms when measured at 3 years[47] and 5 years of age (T.Y.C. Ching TYC, PhD, unpublished data, September 2016). All hearing aids of children have been fitted by AH clinicians according to the national protocol that required real-ear measurements for verifying that hearing devices met prescriptive targets. When standard management included adjustment of hearing aids to match targets of validated prescriptions, amplification provided audibility that supported language development. This reinforces the importance of using real-ear measurements to verify that prescriptive targets are met in hearing aids fitted to children.

<Box 27.4>

Hearing aid prescription

The choice of hearing aid prescription also influences audibility with amplification. Previous studies that compared the relative effectiveness of two widely used prescriptive procedures for children, the NAL and the DSL procedures, were confounded by previous auditory experience.[48] The LOCHI study addressed this question by incorporating a randomised controlled trial of hearing aid prescription in its design. Accordingly, children newly diagnosed with hearing loss were assigned randomly to be fitted with either the NAL or the DSL prescription. The outcomes of children at 3 years of age revealed no significant effect of prescription on their language, speech or functional performance in real-world environments[47]. In line with these results, theoretical modelling of the impact of choice of prescription revealed a difference in loudness, but not in estimated speech intelligibility.[49] Evaluation of the language ability of the cohort at 5 years of age revealed that on average, there were no significant difference in language scores of children between the two groups (see **Fig. 27.2**). After allowing for the effects of age at intervention, nonverbal cognitive ability, severity of hearing loss, maternal education and communication mode, the effect of prescription was not significant (T.Y.C. Ching TYC, PhD, unpublished data, September 2016). The finding lends support to the adoption of a validated prescription for fitting hearing aids to children with PCHL.

<Fig. 27.2 >

<Box 27.5>

Hearing aid processing: nonlinear frequency compression or conventional processing

Differences in processing schemes implemented in hearing devices may also influence audibility. One approach to increasing audibility of high frequency sounds is to use frequency lowering technology, so that information in the high frequencies can be presented at lower frequencies where the hearing loss is less severe. A systematic review on the effectiveness of this technology for children by McCreery et al[50] revealed a lack of high-quality evidence. Bentler et al[51] examined whether children using nonlinear frequency compression (NLFC; one frequency lowering strategy) in their hearing aids had better access to the speech signal than children using conventional processing (CP) schemes. On average, there was no significant difference in audibility estimated by a modified Speech Intelligibility Index between the two groups of children. Consistent with estimated audibility, there were no differences in speech and language abilities between the two groups. As the study did not randomly assign children to the technology options, the effect of the technology remained uncertain. The LOCHI study addressed this question by incorporating a randomised controlled trial of NLFC. Participants who enrolled in this trial were assigned to fitting with either NLFC or CP in their hearing aids. At 3 years of age, there was no significant effect of NLFC on speech and language outcomes[52]. At 5 years of age, audibility of children using NLFC was not significantly different from those using CP in their hearing aids. On average, there was no significant difference in speech and language outcomes between the two groups (see **Fig. 27.3**) (T.Y.C. Ching TYC, PhD, unpublished data, September 2016). The current evidence shows that on average, the use of NLFC processing in hearing aids does not increase audibility, and cannot be expected to provide better support than CP for language development in young children.

<Fig. 27.3 >

<Box 27.6>

FROM EVIDENCE TO PRACTICE: LOCHI FINDINGS AND CLINICAL PRACTICE

The LOCHI study has been tasked to contribute to generating evidence-based guidelines for management of hearing loss in children. The following summarises the findings and illustrates the translation of evidence into clinical practice.

Summary of findings in the LOCHI study

1. Early fitting of hearing aids is effective in improving spoken language outcomes.
2. Early cochlear implantation improves spoken language outcomes.
3. The presence of ANSD was not a significant factor influencing outcomes.
4. The choice of hearing aid prescription was not a significant factor influencing outcomes.
5. The use of nonlinear frequency compression was not a significant factor influencing audibility or outcomes.

These findings, based on population data, reflect what is true on average. Research needs to increase understanding about what is optimal for individual children and families, which may vary with the individual's abilities and the goals of the families.

<Box 27.7>

Evidence-based management

First and foremost is to ensure a streamlined clinical pathway from screening to diagnosis to intervention. In Australia, the loss to follow-up from newborn hearing screening to diagnosis is less than 1%,[53] with a mandatory requirement for the government-funded hearing service provider (AH) to provide services shortly after diagnosis according to a national protocol.[54] The protocol has been updated recently to encompass evidence, including those from the LOCHI study. In order to ascertain that early fitting of hearing aids provides effective audibility to support speech and language development, the key milestones for pediatric amplification over the first 18 months after diagnosis include

1) Fitting of hearing aids according to hearing levels estimated from diagnostic electrophysiological results to match prescriptive targets, within 6 to 8 weeks after diagnosis. The NAL prescription is used to derive real-ear-aided gain targets for each individual, and hearing aids are verified using real-ear measurements to match prescriptive targets. The goal is to achieve audibility through fitting of amplification according to a validated prescription, and evaluation of audibility by measuring aided cortical auditory evoked potentials (CAEPs).[55,56]
2) Secondly, evaluating effectiveness of amplification using the PEACH questionnaire by 3 months after initial fitting of hearing aids or around 6 months of age. The goal is to ascertain functional hearing with amplification in real-world environments.
3) Thirdly, monitoring progress by administering the PEACH at regular intervals. Ear-specific behavioural thresholds are measured using visual reinforcement audiometry, and real-ear measurements are used in verification of hearing aids after adjustments. The goal is to monitor progress through optimising the fitting, and checking that the child uses functional hearing in a way that is commensurate with typically developing children.

Considering the importance of early activation of cochlear implants for optimal outcomes, clinical guidelines for early referral of cochlear implantation have been implemented. When PEACH scores are below 2 SD of the population mean, and measurements of aided CAEPs reveal that responses to high frequency sounds are absent despite fine-tuning hearing aid settings, discussions with parents about cochlear implant candidacy evaluation will be initiated. The goal is to ensure that children for whom cochlear implantation is likely to provide better support than hearing aids will receive cochlear implants before 12 months of age.

The presence of ANSD is not a condition that precludes early fitting of hearing aids. Amplification can be considered on the basis of CAEPs in an unaided condition, with speech sounds presented at conversational levels and soft speech levels. These results, together with PEACH results and information from the family can contribute to determining the need for amplification. Details about management options for ANSD are explained in detail in Chapter 32. If amplification is warranted, the child with ANSD follows through the same clinical pathway as for children with sensorineural hearing loss.

Putting it all together: case studies

Two case studies are presented below to illustrate the application of the protocol in clinical management of a child with sensorineural hearing loss and a child with ANSD.

Case 1. SN1 – Child with sensorineural hearing loss

SN1 was born at 38 weeks gestation, and referred bilaterally through newborn hearing screening. She had jaundice and received phototherapy for 24 hours. As shown in **Fig. 27.4A**, the diagnostic auditory brainstem responses (ABR) to tone bursts showed a mild to moderate sloping hearing loss in both ears. The estimated behavioural audiogram based on ABR results were used to derive prescriptive targets according to the NAL formula, and hearing aids were verified to match targets. The child was aided bilaterally at 6 weeks of age. Aided CAEPs were measured 9 weeks after initial hearing aid fitting. Responses to speech sound stimuli /t/ and /s/ were detected when these sounds were presented at 65 dB SPL. These results combined with a mild degree of hearing loss at low and mid frequencies suggested that speech frequencies at medium conversational levels would be audible to the child when wearing both hearing aids. The PEACH score at 11 months indicated that the child's auditory functional performance in everyday life when aided was within the range for children with normal hearing of the same age (**Fig. 27.4B**). The measurements of PEACH and aided CAEPs were useful for assuring the parents about the effectiveness of amplification for their child.

<Fig. 27.4A and B>

Case 2: AN1 – Child with ANSD

AN1 was born at 36 weeks gestation with cardiopulmonary hypertension, requiring prolonged ventilation and treatment with gentamycin. He also received phototherapy treatment for jaundice. He was referred bilaterally from the newborn hearing screening program, and diagnosed with ANSD. As there were no responses to tone-burst ABR testing at maximum levels, CAEPs were measured in the unaided condition to estimate hearing sensitivity and the need for amplification. At 2 months corrected age, measurement of unaided CAEPs to /t/ /m/ /g/ revealed no detectable responses at stimulus presentation levels of 65 and 75 dB SPL in the sound field. In light sleep, no behavioural responses were observed when broadband, low, mid and high frequency noisemakers were presented at high levels. Combining the unaided CAEP results and behavioural observations, the child was estimated to have hearing loss of at least 80 dB HL in both ears. Accordingly, hearing aids were fitted according to the NAL prescription. After fitting, aided CAEPs were measured showing responses to /m/ and /g/, but not /t/, presented at 75 dB SPL (see **Fig. 27.5A**). No responses were detected for any of the stimuli at lower presentation levels. The PEACH score obtained two weeks after the fitting was at 2SD below the mean of the normative population (see **Fig. 27.5B**). Accordingly, the hearing levels were re-estimated to profound loss and hearing aids were adjusted to match revised prescriptive targets. Four weeks after the readjustment of hearing aids, aided CAEPs were evaluated in the sound field. Ear-specific aided CAEPs revealed responses to /m/ /g/ /t/ at 75 dB SPL in the right ear only. Middle ear problems were reported for the left ear at this time. The PEACH scores continued to show that the child’s functional performance was at 2SD below the normative mean for his age. Speech therapists reported that the child did not show detection of the Ling sounds at one metre when aided, and there was no observable progress. In view of the evaluations of aided CAEPs and PEACH results and the input of the speech therapists, the family was advised to consider referral for cochlear implant candidacy evaluation before it was possible to obtain reliable results with visual reinforcement audiometry. The child received bilateral cochlear implants by 10 months of age.

<Fig. 27.5A and B>

These two cases serve to illustrate a clinical management pathway to ensure early fitting and optimising of amplification for individual children, and early referral for cochlear implant candidacy to ensure that a child who needs a cochlear implant gets it early. The presence of ANSD is not a condition that precludes early amplification or cochlear implantation. The evidence-based protocol uses a combination of objective measurement of CAEPs and behavioural assessment with the PEACH questionnaire to optimise post-diagnostic management of children with hearing loss.

<Box 27.8>

CONCLUSION

Current evidence from the LOCHI study attests to the effectiveness of early intervention for improving outcomes of children with hearing loss. The findings of the study can guide management of childhood hearing loss to maximise the benefit of early intervention. Following early fitting of hearing aids and verification of prescriptive targets, evaluation of the effectiveness of amplification for individual children by using objective cortical measures and the parent-report PEACH scale should be implemented. This ensures that individual progress with intervention is monitored; and, if indicated, that referral for paediatric cochlear implant candidacy evaluation can occur at an early age.

The world for children born with PCHL has changed. With early detection of hearing loss and early intervention, achieving parity of outcomes between children with hearing loss and those with typical hearing is within reach. Post-diagnostic intervention needs to be timely, and be guided by current evidence. The principle with any treatment/intervention is that we must always evaluate its effectiveness for each individual, and if it is not effective, have the courage to change for the better.

CHAPTER SUMMARY

The presence of permanent childhood hearing loss has a negative impact on children's developmental outcomes. The widespread implementation of UNHS makes it possible to detect congenital hearing loss soon after birth, so treatment can begin in infancy. However, the efficacy of early intervention for improving outcomes remained inconclusive. This chapter addresses the question by drawing on findings from a population-based, prospective study – the LOCHI study. On average, children who received earlier fitting of hearing aids or cochlear implants had better spoken language by 5 years of age. Better outcomes were also associated with less severe hearing loss, higher nonverbal cognitive ability, absence of additional disabilities, higher maternal education level, and use of an oral mode of communication during early intervention. The LOCHI study shows that on average, the presence of ANSD did not significantly influence outcomes. Further, hearing aids that were selected according to either the NAL or the DSL prescription and verified to match targets using real-ear measures provided adequate audibility to support language development. This Chapter shows how the evidence can be incorporated in clinical management of childhood hearing loss to maximise the benefit of early intervention. Current best practice includes fitting hearing aids in a timely fashion, verifying that prescriptive targets are matched, and evaluating the effectiveness of amplification for individual children by using objective cortical measures and the parent-report PEACH scale. These processes serve to monitor individual progress with intervention, and ensure that paediatric cochlear implant candidacy referral can occur at an early age if indicated.

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Chapter review questions

1. What are some of the reasons that explain the equivocal findings in the literature on the effectiveness of early intervention?
2. Why might a longitudinal study of a single cohort over time provide stronger evidence than multiple age cohorts in examining the long-term effectiveness of early intervention?
3. What are some of the factors, in addition to age at intervention, that influence outcomes of children with hearing loss?
4. How can the clinical pathway for managing children diagnosed with hearing loss through newborn hearing screening be streamlined to maximise the benefits of early identification?
5. How might the detection of CAEPs and the use of parent-reported functional performance form part of a routine evaluation of hearing aid fitting in young children?
6. How might the detection of CAEPs and the use of parent-reported functional performance form part of standard post-diagnostic management of children with ANSD?
7. Why is evaluation of hearing aid effectiveness crucial to maximising benefits of early intervention?

Pearls (Text boxes)

Box 27.1 The evidence from the LOCHI study supports early fitting of hearing aids and early cochlear implantation to facilitate development of language by children with hearing loss, including those with additional disabilities.

Box 27.2 Early monitoring of auditory behaviour with amplification can assist with identifying children who may be at risk of language development.

Box 27.3 The presence of ANSD should not preclude children from receiving early amplification or cochlear implantation.

Box 27.4 Hearing aids that are verified to meet targets prescribed by validated procedures provide audibility that supports spoken language development.

Box 27.5 The evidence to date supports early fitting with either the NAL or the DSL prescription to facilitate normal development of language by children with hearing loss.

Box 27.6 Current evidence shows that on average, the use of nonlinear frequency compression does not increase audibility or benefit spoken language development.

Box 27.7 Early intervention is effective in improving outcomes of children with hearing loss, including those with additional disabilities and those with ANSD.

Box 27.8 Use objective measurement of CAEPs and behavioural assessment with PEACH to evaluate the effectiveness of hearing aids. Refer for cochlear implant candidacy if indicated. The presence of ANSD should not preclude a child from receiving early amplification or cochlear implantation.

Figure legends

Fig. 27.1 Mean language scores at 5 years of age for children with sensorineural hearing loss (SNHL, indicated by open symbols) or children with auditory neuropathy spectrum disorder (ANSD, indicated by filled symbols). The scores for PLS-4 Auditory comprehension (AC) subscale (depicted by circles), and PLS-4 Expressive communication (EC) subscale (depicted by squares) are shown; separately for children using hearing aids (HA) in the left panel, and those using cochlear implants (CI) in the right panel. The vertical bars denote 95% confidence intervals.

Fig. 27.2. Mean language scores at 5 years of age for children fitted with NAL prescription (depicted by open circles) and children fitted with DSL prescription (depicted by filled squares). The left panel shows scores for the PLS-4 Auditory comprehension (AC) subscale, and the right panel shows scores for the PLS-4 Expressive communication (EC) subscale. The vertical bars denote 95% confidence intervals.

Fig. 27.3. Mean language scores at 5 years of age for children who used nonlinear frequency compression in their hearing aids (NLFC, depicted by open circles) and those using conventional processing (CP, depicted by filled squares). The left panel shows scores for the PLS-4 Auditory comprehension (AC) subscale, and the right panel shows scores for the PLS-4 Expressive communication (EC) subscale. The vertical bars denote 95% confidence intervals.

Fig.27.4A. Case SN1 - Hearing thresholds estimated from auditory brainstem responses to tone bursts, and assessments of aided cortical auditory evoked potentials. Behavioural thresholds obtained at 7 months using visual reinforcement audiometry are also shown.

Fig. 27.4B. Case SN1 - PEACH score obtained at 11 months of age. The solid line represents the relationship between scores and age for normal hearing infants and the broken lines denote ± 2 standard deviations.

Fig. 27.5A. Case AN1 – Auditory brainstem response testing using tone bursts as stimuli showed no detectable responses at maximum presentation levels. Aided cortical auditory evoked potentials testing showed detectable responses to /m/ and /g/ presented at 75 dB SPL. Behavioural thresholds obtained at 6 months (corrected age) using visual reinforcement audiometry are also shown.

Fig.27.5B. Case AN1-PEACH scores at 2.5 and 5 months (corrected age). The solid line represents the relationship between scores and age for normal hearing infants and the broken lines denote ± 2 standard deviations.

Table 27.1. Summary of studies on the effect of age at intervention on outcomes of children with hearing loss

Study	n	Age at fitting (number of children)	Age at evaluation	Outcomes measured	Findings on effect of age at intervention on language outcomes
Yoshinaga-Itano et al, [12] 1998	150	0– 34 months (72 < 6 months)	13 – 36 months	Parent report: Child Development Inventory	On average, children enrolled in intervention before 6 months had scores that were significantly better than those enrolled after 6 months of age.
Moeller,[16] 2000	112	1 – 54 months (24 < 11 months)	5 years	Receptive vocabulary	On average, children enrolled in intervention before 9 months of age had higher scores than those enrolled later.
Wake et al,[17] 2004	89	5 – 54 months (11 < 6 months)	7 - 8 years	Language, speech production	No significant effect of age at intervention.
Kennedy et al,[18] 2006	120	10-40 months (57 < 9 months)	5.4-11.7 years Mean: 8 years	Receptive and expressive language; parent report on speech production	On average, children with hearing loss confirmed by 9 months of age had receptive language scores that were significantly better than those with

					hearing loss confirmed after 9 months of age. There was no significant difference between the two groups in expressive language or speech production.
Fitzpatrick et al.,[23] 2007	65	6.6-18 months (15 < 6 months)	3-5 years	Receptive vocabulary; language; speech production	No significant effect of age at intervention on outcomes.
Korver et al,[20] 2010	130	15.7 months (newborn hearing screening) vs 29.2 months (distraction hearing screening)	4-5 years	Parent reports: Child Development Inventory; Macarthur-Bates Communicative Inventory	No significant effect of age at intervention on language outcomes.
Geers and Nicholas,[26] 2013	60	Age at cochlear implantation: 22 between 12–18 months	4.5 and 10.5 years	Expressive Language Receptive Language Expressive and Receptive Vocabulary	Better language scores were significantly associated with younger age at initial activation of a cochlear implant. The same finding applies to evaluations at 4.5 and 10.5 years of age.

		16 between 19–24 months 22 between 25–38 months			
Pimperton et al.[19] 2016	76 (sub-group of participants in study[18])	(35 ≤ 9 months 41 > 9 months)	13–19 years Mean: 16.8 years	Reading test that measures accuracy, comprehension and summarisation skills.	On average, those with hearing loss confirmed by 9 months had significantly higher scores for reading comprehension and reading summarisation than those with hearing loss confirmed after 9 months of age.
Tomblin et al,[24] 2015	208	(61 <6 months 47 between 6-12 months 12 between 12-18 months 88 > 18 - 65 months)	2-6 years	Expressive and receptive language, parent reports	At age 2 years, early fitted children had better language scores than children who received later fitting. This difference was no longer significant by 6 years of age.