

Development and validation of the Nottingham Auditory Milestones (NAMES) profile for deaf children under two years old, using cochlear implants.

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Abstract

Objectives: Clinicians face considerable challenges in setting appropriate auditory goals for babies and young children who receive cochlear implants. This paper describes the rationale, organisation, implementation and validation of the Nottingham Auditory Milestones profile that was developed to address these challenges.

Methods: The use of the profile has been fully integrated into the post-operative pathway at the Nottingham Auditory Implant Programme since 2009. Data is presented on a cohort of 30 children who received bilateral cochlear implants under the age of two and who have no other diagnosed difficulties. The data was used to validate the profile's structure and characterise the expected development trajectory for this population of children.

Results: The analysis of routine data from the children confirmed that the profile's structure reflected the typical order and rate at which skills emerged and were acquired over the first three years following cochlear implantation. The distribution of profile scores across five assessment time-points established a developmental trajectory for typically-developing children. Three case studies describe the use of the profile to set consistent expectations for progress for a wide range of children.

Discussion: The development trajectory established using the profile provides a mechanism to identify children not making the expected progress, in order to support the need for a review of approach or a differential diagnosis.

Conclusion: The Nottingham Auditory Milestones profile is an accessible and practical tool for identifying, monitoring and appraising the auditory achievements of deaf babies and young children in the first three years following cochlear implantation.

Keywords: paediatric cochlear implantation, auditory achievements, milestones, profile,

Introduction

Providing cochlear implants to profoundly deaf babies and very young children enables them to access sound and voices at a time when their brains are highly receptive to language learning. In the UK, the majority of congenitally deaf children undergo cochlear implantation surgery under the age of two (Raine, 2013). Since the implementation of NICE guidance in 2009, bilateral cochlear implants have been available to all audiological and surgically-appropriate children (NICE, 2009). Studies have indicated that children who have their operations young, and who are given sufficient opportunities, have the potential to learn to listen and talk following a similar route to their hearing peers (Archbold et al., 2008; Archbold et al., 2012; Colletti et al., 2005; Sarant et al., 2014; Verhaert et al., 2008; Yoshinaga-Itano et al., 1998). The opportunity to offer bilateral cochlear operations at an early age was therefore a welcome development in commissioning and raised expectations for an increased rate of progress for this population. However, long term outcomes for this cohort still show considerable variation and not all children develop high level auditory skills and good spoken language (Boons et al., 2012; Harris et al., 2013; Inscoe et al., 2009; Niparko, 2010; Sarant et al., 2008; Sarant et al., 2010; Van Wieringen et al., 2015). A more detailed understanding about the rate at which functional auditory benefits typically develop for this very young population should provide a better understanding of this variability, provide an effective tool for expectation counselling with families and potentially supply valuable information to underpin more individualised approaches to intervention.

Monitoring the progress of very young children following cochlear implantation has generally been based on the auditory, receptive and expressive language developmental norms of hearing babies and children (Ching et al., 2013; Nikolopoulos et al., 2005; Van Wieringen et al., 2015). This model assumes that deaf babies, post cochlear implantation,

will follow a normal but delayed pathway, which over time, may narrow or even close the listening and language gap with their hearing peers. However, there are limitations to this model. A newborn hearing baby differs in many respects to a deaf baby experiencing sound for the first time via sound processors, typically between the ages of nine to fifteen months old. A newborn hearing baby is physically helpless and requires long periods of sleep but hearing is a fully developed sense and she or he is highly attuned to voice. In contrast, the cognitive, physical and oro-motor development of the older deaf baby, alongside their accumulated social experiences, should help him or her make sense of the signal from the processors at a more rapid pace. However, access to sound remains limited to when the processors are in place and working. The equipment has to be managed and maintained by adult caregivers, which means that their commitment, ability, attitudes, expectations and communication choices are vital to success. The effectiveness of the surgery and the programming of the devices, alongside the quality and quantity of professional support are also recognised contributory factors to positive outcomes (Edwards, 2003; Van Wieringen et al., 2015; Yoshinaga-Itano, 2013).

[Studies indicate that deaf babies more likely than the population of hearing babies to have additional problems which may affect their capacity to develop spoken language (Ramirez-Inscoe et al., 2016). For some children, these difficulties will be obvious and well documented at the time of the operation. For example, a significant global developmental delay or certain aetiologies such as congenital cytomegalovirus (cCMV) or meningitis infections put children ‘at risk’ of poorer outcomes but even this is not an inevitable corollary and the extent of the impact is variable (Birman et al., 2012; Lee et al., 2005).

As babies present for initial assessment at only a few months old, clear indicators of underlying difficulties, such as autism or language learning, are rarely evident. Therefore, beyond a small percentage of the assessment population, it is not possible to distinguish with any degree of reliability those children who are likely to gain limited benefit from the signal provided by the cochlear implant from those who will make an easy transition to hearing, listening and talking. Close monitoring in the post implant phase, within a clear framework of expectations, is therefore essential to identify potential problems as early as possible.

In 2006-2007, in the light of the demands of this younger population and with the accumulated clinical experience since the first paediatric operation in 1989 (Nikolopoulos, 2005; Sach et al., 2004), the Outreach Support Team¹ within the Nottingham Auditory Implant Programme (NAIP) reviewed their existing assessment materials and monitoring protocols. With the evidence of success, the client group had widened and expectations for outcomes had increased significantly across those fifteen years. The cochlear implant devices had become more sophisticated, as had the programming options, and bilateral cochlear implants for certain aetiologies were beginning to take place. In general, the patient population was getting younger but included a higher percentage of children with additional needs.

It was of some concern that families could access information which encouraged them to believe that an early operation and better still, two cochlear implants, meant that their child was guaranteed to make rapid progress towards the acquisition of age appropriate spoken language. While the team also had high expectations, this sat alongside the realisation that the picture was complex, and that providing access to sound via the equipment did not of itself, guarantee any particular outcome. There was recognition that

for a number of children, auditory and spoken language progress post implantation was limited, sometimes quite surprisingly given the child's general development and pre-verbal communication, as assessed by the team pre-operatively.

Another key issue was that many professionals in the community were finding it hard to calibrate their expectations for rate of progress in the face of rapid improvements in the technology and the impact of early operations on the reduction in the length of auditory deprivation., The lack of a framework guide expectations on progress meant that the identification and the need for a differential diagnosis and approach for some children, was sometimes slow to be recognised, accepted and implemented.

This paper describes the development, structure, administration and implementation of the Nottingham Auditory Milestones (NAMES) profile tool that has been developed to address these challenges in identifying, monitoring and appraising the auditory achievements of deaf babies and young children in the first three years following cochlear implantation (Datta et al., 2010). It also reports the results of a recent validation study of its use in children implanted under two years of age. Its practical application and usefulness in tracking the progress of individual patients is illustrated through case studies.

Method

Initial development

The development process began with a systematic analysis of the strengths and limitations of materials that were already available, including those profiles developed in-house at NAIP. It identified several priorities for a profile that was designed specifically for the youngest patients, at that time ranging in age between nine months to three years

old at the time of cochlear implantation surgery. It needed to set expectations and chart progress from early to advanced auditory skills. There was recognition that a proportion of children made the expected early progress but that things became much more challenging once there was a requirement to process at a linguistic level, rather than to predominantly respond to very familiar words in context. It had to move beyond the understanding of single words and short phrases through to participation in conversation, given that the ability to listen to others and learn within a group setting, is recognised as essential for good long term language and educational outcomes (Geers et al., 2013; Mayer, 2007).

Although the profile had to be founded on recognised developmental hierarchies, it was also essential to take into account the different auditory starting point and equipment reliance of older, deaf babies. The measures relating to rate of progress were based on an expectation that deaf babies and young children, given access to sound, should be able to make at least a year of listening and spoken language progress for every year of consistent processor use. This seemed a relatively modest expectation for the youngest children, given that research evidence on neuroplasticity indicated that age at the time of surgery correlated strongly with better outcomes (Flexer, 2011).

The profile needed to be flexible enough to record the rate of achievements of those children moving at a faster rate than this predicted average, which was anticipated to be the case for the younger and more able patients. However, it also had to recognise that children with known additional diagnoses or in more complex family situations would be less likely to achieve this pace. Crucially, it was also essential to provide a mechanism to flag up as early as possible, those children with no known complicating factors, who were moving off the expected track.

The developmental expectations were drawn predominantly from established schedules, particularly the work of Mary Sheridan (Sheridan et al., 1997) and the Monitoring Protocol for deaf babies and children (DfES, 2004), a developmental schedule used widely at pre-school level in the UK. The auditory start line was based on the experiences of babies, typically at a minimum of nine months old, coming to sound for the first time via the signal from the implant system. It explicitly acknowledged that the quantity of processor use was central to progress. For example, some early measures of progress linked closely to the acceptance and observed changes in behaviour when the sound processors are worn. Clinical experience suggests a wide variation in average daily wearing times, even for those children whose families describe them as consistent users of the equipment. Auditory skills were ordered to recognise that as children come to hearing at an older age, they have the cognitive capacity to acquire some auditory skills at a faster rate than a hearing baby, particularly in the first few months post cochlear implantation.

NAIP is based within the public health sector and manages a large and diverse population of children who come from families with a wide range of ethnicity, socio-economic backgrounds and educational levels. A central tenet of NAIP philosophy is active family engagement via an Outreach model, working in close collaboration with local professionals. For babies and young children, this means the delivery of sessions jointly with the child's local professionals, typically within the family home or nursery, usually face to face, but sometimes through a telemedicine model. The profile design and content was crucial to its successful implementation. Information needed to be presented in an accessible way to provide the basis for discussions to which families and local professionals could actively contribute. It therefore had to be in a format that would be

easy to understand by non-specialists. The skills identified within the profile are based on observations and the child's participation in everyday age-appropriate activities to which families and local professionals could readily relate. The aim was to use the profile as a conduit to encourage ideas about how to develop new targets based on the child's functional communication and listening needs in daily life, and to make an explicit link with the cognitive and communicative demands which drive language, including the growth of auditory memory and verbal reasoning. The format was designed to be flexible enough to allow the child's progress to be monitored via the main spoken language of the home and to allow for a range of ways to obtain and demonstrate evidence of achievements.

Structure

The NAMES profile and support materials were initially developed in 2006-2007 and were trialled with inter-rater reliability studies within the department in 2007-2008. It was fully implemented as part of the core tracking materials for all the younger children in 2009. Alongside the profile record, available in both detailed and summary form, a range of other related materials was produced which support planning and provide further advice to share with families and local professionals.

The profile consists of five Milestone stages spread over the first three years following cochlear implantation (Table 1). Each new Milestone builds on the skills of the previous set, to create a cumulative profile. Each Milestone stage consists of ten descriptors of the auditory behaviours expected to be achieved within the designated time-scale. In the first year, in acknowledgement of the rapid anticipated changes if progress is on-track, there are three sets of Milestone stages: the first at three months post implant, the second at six months post implant and the third at twelve months post implant. This reduces to a fourth

Milestone stage at two years post implant and a fifth Milestone stage at three years post implant. As the milestones progress, measures of audition are linked to more challenging listening and language situations.

Milestone One was expected to be reached, typically after three months of sound processor use. Key skills expected at this stage were that the equipment is worn consistently for most waking hours with definite behavioural changes observed during use, including the detection of environmental sound, music and voices, alongside an increase in purposeful vocalisation. **Milestone Two** was expected to be reached typically after six months of sound processor use and the expectation was that children should demonstrate recognition of some familiar everyday sounds, show an ability to listen and respond vocally to the voices of others, and that vocalisations would be increasingly influenced by what is heard. By **Milestone Three**, expected to be reached typically after twelve months of sound processor use, the child should give clear signals if the equipment stops working, understand the language of social routines and demonstrate an understanding of familiar phrases within predictable contexts. By **Milestone Four**, expected to be reached typically after two years of sound processor use, the child should understand simple information without relying on context, sing parts of songs, have a two-three item auditory memory and the spoken language skills to contribute to a simple retell of familiar events with adult support. By **Milestone Five**, typically after three years of sound processor use, the child should be conversational, be able to listen and talk as part of a small group, have a three to four item auditory memory, demonstrate the ability to give key information about themselves, be able to recite a short poem or sing the words of a familiar song and provide a simple, independent recount of an event or story.

Method of administration

The profile is completed on the basis of observations and specific ‘probe’ activities with the child, alongside feedback from the family, caregivers or familiar professionals. The participation of both a primary care-giver and a local professional is typically the optimal way to tease out the sorts of behaviours being demonstrated by the child and how consistently these occur. In most instances, it should be possible to observe the child demonstrating behaviours if they are well established but some skills will rely on knowledge of the child’s responses in a range of everyday situations, such as their recognition of a variety of environmental sounds.

At NAIP, parts of the assessment session will usually be filmed so that the child’s performance can be reviewed in conjunction with other team members, to ensure consistency and the benefits of multi-professional review. Probe activities within sessions are individually tailored to the child’s developmental and play level. Wherever possible, the child is provided with the opportunity to demonstrate their skills in real world, functional situations. Books are also generally used at every interval point, moving from responses to symbolic sounds and early words, through to independent retells.

Scoring and interpretation

The scoring system is cumulative in nature. A baseline score is recorded for those children who gain functional benefit from hearing aids pre-implantation. At each subsequent assessment interval, the assessor works through the Milestone targets in the profiles to record all the new skills. A score of 0 is recorded if the behaviour is never observed, 1 if it is emerging but inconsistent and 2 if the child usually demonstrates the behaviour. Observations and recording are not limited to assessing the skills that would be expected at the current Milestone Stage; e.g. it is not limited to assessing the expected skills for Milestone Two after 6 months of sound processor use. Instead, each assessment

begins with the skills previously achieved and moves on through the Milestones until all the achieved skills, either emerging or established, are recorded. This provides a mechanism to record changes relative to the child's previous performance, to record the rate of progress for children who are moving faster or slower than average, and to provide evidence for children who have an unusual profile of skills.

If a child is moving through the profile at the 'average' rate, in accordance with how it was constructed, their scores would be expected to increase by around 20 points at each assessment interval. However, the profile necessarily represents a 'snapshot' at the interval points and many children do not progress at a steady rate but may have spurts of progress and other periods when they plateau for a while. The score obtained at any individual assessment therefore needs to be interpreted with caution. Concerns arise if children with no known mitigating factors are not moving on close to this anticipated rate between interval points, and certainly if that was repeated across two intervals.

Validation

Building the structure of the profile around the five Milestones was based on expectations of the skills that would develop after defined periods of sound processor use; i.e. 3, 6, 12, 24, and 36 months. Data from the routine administration of the profile was used to assess the validity of this structure. Validity in the context of this assessment was defined as whether each skill was recorded as having emerged in a majority of children after these expected durations of usage. The information obtained had already been discussed fully with parents who agreed to its anonymised use, and shared within formal reports circulated to local professionals. No formal ethical approval was required for its use within this study. The assessment time point at which each skill was marked as emerging or acquired, together with the child's score at each assessment, was extracted from the

profiles of a consecutive cohort of 30 children. In order to minimise the impact of factors, such as the potential auditory benefit of bilateral as opposed to unilateral implantation (Sarant et al. 2014), all had bilateral cochlear implant operations under the age of two and had not been diagnosed with an additional difficulty by three years post implant. All children had pre-implant hearing losses compliant with NICE guidance (NICE, 2009); i.e. pure-tone audiometric thresholds greater than 90 dB HL at 2 & 4 kHz and they were all congenitally deaf. Ages at operation ranged from 8-23 months. All lived in families where spoken English was used within the home, although for some children, other spoken languages were also present. All children used spoken language as their main communication mode. However, depending on the philosophy of their family and local education authority, they may also have been encouraged to develop sign skills. Within these parameters, the children represent a wide range of abilities, family circumstances, usage, language learning aptitude, local provision and approach, as would be typically expected for the population supported by a large cochlear implant programme in the UK.

The validity of the profile's structure was assessed by identifying the duration of sound processor use that was required for each skill to emerge and be acquired by a statistical majority of the 30 children; i.e. by determining the 'average' trajectory for the emergence and establishment of each skill. For example, for a sample size of 30 children a skill was considered to have emerged or acquired if it had been marked as such in at least 21 children (70%). This approach was preferable to considering a majority to be anything over 50% as it accounted for the size of the sample being assessed and confirmed statistically whether more than 50% of children had acquired each skill². Each child was assessed on six occasions: the five occasions corresponding to the five Milestones (3, 6, 12, 24, and 36 months) and after 48 months of sound process or usage. This final assessment provided information on the trajectory of any skills that had not been

established after 36 months of sound processor use. The analysis produced a duration of sound processor use for each skill that could be compared against the expected rate of skill acquisition that underpinned the structure of the profile; i.e. to assess whether the skills for Milestone One were established in a majority of children by 3 months of implant use. The results of these analyses were also used to characterise the expected range of the profile scores in a typically-developing cohort at each of the five assessment points.

Results

Validation of profile structure

The duration of sound processor use after which each skill was observed to have emerged or been established by the majority of children was found to be in close agreement with the rates that were assumed when developing the profile structure. All skills in all milestones had been recorded as emerging in a statistical majority of children by the expected assessment time points (Table 2). All skills in Milestones One, Two, and Three had also been recorded as having been established in a majority of children at the expected time points. The analysis identified some skills that emerged in a majority of children earlier than expected: skills 2, 3, and 4 in Milestone 3 emerged by 6 rather than 12 months, skills 2 and 3 in Milestone 4 emerged by 12 rather than 24 months, and skill 1 in Milestone Five emerged by 24 rather than 36 months. Some skills were also established slightly later than expected (Table 2): skills 7-10 in Milestone Four were established in a majority of children by 36 rather than 24 months and skill 10 in Milestone Five was established in a majority by 48 rather than 36 months. Thus, the timing of when all skills emerged and when 46 of the 50 skills were established supported the validity of the profile structure that arranged key skills under five Milestones representing different durations of sound processor use.

Figure 1 shows the expected ranges in which 95% of profile scores fall within across the five primary assessment points; i.e. 3, 6, 12, 24, and 36 months of sound processor use (see Table 3 for the range of expected scores at each assessment point). The scores of the cohort of children followed the expected trajectory in that they increased by an average of approximately 20 points between each assessment (mean increase of 20, 16, 17, and 15 for the change from 3-6 months, 6-12 months, 12-24 months, and 24-36 months respectively). A wide range of scores was observed at each assessment time point, with the difference between the lowest and highest scoring child being over 45 points at the 3, 6, 12, and 24 month time points. The observed ranges shown in Figure 1 and listed in Table 3 provide a useful reference that can be used to determine whether a child's profile score falls within or outside the expected range for scores from typically-developing children without any diagnoses of additional difficulties.

Case studies

The usefulness of the NAMES profile to monitor the progress of individual children is described through the three case-studies below which have been chosen to illustrate the wide range of possible outcomes for children with no identified 'at risk' factors within the assessment phase. Child A and Child B were included in the validation study previously described, but Child C was not included as additional difficulties were diagnosed within the first three years post implant. Individual skills are referenced against the five Milestones of the profile abbreviated as M1 to M5. All three children lived in the same educational authority so had the same level of local professional input and support. In this authority, this meant either weekly or fortnightly visits from a local teacher of the deaf and blocks of sessions from a speech and language therapist who held an additional specialist qualification to work with deaf children. They were all born at

full term following uncomplicated pregnancies and birth histories and in the assessment phase no concerns were raised about their general development or communication. All three children lived in settled home situations, with two parents and supportive extended family networks provided by grandparents and other relatives.

The children all had simultaneous bilateral cochlear implants under the age of 15 months with devices from Cochlear Ltd (Table 4). There were no surgical complications, they attended reliably for programming appointments and their parents were keen that they should develop spoken language. All three children were congenitally deaf; the cause of hearing loss was unknown for two of the children but in the case of child C, the cause was genetic-both parents were hearing but his maternal grandmother was profoundly deaf and communicated via BSL. The parents of children B and C all had some form of post sixteen education but had left college by eighteen. The parents of Child A were educated to postgraduate level and both held professional jobs. Pre-implant, all three families used some sign to support communication.

Child A, was the younger of two siblings. Her general development was on track although some motor milestones, such as sitting independently, were slightly delayed. She had a number of chest infections, particularly during the first year post implant. Despite this, she made a swift transition to processor use and her family were pro-active with care, keeping the equipment securely in place with a headband.

By three months post implant, she was pointing to sounds out of eye line [M1]. She knew which of her toys made a sound and was upset if the batteries were flat and they did not work [M2]. She detected all Ling sounds [M1] and the family were starting to observe responses to voices when she was not looking at the person talking [M1]. She was using

her own voice more purposefully [M1] alongside some signs, which the family then modelled back in words.

By six months post implant, she could recognise if either of her parents was moving around in an adjacent room and would shout for their attention [M2]. She was very responsive to music and would move to early action songs [M2]. She could link some symbolic sounds to actions, such as pretending to sleep when an adult said ‘*sh*’ [M3] and was attempting early social language such as ‘*bye-bye*’ [M3]. Her family had naturally dropped sign as she was responding to their words and checking back visually far less.

By twelve months, she was looking to an adult for help if either processor stopped working [M3]. She was using family names to get attention, she could follow instructions such as to bring a particular toy, could point to items on pictures and could request her favourite actions songs by offering a word and associated action [M3]. She was also developing some auditory closure skills [M4], i.e. if the adult paused, she could provide the final word for a familiar phrase.

By two years post implant, she was attending nursery two days a week and listening well to group stories and making use of a Cochlear MiniMic system. She reported problems with the processors using expressions such as ‘*my ears off*’. She could hold a simple conversation over distance, could follow linked instructions and remember lists of three items [M4]. She was also able to sing parts of favourite songs by herself and join in shared recounts of family events, such as her birthday party [M4].

By three years, she was able to retell a favourite story independently [M5], understand and give verbal instructions for a game [M5] and could correct the mistakes of others

[M5]. She could take her part in a group conversation with familiar adults and talk on the telephone to her grandparents [M5]. Her ability to give independent recounts of her own experiences and to follow more complicated instructions was developing but not fully established but this linked very much to her personality and general maturity.

In this case, Child A developed auditory skills and spoken language as predicted and her NAMES score followed the expected trajectory (see solid line in Figure 1). The profile helped to re-assure the family that all was well, particularly given the periods of illness in the first year and it provided a focus for new expectations, for example the need to listen to the ideas of others within a group rather than adults always following her lead.

Child B was the first child of hearing parents; initial programming went smoothly and the processors were well accepted. By three months post implant, she had made rapid progress and already recognised some familiar sounds [M2] and behaved differently in response to new sounds [M2]. She detected all Ling sounds [M1], rocked in response to music [M2] and had become more vocal [M1]. She knew the difference between a happy voice and a raised, warning tone and reacted accordingly [M1].

Between the three and six month interval, she began to explore and remove her processors more often but her parents were persistent in replacing the devices and extended daily use was the norm. She was highly responsive to sound and could recognise the tune and attempt the actions for her favourite songs [M3]. She recognised a wide number of everyday sounds [M2], listened to others talking and took her turn vocally, even when her visual attention was elsewhere [M2]. She understood some everyday phrases and words linked to routines and her favourite games [M3]. As a result, she continued to progress at a faster than average pace on the profile.

However, between the six and twelve month interval, her younger sibling was born and she had a number of quite severe ear, throat and chest infections. Parental attention was reduced and consistent processor use was interrupted during the periods of illness, although she was happy to accept the equipment once she recovered and would go to an adult for help to replace the coil [M3]. She also began a part-time placement at a local nursery within a large, relatively noisy room. She continued to love action songs and joined in vocally alongside actions but was not yet attempting any of the words. She understood a range of familiar instructions and could bring single items on request [M3]. She joined in with social routines, such as saying '*hiya*' and '*please*' [M3] and could link a wide range of symbolic sounds and early words to objects and people [M3]. Although rate of progress had slowed, it was still in line with average expectations.

After two years, attendance at nursery had increased to two days a week and staff expressed concern that she sometimes arrived without the equipment in good working order. Her mother suffered from periods of extreme exhaustion during which time she found it hard to manage the demands of two young children. Child B also continued with several episodes of relatively minor illnesses but the accumulative impact was noticeable. On at least two occasions this resulted in periods of hospitalisation and at other times, she was generally unwell across periods of several days which affected not only sound processor use but also her general interest in play and communication with others.

Although new auditory skills were developing, many were inconsistent and as a result, progress on the profile slowed. She was able to sing fragments of the words of familiar songs such as '*Twinkle-Twinkle*' [M4]. She could select one and sometimes two items within very familiar games although this was not yet typically generalised into everyday

activities. She understood that when adults talked to her, she needed to make a response but she often simply echoed what she heard. She could answer a few very familiar questions and if adults paused within a familiar language routine, she could sometimes add a final word to a phrase [M4].

The profile allowed us to show family and local staff why our concerns had grown and raised the need for more structured input and support. By three years, episodes of ear and throat infections had reduced in frequency and severity but sleeping remained poor and she was prescribed melatonin. She was listening more actively to conversations around her, particularly if she thought they related to her. At nursery, she followed instructions and some activities within a group [M5] and she needed less adult prompting to understand and respond to simple comments and instructions [M4]. She answered questions but quite often her responses, although related to the topic, were not completely accurate. She noticed when adults make mistakes in very familiar language routines and would correct them [M5]. She could remember three linked items [M4] and with a high level of support could give information or share a simple retell with an adult [M4]. Overall understanding and verbal reasoning was now significantly delayed.

The NAMES profile provided evidence of the likely impact on learning to local professionals who saw Child B as a willing little girl who fitted in well with her peers and had social language. It gave her family evidence to support a request for additional support in the short term and in readiness for school entry the following year. A comparison of the scores of Child B with the expected range of scores from children without diagnoses of additional difficulties, reflect the time points at which concerns started to emerge. While initially within the middle of the range of expected scores, her

score had fallen to the bottom end of that range by 12 months and was well within the lower quartile by 24 months (see dotted line in Figure 1).

Child C was the first child of hearing parents with a deaf maternal grandmother. Although early acceptance of the equipment was good, as programming progressed, he showed intolerance of certain sounds, at which point he would immediately remove the equipment. There were also some concerns expressed by local staff about consistency of processor use and basic maintenance, although parents reported daily use. By three months, he was observed to react to a wide range of speech sounds [M1] and to attempt to copy some simple babble patterns. Parents also reported noticeable changes in his responses to everyday sounds [M1].

By six months, concerns were raised about his attention and he was described as either very distractible or so over-engrossed in activities that it was not possible to develop turn-taking. He would tolerate the processors with adult supervision but without this, they would immediately be removed and pulled apart so their use was not possible in places such as the car, buggy or if an adult had to leave the room. With processors in place, he was immediately much more vocal [M1] and could recognise sounds such as the barking of the family dog in an adjacent room or a knock on the front door [M2]. He responded to his father's voice when he called out on return from work but otherwise auditory awareness of voice was inconsistent. He imitated some word contours but spontaneous vocalisation did not show the influence of access to sound. Although new auditory skills were emerging they were not consistent and this, together with his overall play and attention, flagged up concerns.

By twelve months, he was typically wearing the processors for parts of most days but was still removing and damaging them in frustration or boredom. There was some progress in his interaction and play. He attended a playgroup with additional adult support for two sessions a week where he followed a structured listening and early language programme. His tolerance of the full range of sounds had established and he rocked in response to singing [M2]. His vocalisations were more purposefully communicative and in play activities, he could settle, listen and respond vocally [M2]. He was able to link a few symbolic sounds to activities, such as *'whee'* with a toy going down a slide [M3] and was beginning to understand and attempt some social language such as *'hello'* and *'no'* [M3]. At this stage, in consultation with the family and local professionals, the decision was made to introduce more sign to support communication, alongside a referral to a paediatrician for further advice.

By two years, he was in a daily part-time nursery placement. However, he displayed many sensory intolerances, particularly in noisy environments, and he needed 1:1 adult supervision. Tolerance of the processors was fully established: he would actively seek to replace them if they fell off and was upset if for any reason they were not working [M3]. He could listen with attention to short, structured activities in quiet but could not attend within a larger environment. He could recognise the voices of familiar adults, could imitate a range of symbolic sounds and link them to the correct object [M3] and could follow simple instructions in context [M3]. He could sometimes select a single item from a small group [M3]. He was initiating more communication and spontaneously using some early words such as *'please'*, *'more'* and *'yes'*. His signing was also developing but although vocabulary in this mode was wider, it also remained at single sign level. Rate of progress meant that the gap with typical expectations was widening. The profile provided

a vehicle to record new achievements with his family at the same time as ensuring that the on-going concerns remained highlighted.

By three years, he was in school with full-time support and had received a diagnosis of mild autism, linked to his sensory intolerances, rigid behaviours and language difficulties. Home life was easier as he could accept some changes of routines, understand some simple verbal reasoning and at school there were fewer episodes of unpredictable behaviour. Processor use remained well established and there was a noticeable improvement in his auditory awareness and attention. He was able to identify where an adult was in the house from hearing them move about. He could select single items and with adult scaffolding together with cues, such as photographs, he could provide very simple retells of events [M4]. He was echoing far more fragments of what he heard but also beginning to listen and respond to some simple instructions, comments and questions [M4]. He had a small but growing vocabulary of words and two word combinations were beginning to emerge.

The NAMES profile enabled us to talk to his parents from the early stages about how progress with listening was dependent on processor use. Once use was established and rate of progress remained very slow, it provided evidence to support the need for further investigations and remained a relevant way to record progress and set targets for a child who was not developmentally ready to complete standardised language assessments. A comparison of his profile scores against the expected trajectory was compatible with concerns being raised after only 6 months as his score was already within the bottom quartile of observed scores, and subsequent scores were in the bottom 2.5% of scores or even outside of the range of expected scores (see dashed line in Figure 1).

Discussion

. The validation exercise demonstrated that although there is wide individual variation, the order and rate at which skills are predicted to develop within the NAMES profile is a useful guide for typically developing children implanted under two years of age. As illustrated by the case studies above, the NAMES profile alongside standardised language assessments provides a framework to set consistent expectations, evidence the progress of young children, and a mechanism to log and raise concerns as required. The expected trajectory of the NAMES profile has been validated for the implanted under two population. While the structure of the framework can be used with a wider age range, for children with unilateral cochlear implants and for more complex children, it is with caveats about the anticipated rate of progress.

Even within the criteria set for the validation study, i.e. a group of children who were all congenitally deaf and under two at the time of their operations with no additional diagnosed difficulties, a wide range of scores was observed at each interval point with the deviations becoming more noticeable by Milestones 4 and 5. This may initially appear to be a surprising finding, given that the children were all very young, many being under twelve months of age when they received bilateral cochlear implants. However, they were not specifically selected beyond a few key parameters and therefore represent the wide range of cognitive abilities, language learning aptitudes, family situations, understanding and involvement, as would be typical in any population of very young children managed by a large cochlear implant programme. Children's lives are complicated and impacted upon by many unpredictable factors, such as a change in family circumstance, the loss of a parent, the birth of a sibling, periods of illness, the choice of nursery placement or the amount and quality of professional support, all of which can accelerate or adversely affect overall development, emotional well-being,

opportunities to learn and therefore listening and language progress. Attempts to capture and quantify the inter-relationship between these factors are necessarily simplistic and incomplete and there needs to be an acknowledgement that they can often outweigh and over-ride the advantage conferred by early operations and the best possible technology. Once the full range of children is considered, the impact of these factors becomes even greater.

. The profile is designed to build family understanding and confidence by helping them to identify and validate what their child is already able to do and then encouraging them to formulate their own ideas about how to develop new skills based on their child's functional communication and listening needs in daily life. The accumulated evidence provided by the profile of what are now routine expectations for young children with cochlear implants, has also supported efforts to change professional attitudes and approaches in the wider community.

The profile and accompanying materials² were not designed to replace standardised assessment tools but to supplement them. It is important that assessors do not rely on looking at scores alone but at what the profile shows about current patterns of strengths, and areas for potential future development. At NAIP, children are only reviewed using the profile at the allotted time points. This means only an annual assessment from Milestone Three onwards. It is therefore not possible to state at which point over the next twelve months, skills which had not emerged or were secure, actually developed. However, the profile is flexible enough to allow for more regular updates and this would provide more detail. The flexibility of the profile also makes it more open to individual interpretation by assessors and this could potentially become a weakness if it led to inconsistent judgements and recording. Best practice is therefore to implement an on-

going system of inter-reliability meetings, particularly if the child's case is managed by different professionals over the three year time-scale.

While the profile provides a mechanism to flag up those children not making the expected progress, it is essential that the results are interpreted in line with a broader understanding of the child's social and emotional circumstances. The materials themselves are of limited value unless they are interpreted with sensitivity and with constant re-evaluation, in collaboration with the families and all those involved with the child. This highlights the essential role of professionals with high levels of expertise and sensitivity, who can coach and provide interventions, differentiated as required for the wide range of individual circumstances.

The younger the child at the time of their initial operation, the closer the skills in Milestones 4 and 5 are to their overall developmental age, and the increased level of challenge created by this maturation effect needs to be considered. Many children now have operations at under a year of age and are therefore just four years old by the end of a three year period of monitoring covered by the NAMES profile. This indicates a need to extend the profile to continue the benefits provided by the framework to the monitoring of later stages of development. It would also provide the opportunity to study the ongoing trajectory of skill acquisition for this population in relation to their hearing peers. This work is currently underway within the team.

Conclusion

The NAMES profile has been used consistently by NAIP for the last 8 years and is not only a key way for the team to record and measure progress but also reflects our philosophy that effective intervention should be family, not child, centred. While it is not

possible to predict postoperative outcomes for babies and young children with any sensitivity in the assessment phase, the use of a profile such as NAMES allows judgements about progress to be made from a very early stage post implant. This should lead to the earlier differential diagnosis for some children, together with recommendations for more appropriate support and intervention strategies.

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Footnotes

¹Staff who comprise the Outreach Support Team have professional backgrounds in auditory verbal therapy, deaf education and speech and language therapy.

²21 out of 30 children as a proportion (70%) has a confidence interval from 52% to 83%. 70% was used to signify a majority as it was the smallest proportion whose lower confidence interval was >50%; i.e. the smallest proportion statistically greater than 50%.

³The materials are available on the NAIP website (www.nuh.nhs.uk/naip) and subsequent to their development, have been published by Advanced Bionics Ltd (advancedbionics.com).

References

- Archbold, S., Harris, M., O'Donoghue, G., Nikolopoulos, T., White, A., & Richmond, H. L. (2008). Reading abilities after cochlear implantation: The effect of age at implantation on outcomes at 5 and 7 years after implantation. *International journal of pediatric otorhinolaryngology*, 72(10), 1471-1478.
- Archbold S., Mayer,C. (2012) Deaf Education: The impact of cochlear implantation. *Deafness and Education International*,14(1)
- Barker, B. A., Kenworthy, M. H., & Walker, E. A. (2011). How we do it: Employment of listening-development criteria during assessment of infants who use cochlear implants. *Cochlear implants international*, 12(1), 57-59.
- Birman, C. S., Elliott, E. J., & Gibson, W. P. (2012). Pediatric cochlear implants: additional disabilities prevalence, risk factors, and effect on language outcomes. *Otology & Neurotology*, 33(8), 1347-1352.
- Boons, T., Brokx, J.P., Dhooge, I., et al. (2012) Predictors of spoken language development following pediatric cochlear implantation. *Ear and Hearing*, 33(5):617-39
- Ching, T.Y., Dillon, H., Marnane, V., Hou, S., Day, J., Seeto, M., Crowe, K., Street, L., Thomson, J., Van Buynder, P. and Zhang, V. (2013) Outcomes of early and late-identified children at 3 years of age: Findings from a prospective population-based study. *Ear and Hearing*, 34(5), p.535-52

- Colletti, V., Carner, M., Miorelli, V., Guida, M., Colletti, L., & Fiorino, F. G. (2005). Cochlear implantation at under 12 months: report on 10 patients. *The Laryngoscope*, 115(3), 445-449.
- Datta, G., Odell, A., & Durbin, K. (2010). Nottingham Auditory Milestones: a profile to monitor the growth of active listening, understanding, auditory memory, and sequencing in babies and young children in the first three years after receiving cochlear implants. *Cochlear implants international*, 11(sup1), 282-285.
- Department for Education and Skills(DfES) Monitoring protocol for deaf babies and children 2004, *Dept for Education and Skills*
- Edwards, L. (2003) Candidacy and the Children's Implant Profile: Is our selection appropriate? *International Journal of Audiology*, 42, 426-431.
- Flexer, C. (2011). Cochlear implants and neuroplasticity: linking auditory exposure and practice. *Cochlear implants international*, 12(sup1), S19-S21.
- Geers, A. E., & Nicholas, J. G. (2013). Enduring advantages of early cochlear implantation for spoken language development. *Journal of Speech, Language, and Hearing Research*, 56(2), 643-655.
- Geers, A. E., Nicholas, J. G., & Sedey, A. L. (2003). Language skills of children with early cochlear implantation. *Ear and hearing*, 24(1), 46S-58S.

Harris, M.S., Kronenburger, W.G., Gao, S, Hoen, Miyamoto, R.T., H.N., Pisoni, D.B.

(2013) Verbal short-term memory development and spoken language outcomes in deaf children with cochlear implants. *Ear and Hearing*, 34(2):179-92.

Inscoe J., Odell A., Archbold S. & Nikolopoulos T. (2009) Expressive spoken language development in deaf children with cochlear implants who are beginning formal education. *Deafness and Education International*, 11(1): 39-55.

Lee, D. J., Lustig, L., Sampson, M., Chinnici, J., & Niparko, J. K. (2005). Effects of cytomegalovirus (CMV) related deafness on pediatric cochlear implant outcomes. *Otolaryngology—Head and Neck Surgery*, 133(6), 900-905.

Mayer, C. What really matters in the early literacy development of deaf children. *Journal of Deaf Studies and Deaf Education* 12.4 (2007): 411-431.

Niparko, J.K., Tobey, E.A., Thal, D.J., Eisenberg, L.S., Wang, N.Y., Quittner, A.L., Fink, N.E. and CDaCI Investigative Team. (2010). Spoken language development in children following cochlear implantation. *JAMA*, 303(15), 1498-1506.

National Institute for Health and Care Excellence (NICE). (2009). Cochlear Implants for children and adults with severe to profound deafness: Technology Appraisal Guidance 166. Available from <http://www.nice.org.uk/ta166>.

- Nikolopoulos, T. P., Archbold, S. M., & Gregory, S. (2005). Young deaf children with hearing aids or cochlear implants: early assessment package for monitoring progress. *International journal of pediatric otorhinolaryngology*, 69(2), 175-186.
- Raine, C. (2013). Cochlear implants in the United Kingdom: awareness and utilization. *Cochlear implants international*, 14(sup1), S32-S37.
- Ramirez-Inscoe, J., Bones, C. (2016) Additional difficulties associated with aetiologies of deafness: outcomes from a parent questionnaire of 540 children using cochlear implants *Cochlear Implants International*, 17, (1) 21-30
- Rinaldi, P., Baruffaldi, F., Burdo, S., Caselli, M.C., (2013) Linguistic and pragmatic skills in toddlers with cochlear implants. *International Journal Communication Disorders* , 48(6) 715-25
- Sach, T. H., Whynes, D. K., Parker, P., & Archbold, S. M. (2004). Innovation and funding specialist services: Cochlear implantation. *Journal of health organization and management*, 18(1), 53-63.
- Sarant, J., Harris, D., Bennet, L., & Bant, S. (2014). Bilateral versus unilateral cochlear implants in children: a study of spoken language outcomes. *Ear and hearing*, 35(4), 396.

- Sarant, J. Z., Hughes, K., & Blamey, P. J. (2010). The effect of IQ on spoken language and speech perception development in children with impaired hearing. *Cochlear implants international*, 11(sup1), 370-374.
- Sarant, J. Z., Holt, C. M., Dowell, R. C., Rickards, F. W., & Blamey, P. J. (2008). Spoken language development in oral preschool children with permanent childhood deafness. *Journal of Deaf Studies and Deaf Education*, 14(2), 205-217.
- Sheridan, M., Frost, M., Sharma, A., From Birth to Five Years, Children's Developmental Progress. (1997) *Publ Routledge*
- Van Wieringen, A., & Wouters, J. (2015). What can we expect of normally-developing children implanted at a young age with respect to their auditory, linguistic and cognitive skills?. *Hearing research*, 322, 171-179.
- Verhaert, N., Willems, M., Van Kerschaver, E., & Desloovere, C. (2008). Impact of early hearing screening and treatment on language development and education level: Evaluation of 6 years of universal newborn hearing screening (ALGO®) in Flanders, Belgium. *International journal of pediatric otorhinolaryngology*, 72(5), 599-608.
- Yoshinaga-Itano, C., Sedey, A. L., Coulter, D. K., & Mehl, A. L. (1998). Language of early-and later-identified children with hearing loss. *Pediatrics*, 102(5), 1161-1171.
- Yoshinaga-Itano, C. (2013). Principles and guidelines for early intervention after confirmation that a child is deaf or hard of hearing. *Journal of Deaf Studies and Deaf Education*, 19(2), 143-175.

Figure 1: The distribution of NAMES profile scores at each assessment time point following cochlear implantation in children implanted under two years of age. The shaded regions illustrate the range that covered 95% of profile scores; i.e. the range between the 2.5th and 97.5th percentiles. Solid, dotted, and dashed lines plot the profile scores of the three case studies.

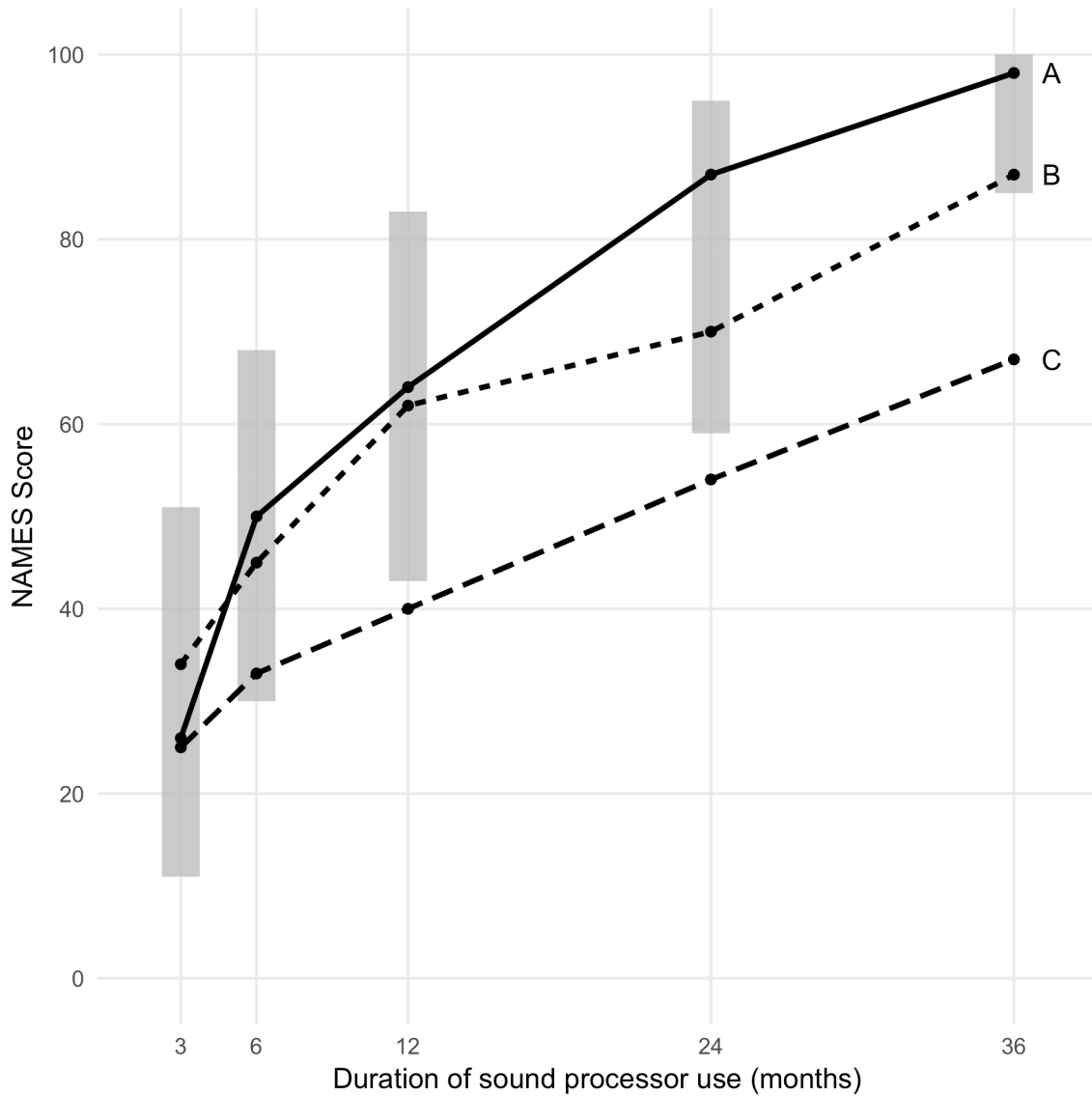


Table 1: The structure of the NAMES profile, comprising five Milestone stages, and the constituent skills of each Milestone.

Skill	Milestone 1 (3 months of use)	Milestone 2 (6 months of use)	Milestone 3 (12 months of use)	Milestone 4 (24 months of use)	Milestone 5 (36 months of use)
1	Wears processor for extended periods	Adults can notice definite changes in child's behaviour if the system stops working.	Child recognises if the system stops working.	Sings fragments of familiar tunes and songs.	Overhears and responds appropriately to conversation not specifically directed towards them.
2	Noticeable changes in behaviour when the processor is worn.	Recognises some familiar everyday sounds.	Joins in with the appropriate actions and later with vocalisations for familiar action songs [without prompting].	Understands simple, one step directions without relying on context or predictable routine.	Able to listen as part of a small group and to pick up the key messages and instructions alongside their peers.
3	Detects and reacts to environmental sound.	Is puzzled/ more interested in new or unusual sounds	Recognises voices.	Constantly imitates fragments of what they hear [and overhear].	Able to have an age appropriate telephone conversation with a familiar adult.
4	Definite turns to locate interesting sounds.	Moves to music	Knows and turns to name even in less favourable conditions.	Can complete simple auditory closure activities.	Enjoys memorising funny expressions, rhymes and messages. Remembers lists of 4-5 items.
5	Responds to music and noise making toys.	Listens to others talking	Links a range of symbolic sounds and early phrases to objects/events.	Can pick out two objects from a set of familiar objects.	Can solve riddles, which involve identifying objects on basis of 3 key features.
6	Attention can be captured by voice only.	Consistently takes a vocal turn.	Joins in with language of social routines.	Responds to simple questions.	Follows more complex instructions and questions.
7	Interested in talk directed towards them.	Non-looking vocal turns established.	Understands a questioning voice.	Can pick out three objects at one time from a set of familiar items.	Can answer simple questions about themselves. Understands simple 'why' questions.
8	Affected by tone of voice.	Vocalisations are influenced by what they hear.	Demonstrates understanding of familiar phrases and simple instructions strongly linked to context.	Can identify an object/person from a minimum choice of four using two key features.	Notices deliberate mistakes in familiar spoken language routines and stories.
9	Use own voice purposefully.	Recognises own name in favourable conditions.	Can select one item from a small set of familiar objects.	Follows two part instructions.	Has favourite stories, often repeated. Remembers and uses phrases from stories.
10	Detects a wide range of sounds across the speech frequencies.	Demonstrates specific responses to wide range of sounds.	Anticipates and enjoys the sequence of familiar stories and songs.	Can complete a simple sequence of known events. Recounts events with prompts.	Retells/recounts simple stories/events independently.

Table 2: The proportion of children for whom each skill was recorded as emerging or established by the expected time point for each Milestone. Values highlighted in bold font indicate that a statistical majority of the sample did not reach the expected milestone at the expected time point.

Skill	Proportion of children (% skill emerged / % skill established)				
	Milestone 1 (3 months of use)	Milestone 2 (6 months of use)	Milestone 3 (12 months of use)	Milestone 4 (24 months of use)	Milestone 5 (36 months of use)
1	100% / 93%	97% / 90%	97% / 93%	100% / 90%	100% / 90%
2	97% / 97%	97% / 97%	97% / 93%	93% / 93%	100% / 86%
3	100% / 90%	97% / 93%	97% / 93%	97% / 93%	93% / 86%
4	97% / 97%	97% / 97%	97% / 93%	93% / 77%	90% / 81%
5	97% / 97%	97% / 90%	97% / 97%	90% / 83%	86% / 75%
6	97% / 90%	100% / 93%	97% / 93%	97% / 83%	89% / 83%
7	93% / 93%	93% / 76%	93% / 87%	76% / 52%	100% / 84%
8	86% / 72%	97% / 86%	97% / 87%	73% / 59%	97% / 74%
9	100% / 90%	97% / 97%	93% / 90%	80% / 60%	97% / 88%
10	93% / 83%	97% / 83%	97% / 93%	80% / 53%	96% / 65%

Table 3: Ranges of expected NAMES profile scores at the five key assessment periods that align with the five milestones of the profile.

50% of children	95% of children	All children	Duration of sound processor use (months)				
			3	6	12	24	36
		Minimum	6	10	12	49	84
	2.5 th %ile		11	30	43	59	85
25 th %ile			20	42	62	79	94
75 th %ile			33	54	67	87	100
	97.5 th %ile		51	68	83	95	100
		Maximum	52	68	84	98	100
Full range (minimum to maximum)			46	58	72	49	16
Interquartile range (25 th to 75 th %ile)			13	12	5	8	6

Table 4: Characteristics and NAMES scores of the three case studies.

	Gender	Age at implantation (months)	Duration of sound processor use (months)				
			3	6	12	24	36
Child A	Female	12	26	50	64	87	98
Child B	Female	10	34	45	62	70	87
Child C	Male	15	25	33	40	54	67