OPPORTUNITIES WITHIN AND AFTER REHABILITATION

for patients with hearing loss

Tirza van der Straaten

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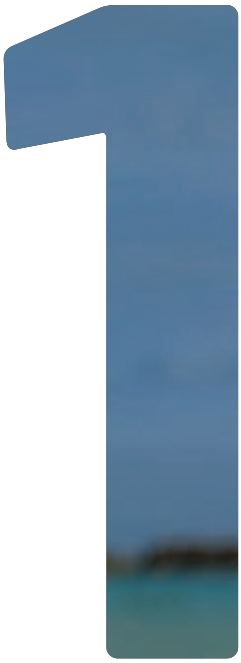
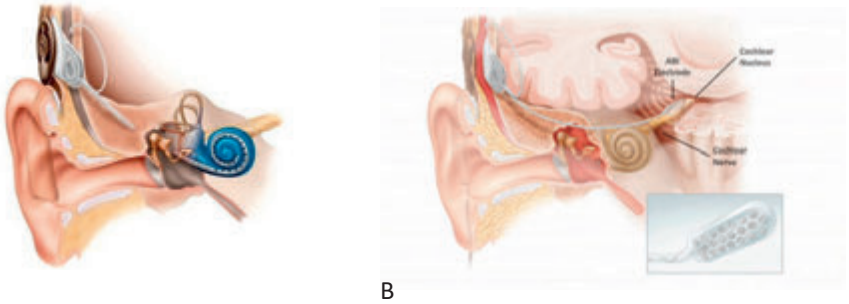
Door de vrolijke kleuren van Curaçao gaat men vanzelf dansen



Introduction

Cochlear implantation procedures first started in the Netherlands in 1985, under scientific restrictions which led to the reimbursement and implementation of cochlear implants (CI) as a standard of care for the severely hearing-impaired in 2000. Until now, more than 7.500 individuals have received CI in the Netherlands and almost 600 implantations are performed every year (CI-ON, 2019). More recently, auditory brainstem implantation (ABI) became available for children with non-functional cochlea's or cochlear nerves (Figure 1). Over the past couple of years, 12 deaf children have undergone this new procedure at the Leiden University Medical Center (The Netherlands). These implantations and other developments in rehabilitation have significantly changed the lives and future prospects of hearing-impaired individuals. Yet, the actual impact of these recent implementations and expansions in rehabilitation remains unclear. What are current patients' expectations when it comes to rehabilitation? May an individual with hearing loss (HL) expect to fully participate in a world driven by sound and verbal communication after rehabilitation, or should he or she accept the consequences of a chronic handicap?



A B

FIGURE 1. The electrode array of the cochlear implant **(A)** is inserted circa 1.5 turns in the scala tympani of the cochlea and lays alongside nerve endings of the cochlear nerve. The array of an auditory brainstem implant **(B)** is inserted alongside the cochlear nucleus of the brainstem in the lateral recess of the fourth ventricle.

Developments in rehabilitation, a short history

In the early days of rehabilitation, individuals with HL were allocated to deaf communities until CI became mainstream in 2000 (Tijsseling, 2014). This new technique achieved several improvements in pediatric and adult rehabilitation, starting with auditory input and speech understanding. Initially, only profoundly deaf individuals were eligible for implantation (Frijns et al., 2002), but criteria gradually expanded to include severely hard of hearing individuals who could understand 30-40% of monosyllabic words. Nowadays, selection criteria have been expanded even further. At the Leiden University Medical Center, CI is already an option for individuals with residual hearing, who can still understand more

than 60% of words but have severe problems in complex listening environments with background noise (CI-ON, 2013; Snel-Bongers et al., 2018).

Another important milestone in the development of rehabilitation for patients with HL, is the implementation of early identification of congenital HL. Initially, children were diagnosed with HL and rehabilitated with hearing aids (HAs) at around 2 years of age (Yoshinaga-Itano, 2004) and received their CI at 3 to 4 years of age (Lammers et al., 2015). However, the plasticity of the brain decreases with age, making the brain less susceptible to auditory and language input as children grow older (Niparko, 2010). Between 2003 and 2005, newborn hearing screening was implemented in the Netherlands, changing the lives of children with congenital HL (Korver et al., 2013). Today, early identification of HL in the first days after birth results in early intervention and is CI implantation possible at 6 months to 1 year of age (CI-ON, 2013). For these children, early implantation enables early auditory input during the linguistic development phase of the brain and significantly improves language skills (Boons et al., 2013; Niparko, 2010; Yoshinaga-Itano, 2003a).

Improved spoken language skills offer major benefits for children's development in many areas, as it enhances their possibility to participate in a world driven by verbal communication (Boons et al., 2013; Korver et al., 2010; Niparko, 2010; Pimperton & Kennedy, 2012; Yoshinaga-Itano, 2003a). Nowadays, a large proportion of children with CI are able to attend mainstream instead of special schools.

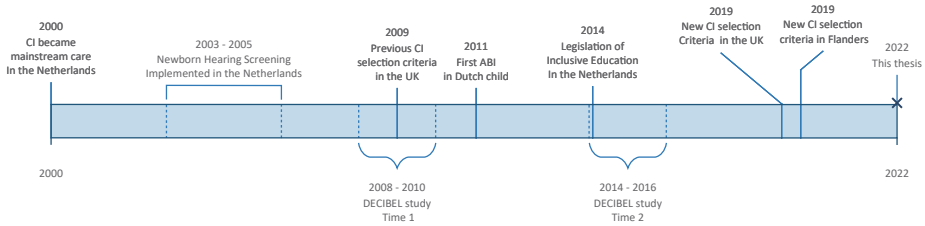


FIGURE 2. Timeline of important milestones in rehabilitation for patients with HL

In addition, new rehabilitation techniques have evolved, resulting in ABI implantation when CI is not suitable. Initially, only deaf adults with neurofibromatosis type 2 were candidates for this procedure (Schwartz et al., 2003). However, research by Sennaroglu et al. and Colletti et al. have shown that prelingually deaf children were also good candidates for this type of implant (Colletti et al., 2014; Sennaroglu et al., 2009). This led to the first pediatric auditory brainstem implantation in 2011 in the Netherlands. Indications for ABI implantations are congenital malformation of the cochlea or cochlear nerve (Figure 3) and

ossification of the labyrinth after meningitis or after a base skull fracture (Sennaroğlu et al., 2016). Prelingually deaf children with ABI are now able to identify environmental and speech sounds and even develop intelligible speech within 5 years after implantation (Noij et al., 2015).

Impact of expanded rehabilitation for patients with HL

Rehabilitation for patients with HL has matured, with many improvements made over the past 20 years. However, the impact of these different developments in rehabilitation is still unclear. What would happen if we broadened the selection criteria for CI and ABI even further? Can one expect to fully participate in society after rehabilitation? To what extent can we assume changes in the social-emotional well-being and educational attainment of hearing-impaired individuals? In other words, what can we expect from rehabilitation nowadays? These are the questions patients and parents currently ask when faced with the choice between CI or ABI. In order to answer these questions, one needs to understand that the hearing-impaired population is very heterogenous. This requires examining different outcomes taking into account these individual differences within the population, in order to investigate the impact of rehabilitation. Therefore, this thesis aimed to investigate different aspects of outcomes of current rehabilitation for patients with HL. The next sections will further discuss current knowledge and missing links in rehabilitation. The sections are divided into speech perception after adult CI, language development after pediatric ABI, and developmental outcomes after pediatric rehabilitation, such as social-emotional functioning and level of education.

Current selection criteria for cochlear implantation

The main goal of CI is to enable individuals with HL to participate more easily in a aurally oriented society, and therefore to improve their speech understanding. This is difficult to measure in children because they are mostly implanted before speech understanding has developed, but it can easily be measured in post-lingually deafened adults. Moreover, CI should be provided to candidates who are likely to benefit from the implant, while avoiding unnecessary costs and medical interventions for patients for whom acoustic HAs are sufficient. Technical developments of the implant and changes in surgical techniques have allowed postoperative speech perception outcomes to improve and for residual hearing

In the Netherlands, selection criteria for pediatric CI are based on the degree of HL (>70-80 dB) and the auditory response with HAs. Prerequisites for adults are primarily based on speech understanding (<60% word score or difficulty with listening in noise).

to be preserved (Blamey et al., 2013; Snel-Bongers et al., 2018). However, candidates with residual hearing exhibit relatively high preoperative speech scores which limit a 'remarkable' improvement of speech understanding postoperatively. Defining selection criteria therefore remains difficult, requiring further investigation.

Selection criteria are mainly based on preoperative audibility. Yet there is no golden standard worldwide and pre-implant prerequisites vary widely. Some countries only use the degree of HL as measured by pure-tone audiometry, since this test is easily available. Others use the level of preoperative speech understanding with or without taking into account the degree of HL. There are also different speech perception tests available and pure tone audiometry can be measured in various ways. Furthermore, the cut-off values of selection criteria vary from relatively strict in England and Flanders (Table 1) to lenient in the Netherlands (e.g., 80% of speech understanding), Germany, and Australia (Deborah Vickers et al., 2016a). These diverse type of measurements and selection criteria are remarkable as all countries pursue the same goal: rehabilitation for individuals with severe hearing problems, resulting in their increased participation and the improvement of their quality of life in a cost-effective way.

Therefore, we were interested in which preoperative measure would be most effective in indicating which post-lingually deafened candidate would improve after CI implantation. Previous research has found that the preoperative speech perception score is a valuable indicator for postimplant performance through prediction models (Cullen et al., 2004; Gomaa et al., 2003; Kraaijenga et al., 2016). However, the diagnostic value of different preoperative tests (the various speech understanding tests and pure tone audiometry) has not yet been analyzed. This was therefore our research aim in **Chapter 2**.

Selecting candidates for CI also involves the evaluation of patient-related characteristics. For example, the time at which HL is acquired: at birth, in early childhood (pre-lingual) or at an older age (post-lingual). In a post-lingually deafened adult, the level of speech understanding facilitates prediction of the possible postoperative benefit with CI (Cullen et al., 2004; Gomaa et al., 2003; Kraaijenga et al., 2016). This is different in pre-lingually deafened adults where the intelligibility of their speech production relates to the acquired auditory speech input and the potential postoperative outcome (van Dijkhuizen et al., 2016). Which patient-related factors are used varies widely and each country evaluates different factors such as the duration of deafness, age at implantation, and etiology of HL (Gomaa et al., 2003; Kraaijenga et al., 2016; Zhao et al., 2020). The socio-economic status of a candidate is also important in some countries where implants are only available through self-funding (Deborah Vickers et al., 2016a).

The United Kingdom and Flanders (Dutch-speaking part of Belgium) have recently broadened their selection criteria for CI in post-lingually deafened adults (Table 1)(National Institute for Health and Clinical Excellence, 2009, 2019; Raeve, de & Wouters, 2013). This enabled us to examine the possible increase in candidates who are able to improve their speech perception postoperatively. This is investigated in **Chapter 3,** alongside the sensitivity and specificity rate of the new selection criteria in both countries.

TABLE 1. Selection Criteria for Cochlear Implant Candidacy in the United Kingdom and Flanders

	United Kingdom	Flanders
Old criteria	>90 dB at 2 and 4 kHz and <50% sentence score (2009)	Average of >85 dB at 0.5, 1, and 2 kHz and <30% phoneme score (2013)
New criteria	≥80 dB at ≥2 frequencies (0.5, 1, 2, 3, and 4 kHz) and <50% phoneme score (2019)	Average of >70 dB at 0.5, 1, 2, and 4 kHz and <50% phoneme score (2019)

Language development after auditory brainstem implantation in congenital HL

ABI is a new and complex procedure that continues to develop. Implantations in young children with ABI are particularly challenging as they have never heard sounds before. They will experience the variety of frequencies and patterns of sounds after stimulation of the electrode array on the auditory brainstem. Additionally, the tonotopy of the auditory brainstem in the pediatric population remains nearly unknown and is difficult to mimic (Long et al., 2005). Nevertheless, the plasticity of the pediatric brain is assumed to adjust the auditory pathway according to the stimuli received from the ABI. The first results indicated that children with ABI can develop speech perception and speech intelligibility (Noij et al., 2015). Their auditory skills develop relatively slowly and reach lower levels compared with children with CI (Colletti et al., 2014; Sennaroglu et al., 2009). However, a direct comparison between pediatric CI and ABI users has not been made. When doing so, one should consider the differences between children who are eligible for CI or ABI. The presence of congenital cochlear malformations and cochlear nerve deficiencies (Figure 3) may also imply an impaired auditory pathway in the brainstem and further on in the brain (Sennaroğlu et al., 2016) and are most often present in the context of complex syndromes and/or additional comorbidities.

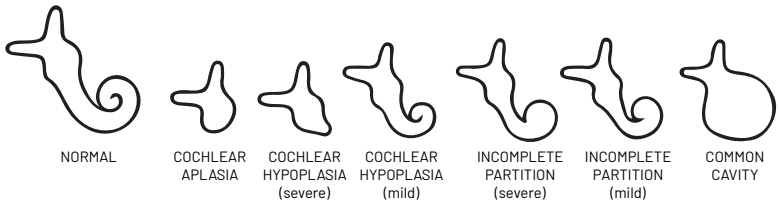


FIGURE 3. A normal cochlear anatomy and several congenital malformations of the cochlea (Jackler et al., 1987). CI is first attempted in some cases with mild cochlear hypoplasia. The other cases are not eligible for CI, but possible candidates for ABI.

Therefore, the prevalence of additional disabilities is expected to be higher in these children compared with children who receive CI (Sennaroğlu et al., 2016). Severe additional disabilities are related to lower levels and a broader variety of expected developmental outcomes in children with CI (Eze et al., 2013). Yet, the relation between additional disabilities and developmental outcomes in children with ABI has not yet been investigated. We will, therefore, examine the long-term auditory development of children with ABI and compare this with the auditory development of pediatric CI users in **Chapter 4**, taking additional disabilities into account.

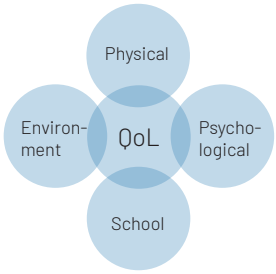
Developmental outcomes after pediatric rehabilitation

The direct benefit of rehabilitation in children with a HA or CI is difficult to capture. Children eligible for CI are unable to understand speech preoperatively which prevents us from examining direct benefits to speech understanding after implantation. Instead, the development of speech understanding and expressive language are closely monitored. Language development is therefore the first step in examining the impact of pediatric rehabilitation and has been studied extensively (Moeller et al., 2007; J. Bruce Tomblin & Moeller, 2015; van Schoonhoven et al., 2013). Nowadays, most children with CI or HA are expected to eventually acquire language skills comparable to normal hearing children (J. Bruce Tomblin et al., 2018). The age at intervention (implantation or amplification with HA) is one of the most important factors contributing to optimal and early language development in children with HL (Yoshinaga-Itano, 2004). It is clear that a child with HL can participate in a world driven by verbal communication, however, it is unclear to what extent. Research that examines the impact of HL on other areas of a child's development is relatively new and remains scarce. Two important pointers for examining the impact of rehabilitation are wellbeing and educational attainment of children with HL.

Wellbeing

Growing up in a world driven by sound and verbal communication can have a considerable impact on the development and identity of children with HL. Hearing impairment interferes with unplanned ('incidental') learning opportunities as not every conversation can be overheard and learned from (Luckner & Cooke, 2010), especially in social situations with a lot of background noise or voices coming from different directions, like at a playground or sport club. Studies have found that children with HL engage less in peer relationships and friendships than hearing children (Rieffe et al., 2018; Stevenson et al., 2015; S.C.P.M. Theunissen, Rieffe, Kouwenberg, et al., 2014). It might be due to the inability to keep up with their peers in conversations (Luckner & Cooke, 2010) or the incapacity of knowing how to socially communicate and interact (Netten et al., 2015; Rieffe et al., 2018; Anat Zaidman-Zait & Dotan, 2017). Misinterpretation of social situations can lead to feelings of exclusion, social isolation, and consequently, a lower quality of life (Contrera et al., 2017; Lin et al., 2013; Mathers et al., 2000). Elevated levels of psychopathologic symptoms (depression, anxiety, aggression, and behavioral problems) are found in children and adolescents with HL (S.C.P.M. Theunissen, Rieffe, Netten, et al., 2014). It is therefore unsurprising that children with HL appear to have a lower quality of life compared to their hearing peers (Roland et al., 2016). Within the quality of life domains, social interactions and school activities appear to be the principal problems these hearing-impaired children face.

Previous studies examining quality of life and its relation to HL related factors were performed in cross-sectional designs. These designs lack information relating to the effect of time and, consequently, the direction of causality. Therefore, we also need to study the development of children over time in order to identify causal factors. This research focus formed the basis for **Chapter 5,** where we examined the longitudinal development of quality of life in children with CI and HA. By studying the extent to which the quality of life of children with HL changes over time, the influence of language skills, type of hearing device, degree of HL, and type of education may be analyzed as possible risk or protective causal factors for a lower quality of life among hearing-impaired children.



The World Health Organization defines quality of life as an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards, and concerns (1995). This illustrates the main value systems for developing children.

Education

Great enhancements for the development of children with HL were brought about by the various improvements in rehabilitation. Pre-school and extra guidance at school have also enormously contributed to their development (Marschark & Spencer, 2011; J. Bruce Tomblin & Moeller, 2015). Children with HL and their parents are nowadays able to choose between special education for the deaf or mainstream education. When a child is able to keep up with mainstream education, it does not directly imply that school is not challenging for children with HL. They often need extra assistance in class, and face the challenge of misunderstanding teachers due to background noise or the direction in which they speak (Curle et al., 2017; Anat Zaidman-Zait et al., 2019). Learning to read is one of the biggest challenges for many hearing-impaired children (A. E. Geers & Hayes, 2011; Trybus & Karchmer, 1977; Worsfold et al., 2010) and they appear to underachieve in mathematical subjects (Gottardis et al., 2011). Consequently, pupils with HL frequently fail to pass grades (Gilani et al., 2017). It is therefore expected that pupils with HL are at risk of lower educational attainment and unemployment later in life (Dammeyer & Marschark, 2016; Qi & Mitchell, 2012). There is a lack of knowledge regarding the educational level of adolescents with HL during secondary education and the longitudinal effect of the different types of primary education (special and mainstream). In **Chapter 6**, this issue is addressed.

Outline of this Thesis

The PhD-project described in this thesis has investigated the various potentials of current rehabilitation, including pre-lingual and post-lingual HL, a direct benefit of speech perception after implantation, and the long-term effect on child development. This was performed in both cross-sectional (**Chapter 4**) and longitudinal studies (**Chapter 2, 3, and 5**), including a nation-wide study that included all individuals with HL born between 1995 and 2013 in the Netherlands (**Chapter 6**). The main outcomes of the studies are discussed in **Chapter 7**. In this chapter, we reviewed what one can expect from rehabilitation for patients with HL nowadays. We discussed the questions raised by patients and parents when faced with rehabilitation and further elaborated on prospective studies in future perspectives. A Dutch summary of this thesis can be found in **Chapter 8**.

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Diagnostic Value of Preoperative Measures in Selecting Post-lingually Deafened Candidates for Cochlear implantation: a Different Approach

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Under review

ABSTRACT

Objectives: We examined which preoperative diagnostic measure is most suited to serve as a selection criterion to determine adult cochlear implantation (CI) candidacy.

Design: 552 post-lingually deafened adults with CI underwent pure tone audiometry (PTA; 0.5, 1, 2, 4 kHz), speech perception tests (SPT) unaided with headphones and with best-aided hearing aids (in quiet and in noise). Gain in speech perception was used as outcome measure. Performance of preoperative measures was analyzed using the area under the curve (AUC) of receiver operating characteristic (ROC) curves.

Results: Best-aided SPT in quiet was the most accurate in defining which CI candidates improved their speech perception in quiet postoperatively. For an improvement in speech perception in noise, the best-aided SPT in noise was the most accurate in defining which adult would benefit from CI. PTA measures performed lower compared to the SPT measures.

Conclusions: SPT is better than PTA for selecting CI candidates who will benefit in terms of speech perception. Best-aided SPT in noise was the most accurate for indicating an improvement of speech perception in noise but was only evaluated in high performers with residual hearing. These insights will assist in formulating more effective selection criteria for CI.

INTRODUCTION

Inclusion criteria, to accurately select hearing-impaired patients for cochlear implantation, have been investigated extensively to determine the optimal pre-implant audiometric threshold values (Gubbels et al., 2017; Hoppe et al., 2015; Huinck et al., 2019; J. R. Leigh et al., 2016; Maeda et al., 2018; McRackan et al., 2018; Snel-Bongers et al., 2018; Deborah Vickers et al., 2016a). Typically, readily available clinical tests are used to evaluate the level of hearing loss (HL). This has resulted in many alternative tests being used across different countries. However, to date, no studies have focused specifically on which pure tone audiometric or speech measure(s) would be the most accurate in defining which post-lingual adult will improve their speech perception after receiving cochlear implants (CIs). In this retrospective study, we evaluated the accuracy of different preoperative measures in determining which post-lingually deafened adult will benefit from CI and to what extent they can function as a measure to determine candidacy for CI.

The intention of CI candidacy criteria is to ensure that a large proportion of patients will hear better with a CI than they do with a hearing aid (HA). The gradual shift in criteria for cochlear implantation has resulted in the consideration of patients with more residual hearing. These patients often have far higher pre-implant speech understanding abilities, which makes it more difficult to demonstrate substantial benefit in post-implantation speech perception.

The degree of rigidity for inclusion criteria and mechanisms for setting them differ per country and is mainly driven by reimbursement policies within each country (Deborah Vickers et al., 2016a). A cost element is often applied using a cost-benefit evaluation, or by limiting the number of implantations. Previous research has suggested that candidacy criteria should be based on the post-implant outcomes from the lowest 10th to 25th percentile (p10-p25). The associated cut-off values for preoperative pure tone audiometry (PTA) (Gubbels et al., 2017; Hughes et al., 2014) and speech perception tests (SPT) (Gubbels et al., 2017; Snel-Bongers et al., 2018; Verhaegen et al., 2008) were then used to define the pre-operative criteria. However, these criteria are based on the locally adopted speech tests (i.e., consonant-vowel-consonant words or sentence list unaided with headphones or best-aided, in quiet or noise) and/or audiometric frequencies evaluated (i.e., degree of HL at 2 or more frequencies), which vary greatly from country to country. No research to date has considered which preoperative PTA or SPT may be more appropriate to determine candidates who will benefit from CIs.

PTA relates to SPTs because the audibility of the speech signal affects its perception (Firszt et al., 2018; Hoppe et al., 2015; Lovett et al., 2015; Maeda et al., 2018). However, research has shown a weak to moderate correlation between PTA and unaided maximum monosyllabic word score indicating that outcomes of one measure cannot completely predict the other (Hoppe et al., 2015). Moreover, this latter unaided speech perception score with headphones frequently underestimates patients' best-aided speech perception in the free-field as the real-world communication abilities are not accurately reflected (McRackan et al., 2018).

Previous research identified predictors of post-implant outcomes that can be used to inform patients about their chances of improvement after CI (Blamey et al., 2013; Cullen et al., 2004; Gomaa et al., 2003; Gubbels et al., 2017; Hoppe et al., 2015; Kraaijenga et al., 2016; Rubinstein et al., 1999a). Some of these studies found that either the preoperative degree of HL (Gubbels et al., 2017; Hoppe et al., 2015; Rubinstein et al., 1999b) or preoperative speech scores (Cullen et al., 2004; Firszt et al., 2018; Gomaa et al., 2003; Gubbels et al., 2017; Hoppe et al., 2015) were valuable for predicting postoperative outcomes by using a multi linear regression analysis, correlation or pairwise comparison.

However, no research to date has compared the diagnostic performance of preoperative PTA and SPT for CI-candidates by calculating the predictive values with a binary classification. One study reported a sensitivity of 87% and specificity of 91% when an average PTA (0.5, 1, 2, 4 kHz) and the maximum monosyllabic word score with headphones were used as preoperative measures to predict the word score with HAs (Hoppe et al., 2015). They calculated this based on a simple linear formula where CI could be considered when the average PTA and unaided word score differed from each other. However, a comparison between the two different preoperative audiometric and speech measures was not conducted. It is still not known which PTA approach (average or threshold of one or more frequencies) is more effective at indicating which CI candidates will clinically improve their speech perception following implantation. Frequencies between 1 and 4 kHz are important for the discrimination of speech, especially for patients with high frequency loss who often fail to detect the consonant cues (Maeda et al., 2018). For example, the United Kingdom recently changed their CI candidacy criteria and now use PTA differently (National Institute for Health and Clinical Excellence, 2009, 2019). Previously, the level of HL was evaluated on the 2 and 4 kHz frequencies, which were changed to two or more frequencies between 0.5 and 4 kHz without solid evidence (Lovett et al., 2015; D. Vickers et al., 2016).

Performance analysis of screening methods with a binary outcome has never been used in CI evaluation but is commonly used in biomedical decision-making (Lasko et al., 2005). The binary outcome for CI candidacy used in the context of the present study, is improvement versus no improvement of speech perception after implantation. The proportion of patients selected correctly by the preoperative measure i.e., who improve their speech perception postoperatively (sensitivity) is compared to the proportion of patients (hypothetically) rejected by the preoperative measure who showed no improvement in their speech perception scores after CI (specificity). Subsequently, these proportions of sensitivity and specificity for each cutoff value can be plotted on a receiver operating characteristic (ROC) curve (Fawcett, 2006; Lasko et al., 2005). The larger the area under the curve (AUC) of a measure, the higher its performance in selecting appropriate patients and rejecting patients who will not have improved speech perception after receiving the CI. Three conditions are required for such an analysis: (1) a large number of patients who have been implanted with a CI, (2) based upon relatively lenient candidacy criteria (80% best-aided phoneme score or 60% word score), and (3) the availability of a broad range of preoperative measures (e.g., different PTAs, including an average or threshold of different frequencies, and unaided or best-aided SPT in quiet or noise). These conditions ensure that there is a discrimination value based on the number of patients who will not improve their speech perception postoperatively.

Present study

The main aim of this retrospective study was to determine which preoperative measure is the most effective in selecting CI candidates who will improve their speech perception postoperatively. Different preoperative measures used in various countries were compared, including PTA with different combinations of frequencies (e.g., average or threshold, high vs. low or 2 vs. 3 or 4 frequencies) and SPT (e.g., unaided or with best-fitted HAs, scored as words or phonemes correct, either in quiet or in noise). This was evaluated by defining the correlation between the measures and comparing the AUC of the ROC curves. The study included a large group of post-lingually deafened patients who were implanted with a CI at Leiden University Medical Center (LUMC) in the Netherlands.

MATERIALS & METHODS

Procedure

This retrospective study reviewed all patients with post-lingually occurring HL implanted with a CI at LUMC (ethical approval was obtained through the Medical Ethics Committee of the LUMC). Post-lingual HL was defined as the onset of moderately severe to profound HL (Clark 1981) after 4 years of age. Records were reviewed for a total of 566 adult patients (≥18 years of age at time of implantation) with bilateral post-lingual onset of HL who were implanted with CI between 2000 and 2017. Four patients were sequentially implanted (the second-side was excluded from the analysis). All patients had to have at least 1 year of postoperative follow-up. Fourteen patients were consequently excluded, of whom five were explanted within one year (because of partial luxation or migration of the electrode, implant failure, wound infection, or removal of vestibular schwannoma), seven died (due to causes unrelated to implantation) during the first year, and two (one of them a marginal performer) were lost to follow-up after 3 months, precluding conclusions about their final outcomes. After exclusions, 552 patients with post-lingual onset of HL were included in the study (Table 1).

Selection criteria of the LUMC

Based on the good outcomes with CI, selection criteria became more relaxed over the years in LUMC. Current criteria are based on a detailed analysis as described in Snel-Bongers et al. (Snel-Bongers et al., 2018). In summary, the current selection criteria for adults require candidates to score less than 80% on a CVC phoneme test (approximately equivalent to 60% CVC word score) with best-fitted HAs at 65 dB SPL in quiet . Additionally, for patients with best-aided phoneme scores above 50%, their best-aided phoneme score with speech at 65dB SPL in a +5dB SNR condition must be less than 50% in order to be eligible for CI. There are no explicit minimum inclusion criteria in our center, e.g. duration of deafness is no reason not to implant as long as it concerns patients with post-lingual HL. In our center, without contraindications, it is standard of practice to implant the worst-performing ear to preserve the best-performing ear for HA usage.

TABLE 1. Characteristics of the study population (*n* = 552)

Age at implantation in years, mean (SD)	60.6 (14.6)
Duration of hearing loss in years, mean (SD)	33.9 (18.2)
Duration of severe bilateral hearing loss in years, mean (SD)	19.4 (17.5)
Sex, *n* (%)	
Male	241 (43.7%)
Female	311 (56.3%)
Cause of deafness, *n* (%)	
Hearing loss with unknown cause	193 (34.3%)
Genetic hearing loss	185 (32.9%)
Infections	84 (14.9%)
Sudden deafness	48 (8.5%)
Middle ear problems	31 (5.5%)
Other	21 (3.7%)
Number of hearing aids prior to implantation, *n* (%)	
Two	405 (73.4%)
One	102 (18.5%)
None	40 (7.2%)
Number of patients with asymmetric hearing loss, *n* (%)	
50% difference in unaided phoneme scores	70 (12.9%)
- without	472 (87.1%)
30% difference in unaided phoneme scores	161 (29.7%)
- without	381 (70.3%)
Implantation side, *n* (%)	
Right	295 (53.4%)
Left	248 (44.9%)
Bilateral	9 (1.6%)
Manufacturers & implant electrode types, *n* (%)	
Advanced Bionics (Los Angeles, California)	460 (83.3%)
Clarion II implant with HiFocus1 electrode	49
HiRes 90K implant with HiFocus1J electrode	233
HiRes 90K implant with HiFocus MS electrode	178
Cochlear (Sydney, Australia)	49 (8.9%)
Nucleus Freedom with Contour Advance electrode	24
Nucleus Freedom with Hybrid-L24 electrode	25
MED-EL (Innsbruck, Austria)	43 (7.8%)
Concerto implant with Medium electrode	36
Concerto implant with Flex electrode	7

TABLE 1. Continued

Pure tone audiogram in dB HL, mean (SD) | range	
Best ear (*n*=551)	
– 250 Hz	73.96 (27.3) | 0-130
– 500 Hz	82.97 (23.1) | 0-130
– 1000 Hz	93.30 (20.0) | 15-130
– 2000 Hz	102.20 (21.6) | 30-130
– 4000 Hz	109.95 (21.6) | 10-130
– Average of 1, 2 kHz	97.87 (18.8) | 23-130
– Average of 0.5, 1, 2 kHz	92.82 (18.1) | 18-130
– Average of 0.5, 1, 2, 4 kHz	97.21 (17.2) | 21-130
Ear-to-be-implanted (*n*=543)	
– 250 Hz	83.0 (29.3) | 5-130
– 500 Hz	93.5 (23.6) | 5-130
– 1000 Hz	105.2 (19.2) | 25-130
– 2000 Hz	112.8 (19.2) | 5-130
– 4000 Hz	118.7 (17.4) | 55-130
– Average of 1, 2 kHz	109.0 (17.6) | 30-130
– Average of 0.5, 1, 2 kHz	103.8 (17.6) | 37-130
– Average of 0.5, 1, 2, 4 kHz	107.5 (16.2) | 41-130
Speech perception scores, mean (SD) | range	
Preoperative	
Maximum unaided phoneme score of best ear (*n*=551)	38.9% (27.1) | 0-97%
Maximum unaided phoneme score of ear-to-be-implanted (*n*=543)	21.0% (22.2) | 0-87%
Maximum phoneme score of the other ear (*n*=542)	35.5% (27.9) | 0-97%
Best-aided phoneme score at 65 dB SPL in quiet (*n*=485)	39.9% (23.7) | 0-97%
Best-aided phoneme score with +5 dB signal to noise ratio (*n*=201)	36.6% (15.5) | 0-84%
Best-aided word score at 65 dB SPL in quiet (*n*=482)	19.8% (19.3) | 0-91%
Best-aided word score with +5 dB signal to noise ratio (*n*=201)	13.7% (10.7) | 0-52%
Postoperative at 1 year	
Phoneme score with CI only at 65 dB SPL in quiet (*n*=416)	78.7% (15.8) | 20-100%

Preoperative measures

PTA was performed using the frequencies 0.25, 0.5, 1, 2, and 4 kHz to calculate the degree of HL. Different types of PTA were established based on either an average of two to five frequencies. In some countries a more binary criterion based on the PTA is used (e.g., two or more thresholds in the audiogram above 85dB). These criteria for the degree of HL were assessed by individually evaluating each frequency that exceeded a varying value or threshold.

SPT was conducted using the standard Dutch Society of Audiology test, consisting of phonetically balanced monosyllabic consonant-vowel-consonant (CVC) words (Bosman & Smoorenburg, 1995). The testing procedure comprised four lists per condition, each containing 11 words of one syllable (total 44 words and 132 phonemes). First, the maximum unaided phoneme score with headphones was obtained. The maximum percentage of phonemes correct at presentation levels between 30 and 130 dB SPL was reported for each ear separately. Hereafter, the phoneme and word score with best-fitted HAs in the free-field were obtained to measure the real-world speech perception abilities. The difference between word and phoneme scores is based on the scoring method. For word scores (p_w) the percentage of correct 'whole' words is scored while for the phoneme scores (p_{ph}) the percentage in correct phonemes is scored (e.g., the response "tip" when "ship" is presented, will give 0% word score, and 66% phoneme score). The scores are highly correlated (for the Dutch CVC test: $p_w=p_{ph}^{2.3}$) giving the word score a higher specificity in the high performance range (>70%) and the phoneme score in the lower one (Gelfand et al. 2013).

Within the population, 405 patients used two, 102 patients used one, and 40 patients used no HAs (5 subjects with missing data). The latter patients had either profound HL due to meningitis (n=14), progressive HL (n=9), sudden deafness (n=8), trauma (1), or no measurable hearing without specific etiology (n=8) that impeded them from using amplification. Words were presented at 65 dB SPL over a loudspeaker placed 1 m in front of the patient (calibrated with a Rion Class 1 NA-28 Sound Level Meter). If a phoneme score in quiet of >50% was achieved, a speech-in-noise test was conducted in speech-shaped noise at a +5 dB signal to noise ratio.

Postoperative outcome measure

During the first 3 months of CI use, patients received intensive hearing training from professional speech therapists (daily in the first four weeks, decreasing to weekly in the last weeks) and approximately 5 fitting sessions. The postoperative SPT took place at 1 and 2 weeks, 1, 3, and 6 months, and 1, 2, and 3 years after initial stimulation. Tests were performed under the same conditions as the preoperative tests (65dB SPL in quiet and +10dB, +5dB, 0dB SNR). The one-year postoperative speech scores were in some cases collected on a slightly earlier or later time-point due to logistics (range 1-3 months). This deviation, in our opinion, does not influence the outcomes as postoperative speech scores at 1 year are nearly similar to the 6 months and 2nd year follow-up scores (Snel-Bongers et al., 2018). Postoperatively and during follow-up, only the implanted ear was tested with an unaided or plugged contralateral ear to examine the actual progress with the CI. Of course,

in a couple of patients (esp. with ski-sloped audiograms) some minor benefit of the plugged ear could still be present. Masking would however be overheard via the CI in many cases, and the difference of the plugged ear relative to the aided condition was deemed large enough that the CI performance would dominate the scores. Postoperative improvement in speech perception was analyzed at the level of the patient by subtracting the best-aided preoperative phoneme scores (both in quiet and in noise) from the postoperative phoneme scores with the CI at 1 year after initial stimulation.

For the ROC analysis a binary outcome is needed relative to the variable inclusion criteria. For this study the binary outcome of no improvement (<0%) and improvement (≥0%) was chosen to indicate that the patients have reached the same speech perception level with CI (either in quiet or in +5 dB SNR noise) as preoperatively with optimally fitted HAs. In addition, a third condition was included, focusing on the benefit of only the implanted ear. This criterion was obtained by subtracting the maximum phoneme score (irrespective of the level) with headphones of the implanted ear from the postoperative phoneme scores with the CI at 1 year after initial stimulation. In this case, the criterion for improvement was that the phoneme score in the implanted ear had increased by at least 20%.

Statistical analysis

The preoperative measures used in LUMC were adapted as far as possible to correspond to internationally used preoperative measures discussed in the literature (Gubbels et al., 2017; Hughes et al., 2014; Huinck et al., 2019; National Institute for Health and Clinical Excellence, 2009, 2019; Raeve, de & Wouters, 2013; Snel-Bongers et al., 2018). The accuracy of the preoperative measures were evaluated with a ROC curve analysis. This method is extensively used in medicine to describe the diagnostic accuracy of a test (Fawcett, 2006; Hoo et al., 2017; Obuchowski, 2005; Obuchowski & Bullen, 2018). A clinical test based on a continuous outcome uses different cut-off points to predict the presence of a disease which is associated with a sensitivity and specificity (Obuchowski & Bullen, 2018). In case of our study, we are not interested in the presence of disease, but in the presence of improved speech perception postoperatively. All possible cut-off points are chosen and the sensitivity/specificity pairs are used to generate a curve. Each coordinate (x, y) on the curve represents the true-positive (sensitivity) and the false-positive rate (1-specificity) associated with a cutoff-point of the test (0-120 dB for PTA and 0-100% for SPT). This ROC curve is thus a graphical plot that exemplifies a diagnostic test's accuracy and can be used on both paired and unpaired data (Fawcett, 2006; Lasko et al., 2005; Obuchowski & Bullen, 2018). If the curve crosses the plot as a diagonal line, the test has no distinctive

capability but uses random decision-making. The ideal test has a ROC curve that bends to the upper left corner which illustrates a high true-positive rate against a low false-positive rate. The area under the ROC curve (AUC) is a measure of discriminatory power of the test, irrespective of a specific cut-off point. The AUC of a test with no diagnostic ability is 0.5 while a measure that perfectly discriminates between two conditions has an AUC of 1.

We checked if the outcomes of the ROC-curves were different when the target of postoperative improvement of speech perception was changed (less than 0%, 5%, 10%, 15%, or 20% speech improvement as a negative outcome) to examine if the accuracy of each measure differs when the number of true negatives increases. This did not yield a difference in the order of which preoperative measure had the highest AUC (Fawcett, 2006; Lasko et al., 2005; Obuchowski & Bullen, 2018). For clarity only the analysis where we compared the postoperative speech scores with CI only with the preoperative best-aided condition, with speech improvement of ≥0% as a positive outcome and <0% speech improvement after CI as a negative outcome will be presented. In the condition where we compared the postoperative with the preoperative speech perception score of the ear-to-be-implanted, only 7 patients did not improve their speech perception at the implanted side by more than 0% for this reason the criterium for this condition was set at >20%. Data analyses were performed using the IBM SPSS Statistics 26.0 software package which enabled us to compare the AUC of the preoperative measurements with paired sample *t*-tests. The multiple comparisons were corrected with a Bonferroni correction resulting in a significance level of $p < .0003$.

Missing data

Little's missing completely at random test was significant ($p < .001$), meaning that the missing data were either missing at random or missing not at random rather than missing completely at random. Missing at random means that the reason for missingness is related to other factors that are measured within the dataset, see for a detailed explanation of terms Netten et al. 2017 (Buuren, 2012; Netten, Dekker, et al., 2017). In the case of our study, we therefore argue that the missing data was missing at random as the reason for missingness was held in the dataset: most patients with missing data were either good or poor performers (based on the measurements at 6 months, 2 or 3 years postoperative) which might made them think that their yearly appointments deemed unnecessary. Postoperative 1 year SPT in quiet and noise were unavailable for 136 and 221 patients, respectively. When conducting standard analyses, such as ROC curves, incomplete cases are automatically excluded (Madley-Dowd et al., 2019; Netten, Dekker, et al., 2017). Excluding the poor and

good performers would bias the findings and potentially lower the statistical power due to loss of participants. To adequately deal with these missing data, the multiple imputation technique was used (Buuren, 2012; Madley-Dowd et al., 2019; Schafer & Graham, 2002; Sterne et al., 2009). With this technique, missing data are imputed based on the known characteristics of the patients (gender, age at implantation, implantation side, duration of deafness, cause of deafness, preoperative and postoperative measures at 1 and 2 weeks, 1, 3, and 6 months, and 1, 2, and 3 years after initial stimulation). We used 10 imputed datasets and pooled the 10 outcomes. All analyses were performed on the imputed and original data, which did not yield different outcomes.

RESULTS

The ROC curves with imputed data were plotted for a selection of preoperative measures (Figure 1). Table 2 reports the AUC of all preoperative measures. Within the best-aided condition, 28 patients (5.1%) did not improve while 524 patients did improve their speech perception after cochlear implantation based on the binary threshold of 0% improvement of phoneme scores. The 28 patients scored preoperatively on average 67% (range 21-97%) with the best-aided SPT in quiet and 41% (20-84%) in noise. Most of them had an asymmetrical HL (n=19 had more than 30% phoneme score difference between ears).

Figure 1A shows that most preoperative measures performed nearly similar when using improvement in a best-aided condition. The best-aided phoneme score presented in quiet at 65 dB SPL in the free-field had the highest AUC of all preoperative measures, followed by the best-aided word score in quiet. The best-aided phoneme and word score significantly differed from the maximum unaided phoneme score at the implanted side, individual evaluation of 0.5, 1, and 2 kHz, individual evaluation of 2 or more frequencies (0.5, 1, 2, 4 kHz), individual evaluation of 2 or more frequencies (0.25, 0.5, 1, 2, 4 kHz), and individual evaluation of 0.5 and 1 kHz ($p < .0003$). The maximum unaided phoneme score of the ear-to-be-implanted had a ROC curve that was smaller than the reference line (also Table 2). The average PTA of 1 and 2 kHz had the highest AUC of all PTAs, but was not significantly different ($p = .005$-$.631$). Evaluating two or more frequencies individually did not significantly differ from the reference line, which indicated that this preoperative measure did not have a distinctive capability (Table 2).

Twenty percent of improvement at the implanted ear also resulted in 28 patients who did not and 524 patients who did improve. The 28 candidates scored preoperatively on average 53% (range 5-87%) phonemes correct at the ear that would be implanted. Only four of these patients had asymmetrical HL of more than 30% phoneme score difference between ears.

After using improvement of phoneme scores in the implanted ear in a ROC analyses, we found that the maximum unaided phoneme score with headphones of the ear-to-be-implanted had the highest AUC of all preoperative measures (Figure 1B and Table 2) and significantly differed compared to all other SPTs and PTAs ($p < .0003$). The maximum unaided phoneme score with headphones of the best ear performed second-best, but did not differ significantly compared to other SPTs ($p = .003$-$.075$). Average of five frequencies

(0.25, 0.5, 1, 2, 4 kHz) had the highest AUC of all PTAs, but was not significantly different compared to other PTAs (p = .019-.664).

When analyzing improvement with best-aided phoneme scores in noise, we found that 33 patients did not and 519 patients did improve. The 33 patients scored preoperatively a mean phoneme score of 50% (range 26-84%) correct in a best-aided condition with +5 dB signal-to-noise ratio. The best-aided phoneme score at +5 dB signal to noise ratio had the highest AUC compared to all other preoperative measures (p < .0003), except for no significant difference compared to the best-aided phoneme and word score in quiet and word score in noise (p = .002-0.069)(Figure 1C).

Analysis showed that the order of AUC-outcomes of the preoperative measures did not change when the threshold for improvement was set to 5% or 20% improvement in phoneme scores instead of 0% (in all three conditions). The number of patients with no improvement in for example the best-aided condition increased from 28 to 41 and 107, respectively (i.e., 7.4% to 19.4% of the total population).

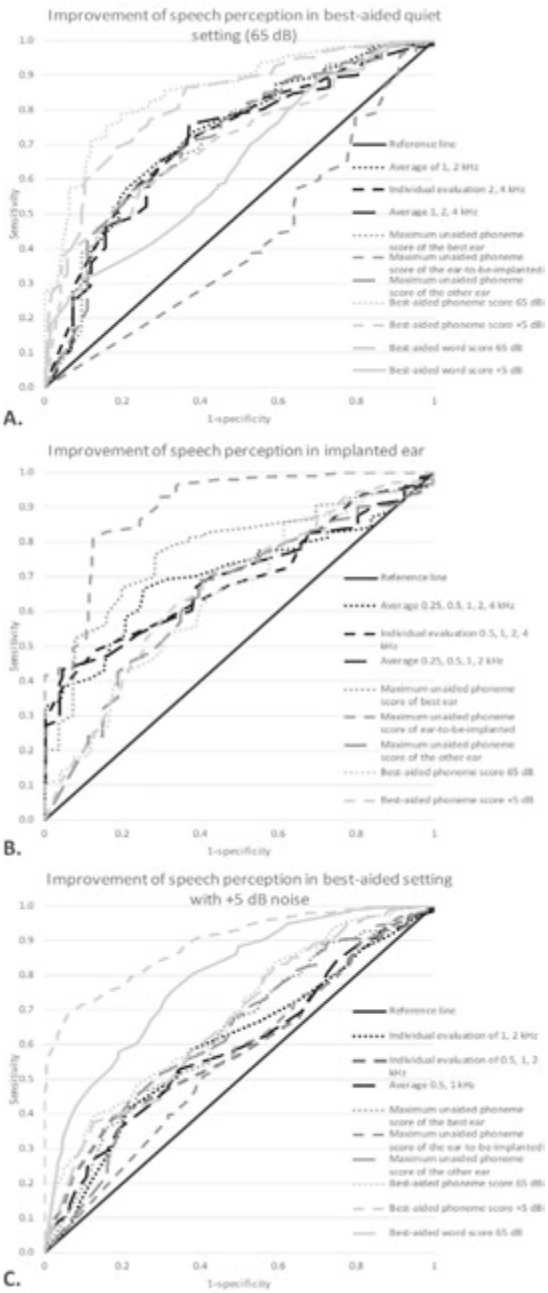

FIGURE 1. Receiver operator characteristic curves of preoperative pure tone audiometry and speech perception measures with imputed data (n=552).
Diagnostic performance was analyzed for pure tone audiometry and speech perception tests using improvement of phoneme score in a best-aided quiet setting at **(A)**, in the implanted ear **(B)**, or in a best-aided setting with +5 dB signal to noise ratio as a positive outcome **(C)**. Only pure tone audiometry with the highest area under the curve were added to the ROC curve.

TABLE 2. Area under the ROC curve of each preoperative measure for improvement of postoperative speech perception (imputed data; *n* = 552)

		Area under the curve	Asymptomatic 95% Confidence Interval	
			Lower Bound	Upper Bound
Improvement of phoneme score in best-aided quiet setting (65 dB SPL)	Average PTA 1, 2 kHz	0.724***	0.625	0.822
	Average PTA 1, 2, 4 kHz	0.700***	0.601	0.799
	Average PTA 0.5, 1, 2 kHz	0.688**	0.587	0.789
	Average PTA 0.5, 1, 2, 4 kHz	0.685**	0.585	0.787
	Average PTA 2, 4 kHz	0.680**	0.586	0.774
	Average PTA 0.5, 1 kHz	0.634*	0.532	0.735
	Individual evaluation of 2, 4 kHz	0.718***	0.623	0.813
	Individual evaluation of 1, 2 kHz	0.696**	0.594	0.798
	Individual evaluation of 0.5, 1, 2 kHz	0.619*	0.524	0.715
	Individual evaluation of 2 or more frequencies (0.5, 1, 2, 4 kHz)	0.612	0.503	0.720
	Individual evaluation of 2 or more frequencies (0.25, 0.5, 1, 2, 4 kHz)	0.611	0.502	0.721
	Individual evaluation of 0.5, 1 kHz	0.598	0.500	0.696
	Best-aided phoneme score at 65 dB SPL	0.853***	0.788	0.918
	Best-aided word score at 65 dB SPL	0.821***	0.746	0.897
	Maximum unaided phoneme score best ear	0.715***	0.616	0.817
	Maximum unaided phoneme score other ear	0.713***	0.613	0.816
	Best-aided phoneme score with +5 dB noise	0.709**	0.624	0.796
	Best-aided word score with +5 dB noise	0.641*	0.539	0.745
	Maximum unaided phoneme score ear-to-be-implanted	0.423	0.314	0.535
Improvement of phoneme score in implanted ear	Average PTA 0.25, 0.5, 1, 2, 4 kHz	0.706***	0.633	0.778
	Average PTA 0.25, 0.5, 1, 2 kHz	0.691***	0.615	0.767
	Average PTA 0.25, 0.5, 1 kHz	0.686***	0.604	0.767
	Average PTA 0.5, 1, 2, 4 kHz	0.675***	0.587	0.762
	Average PTA 0.5, 1 kHz	0.673***	0.584	0.762
	Average PTA 0.5, 1, 2 kHz	0.672***	0.586	0.758
	Individual evaluation of 0.5, 1, 2, 4 kHz	0.692***	0.616	0.768
	Individual evaluation of 0.5, 1, 2 kHz	0.687***	0.608	0.767
	Individual evaluation of 0.25, 0.5, 1, 2, 4 kHz	0.680***	0.595	0.765
	Individual evaluation of 0.25, 0.5, 1, 2 kHz	0.675***	0.587	0.764
	Individual evaluation of 1, 2, 4 kHz	0.675**	0.570	0.780
	Individual evaluation of 0.5, 1 kHz	0.674***	0.593	0.756
	Maximum unaided phoneme score ear-to-be-implanted	0.899***	0.834	0.965
	Maximum unaided phoneme score best ear	0.775***	0.698	0.851

TABLE 2. Continued

		Area under the curve	Asymptomatic 95% Confidence Interval	
			Lower Bound	Upper Bound
	Best-aided phoneme score with +5 dB noise	0.665**	0.559	0.771
	Maximum unaided phoneme score other ear	0.642*	0.539	0.746
	Best-aided phoneme score at 65 dB SPL	0.642*	0.537	0.747
	Best-aided word score at 65 dB SPL	0.639	0.531	0.747
	Best-aided word score with +5 dB noise	0.636	0.529	0.743
Improvement of phoneme score (65 dB SPL) in best-aided setting with a signal to noise ratio of +5 dB	Average PTA 0.5, 1 kHz	0.592	0.490	0.694
	Average PTA 0.5, 1, 2 kHz	0.587	0.487	0.687
	Average PTA 1, 2 kHz	0.570	0.466	0.674
	Average PTA 0.5, 1, 2, 4 kHz	0.568	0.469	0.668
	Average PTA 1, 2, 4 kHz	0.546	0.442	0.650
	Individual evaluation of 1, 2 kHz	0.598	0.495	0.702
	Individual evaluation of 0.5, 1, 2 kHz	0.593	0.497	0.690
	Individual evaluation of 0.5, 1 kHz	0.583	0.483	0.684
	Individual evaluation of 0.25, 0.5, 1, 2,	0.581	0.477	0.684
	4 kHz	0.525	0.413	0.637
	Individual evaluation of 2, 4 kHz	0.494	0.381	0.607
	Individual evaluation of 2 or more frequencies (0.25,0.5, 1, 2, 4 kHz)			
	Best-aided phoneme score with +5 dB noise	0.887***	0.841	0.933
	Best-aided word score with +5 dB noise	0.784***	0.695	0.874
	Best-aided phoneme score at 65 dB SPL	0.680	0.581	0.779
	Best-aided word score at 65 dB SPL	0.664	0.560	0.768
	Maximum unaided phoneme score best ear	0.631	0.524	0.738
	Maximum unaided phoneme score other ear	0.625	0.512	0.738
	Maximum unaided phoneme score ear-to-be-implanted	0.559	0.442	0.675

*p<0.05; ** p<0.01; ***p<0.001 compared to the reference line or AUC=0.5; PTA = pure tone audiometry;

DISCUSSION

This study compared the diagnostic performance of different preoperative measures in selecting post-lingually deafened adult CI candidates who had better speech perception with a CI than preoperatively and rejecting candidates who would not improve. We found that the diagnostic performance of the preoperative measures depended on which outcome objective was used and that the evaluated preoperative measures did not differ considerably in their efficacy as a diagnostic test. The best-aided SPT in quiet at 65 dB had the highest diagnostic performance to select candidates who improve their best-aided speech perception in quiet. For an improvement of 20% in the ear-to-be-implanted, the preoperative maximum unaided phoneme score with headphones of the ear-to-be-implanted had the highest performance. The best-aided SPT with a +5 dB signal to noise ratio had the highest performance for indicating a postoperative improvement in noise but was only evaluated for those at the upper end of the performance range, as they have to reach at least 50% phoneme scores in quiet in order to be tested in noise in our center. The results of this study could help different authorities, such as healthcare commissioners and implant centers, improve adult CI selection criteria by changing the preoperative measures to the most effective one.

Most studies have examined which inclusion criteria should be followed by using the standard preoperative measures available (Gubbels et al., 2017; Hoppe et al., 2015; Huinck et al., 2019; J. R. Leigh et al., 2016; Maeda et al., 2018; McRackan et al., 2018; Snel-Bongers et al., 2018; Deborah Vickers et al., 2016a). This study found that preoperative SPT had the highest performance as a classifier to indicate speech improvement in CI candidates. This result was not completely unexpected as the main goal of a CI is to improve speech perception.

The patients with no improvement often had residual hearing, performed at the better end of the performance range, or had an unusual HL (Huinck et al., 2019; J. R. Leigh et al., 2016; Snel-Bongers et al., 2018). This allowed them to only marginally improve their speech perception in quiet after CI. Making accurate and informed decisions for these high-performers is the most critical. This evidence-based study reasoned that for candidates with residual hearing or unusual HL (e.g., shape of the PTA), a SPT in noise is a more appropriate preoperative test to indicate which candidates would improve their speech perception in noise with CI.

One can argue about the ideal postoperative measure to indicate improvement. A best-aided condition, including the contralateral HA, represents actual clinical progress. However, such a measure can also mask the benefit of the implant. This can be the case in a patient with asymmetric HL where one ear meets CI-criteria (e.g., speech perception of 30% correct) while the better ear still reaches speech perception of 80% correct. Measuring postoperative speech perception in these patients in a best-aided condition will probably only demonstrate the progression of the non-implanted ear and not the effect of CI. These patients will also rate the effect of the implant rather poorly as it is underperforming relative to the HA. Therefore, in line with earlier research (J. R. Leigh et al., 2016; Snel-Bongers et al., 2018), we argue that postoperative improvement should be calculated as the speech perception scores with CI only in relation to the preoperative level of performance in a best-aided condition. This is probably the most stringent criterion of improvement that can be defined. However, when improvement is achieved with this method, actual clinical progress with the CI is the case. Of course showing progress using this definition in single sided deaf patients will be impossible and makes this criterion less than optimal in these populations.

At our center, the number of patients who obtained poorer speech perception after CI was very low (5%) giving a small group of patients a large influence on the outcomes. Therefore, we also analyzed the performance of preoperative measures with 5-20% improvement in speech perception (instead of the 0% as reported above), which led to an increased number of patients with a negative outcome (7-19% of the total population). However, this did not influence the order of efficacy of preoperative measures. Apparently, the type of preoperative measure, rather than the amount of improvement, was important for selecting CI candidates with improved postoperative speech perception.

PTA is easily accessible worldwide and often used as a preoperative criterion for cochlear implantation. However, this study showed that the PTA did not perform as well as SPT in predicting speech improvement after CI. Even using PTA differently, for example, by the individual evaluation of each frequency, or an average of different frequencies, did not lead to a better performance. PTA and SPT are often combined for selecting CI candidates. Yet, calculating the performance of these two preoperative measures together was not possible with the ROC analysis as PTA and SPT have different scales. This would require the cut-off point of one of the measures to be fixed (e.g., PTA with 85 dB HL or SPT with 50% phonemes correct as cut-off point). Future studies examining the selection criteria by expanding the cut-off values of their preoperative measures should add ROC analysis (van der Straaten et al., 2020).

Furthermore, policy-makers can debate inclusion criteria when knowing which preoperative measure has the best performance. The cut-off value of this measure can then be chosen by shifting between the sensitivity and specificity rates. One could, e.g., require that the preoperative measure selects 90% of the candidates who improve their speech perception after CI instead of the requirement that candidates must have at least a 90% chance that their speech perception with CI exceeds their performance with conventional HAs (sensitivity vs. positive predictive value)(Snel-Bongers et al., 2018; Verhaegen et al., 2008).

Strengths & limitations

This study used a large group of post-lingually deafened adults who had been implanted with CIs under relatively lenient candidacy criteria. These lenient criteria resulted in some patients not showing improvements in speech perception after implantation, allowing us to calculate which preoperative measure rejected these patients based on the postoperative improvement. The possibility of using speech in noise as a preoperative measure was explored, and was demonstrated to be an important measure for assessing the borderline candidates at the upper end of the performance spectrum. It would be interesting to validate our results in populations using even more lenient candidacy criteria, such as in Germany (Deborah Vickers et al., 2016a).

It is important to mention and take note of a confounder in the presented data. The success criteria used in all ROC-curves is based on improvement in speech scores. This variable includes the pre-operative speech score that is also used as a predictor. At the same time there is a covariate in the known fact that the postoperative performance is correlated with the preoperative scores. A direct consequence hereof is an increased AUC for the speech-based predictors. We have, however, chosen for these variables because they agree with clinical practice of CI candidacy, counseling, the predictive values used in literature, and the way policies are made. As described before, CI criteria research takes place on speech scores while success is measured in improvement or the attained score in the same domain. The presented data, although inevitably statistically biased, are a reflection of the standard clinical considerations. A statistically more accurate method would have been to use more independent variables (e.g., quality of life) as success criteria, a more or less independent measure relative to all audiological measures. Although valuable, such an approach would be less fitting to the daily situations. In the recent paper by Reddy et al., the selection of candidates was used as the criterion in the ROC curve (Reddy et al., 2022). This would allow us to include the rejected candidates in our center as well. It would, however, not have solved the

issue of the confounder, probably even have enhanced it, as in the latter paper, the criteria are also based on speech scores (Aided AzBio sentences in quiet <60% or Aided AzBio +10 <60%).

We are aware that measuring the postoperative performance with CI only is rather strict. It could be argued that postoperative improvement should be examined by using the same SPT pre- as postoperatively (e.g., in a best-aided condition). However, in our opinion it is much more relevant to examine the actual progress of the CI rather than the best-aided condition. Patients with residual or asymmetric HL for example obtain high preoperative best-aided speech perception scores with their better contralateral amplified ear. Measuring the postoperative speech perception in a best-aided condition would not be informative regarding the CI-performance, especially when the poorer performing ear is implanted, as is most often the case in our center.

Importantly, the findings of this study only apply to post-lingually deafened adults and not pre-lingually deaf children, as they differ considerably in preoperative characteristics (e.g., etiology and age at implantation) (Peterson et al. 2010). Children are also not able to complete a preoperative SPT and therefore the only available preoperative measure would be a PTA (Lovett et al. 2015). The outcomes of the PTA and SPT in this study were adapted in order to correspond to internationally available tests. In addition, the SPT was only validated in Dutch and not in other languages (Bosman & Smoorenburg, 1995). Therefore, the conclusions of this study should be validated in other countries which have different preoperative outcome measures. Future studies should also consider other preoperative measures, such as sentence tests, or quality of life questionnaires, as potential measures for selecting adult CI candidates, although these are more likely to be influenced by cognitive function (Lee et al., 2016). In addition, other factors (e.g., subjective, spatial hearing, or pitch discrimination) that could contribute to an improvement of listening experience after CI have not been taken into account in this study.

To conclude, this study examined which preoperative measures should be used to appropriately determine which post-lingually deafened adults will improve their speech perception after CI. The findings showed that SPTs in quiet and in noise, rather than PTA-based criteria, have a higher performance for indicating which CI candidates will most likely show post-operative improvement in speech perception in quiet or in noise. Implementation of these insights could improve the approach for selecting candidates and help commissioning bodies formulate more effective selection criteria for CI in post-lingually deafened adults.

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Selection Criteria for Cochlear Implantation in the United Kingdom and Flanders: Toward a Less Restrictive Standard

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ABSTRACT

Objectives: The impact of the newly introduced cochlear implantation criteria of the United Kingdom (UK) and Flanders (Dutch speaking part of Belgium) was examined in the patient population of a tertiary referral center in the Netherlands. We compared the patients who would be included/excluded under the new vs. old criteria in relation to the actual improvement in speech understanding after implantation in our center. We also performed a sensitivity analysis to examine the effectiveness of the different preoperative assessment approaches used in the UK and Flanders.

Design: This retrospective longitudinal cohort study included 552 postlingually deafened adults with cochlear implants (CI). The selection criteria were based on preoperative pure tone audiometry (PTA) at 0.5, 1, 2, and 4 kHz, and a speech perception test (SPT) with and without best-aided hearing aids. Postoperatively, the same SPT was conducted to assess the benefit in speech understanding.

Results: The newly introduced criteria in Flanders and the UK were less restrictive, resulting in greater percentages of patients implanted with CI (increase of 30%), and sensitivity increase of 31%. The preoperative best-aided SPT, used by both countries, had the highest diagnostic ability to indicate a postoperative improvement of speech understanding. We observed that patient selection was previously dominated by the PTA criteria in both countries, whereas speech understanding became more important in their new criteria. Among patients excluded by the new criteria, seven of eight (UK and Flanders) did exhibit improved postoperative speech understanding.

Conclusions: The new selection criteria of the UK and Flanders led to increased numbers of post-lingually deafened adults benefitting from CI. The new British and Flemish criteria depended on the best-aided SPT with the highest diagnostic ability. Notably, the new criteria still led to the rejection of candidates who would be expected to gain considerably in speech understanding after implantation.

INTRODUCTION

In post-lingual adults with severe-to-profound hearing loss (HL), the general goal of a CI is to improve speech understanding. When setting selection criteria, the aims are to ensure that CIs are provided to candidates who are likely to benefit in speech understanding, while avoiding unnecessary costs and medical intervention for patients for whom acoustic hearing aids (HAs) are sufficient. Over recent years, technological developments and changes in surgical techniques have enabled the preservation of residual hearing and improved postoperative speech outcomes (Blamey et al., 2013; Snel-Bongers et al., 2018). However, improvement of speech understanding remains challenging when CI candidates have residual hearing and exhibit relatively high preoperative scores. For these borderline candidates, defining selection criteria for CI is a difficult process and is often based on expert opinions.

CI selection criteria show substantial variation at the international level (Cullen et al., 2004; Dowell et al., 2004; Friedland et al., 2003; Gubbels et al., 2017; Hughes et al., 2014; Huinck et al., 2019; J. R. Leigh et al., 2016; Maeda et al., 2018; McRackan et al., 2018; Raeve, de & Wouters, 2013; Snel-Bongers et al., 2018; Verhaegen et al., 2008; Deborah Vickers et al., 2016a). Such candidacy criteria are commonly based on the anticipated post-implant speech outcomes, with cut-off values for preoperative criteria defined using the lowest 10^{th} to 25^{th} percentile (p10–p25) (Dowell et al., 2004; Snel-Bongers et al., 2018; Verhaegen et al., 2008) or the proportions of patients with and without postoperative improvement in speech understanding (e.g., 1/4 patients may have no benefit post-implantation). However, there is also still a tendency to use conservative CI selection criteria to preserve a benefit in speech understanding post-implantation. For example, a conservative selection criterion would be an average of 85 dB or higher at 0.5, 1, and 2 kHz, and a maximum phoneme score of 30% with HAs (Huinck et al., 2019).

When aiming to improve speech understanding after CI, the preoperative level of speech understanding is the most valuable indicative measure to use for CI selection criteria (T. F. K. van der Straaten et al., unpublished data). However, the types of preoperative audiometric and speech measures used to assess CI candidacy vary widely. For example, the United States of America applies a broad spectrum of preoperative measures (e.g., sentence or word tests) and selection criteria across the country (Cullen et al., 2004; Friedland et al., 2003; Gubbels et al., 2017; Holder et al., 2018; Hughes et al., 2014). Developing countries tend to exclusively use pure tone audiometry (PTA) due to the accessibility. In general, a patient's degree of HL and benefit from acoustic HAs is frequently determined via a combination of PTA and speech perception tests (SPTs) (Deborah Vickers et al., 2016a).

Until recently, the United Kingdom (UK) and Flanders (the Dutch speaking part of Belgium) used relatively conservative criteria compared to the Netherlands, Germany, and Australia (Deborah Vickers et al., 2016a), but both have recently developed new criteria (Table 1) (National Institute for Health and Clinical Excellence, 2009, 2019; Raeve, de & Wouters, 2013). The new British criteria were driven by a panel of experts who reviewed the available evidence and provided recommendations. To determine the best SPT, a National Service Evaluation was conducted to collect SPT and PTA scores from adults with CIs, both preoperatively and up to 1 year postoperatively (Deborah Vickers et al., 2016a). Additionally, research comparing outcomes of children with CIs vs. HAs provided evidence for shifting thresholds to an 80-dB level of HL (Lovett et al., 2015); however, this was based on children who were implanted under the prior conservative criteria (90 dB HL at 2 and 4 kHz). The National Institute for Health and Clinical Excellence has stated that the newly introduced criteria will help to better identify CI candidates. They predicted a 70% increase of patients under the updated recommendation. The new Flemish criteria were also recently implemented, to replace their outdated and conservative previous criteria (Raeve, de & Wouters, 2013).

In the present retrospective study, we aimed to evaluate the effects of using new selection criteria for CI patient selection in the UK (January 2019) and Flanders (August 2019), and to compare the postoperative gains in speech understanding among CI candidates, based on the outcomes of a large group of patients (n = 552) implanted under relatively lenient criteria at the Leiden University Medical Center (LUMC), a tertiary referral center in the Netherlands. Each country uses a different combination of PTA and SPT to evaluate the degree of HL and the benefit from acoustic HAs. It is of interest to determine the extent to which both selection criteria contribute to identifying the candidates who will benefit most from implantation. We expected to find that higher percentages of patients, who exhibited improved speech understanding postoperatively, were accepted under the new criteria compared to the old criteria. Additionally, we examined the diagnostic values of the different preoperative measurement approaches used by the UK and Flanders, using sensitivity and specificity analyses.

TABLE 1. Selection Criteria for Cochlear Implant Candidacy by the United Kingdom and Flanders

	Old criteria	New criteria
United Kingdom	>90 dB at 2 and 4 kHz and <50% sentence score (2009)	≥80 dB at ≥2 frequencies (0.5, 1, 2, 3, and 4 kHz) and <50% phoneme score (2019)
Flanders	Average of >85 dB at 0.5, 1, and 2 kHz and <30% phoneme score (2013)	Average of >70 dB at 0.5, 1, 2, and 4 kHz and <50% phoneme score (2019)

MATERIALS & METHODS

Procedure

In this retrospective study, we reviewed all adults with post-lingual HL who were implanted with CI at the LUMC (ethical approval was obtained through the Medical Ethics Committee of the LUMC). Post-lingual HL was defined as the onset of moderate-to-profound HL (>40 dB) after 4 years of age. In total, we reviewed the records of 566 patients with bilateral post-lingual HL, who had CI implanted between 2000–2017, and who were ≥18 years of age at the time of implantation. The second side of patients with sequential bilateral implantation were excluded from analysis (n = 4). All patients had to have a postoperative follow-up of at least one year. Fourteen patients were consequently excluded, of whom five were explanted within one year (because of partial luxation or migration of the electrode, implant failure, wound infection, or removal of vestibular schwannoma), seven died (due to causes unrelated to implantation) during the first year, and two (one of them a marginal performer) were lost to follow-up after 3 months, precluding conclusions about their final outcomes. After exclusions, our analysis included 552 post-lingual patients. Table 2 presents descriptive statistics of this study population.

Preoperative Measures

PTA was performed using frequencies of 0.5, 1, 2, and 4 kHz to calculate the preoperative degree of HL. Additionally, speech understanding scores were conducted using the standard Dutch SPT of the Dutch Society of Audiology, which comprises phonetically balanced monosyllabic consonant-vowel-consonant words (Bosman & Smoorenburg, 1995). First, we determined the maximum unaided phoneme score (over headphones). Next, we determined the phoneme and word score using best-fitted HAs in the free field at 65 dB and 75 dB SPL, or with a +5 dB signal-to-noise ratio. The standard testing procedure comprised four lists, containing 11 words per condition (a total of 44 words and 132 phonemes). In the free field, words were presented through a loudspeaker set 1 meter in front of the patient. If a patient achieved a phoneme score above 50% in a quiet setting, a speech-in-noise test was conducted in speech-shaped noise at a +5 dB signal-to-noise ratio.

Selection Criteria of the LUMC

At the start of the CI program in 2000, the auditory candidacy criteria included a pure-tone average HL of >90 dB at 0.5, 1, 2, and 4 kHz in the better ear, and best-aided (with one or two HAs) speech understanding of ≤30% phonemes correct in a quiet setting, corresponding to a 10% word score. These criteria changed over time. Since 2012, the selection criteria

have been based only on SPT, with patients having phoneme scores in a quiet setting of ≤60%, and from 2016 onwards of ≤80% (≤60% word scores), considered as CI candidates (Snel-Bongers et al., 2018). An additional criterion for candidates with >50% phoneme score in quiet was that they should have a phoneme score <50% in a +5 dB signal-to-noise ratio. The worst-performing ear was often implanted to preserve the best-performing ear for HA usage.

TABLE 2. Descriptive Statistics of the Study Population (*n* = 552)

Age at implantation, years, mean (SD)	60.6 (14.6)
Duration of hearing loss, years, mean (SD)	33.9 (18.2)
Duration of severe bilateral hearing loss, years, mean (SD)	19.4 (17.5)
Sex, *n* (%)	
Male	241 (43.7%)
Implantation side, *n* (%)	
Right	295 (53.4%)
Left	248 (44.9%)
Bilateral	9 (1.6%)
Manufacturer & Electrode type, *n* (%)	
Advanced Bionics (Los Angeles, California)	460 (83.3%)
Clarion II implant with HiFocus1 electrode	49
HiRes 90K implant with HiFocus1J electrode	233
HiRes 90K implant with HiFocusMS electrode	178
Cochlear (Sydney, Australia)	49 (8.9%)
Nucleus Freedom with Contour Advance electrode	24
Nucleus Freedom with Hybrid-L24 electrode	25
MED-EL (Innsbruck, Austria)	43 (7.8%)
Concerto implant with Medium electrode	36
Concerto implant with Flex electrode	7
Cause of deafness, *n* (%)	
Hearing loss with unknown cause	193 (34.3%)
Genetic hearing loss	185 (32.9%)
Infectious	84 (14.9%)
Sudden deafness	48 (8.5%)
Middle ear problems	31 (5.5%)
Other	21 (3.7%)

n, number of patients; SD, standard deviation

Postoperative Outcome Measure

During the first three months following implantation, patients received intensive hearing rehabilitation from professional speech therapists. Postoperative follow-up occurred at 1 and 2 weeks; 1, 3, and 6 months; and 1, 2, and 3 years after surgery. Postoperative and follow-up examinations included testing of only the implanted ear, with an unaided or plugged contralateral ear, to examine the actual CI progress. Improvement in speech understanding was analyzed by subtracting the best-aided preoperative phoneme and word score at 65 dB and 75 dB SPL from the postoperative phoneme and word scores with CI at the same presentation level.

Statistical Analysis

We modified the preoperative PTA and SPT to be comparable to the selection criteria of the UK and Flanders (National Institute for Health and Clinical Excellence, 2009, 2019; Raeve, de & Wouters, 2013). Different PTA frequencies were utilized, and we calculated the average of SPT at 65 dB and 75 dB to approximate the SPT at 70 dB used in these two countries. Using the conversion formula of Vickers et al. (2013), we converted the 50% score on the Bamford-Kowal-Bench sentence test from the old British criterion to a 30% phoneme score on the Arthur Boothroyd word test (D. Vickers et al., 2013). This test provides a phoneme score that is highly comparable to Dutch phoneme scores. Flanders uses the same Dutch CVC word list for evaluating speech understanding, ensuring direct comparison with our data (Bosman & Smoorenburg, 1995).

We used the old and new CI selection criteria of the UK and Flanders to separate the study sample into different groups: excluded or included according to the old and new criteria (Table 1). Descriptive analyses were performed, and a graphical scatter plot was generated with the included and excluded patients plotted against the improvement of speech understanding after CI. We evaluated the performance of preoperative measurements for predicting benefit using receiver operator characteristic (ROC) curve analysis. A ROC curve is a graphical plot that illustrates the diagnostic ability of a measurement with a binary outcome (improvement of speech understanding ≥0% or no improvement after CI), as its discrimination threshold is varied (Fawcett, 2006; Lasko et al., 2005). Data analyses were performed using the IBM SPSS Statistics 26.0 software package.

Missing Data

Missing data were analyzed with Little's missing completely at random (MCAR) test (Little, 1988). The result was significant ($p < .001$) meaning that the missing data were either missing at random or missing not at random. Missing at random would indicate that the underlying reason for missing data was related to known patient characteristics, which was the case in our study. Most patients with missing data were either good or poor performers, such that their yearly appointments were deemed unnecessary. We were missing 1-year postoperative SPT results from a quiet setting from 136 patients, and from a setting with noise from 221 patients. Incomplete cases are automatically excluded from standard analyses, such as ROC curves (Netten, Dekker, et al., 2017). However, excluding these patients might bias the findings and potentially lower the power of the results. Thus, we applied a multiple imputation technique to impute the missing data based on known patient characteristics (gender, age at implantation, implantation side, deafness duration, deafness cause, preoperative PTA, preoperative SPT, and postoperative SPT at other follow-up evaluations)(Buuren, 2012). Ten datasets with imputations were produced. All ROC curves were generated using both the imputed and original datasets, revealing no differences in outcomes. The original dataset, including 416 patients with CI and postoperative scores at 1 year, was used for descriptive analyses and scatter plots.

RESULTS

Postoperative Speech Understanding Scores

At one year postoperatively, 396 patients (95.2%) exhibited improvement and 20 patients (4.8%) did not show improvement of their speech understanding in a quiet setting (70 dB SPL) with their CI. The postoperative improvement in speech understanding exhibited a ceiling effect, indicating that abundant improvement was not possible in patients with high preoperative best-aided phoneme scores (reference line in Fig. 1). Among the 20 patients without improved speech understanding in a quiet setting, two patients exhibited improved speech understanding in the setting with noise (difference in phoneme score of 5% and 18% at the +5 dB signal-to-noise ratio), and 15 patients exhibited improved speech understanding on the side of implantation (mean improvement from maximum phoneme score: 18%; SD, 28%). Three patients (0.7% of the total population) exhibited no improvement of speech understanding at any level after implantation (difference in phoneme score at 70 dB: –1%, –14%, and –16%).

Included or Excluded by the Selection Criteria of the UK and Flanders

Figure 1 shows the range of preoperative degree of HL and best-aided phoneme scores plotted against the postoperative improvement of speech understanding for each criteria.

The new selection criteria of the UK led to the inclusion of 30% more patients, of whom 0.2% did not exhibit postoperative improvement of speech understanding. This new group exhibited a 41% improvement of speech understanding, in contrast to the 59% improvement within the group accepted based on the old criteria (Table 3). Among all analyzed patients, 34.4% were excluded by both the old and new selection criteria. In this excluded group, one of eight patients (4.3%) did not exhibit postoperative improvement, and this group improved their speech understanding by an average of 17%. Overall, our findings indicated that the new British criteria result in the selection of patients who will have a postoperative improvement, excluding two patients who showed a >50% improvement postoperatively.

The new selection criteria of Flanders led to the inclusion of 30.2% more patients, of whom 0.2% did not exhibit postoperative improvement of speech understanding. This group of newly included candidates exhibited a 40% improvement of speech understanding on average, as opposed to an improvement of 60% among patients who would be included by both the old and new criteria (Table 3). Among all analyzed patients, 33.4% would be excluded by both the old and new selection criteria of Flanders. Within this excluded

group, one of eight patients (4.3%) exhibited no postoperative improvement, and this group exhibited a 16% postoperative increase of speech understanding. On the other hand, the new criteria of Flanders included all patients who showed >50% improvement of speech understanding postoperatively.

Next, the selection criteria were separated by the PTA and speech prerequisites (Fig. 1B, D). The new criteria of the UK relied more on the speech criterion than the PTA criterion, since the amount of patients excluded almost corresponded when following both PTA and speech criteria versus following only the speech criteria. There were five instead of 12 additional patients excluded based on the PTA prerequisites of the new and old criteria, respectively, on top of the patients excluded based on the speech criteria (Fig.1A). Moreover, following only the PTA prerequisites of the UK resulted in a considerable amount of patients (65.4%) included by both the old and new criteria (Fig. 1B). Notably, a small group of patients (7.5%) were excluded by the new PTA criterion but were included by the old PTA criterion.

The new criteria of Flanders were also predominantly based on the speech criterion, since the amount of patients excluded nearly resembled the amount when following both PTA and speech criteria or the speech criteria alone. Two instead of 11 additional patients were excluded based on the PTA conditions when following the new instead of the old criteria on top of the patients excluded based on the speech criteria (Fig. 1C). Following only the PTA condition of Flanders resulted in a substantial amount of patients (70.4%) included by both the old and new criteria (Fig. 1D).

TABLE 3. Postoperative Improvement of Speech Understanding Among the Candidates Included or Excluded by the Selection Criteria of the United Kingdom and Flanders (Raw Data, *n* = 416)

	United Kingdom			**Flanders**		
	Included by both criteria	Additionally included by new criteria	Excluded by both criteria	Included by both criteria	Additionally included by new criteria	Excluded by both criteria
n (%)	148 (35.6)	125 (30.0)	143 (34.4)	151 (36.34)	126 (30.24)	139 (33.42)
Mean improvement (SD)	59.1% (18.8%)	41.2% (16.6%)	16.7% (17.3%)	59.5% (18.6%)	40.4% (15.9%)	15.8% (16.7%)
Range	−3% to 96%	−29% to 86%	−53% to 58%	−3% to 96%	−28% to 81%	−52% to 46%

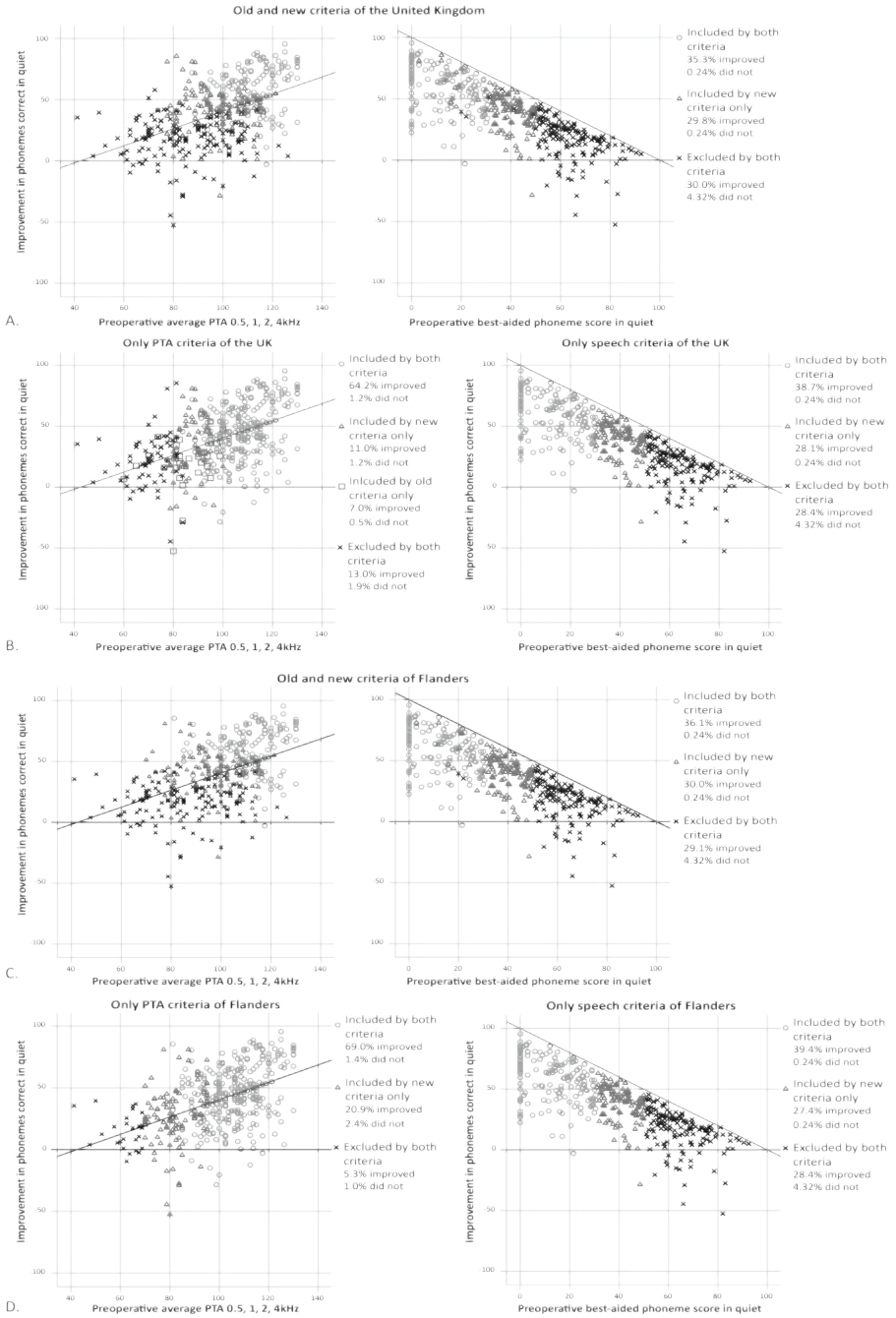

FIGURE 1. Numbers of patients included or excluded by the selection criteria in the United Kingdom and Flanders (raw data, *n* = 416). **A** and **C** shows the combination of preoperative pure-tone audiometry (PTA) and speech perception test as selection criteria. **B** and **D** illustrates each individual preoperative measure as the selection criterion.

Performance of Preoperative Measures

The change of British criteria led to a sensitivity increase from 37.1% to 68.4% (respectively, 147 and 271 patients who were included and improved postoperatively) and a specificity decrease from 95% to 90% (respectively, one and two patient(s) who were excluded and did not improve postoperatively). The change of Flemish criteria had an identical decrease of specificity and a similar increase of sensitivity from 37.9% to 69.4% (respectively, 150 with the old criteria and 275 patients with the new criteria who were included and improved postoperatively).

We constructed ROC curves to compare the performance of all preoperative measurements used by the Netherlands, UK, and Flanders (Fig. 2). Improved (≥0%) or diminished (<0%) speech understanding after CI was used as a binaural outcome, and the discrimination thresholds of the different preoperative measures were varied to calculate the sensitivity and 1-specificity of each threshold. The best-aided phoneme score in a quiet setting had the highest diagnostic ability for the improvement of speech understanding in a quiet setting, with an area under the curve (AUC) of 0.853, which was significantly higher ($p <$ .001) than all other preoperative measures, except best-aided word score in a quiet setting (AUC = 0.830; $p = .055$). Compared to the new British criteria, the old British criteria used a better approach to the PTA for predicting improved speech understanding in a quiet setting, with an AUC of 0.707 for evaluation of degree of HL at 2 and 4 kHz being significantly larger than an AUC of 0.623 for evaluation at 2 or more frequencies (0.5, 1, 2, 3, and 4 kHz) ($p = .046$). In contrast, the approach to PTA in the old and new Flanders criteria did not differ from each other (AUC of 0.688 for evaluation of the degree of HL at 0.5, 1, and 2 kHz; AUC of 0.687 for evaluation at 0.5, 1, 2, and 4 kHz; $p = .668$).

The best-aided phoneme score in a setting with noise had the highest diagnostic ability for improvement of speech understanding in noise after CI, with an AUC of 0.887, which was significantly higher ($p < .001$) than all other preoperative measures, except best-aided word score in a setting with noise (AUC = 0.784; $p = .069$). The best-aided phoneme and word score in a quiet setting (AUC = 0.684 and 0.678, respectively) had higher diagnostic abilities than the old and new PTA criteria of the UK (AUC = 0.525 and 0.491, respectively; $p = .035$ and .011), but did not differ from the old and new PTA criteria of Flanders (AUC = 0.586 and 0.567, respectively; $p = .135$ and .064).

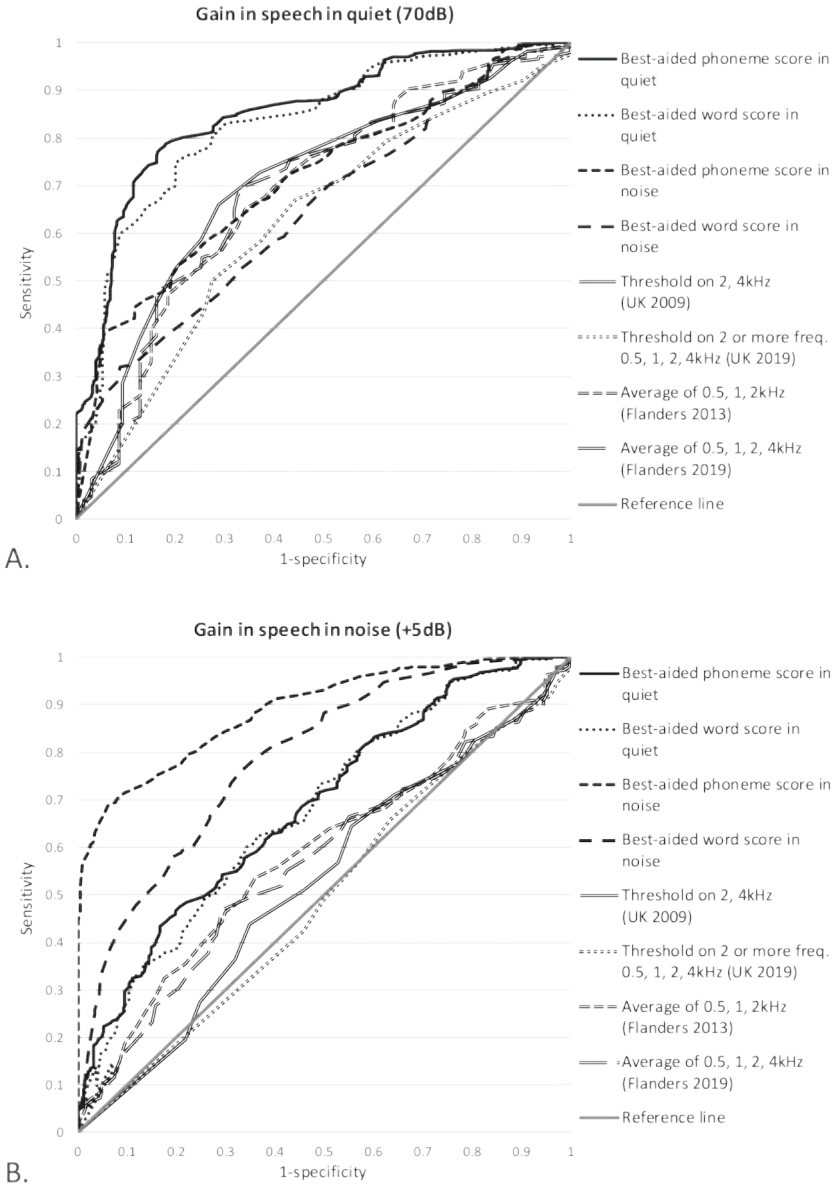

FIGURE 2. Receiver operator characteristic curves of the preoperative measures used by the Netherlands, United Kingdom, and Flanders. Diagnostic value was analyzed with the binaural outcome of improvement or no improvement (≥0% or <0%) in speech understanding in a quiet setting **(A)** or in a setting with noise **(B)** after cochlear implantation (imputed data, $n = 552$). Thresholds in pure tone audiometry represent the individual evaluation of each frequency that exceeds a certain cut-off value

DISCUSSION

In this retrospective study, we evaluated the selection criteria used for adult CI candidacy in the UK and Flanders. The new criteria introduced in 2019 resulted in a 30% increase of the inclusion of patients with improved speech understanding after CI, as well as sensitivity increase of 31% in both countries. However, the specificity of the new criteria of both countries slightly decreased from 95% to 90%. We found that preoperative best-aided SPT had the highest diagnostic ability for postoperative improvement of speech understanding. This preoperative measurement was dominant for patient selection in the new British and Flemish selection criteria, whereas the PTA prerequisites were more dominant in the old criteria. Notably, the new criteria still resulted in rejection of candidates who would be expected to gain considerably in speech understanding after implantation. Within the excluded groups, only one out of every eight patients did not exhibit postoperative improved speech understanding.

Both Flanders and the UK recently implemented less restrictive selection criteria for adult CIs (National Institute for Health and Clinical Excellence, 2009, 2019; Raeve, de & Wouters, 2013). Surprisingly, we found that the old PTA criterion of the UK was more accurate for selecting CI candidates compared to their new PTA criterion, since higher frequencies (e.g., 2 and 4 kHz) were more accurate for defining which candidates will benefit from a CI than the evaluation of more frequencies (0.5–4 kHz). This finding may be explained by the fact that the patients who lacked postoperative improvement often had residual hearing in the lower frequencies (Francis et al., 2004). However, a previous study revealed that the preoperative best-aided SPT should be used as the selection criterium, since it showed the highest diagnostic ability of all preoperative measures (Reference note 1). Both the UK and Flanders selection criteria changed from being dependent on the degree of HL to being more reliant on preoperative SPT. In general, and also in the UK and Flanders, it would be beneficial to stop using PTA criteria, and to instead use only preoperative best-aided SPT for candidacy selection.

CIs should be provided to candidates who are likely to benefit in terms of speech understanding. Policy-makers frequently discuss the degree of benefit in postoperative speech understanding; however, subjective improvement of speech understanding in daily life varies between patients. While some patients experience a substantial improvement in daily speech understanding with a postoperative score improvement of 5%, other candidates might experience almost no difference in daily listening with a postoperative

score improvement of 10%. Moreover, nowadays, candidates with progressive HL are implanted at an earlier stage, while they still have residual hearing (Cullen et al., 2004; Francis et al., 2004; Friedland et al., 2003; Gomaa et al., 2003; Summerfield & Marshall, 1995). In this scenario, the anticipated decrease of preoperative speech understanding will be eliminated by the earlier implantation, and the postoperative speech understanding would be stable, albeit sometimes comparable to their preoperative speech understanding while they still had residual hearing. It remains challenging to define selection criteria for these patients of borderline candidacy.

In the present study, we focused only on the CI selection criteria for adults with post-lingual HL, not for children with prelingual HL. These groups differ considerably in etiology, age at implantation, and the time during which they could develop language and speech with sufficient auditory input preimplantation (Peterson et al., 2010). In addition, preoperative selection criteria for children are often based on PTA due to the fact that they are not able to complete a preoperative SPT (Lovett et al., 2015). Using PTA criteria in adults resulted in the inclusion of a higher proportion of candidates without postoperative improvement, and thus had lower diagnostic ability. It would be interesting to assess the performance of PTA criteria in children with CI. Of course, one should use other measures than in the present study when pre-operative speech understanding data are not available.

The relatively lenient criteria and the large number of implantations in the LUMC enabled our present evaluation of the CI selection criteria of the UK and Flanders. Countries with more lenient CI criteria, such as Germany and Australia, could check the performance of the Dutch selection criteria (Hoppe et al., 2019; J. R. Leigh et al., 2016; Deborah Vickers et al., 2016a). These countries use selection criteria for each individual ear, which enables them to additionally implant patients with asymmetrical or unilateral HL and leads to more bilateral implantations. In the Netherlands bilateral implantation is not reimbursed for adults. Therefore, the number of adult bilateral CI users in the Netherlands is small, making it hard to analyze the effect on speech understanding. However, the current dataset does allow us to identify the group of users that obtained considerable benefit of implantation in their worst-performing ear relative to their preoperative performance. For example, patients with preoperative best-aided phoneme scores less than 50% who improved this after implantation with more than 20-30% in their worst-performing ear. Considering the correlation with preoperative speech understanding, one could expect an even better performance if the best-performing ear was implanted (Hoppe et al., 2019). This would allow to carefully select the patients who will benefit of a second implant, irrespective of the

bilateral benefits. Testing the bimodal speech understanding with an additional HA prior to sequential bilateral implantation would make this selection process even more robust.

The SPT used in our center was identical to the one used in Flanders (Bosman & Smoorenburg, 1995), but differed from the one used in the UK wherefore an estimate of the old sentence and new phoneme criteria was used (D. Vickers et al., 2013). Notably, the number of candidates who did not exhibit improved speech understanding after implantation in this study may differ from other CI centers depending on multiple factors, such as the surgeon, the amount of preserved residual hearing, type of device, the effort towards rehabilitation, and so on (Peterson et al., 2010).

In conclusion, the criteria newly introduced in Flanders and the UK resulted in increased sensitivity and increased numbers of patients who will exhibit improved speech understanding after CI. These criteria still resulted in the rejection of candidates who would be successfully implanted in the Netherlands, with only one out of eight of the rejected candidates showing no postoperative improvement. The best-aided SPT had the highest diagnostic ability and would, therefore, be the ideal instrument for CI selection criteria. These findings will improve appropriate selection of CI candidates, and help authorities and CI centers to effectively formulate selection criteria for adults with post-lingual HL.

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Pediatric Auditory Brainstem Implant Users Compared With Cochlear Implant Users With Additional Disabilities

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ABSTRACT

Objectives: To evaluate long-term language development in children with prelingual deafness who received auditory brainstem implants (ABIs) compared with children who received cochlear implants (CIs) at the same hospital. Additional non-auditory disabilities were taken into account. Study Design: Retrospective cohort study.

Setting: Tertiary referral center.

Patients: Ten children with bilateral malformations of the cochlea and/or cochlear nerve who received ABIs, including seven with additional disabilities, and 147 children with CIs as a reference group, including 22 children with additional disabilities.

Intervention: ABIs were implanted at 1.3 to 6.2 years of age. Follow-up ranged from 1.1 to 7.7 years.

Main Outcome Measures: Receptive and expressive language abilities were assessed using the Infant Toddler Meaningful Auditory Integration Scale (IT-MAIS), the Categories of Auditory Performance (CAP), the Meaningful Use of Speech Scale (MUSS), and the Speech Intelligibility Rate (SIR).

Results: Of the 10 children with ABIs, seven had long-term follow-up data. Within 1 year, six of the seven children with ABIs could identify sounds, respond to speech, and use their voice to attract attention. Language skills developed at a slower rate than in children with CIs and reached the same competence level when additional disabilities were absent. These language skills matched, on average, those of children with CIs with additional disabilities.

Conclusion: For deaf children with bilateral inner ear malformations, ABIs provide satisfactory auditory input. Children with ABIs are able to develop receptive and expressive language skills comparable to those of children with CIs with additional disabilities. Using this knowledge, preoperative parent counselling can be refined.

INTRODUCTION

The auditory brainstem implant (ABI) was initially developed for patients with neurofibromatosis type 2 (NF2) (Hitselberger et al., 2001). Later, indications for ABI expanded to children with profound hearing loss (HL) who were not eligible for cochlear implants (CIs) (Sennaroğlu, Colletti, et al., 2016). Several studies have reported that ABIs enable children to develop variable levels of receptive and expressive language skills (Noij et al., 2015; Sennaroğlu, Sennaroğlu, et al., 2016). However, to date, no studies have determined whether the skills these children develop are equivalent to the skills observed in children who receive CIs after taking into account additional disabilities. Therefore, the present study aimed to describe the long-term developmental outcomes of Dutch children with ABIs to compare their outcomes to those of a group of children who received CIs at the same institution while accounting for additional disabilities.

In recent years, the indications for ABI have gradually expanded to pediatric patients with congenital cochlear malformations, congenital cochlear nerve deficiency, cochlear trauma, and cochlear ossification after meningitis (V. Colletti et al., 2005; Kaplan et al., 2015; Merkus et al., 2014; Noij et al., 2015; Puram & Lee, 2015; Sennaroğlu, Colletti, et al., 2016; Sennaroğlu & Bajin, 2017; Shannon, 2015). Studies have reported that some children with ABIs can identify speech sounds and develop intelligible speech within 5 years after implantation (L. Colletti & Zoccante, 2008; Goffi-Gomez et al., 2012; Lundin et al., 2016; Noij et al., 2015; Puram et al., 2016; Schwartz & Wilkinson, 2017; Sennaroğlu, Sennaroğlu, et al., 2016; Shah et al., 2016). These results were promising, but ABI implantation remains a relatively new procedure, and developmental outcomes vary considerably. Therefore, further controlled long-term studies with a wide range of developmental outcomes in non-NF2 children with ABIs are warranted.

It is generally accepted that children with ABIs will develop receptive and expressive language abilities at a slower rate and achieve lower maximum performance levels than children with CIs (Eisenberg et al., 2018; Sennaroğlu, Colletti, et al., 2016; Sennaroğlu, Sennaroğlu, et al., 2016; Sung et al., 2018). To date, one study has directly compared tonal language development in pediatric recipients of CIs and ABIs that were implanted in the same center. Sound detection was achieved in a comparable timeframe for ABI recipients as their age-matched CI recipients. However, slower development of tone imitation and production was found for the pediatric ABI recipients (Sung et al., 2018).

It is important to bear in mind that children who receive CIs or ABIs are different in many ways, which makes comparisons difficult. First of all, children with CIs receive implants at an early age, whereas children with ABIs generally receive implants at an older age (e.g., after first attempting a CI, after an extensive work-up to determine the correct indication, or after treating another comorbidity that was considered a priority)(Sennaroğlu, Colletti, et al., 2016). It is well known that early (<24 months of age) rather than late implantation provides better developmental outcomes (Boons et al., 2013; Niparko, 2010; Yoshinaga-Itano, 2003). Second, the type of implant is different in design and placement. Electrodes of CIs are implanted along the tonotopical arrangement of the cochlea, whereas the electrodes of ABIs are implanted in the nucleus cochlearis with a nearly unknown tonotopic arrangement in young deaf children (Vesseur et al., 2018). Third, the etiology of HL may influence the developmental outcomes of children. Presence of a cochlea and/or cochlear nerve often indicate an intact auditory pathway in the brainstem, whereas congenital cochlear malformations and cochlear nerve deficiencies may imply an impaired auditory pathway in the brainstem (Sennaroğlu, Colletti, et al., 2016). Lastly, the prevalence of additional disabilities is expected to be higher in children who receive ABIs compared to children who receive CIs due to the etiology of HL (Sennaroğlu, Colletti, et al., 2016). The complex inner ear malformations of patients who receive ABIs are most often present in the context of complex syndromes and/or additional comorbidities. Severe additional disabilities are related to lower levels and a broader variety of expected developmental outcomes in children with ABIs or CIs (Behr et al., 2007; V. Colletti et al., 2002; Medel, n.d.; Sennaroglu et al., 2012; Sennaroğlu, Sennaroğlu, et al., 2016). Therefore, for a clinically relevant study, children with ABIs and CIs should be compared after taking into account their additional non-auditory disabilities.

ABI implantation is a relatively new procedure in young children with profound HL. Therefore, in this retrospective cohort study conducted at the first academic center for ABI in the Netherlands, we aimed to describe the entire process of ABI implantation in 10 children and their receptive and expressive outcomes. The second aim was to place the long-term developmental outcomes of children with ABIs into a broader context by comparing them to the outcomes of children who received CIs at the same center. The third aim was to stratify children according to the presence/absence of additional disabilities and explore developmental patterns in these subgroups. In line with previous findings, we hypothesized that children with ABIs develop language at a slow pace and eventually have lower scores than children with CIs (Sennaroğlu, Colletti, et al., 2016).

MATERIALS & METHODS

Children with ABIs

Ten children with prelingual deafness were implanted with ABIs at our tertiary referral center between 2011 and 2017 and followed until November 2018 (Table 1). The age at implantation ranged from 1 year 4 months to 6 years 2 months (median 2 years 9 months, mean 3 years 0 months). Three children received implants at a relatively older age due to uncertain etiology of HL or variable audiological outcomes with CIs. These children had previously received CIs at another center under the presumption of an implantable cochlea and a functional cochlear nerve based on radiological findings. However, they received very limited benefit with CIs. Radiological images from the referring hospitals were reassessed and imaging repeated when necessary. Follow-up ranged from 1 year 1 month to 7 years 7 months (median 4 years 1 month). Child no. 7 was previously described in detail by Vesseur et al. due to her cochlear nerve deficiency (Vesseur et al., 2018). Developmental outcomes were available for the first seven children with ABIs only. Child no. 8 could not use his ABI due to severe behavioral problems that prevented reliable fitting and use of his ABI. For the last two children, the follow-up after implantation was too short to record expressive language outcomes. Nevertheless, preliminary receptive language skills are described.

Children with CIs

Between 2002 and 2017, 147 children received CIs for various indications. The age at implantation varied widely (6 months to 10 years 6 months; median 2 years 6 months). Sixty-two of these children received bilateral CIs and 85 received unilateral CIs. Children with CIs were divided into two groups based on the presence (n = 22) or absence (n = 125) of one or more additional disabilities, including mild physical impairments (n = 3), severe physical impairments (n = 6), behavioral problems (n = 1), mental retardation or low intelligence quotient (n = 7), impaired communicative intentions (n = 2), severe visual impairments (n = 1), and CHARGE (Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of growth and/or development, Genital and/or urinary defects, Ear anomalies and/ or deafness) syndrome (n = 2). Children with more than one additional disability are described as having the disability with the greatest impact. Children without disabilities received CIs at a mean age of 2 years 10 months, whereas children with disabilities received CIs at a mean age of 3 years 8 months ($t[145] = -1.55$, $p>0.05$). Mean age of implantation of ABIs was not significantly different from the CI groups ($t[133] = -0.11$, $p>0.05$ and $t(30) = 0.96$, $p>0.05$).

Procedure

The present study was approved by the Medical Ethics Committee of Leiden University Medical Center (LUMC, ref. G18.001). In 2011, LUMC, the largest center for skull base surgery in the Netherlands, commenced implanting ABIs in young children. This multidisciplinary team consisted of otorhinolaryngologists, specialized audiologists, neurosurgeons, a head and neck radiologist, a pediatric psychologist, and language therapists. In our opinion, the valuable experience of intensive and prolonged collaboration between neurosurgeons and otorhinolaryngologists was an essential prerequisite for achieving successful ABI implantations in children. For each child, the multidisciplinary team evaluated whether an ABI was indicated and may provide more benefit than a CI based on anatomical variations in the cochlea and auditory nerve, audiological abilities, and cognitive abilities for hearing rehabilitation (Table 1) (Sennaroğlu & Bajin, 2017). The temporal bone was evaluated by high-resolution computerized tomography (CT) and magnetic resonance imaging (MRI). Non-verbal functioning was evaluated to assess cognitive intelligence, providing basic insight into the developmental capabilities of the child. In accordance with the Dutch Civil Code regulations relating to medical treatment contracts (WGBO), parents were informed extensively about the procedure, the risks (intra-cranial bleeding or cerebrospinal fluid (CSF) leakage, meningitis, and (temporary) brain damage), and the potential benefits of the ABI (auditory input, speech understanding, and speech production). All parents provided informed consent. All children were implanted with ABI models manufactured by MedEl (Med-El, Innsbruck, Austria). One child received the ceramic Pulsar CI100, five children received the titanium Concerto Pin, and the remaining four children received a titanium Synchrony Pin implant. A retro-sigmoidal approach was used for all operations, with a wide retro-auricular skin incision (Behr et al., 2007; Brackmann et al., 1993; V. Colletti et al., 2002; Sennaroglu et al., 2012). Intra-operative eABRs were recorded during bipolar stimulation with a positioning electrode. This intra-operative eABR process is challenging because prolonged stimulation and repositioning of the implant can lead to reduced responses, most likely due to brain tissue swelling. When an accurate position was ascertained, the final electrode array was implanted and the eABR measurements repeated. Postoperative CT was performed to confirm that the ABI electrode was positioned correctly in the lateral recess. After surgery, children were admitted to the Pediatric Intensive Care Unit for intensive monitoring during the first 24 to 48 hours. No permanent or major postoperative complications were encountered. Seven children (no. 1, 2, 3, 5, 8, and 9) endured minor postoperative complications (Table 1). The eABR measurement was repeated 4-6 weeks after implantation under general anesthesia to determine which electrode contacts elicited auditory responses and to determine the auditory thresholds. After this second eABR

measurement, the ABI was fitted while the child was awake by a specialized audiologist with broad expertise in determining the most comfortable thresholds for both ABIs and CIs. Contacts were deactivated when side effects emerged during electrode fitting (e.g., from the neighboring facial, glossopharyngeal, and accessory nerves)(Table 1). After their first fitting, an intensive rehabilitation program was followed. The training and guidance through rehabilitation with ABIs were based on clinical expertise identical to the successful program designed for children with CIs at our clinic. The rehabilitation consisted of six 1-week training sessions every 2-3 months including fitting the ABI, training to acquire auditory, receptive, and expressive language skills, and play therapy with the pediatric psychologist. After the six comprehensive weeks, the children attended regular follow-up appointments at 6-month intervals until 5 years after implantation, were after the appointments changed to once per year. Reproducible reactions were observed in six out of seven children during free-field measurements. Auditory thresholds ranged from 30 to 45 dB at frequencies between 250 and 6000 Hz.

Assessment materials

The four validated questionnaires used in this study comprised part of the standard assessments for children with CIs at our center and were identical for children with ABIs. Receptive and expressive language abilities were assessed from the parents' perspective based on an interview with a speech and language therapist. The Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) is designed to assess the child's spontaneous responses to sound in their everyday environment (Ben-Itzhak et al., 2014; Zimmerman-Phillips et al., 2000). It is a 10-item structured interview that measures vocalization behavior, alerting to sounds, and deriving meaning from sounds. Answers were scored on a 4-point Likert scale based on the occurrence of each behavior (0 = did not occur to 4 = most likely to occur). The Categories of Auditory Performance (CAP) score is a general index for indicating the hearing level of the child and consists of a 7-point rating scale arranged in order of increasing difficulty (0 = no awareness of environmental sounds, 4 = discrimination of speech sounds, 7 = use of telephone with known speaker) (Archbold et al., 1998; Nikolopoulos et al., 2005). The Meaningful Use of Speech Scale (MUSS) investigates speech production behavior, including voice control, use of spontaneous speech, and the child's ability to change his or her communication strategy to improve intelligibility (Zhong et al., 2017). It consists of a 10-item rating scale and is based on the occurrence of each behavior (0 = did not occur to 4 = most likely to occur). The Speech Intelligibility Ratings (SIR) scale evaluates the child's speech production (Allen et al., 2001). The questionnaire consists of a 5-point rating scale (1 = unintelligible, pre-recognizable

words in spoken language, 3 = intelligible to a listener who concentrates and lip-reads, 5 = intelligible to all listeners and easily understood in everyday context). All tests were translated according to international guidelines and validated in Dutch (Nottingham Early Assessment Package 2.0, The Ear Foundation)(Schaaij-Gulpen et al., 2011).

Statistical analysis

Descriptive statistics are presented as the mean, median, and percentile scores for quantitative data. As a result of the ordinal scales of the CAP and SIR, we used the median to indicate the outcomes of CI groups with and without disabilities. We used independent t-tests to assess differences between the age at implantation of CIs for children with and without additional disabilities. Missing data were imputed with the last observation carried forward for children with CIs. Data analyses were performed using the IBM SPSS Statistics 23.0 software package (IBM Corp., Armonk, NY).

TABLE 1. Characteristics of children with auditory brainstem implants (ABIs)

Child no.	Sex	Radiology	Pre-op. device	Age at detection	Age at implantation	Duration of ABI use	ABI side	Electrodes activated
1	F	Aplasia of cochlea and cochlear nerve ADS; Absent cochleovestibular nerve AD	HA ADS	0;1	1;4	7;7	AS	6
2	M	Cochlear hypoplasia (type I) and aplasia cochleovestibular nerve AS; Aplasia cochlear nerve AD	CI AD*	0;1	2;8	6;5	AS	10
3	M	Aplasia cochlear nerve ADS	-	1;6	2;9	5;4	AD	6
4	M	Cystic cochleovestibular dysplasia; Aplasia cochlear nerve AD & cochleovestibular nerve AS	CI AD	0;1	6;2	5;0	AS	5
5	F	X-linked deafness (type III hypoplasia) with ossifying labyrinthitis	HA ADS	0;1	1;11	4;2	AS	5
6	F	Aplasia cochlea and cochlear nerve ADS	-	0;1	4;1	4;0	AD	6
7	F	Cochlear hypoplasia (type III) and aplasia cochlear nerve ADS	CI ADS	0;1	4;5	3;10	AD	8
8	M	Ossifying labyrinthitis ADS	-	1;10	3;0	****	AD	7
9	F	Cochlear hypoplasia (type IV) and aplasia cochlear nerve ADS	HA ADS	0;1	1;11	1;4	AS	7
10	F	Cochlear hypoplasia (type III AD & type I AS) and aplasia cochlear nerve AD	HA ADS	0;1	1;11	1;1	AS	10

Notes: Ages are given as years;months; F=female; M=male; Pre-op. device=pre-operative device; HA=hearing aid; ABI=auditory brainstem implant; CI=cochlear implant; AD=right ear; AS=left ear; ADS=both ears; Comm. Mode=communication mode of child at the time of last follow-up; Total comm.=different modes of communication are offered, starting with visual aids; Age at detection of hearing loss, implantation, and duration of ABI use are indicated as years;months; Children are numbered based on the chronological order of the date of ABI implantation. † The nystagmus resolved completely in all children 2 days post-operatively. The subcutaneous pouches located around the surgical field contained CSF or serous fluid and spontaneously resolved in 1 to 4 months post-implantation. The one sided cerebellar syndrome of child no. 2 mostly

Complications†	Side effects	Additional disabilities	Cognition level††	Comm. mode	Education
Small subcutaneous pouch; Nystagmus	-	None	Average	Spoken language supported with sign language	Special education for hearing impairment
One-sided cerebellar syndrome (left-hand tremor); Nystagmus	Involuntary movements	None	Average	Spoken language occasionally supported with sign language	Mainstream education
CSF pouch**; Nystagmus	Vestibular stimulation	None	Average	Sign language	Special education for the deaf
-	Discomfort in shoulder	Ventricular septum defect	Average	Sign language occasionally supported with spoken language	Special education for the deaf
Small subcutaneous pouch; Nystagmus	Vestibular stimulation and itching of the cheek	Probable autism spectrum disorder	Severe delay	Total comm.	Special education for the deaf
-	-	Physical disabilities***	Severe delay	Sign language	Special education for the deaf
-	Itching with discomfort of the body and/ or limb	CHARGE syndrome	Below average	Sign language	Special education for the deaf
Nystagmus	Vestibular stimulation	Behavioral problems and epilepsy	Unreliable ****	Sign language	Special education for the deaf
Subcutaneous pouch	-	CHARGE syndrome	Average	Sign language	Special pre-school for the deaf
-	-	Multiple dysmorphic anomalies *****	Severe delay	Total comm.	Not yet

resolved after 1 week and completely after 3 months. †† Cognition level is indicated as Average (90-110), Below average (80-89), Severe delay (≤ 80). * Child no. 2 still uses his CI AD simultaneously with his ABI AS. Others did not use hearing aids/CIs on the contralateral side. ** Child no. 3 developed an intracranial CSF pouch along the right convexity, which required Ommaya drain insertion (removed after 9 months). *** Physical disabilities that were not diagnosed as a syndrome: cardiac malformations, spinal and costal anomalies, and a behavioral problem. **** Child no. 8's ABI was switched off due to other priorities, such as treatment of his behavioral problems. The severity of his behavioral problems resulted in an unreliable cognition level. ***** Multiple dysmorphic anomalies: dysmature, microcephaly, atrial septum defect type II, and failure to thrive.

RESULTS

Clinical framework of children with CIs

To determine a clinical reference of developmental outcomes for children with ABIs, this study implemented a comprehensive clinical framework based on the receptive language outcomes (IT-MAIS) of children with CIs (Figure 1). The other language outcomes of children with CIs (CAP, SIR, and MUSS) were also available (Figure 2), but it is beyond the scope of this study to discuss in detail all four language outcomes of children with CIs. Figure 1 illustrates the developmental trajectories of two subgroups of children with CIs, those with additional disabilities and those without additional disabilities.

On average, children without disabilities out-performed children with disabilities (mean IT-MAIS scores: 26.4 vs. 37.3 points after 72 months). For children without disabilities, the median score was 27.9 points 6 months after hook-up, and it reached a maximum score 48 months after hook-up (ceiling effect). For these children, the median score was higher than the mean score, indicating that the score distribution was negatively skewed (mean skewness for all nine time points = -2.32). A ceiling effect was clear after 36 months for the 90th percentile. After 60 months, the 10th percentile of children without disabilities developed a maximum score of 31.7 points.

For children with additional disabilities, the median score was 17.5 points 6 months after hook-up; it increased to 23 points at 12 months and then slightly declined at 18 months. This group reached a maximum score of 33 points after 36 months. For children with disabilities, the median and mean scores were approximately the same over time, with a normal distribution (mean skewness of all nine time points = -0.27). A ceiling effect became clear after 36 months for the 90th percentile of children with disabilities. The 10th percentile developed a maximum score of 19.4 points after 48 months.

Free-field speech audiometry of two children with ABIs

The first two children implanted with ABIs were able to perform free-field speech audiometry with the standard, open-set Dutch monosyllabic (consonant-vowel-consonant) word test (Bosman & Smoorenburg, 1995). Seventy-two and 87 months after ABI hook-up, child no. 1 scored 54% and 66% of phonemes correct at 65 dB SPL. Child no. 2 scored 70% with the ABI and 24% with the CI after 48 months. Wearing both implants simultaneously resulted in a phoneme score of 76% after 48 months of auditory rehabilitation.

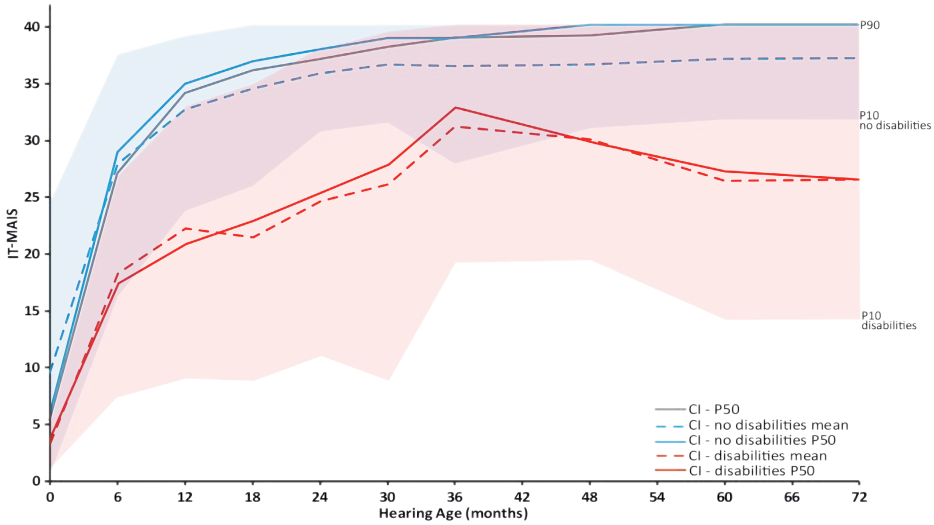


FIGURE 1. Receptive language development of children with CIs. Changes in auditory performance were measured by the Infant Toddler Meaningful Auditory Integration Scale (IT-MAIS) over time in children with CIs with and without additional disabilities. Hearing age is defined as the number of months after hooking up the implant (0=preoperative). P10=10th percentile, P50=median, P90=90th percentile.

Language outcomes of children with ABIs compared to children with CIs

Receptive language

Children no. 1 and 2 exhibited increases in receptive language over time (Figure 2A, B). The other five children had more variability (ups and downs) in outcome over time, particularly 1 year after ABI hook-up. Six out of seven children could identify environmental sounds within 1 year (i.e., 3 points on the CAP). On average, children with ABIs achieved IT-MAIS scores similar to the mean score for children with CIs with disabilities after 36 months of implant use.

The two best performing children (children no. 1 and 2) achieved open-set speech recognition after 42 and 60 months (5 and 6 points on the CAP). They even achieved similarly high scores on the IT-MAIS as the average child with CIs without additional disabilities, though it was 12 to 30 months later. Thus, these two children could understand phrases or conversations without lip-reading and recognized changes in emotion conveyed by voice. The lowest performer (child no. 5) achieved only 5 points on the IT-MAIS after 12 months, followed by a deterioration of outcomes. Autism spectrum disorder was suspected and uncertainty persisted concerning her reactions to calling her name or hearing environmental sounds. Therefore, this child was eventually categorized as a non-user. The reactions of child no. 9 varied but started to develop 9 months after hook-up (3 points, IT-

MAIS). After 11 months, the parents observed from time to time a reaction to environmental sounds and after calling her name in a quiet environment (5 points, IT-MAIS). Child no. 10 started to occasionally show a reaction to environmental sounds 6 months after hook-up (2 points, IT-MAIS).

Expressive language

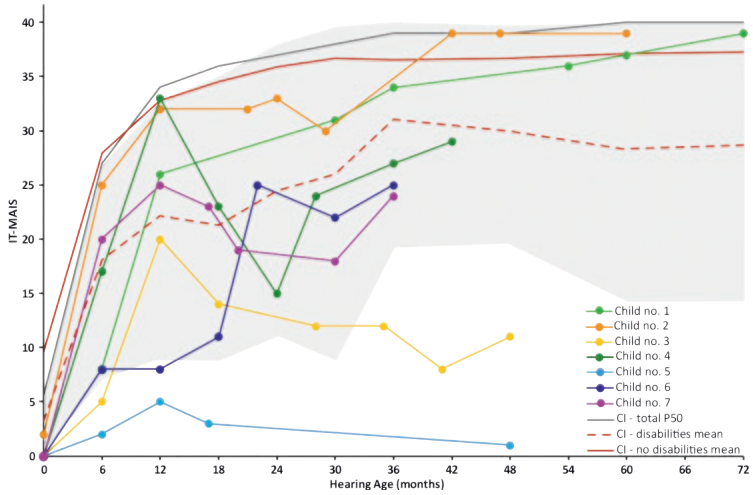
Children no. 1 and 2 demonstrated growth in expressive language over time (Figure 2C,D). The other five children exhibited more variability in outcomes, particularly 1 year after ABI hook-up. Six children produced pre-recognizable words in spoken language and could use their voice to draw attention from a listener (1 point on the SIR and 5 points on the MUSS). These scores were achieved within 1 year after ABI hook-up and equaled the scores of children with CIs with disabilities.

The two best performing children (children no. 1 and 2), after 72 and 60 months, could produce intelligible speech for listeners with little experience interpreting the speech of individuals with deafness and were able to produce 4-word sentences (4 points on the SIR and 34 and 37 out of 40 points on the MUSS). These two children developed expressive language slowly; the average child with CIs without disabilities obtained their maximum score on the MUSS after approximately 30 months, whereas the two best performing children with ABIs achieved that same level 12 to 42 months later. Child no. 2 was also able to complete the Clinical Evaluation of Language Fundamentals (CELF) – Fourth edition and achieved scores with an age-equivalent of 5-7 years at an age of 9 years (Kort et al., 2008). Child no. 4 started to develop expressive language with single words after 12 months, though his speech remained unintelligible and he did not develop further after 12 months (2 points, SIR). The lowest performer (child no. 5) produced sounds unintentionally and arbitrarily.

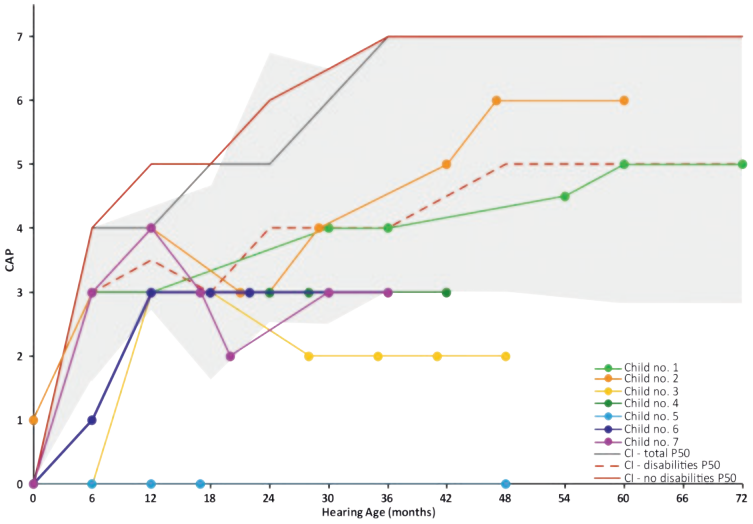
Outcomes of children with comparable non-auditory disabilities

The highest achievable outcomes of children with ABIs were compared to the outcomes of children with CIs with similar non-auditory disabilities at the same time point, if available (Table 2).

A


IT-MAIS
Hearing Age (months)
Child no. 1
Child no. 2
Child no. 3
Child no. 4
Child no. 5
Child no. 6
Child no. 7
CI - total P50
CI - disabilities mean
CI - no disabilities mean

B


CAP
Hearing Age (months)
Child no. 1
Child no. 2
Child no. 3
Child no. 4
Child no. 5
Child no. 6
Child no. 7
CI - total P50
CI - disabilities P50
CI - no disabilities P50

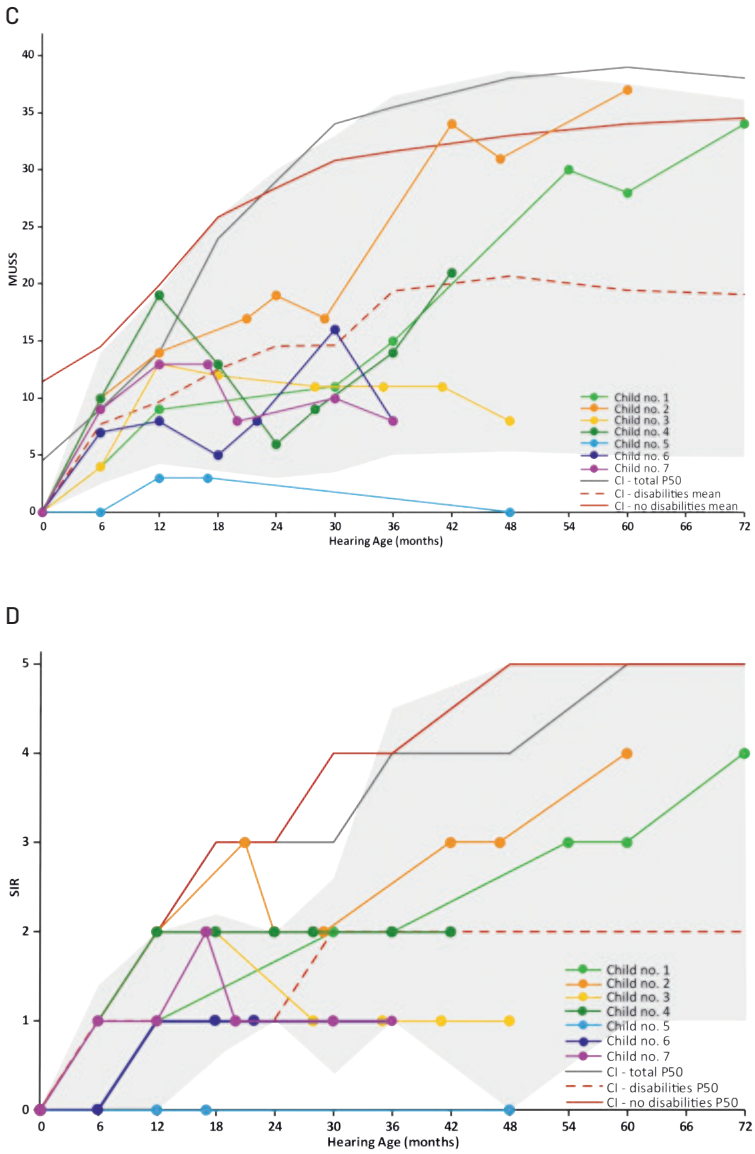


FIGURE 2. Language outcomes in children with ABIs compared to children with CIs. Receptive language was measured by the **(A)** Infant Toddler Meaningful Auditory Integration Scale (IT-MAIS) and **(B)** Categories of Auditory Performance (CAP). Expressive language was measured by the **(C)** Meaningful Use of Speech Scale (MUSS) and **(D)** Speech Intelligibility Rating Scale (SIR). Children no. 1-7 were implanted with ABIs; CI-Total P50 = the median of the total group of children with CIs. CI- disabilities/no disabilities mean/P50 = average or median scores of children with CIs, with or without additional disabilities. The grey area indicates the 10th to 90th percentiles of the children with CIs with additional disabilities. Hearing age is defined as the number of months after hooking up the implant (0=preoperative).

TABLE 2. Comparison of children with auditory brainstem implants (ABIs) and cochlear implants (CIs) based on their non-auditory disability

	ABI							CI						
	Child no.	Age at implantation	FU (months)	IT-MAIS	CAP	MUSS	SIR	*n*	Age at implantation	FU (months)	IT-MAIS	CAP	MUSS	SIR
Mild physical impairment	4	6;2	42	29	3	21	2	2	1;1-1;11	48	40	7	37-39	5
Behavioral problems	5	1;11	48	1	0	0	0	1	1;6	48	40	7	39	5
Severe physical impairment	6	4;1	36	25	3	8	1	5	1;10-5;4	36	19-30	3-5	3-29	0-4
CHARGE syndrome	7	4;5	36	24	3	8	1	2	2;7-8;7	36	29-33	3	8-16	2

Notes: Outcomes (range) indicate the highest achievable scores after the last follow-up (FU) for children with ABI; Age at implantation is given in years;months.

DISCUSSION

This longitudinal cohort study described the first 10 children implanted with ABIs in the Netherlands. This study was unique in its comparison of receptive and expressive language skills between 7 children with ABIs and 147 children with CIs who received implants at the same tertiary referral center. In general, children with ABIs vary considerably but eventually develop language scores similar to children with CIs with additional disabilities. The two best performing children (no. 1 and 2) had no additional disabilities and developed language scores similar to the average scores of children with CIs without additional disabilities 3 years after ABI hook-up. One child with ABI and no additional disabilities (no. 3) had thresholds around 40 dB but did not use his ABI for receptive and expressive language. Children with ABIs with additional non-auditory disabilities performed lower on all outcomes than children with CIs with comparable additional disabilities. However, one child with severe physical impairments had outcomes within the range of outcomes of children with CIs and comparable disabilities.

This study corroborated the notion that ABI implantation is a safe procedure with no major complications or long-term side-effects. Nevertheless, ABIs require adequate preoperative radiological and audiological evaluations by a multidisciplinary team (Sennaroğlu, Colletti, et al., 2016). In our opinion, this multidisciplinary team should have elaborate expertise in pediatric CIs and associated skull base surgery. Moreover, close cooperation between the otorhinolaryngologist and neurosurgeon is essential. Fitting an ABI is a complex procedure that is strikingly different from CI fitting. Considering these crucial factors and the small number of ABIs implanted annually, we highly recommend performing pediatric ABI implantations in a specialized academic center to ensure a safe and successful procedure.

This study added longitudinal data from multiple language assessment scales to the growing body of evidence on pediatric ABI implantations and the efficacy of ABIs for receptive and expressive language development (V. Colletti et al., 2009; Eisenberg et al., 2008; Goffi-Gomez et al., 2012; Noij et al., 2015; Sennaroğlu, Colletti, et al., 2016). In this study, six out of seven children with ABIs could ultimately respond to speech, identify environmental sounds, and use their voice to draw attention within 1 year of ABI rehabilitation. The two best performing children with ABIs could even use spoken language. These outcomes can be ascribed to the absence of additional disabilities, but may also be related to the relative early implantation and longer follow-up of these children or the

contralateral CI of the second child. One out of seven children did not appear to integrate sounds from the ABI to develop receptive and expressive language after 12 months of rehabilitation, indicating a probable non-user (Sennaroğlu, Colletti, et al., 2016).

The clinical relevance of this study in children with ABIs is further enhanced by the direct comparison with children with CIs with and without additional disabilities from the same academic center. Therefore, all children underwent the same rehabilitation program, which allows for a more equitable comparison between the groups. Our results confirmed the previous finding that children with CIs have better language outcomes in a shorter time period than children with ABIs (Sennaroğlu, Colletti, et al., 2016; Sung et al., 2018). In fact, this study showed that, on average, children with ABIs performed at similar levels as children with CIs with additional disabilities. This finding may be explained by the high prevalence of additional disabilities in our children with ABIs. These severe additional disabilities, including cognitive delays and behavioral problems, can influence language development in children (Behr et al., 2007; V. Colletti et al., 2002; Medel, n.d.; Sennaroglu et al., 2012; Sennaroğlu, Sennaroğlu, et al., 2016). In addition, multiple other aspects influence the language outcomes of children with ABIs, which are extremely different from CIs, including the etiology of HL, age at implantation, and surgical and audiological conditions, such as the tonotopic arrangement (Long et al., 2005).

The subjective language assessments depended on parental observations, which led to great variability. A decline in performance occurred mostly 1 year after ABI hook-up, when training intensity had decreased. Consequently, the lower scores after the first year could have been due to less guidance or fewer positive observations by the parents. This finding points to an opportunity to reassess rehabilitation; extended intensive guidance may be necessary for children with ABIs. In addition, individuals in the environment of children with ABIs (e.g., parents and teachers in early intervention groups) often observed limited growth in speech development after 1 year. Therefore, the verbal communication strategy for children with ABIs was switched from Dutch-supported sign language to sign language alone. This switch may have caused a decline in receptive and expressive language development after 1 year (e.g., child no. 3). Aware of this phenomenon, we now actively counsel parents and teachers during the rehabilitation phase. It is important to continue to stimulate the use of spoken language besides sign language with and without lip-reading in order to enrich the overall expressive language skills in children with ABIs. Moreover, because pediatric ABI surgery is a new approach, prospective studies with a clinical control group are necessary to ascertain the long-term efficacy of ABIs in children.

Specific questionnaires that address sign language development may be useful in future studies.

In conclusion, ABIs should be considered in children with severe bilateral malformations of the auditory nerve or cochlea. This Dutch cohort study showed that pediatric ABI implantation is a safe procedure and enables the development of receptive and expressive language skills similar to those of children with CIs with additional disabilities. Overall, this clinical comparison yielded valuable information for counseling individuals in the environment of the child, particularly for mitigating overly optimistic expectations associated with ABI implantation.

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Quality of Life of Children with Hearing Loss in Special and Mainstream Education: a Longitudinal Study

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ABSTRACT

Objectives: To compare the quality of life (QoL) of children with hearing loss (HL) and children with normal hearing (NH) and to examine how the QoL of children with HL changes over time, considering language skills, type of hearing device, degree of HL, and type of education.

Materials & Methods: This longitudinal study included 62 children with HL and their parents. Developmental outcome data were collected at two time points, when the mean ages of the children were 4 and 11 years. The Pediatric Quality of Life (PedsQL™) questionnaire, which includes assessments of Physical, Emotional, Social, and School functioning, was completed by parents at both time points and by the children with HL at the second time point. Receptive and expressive language skills at 4 years were assessed by the Reynell Developmental Language Scale. Results were compared with a Dutch normative sample.

Results: The QoL of children with HL was similar to that of children with NH at both time points on two of the four QoL scales, Emotional and Physical functioning. On the other two scales, Social and School functioning, children with HL who attended special education and children who switched to mainstream education showed lower scores than children with HL who were consistently in mainstream education and lower scores than children with NH. The School QoL of children with HL decreased over time, as did the School QoL of children with NH. Social QoL of children with cochlear implants decreased over time, but this was not the case in children with hearing aids. Language skills and the degree of HL did not clinically improve the QoL over time of preschool children with HL.

Conclusions: The QoL of children with HL in mainstream education and the Physical and Emotional QoL of all children with HL were satisfactory. It is essential to develop specific guidance regarding school activities for children with HL in special education and for children with HL who switch to mainstream education in order to increase their social QoL.

INTRODUCTION

Hearing loss (HL) greater than 25 dB HL is a serious condition that affects 1–1.7:1000 infants worldwide at birth and this number increases with age due to progressive or late onset hearing loss (Korver et al., 2010; Mehra et al., 2009; van der Ploeg et al., 2015). Children who have been identified with permanent childhood hearing impairment which require auditory amplification must cope with their HL in everyday situations. They experience language and communication problems that are consequences of their diminished auditory input (Moeller et al., 2007; Stevenson et al., 2015; Yoshinaga-Itano, 2003b). In noisy environments, such as classrooms or school playgrounds, they regularly misperceive crucial information (A. E. Geers et al., 2013; McCreery et al., 2015; Nittrouer et al., 2013; Picard & Bradley, 2001). The misunderstanding and/or misinterpreting of social situations can lead to feelings of exclusion and eventually to social and emotional difficulties (Fellinger et al., 2012; Moeller et al., 2007; Netten et al., 2015; Stephanie C P M Theunissen et al., 2014). Meta-analyses show that HL is associated with a lower quality of life (QoL) for social interactions and school activities (Nordvik et al., 2018; Roland et al., 2016). Although factors such as hearing devices (Liu et al., 2016; Roland et al., 2016; Schorr et al., 2009) and better language skills (Clark et al., 2012; Kushalnagar et al., 2014; Netten et al., 2015) contribute positively to the development and QoL of children with HL, these studies are cross-sectional, which prevents us from drawing conclusions about the causality of these relationships. Therefore, the present longitudinal study investigated the extent to which QoL of children with HL changed over time and whether language ability, type of hearing device, degree of HL, and type of education were associated with changes in QoL of these children.

Health-related QoL, which we refer to as QoL, encompasses the physical and psychosocial aspects of an individual's perception of their position in life (Whoqol Group, 1994). QoL is an important outcome measure that is widely used for clinical and research purposes to assess the impact of acute and chronic diseases, to compare affected individuals with healthy individuals, and to measure progress after treatment. It is known that QoL of children with HL increases after receiving auditory rehabilitation alongside their hearing device such as a hearing aid (HA) or cochlear implant (CI) (Liu et al., 2016; Roland et al., 2016; Schorr et al., 2009). However, there appears to be a lack of consistency within the literature regarding the comparison of QoL of children with and without HL. Some studies reported no difference (Borton et al., 2010; M Wake et al., 2006) and a number of studies showed that children with HL had a lower QoL compared to the children without HL (Rachakonda et al., 2014; Schick et al., 2013; Melissa Wake et al., 2004). When considering the different

domains of QoL, the outcomes of a meta-analysis showed that children with HL had lower general QoL in terms of school and social domains than their peers with normal hearing (NH), although children with and without HL did not differ in physical and emotional domains (Roland et al., 2016). The lower QoL with regard to school and social domains is often assumed to be related to the diminished auditory input received by children with HL. However, various other risk and protective factors affecting the QoL of individuals with HL have been identified.

Many studies emphasize the importance of language for the development of children with HL (Clark et al., 2012; Kushalnagar et al., 2014; Netten et al., 2015). Language delays are relatively common in children with HL and affect their communication, academic outcomes, and social-emotional functioning since they face more difficulties in expressing themselves and understanding others (Clark et al., 2012; Fellinger et al., 2012; Moeller et al., 2007; Stevenson et al., 2015; S.C.P.M. Theunissen, Rieffe, Kouwenberg, et al., 2014; Yoshinaga-Itano, 2003a). In addition, the type of educational setting is reported to be related to the QoL of children with HL. Children in special education report a lower QoL than children with and without HL in mainstream settings. This is associated with IQ level, additional disabilities, degree of HL, and communication abilities (Hintermair, 2011; Keilmann et al., 2007; Schick et al., 2013). Inclusive educational settings have made it possible to include children with HL without additional severe disabilities and who have adequate speech and language skills into mainstream schools with or without extra support (Chorozoglou et al., 2018; Marlatt, 2014; Raeve, de, 2010; Sontag, 2006; Xie et al., 2014). No studies to date have examined whether switching from special to mainstream education has an impact on the QoL of children with HL in comparison to children with HL who remain in special or mainstream education.

To the best of our knowledge, this nationwide study is the first to examine longitudinal changes of QoL outcomes of children with HL. Longitudinal studies can identify causal relationships and define developmental trends between groups. Data of this study were collected at two time points, when the mean ages of the children with HL were 4 and 11 years. These time points captured the beginning and end of their primary school years, allowing us to obtain an impression of the development of QoL of school-aged children with HL.

First, we compared the QoL of children with HL with the QoL of a normative group of Dutch children with NH (Roland et al., 2016). Second, we examined changes in the QoL of children with HL over time. Given the lack of research in children with HL, we based our expectations on research in children with NH and expected a decrease of QoL over time as life becomes more challenging with age (Bisegger et al., 2005; Meade & Dowswell, 2016). Third, we aimed to identify the risk and protective factors associated with changes in the QoL over time of children with HL. Based on existing literature, we expected that higher language skills and attending mainstream education would have a positive effect on the QoL (Hintermair, 2011; Keilmann et al., 2007; Moeller et al., 2007; Netten et al., 2015; Schick et al., 2013; Yoshinaga-Itano, 2003b). This study also considered the QoL of a novel group of children with HL, namely those who switched from special to mainstream education and compared them with those who remained in their educational setting between the ages of 4 and 11 years. Given the inconclusive results in terms of the level of QoL of children with either HAs or CIs (Anmyr et al., 2011; Looi et al., 2016) and the degree of HL (Patrick et al., 2011; Smith-Olinde et al., 2008), no specific expectations could be formulated in this respect.

METHODS & MATERIALS

Procedure

This longitudinal study is part of the DECIBEL study (Developmental Evaluation of Children: Impact and Benefits of Early hearing screening strategies Leiden). In this nationwide study, the parents of 204 children with HL aged 2 to 6 years agreed to participate in the first measurement, which took place from 2008 to 2010 (Time 1). After providing informed consent, the parents completed a QoL questionnaire (at this time children were too young to complete a self-report) and a general background questionnaire (characteristics of children e.g., mode of communication). With the parents' permission, the children's audiological and medical records were reviewed to collect background information and information on language skills. These outcomes were published previously (Korver et al., 2010; Netten et al., 2015; Netten, Rieffe, et al., 2017).

All 204 children who participated in the first study were invited to participate in a follow-up study 7 years later, just before they went to secondary school (Time 2). At this time point, 62 children with HL and their parents provided informed consent (a response rate of 30.4%). The main reasons for not participating at Time 2 were; additional non-auditory disabilities (*n*=6), already participating in other research or medical/audiological assessments (*n*=2), and the burden of the study along with exams during the last year of primary school together with switching to secondary school (*n*=2). The remaining 132 children did not provide a reason for non-participation. Children were visited at home between 2015 and 2016 when they were 10 to 13 years old. At this age, they reported their QoL via a self-report questionnaire and completed a language task. The parents also completed questionnaires about their child's QoL and provided additional background information (e.g., preferred communication mode). Audiological and medical records were reviewed again. Ethical approval for this study was obtained from the Medical Ethics Committee of Leiden University Medical Center (LUMC, ref. P14.270 20-01-2015).

Participants of this study compared to the non-responders at Time 2

The final study group consisted of 62 children with bilateral HL (Table 1). The 62 children with HL who participated at Time 2 and the 142 children who did not participate at Time 2 were not significantly different in terms of sex, degree of HL, or type of hearing device. The level of education of the mother, the Total QoL, and the Physical QoL of the child at Time 1 was higher in the follow-up group than in the group that participated only at Time 1 (for further information please see the supplementary table).

TABLE 1. Demographic characteristics of the children with hearing loss in this study (n=62).

	Time 1	Time 2
Age at time of assessment		
Mean, years;months (SD)	4;5 (0;9)	11;10 (0;10)
Range, years;months	2;6–6;0	10;5–13;6
Sex, *n* (%)		
Male	40 (64.5)	
Hearing amplification type, *n* (%)*		
Hearing aid	50 (80.6)	46 (74.2)
Cochlear implant	11 (17.7)	16 (25.8)
Bone-anchored hearing aid	1 (1.6)	0
Degree of hearing loss, *n* (%)**		
<40 dB (mild)	7 (11.3)	10 (16.1)
41–60 dB (moderate)	28 (45.2)	19 (30.6)
61–80 dB (severe)	14 (22.6)	14 (22.6)
>80 dB (profound)	13 (21.0)	19 (30.6)
Mean age at detection, months (SD)	13.40 (16.2)	
Age range at detection, months	0–50	
Mean age at amplification, months (SD)	21.44 (15.0)	
Age range at amplification, months	2–55	
Education, *n* (%)***		
Mainstream	20 (32.3)	47 (75.8)
Special	42 (67.7)	15 (24.2)
Preferred mode of communication, *n* (%)		
Oral language only	32 (51.5)	55 (88.7)
Spoken and sign-supported	18 (29)	7 (11.3)
Spoken, sign, and sign-supported	3 (4.8)	
Sign language only	2 (3.2)	
Sign-supported	2 (3.2)	
Sign and sign-supported	1 (1.6)	
Missing	4 (6.5)	
Receptive Language Skills, *n* (%)		
One standard deviation below average < 85	28 (52.8)	22 (35.5)
Average 85-100	14 (26.4)	18 (29.0)
Average >100	11 (20.8)	22 (35.5)

TABLE 1. Continued

	Time 1	Time 2
Expressive Language Skills, *n* (%)		
One standard deviation below average < 85	23 (37.1)	16 (25.8)
Average 85-100	14 (22.6)	23 (37.1)
Average >100	11 (17.7)	23 (37.1)
Maternal education, *n* (%)		
Primary/lower general secondary education	4 (6.4)	
Secondary vocational education	20 (32.3)	
Higher general secondary education	6 (9.7)	
College/university	32 (51.6)	

Time 1: 2008 to 2010; Time 2: 2015 to 2016. One child had a diagnosis of autism spectrum disorder, and another had a developmental delay with severe physical impairment. *After Time 1, five children received cochlear implants, and one child used a hearing aid instead of a bone-anchored hearing aid. **The degree of hearing loss was calculated by averaging unaided hearing thresholds at 500, 1000, 2000, and 4000 Hz. Between Time 1 and 2, three children changed from having moderate to having mild hearing loss because their middle ear problems resolved spontaneously or after surgery. Six children deteriorated from having moderate to having profound hearing loss from Time 1 to Time 2 due to progressive hearing loss. ***29% of the children with HL attended mainstream education at both time points, and 24.2% attended special education at both time points. Between 4 and 11 years of age, 47.8% of the children switched from special to mainstream education due to adequate speech and language skills. Of all the children in mainstream education, 44.7% received remedial teaching during school hours and 12.7% still used speech therapy at time 2.

Quality of life

The Pediatric Quality of Life Inventory (PedsQL™) (James W. Varni et al., 1999; James W. Varni & Limbers, 2009) incorporates four domains: Physical functioning (e.g. "I have problems with running"; 8 items), Emotional functioning ("I feel sad"), Social functioning ("Other children are teasing me"), and School functioning ("It is difficult to pay attention in class")(the last 3 domains have 5 items each for a total of 15 items). Each of the 23 items are scored on a 5-point Likert scale: never, 0 points; almost never, 1 point; sometimes, 2 points; often, 3 points; almost always, 4 points. Each answer is reverse-scored and rescaled to a 0 to 100 scale, where higher scores indicate better QoL. The parent questionnaires are parallel versions of the children's self-reported questionnaires, with differences in the use of age-appropriate language and first- or third-person tense. In this study, the questionnaire was completed by parents at both time points and by children with HL at the second time point. The mean QoL as reported by the parents at Time 1 and by the children with HL themselves at Time 2 were compared with the available QoL outcomes of Dutch children with NH within the same age range (mean differences presented in Table 2)(Engelen et al., 2009; Schepers et al., 2017). A clinically significant difference was considered when the reported QoL was exceeded by the absolute value of 4 (Roland et al.,

2016). Both the English and Dutch versions of the questionnaire have shown good reliability and validity (Engelen et al., 2009; Schepers et al., 2017; J W Varni et al., 2001).

Language skills

Both receptive and expressive language skills were measured with age-appropriate tests. The Dutch version of the Reynell Developmental Language Scale was administered at Time 1 (appropriate for children aged 1;2-6;3 years and language levels of 55-145) (van Eldik, 1998) and the Clinical Evaluation of Language Fundamentals - Fourth Edition (CELF-4^{NL}) at Time 2 (appropriate for children aged 5-15 years and language levels of 40-160) (Kort et al., 2008; Semel et al., 1987). Receptive language abilities were assessed with a verbal comprehension scale and expressive language abilities were assessed with word and sentence development scales. All language outcomes are standardized to norm scores according to age, using quotients in which the population mean for hearing children is 100 with a minimal clinical important difference of one standard deviation (SD) of 15 (e.g., 85 is below average and indicates language difficulties).

Intelligence

At Time 1, the nonverbal intelligence quotient (IQ) was derived from the child's medical files (either the Snijders-Oomen nonverbal intelligence tests or the Bayley Scales of Infant and Toddler Development-III) (Tellegen & Laros, 1993). Nonverbal IQ at Time 2 was assessed at home using the block design and picture concepts components of the Wechsler Intelligence Scale for Children-Third Edition (WISC-III) (Kort, W., Schittekatte, M., Compaan, E.L., Bosmans, M., Bleichrodt, N., Vermeir, G., Resing, W.C.M., Verhaeghe, 2002; Wechsler, 1991).

Statistical analysis

Statistical analysis was performed on the final study group consisting of 62 children with bilateral HL. To compare the QoL of children with HL with Dutch normative data, summary independent sample *t*-tests were performed for the Total QoL score and for each domain separately (Engelen et al., 2009; Schepers et al., 2017). To compare self-reported QoL with parent-reported QoL at Time 2, we used a dependent sample *t*-test. To evaluate whether QoL of children with HL had changed after 7 years, linear mixed models were used. Because we were interested in the development of QoL over time, parent-reported data of the final 62 children with HL were used as they reported the QoL of their children with HL at both time points. To control for confounders, sex and age at Time 1 were added as fixed effects in these linear mixed models (Bisegger et al., 2005). Next, we examined the

effects of the following factors on changes in the QoL over time: language skills at Time 1, type of hearing device, degree of HL, and educational settings (mainstream education, special education, or switched from special to mainstream education between the two time points). Accordingly, each variable was sequentially added (first main effect and second interaction effect with Time). In addition to sex and age at Time 1, level of IQ was added as a confounder to the model with educational settings. Due to the large number of missing IQ scores at Time 1, the IQ-score at Time 2 was used in the analyses (Pearson's correlation between IQ Time 1 and Time 2 = 0.385, $p = 0.027$)(Neisser et al., 1996). All linear mixed models contained a single random effect for each subject and fixed effects for the independent variables. Statistical analyses were performed using the IBM SPSS Statistics 23.0 software package.

Missing data

In our final study sample of 62 children, receptive language, expressive language, and IQ scores at Time 1 were missing for 9, 12, and 28 children with HL, respectively (Table 2). At Time 2, one child was unable to complete the QoL-questionnaire and IQ measure due to her additional non-auditory disability, one child lost her focus while completing the IQ measure at the end of the testing session, and six parent-reported QoL outcome questionnaires were incomplete. The pattern of missing data was examined using Little's MCAR test ($c^2 = 483.47$, DF = 529, $p = 0.92$), which indicated that the data were missing at random. When conducting standard analyses, such as independent *t*-tests, incomplete cases will automatically be excluded (Netten, Dekker, et al., 2017). This can introduce bias and lower statistical power if these participants were excluded from the analyses. This type of missing data can be reconstructed using multiple imputations (Buuren, 2012; Netten, Dekker, et al., 2017; Sterne et al., 2009). We used 10 imputations to create good estimates of the missing data (Sterne et al., 2009). The imputations were based on the child's age at Time 1 and Time 2, language skills, IQ, sex, educational status of the parents, and QoL outcomes. Ten imputations were performed, and the pooled results are reported in Tables 3 and 4 (Sterne et al., 2009). There were no differences between outcomes with the original data and the imputed data.

RESULTS

The outcomes are reported in order of the three aims of this study.

Comparison of the QoL of children with HL versus normative QoL data from Dutch children with NH

The psychometric properties and mean QoL results of the final study sample of 62 children with HL are shown in Table 2. At Time 1, parents reported a clinically lower Total QoL for children with HL compared to the parent-reported normative data from Dutch children with NH. When considering the different subscales reported by parents, QoL scores among children with HL were clinically lower compared to children with NH in the Social and School domains at Time 1. At Time 2, the children with HL self-reported a clinically lower Total QoL compared to the self-reported normative data from Dutch children with NH. Concerning the subscales, the School QoL scores among children with HL were clinically lower compared to children with NH at Time 2. Parent-reported and self-reported QoL scores of children with HL were not significantly and clinically different at Time 2, except for the Physical QoL, which was reported more positively by the parents.

Changes in QoL over time and the relation with risk and protective factors

Changes in QoL over time were analyzed using the parent-reported data of 62 children with HL and a linear mixed model with Time as the time-dependent variable. A positive coefficient of time indicates an increase in QoL over time and a negative coefficient indicates a decrease in QoL over time (Table 3).

The parent-reported Total QoL of children with HL decreased significantly from Time 1 to Time 2, but this was not clinically different as the absolute value of 4 was not exceeded (Roland et al., 2016). When considering the different subscales, no clinical differences were observed in parent-reported Physical QoL and Emotional QoL between Time 1 and Time 2, but the scores on the School QoL and Social QoL subscales had significantly and clinically declined at Time 2. Notably, the decrease in parent-reported Social QoL was found only in children with CIs (Figure 1A and Table 4), while children with HAs had similar parent-reported Social QoL outcomes at both time points. Post-hoc analyses showed that 75% of children with CIs (12 of 16 children), but only 37% of children with HAs (17 of 46 children), had switched from special to mainstream education ($p<0.05$). Changes in parent-reported Total QoL, Physical QoL, Social QoL, and School QoL were not influenced by language or degree of HL. Only parent-reported Emotional QoL was influenced by receptive language (Figure 1B and Table 4). Children with HL with average receptive language skills (100) at Time 1 had significantly but not clinically higher Emotional QoL at Time 2 (Figure 1B).

TABLE 2. Psychometric properties and mean scores for quality of life, language skills, and intelligence quotient of the children with hearing loss in this study (*n*=62).

					Time 1			Time 2		
			Cronbach's alpha		Mean (SD)	Mean (SD)	Mean difference compared to norm (95% confidence interval)	Mean (SD)	Mean difference compared to norm (95% confidence interval)	Mean (SD)
	Number of items	Answer range	Child	Parent	Child	Parent		Child		Parent†
Pediatric Quality of Life Inventory 4.0						*n*=61		*n*=61		*n*=56
Total score	23	0–4	0.88	0.89		**84.43 (10.77)**	**-4.1 (-6.9;-1.3)**	**78.05 (13.67)**	**-4.06 (7.7;-0.5)**	80.61 (10.99)
Physical	8	0–4	0.79	0.80		90.52 (12.86)	-1.3 (-4.5;1.9)	84.52 (14.34)*	-0.35 (-4.1;3.4)	91.05 (10.81)*
Emotional	5	0–4	0.81	0.79		76.33 (16.05)	-2.1 (-6.3;2.0)	71.81 (20.90)	-5.24 (-10.7;0.2)	75.71 (16.38)
Social	5	0–4	0.71	0.88		**81.33 (15.56)**	**-8.76 (-12.6;-4.9)**	82.21 (16.33)	-3.93 (-8.3;0.4)	76.88 (21.48)
School	5	0–4	0.68	0.74		**86.43 (14.57)**	**-7.63 (-11.2;-4.1)**	**69.57 (16.30)**	**-9.13 (13.5;-4.8)**	73.05 (15.30)
Language skills										
Receptive language	67-70				84.70 (19.83) (*n*=53)			91.44 (18.68) (*n*=62)		
Expressive language	102-77				87.53 (14.05) (*n*=50)			94.02 (18.33) (*n*=62)		
Non-verbal intelligence	26				105.21 (13.40) (*n*=34)			101.18 (17.71) (*n*=60)		

SD, standard deviation; n, number of participants who completed the questionnaire or task; Mean difference and 95% confidence interval indicates differences (**Bold=***p*<0.05) between participants and Dutch normative samples (Engelen et al., 2009; Schepers et al., 2017); †, no normative data available for parent-reported measures at this age; **p*<0.05 differences between parent- and self-reported data at Time 2. Language skills and intelligence are shown as standard scores (mean of 100 and SD of 15).

Level of QoL differs according to sex and type of education

To appraise whether QoL of children with HL had changed after 7 years, linear mixed models were used with parent-report data. Based on these parents' reports, sex and the educational setting of children with HL influenced the level of QoL of these children at both time points. When controlled for age and time, linear mixed models showed that boys had a higher Total QoL and Social QoL than girls at both 4 and 11 years of age (coefficient of sex (boys=1 girls=0) for Total QoL = 5.88, [0.93, 10.83], $p < 0.05$; coefficient of sex (boys=1 girls=0) for Social QoL = 13.27, [5.31, 21.22], $p < 0.001$). When corrected for sex, age, IQ, and time, linear mixed models revealed that children who attended special education at one or at both time points had significantly and clinically lower Total QoL, School QoL, and Social QoL than children in mainstream education (Figure 1C and Table 4). Children with HL in mainstream education had similar levels of School QoL and Social QoL to children with NH at both time points.

TABLE 3. Changes of quality of life over time of children with hearing loss (*n*=62) analyzed with linear mixed models.

	Time		**Time**	
	Uncorrected		Corrected for sex and age at Time 1	
	Coefficients	95% Confidence interval	Coefficients	95% Confidence interval
Total QoL	**-3.59***	**[-6.47, -0.70]**	**-3.86****	**[-6.74, -0.98]**
Physical QoL	0.60	[-2.60, 3.80]	0.39	[-2.82, 3.60]
Emotional QoL	0.10	[-5.21, 5.40]	-0.10	[-5.47, 5.27]
Social QoL	-4.19	[-9.54, 1.16]	-4.64	[-9.98, 0.69]
School QoL	**-13.49*****	**[-18.18, -8.80]**	**-13.73*****	**[-18.44, -9.02]**

Bold *$p \leq 0.05$, **$p \leq 0.01$, ***$p \leq 0.001$; Time: 0 = Time 1, 1 = Time 2; QoL, quality of life

TABLE 4. The association between hearing loss related factors and changes of quality of life over time of children with hearing loss (n=62) analyzed with linear mixed models.

	Receptive language			**Hearing device**			**Educational setting**		
		Coeffi-cients	95% confidence interval		Coeffi-cients	95% confidence interval		Coeffi-cients	95% confidence interval
Total QoL	Receptive language	0.02	[-0.15, 0.18]	Hearing device	-3.92	[-10.49, 2.66]	Special education	**-7.79***	**[-13.82, -1.77]**
	Time* Receptive language	0.05	[-0.10, 0.20]	Time* Hearing device	4.67	[-1.76, 11.11]	Switched from education	**-6.27***	**[-11.54, -1.00]**
							Mainstream education	**0**	
Physical QoL	Receptive language	0.02	[-0.16, 0.20]	Hearing device	-4.77	[-8.28, 1.27]	Special education	-5.22	[-12.15, 1.70]
	Time* Receptive language	-0.04	[-0.21, 0.13]	Time* Hearing device	6.19	[-0.92, 13.31]	Switched from education	-4.38	[-10.43, 1.66]
							Mainstream education	0	
Emo-tional QoL	Receptive language	-0.06	[-0.27, 0.16]	Hearing device	-3.22	[-7.99, 1.55]	Special education	-1.07	[-9.66, 7.52]
	Time* Receptive language	**0.33***	**[0.04, 0.62]**	Time* Hearing device	5.32	[-6.85, 17.49]	Switched from education	-6.64	[-14.15, 0.86]
							Mainstream education	0	

TABLE 4. Continued

	Receptive language			Hearing device			Educational setting		
		Coeffi-cients	95% confidence interval		Coeffi-cients	95% confidence interval		Coeffi-cients	95% confidence interval
Social QoL	Receptive language	0.04	[-0.21, 0.30]	Hearing device	-6.73	[-17.20, 3.70]	Special education	**-12.99***	**[-23.00, -2.98]**
	Time* Receptive language	0.15	[-0.13, 0.43]	Time* Hearing device	**14.04***	**[2.45, 25.63]**	Switched from education	**-8.83***	**[-17.58, -0.08]**
							Mainstream education	**0**	
School QoL	Receptive language	0.07	[-0.15, 0.30]	Hearing device	0.32	[-4.18, 4.81]	Special education	**-14.22*****	**[-21.85, -6.59]**
	Time* Receptive language	-0.11	[-0.37, 0.15]	Time* Hearing device	-6.10	[-16.73, 4.54]	Switched from education	**-6.90***	**[-13.55, -0.26]**
							Mainstream education	**0**	

Bold *$p \leq 0.05$, **$p \leq 0.01$, ***$p \leq 0.001$; Time: 0 = Time 1, 1 = Time 2; hearing device: 0 = cochlear implant, 1 = hearing aid; QoL, quality of life; All models are corrected for at least sex and age at Time 1, educational setting also for IQ.

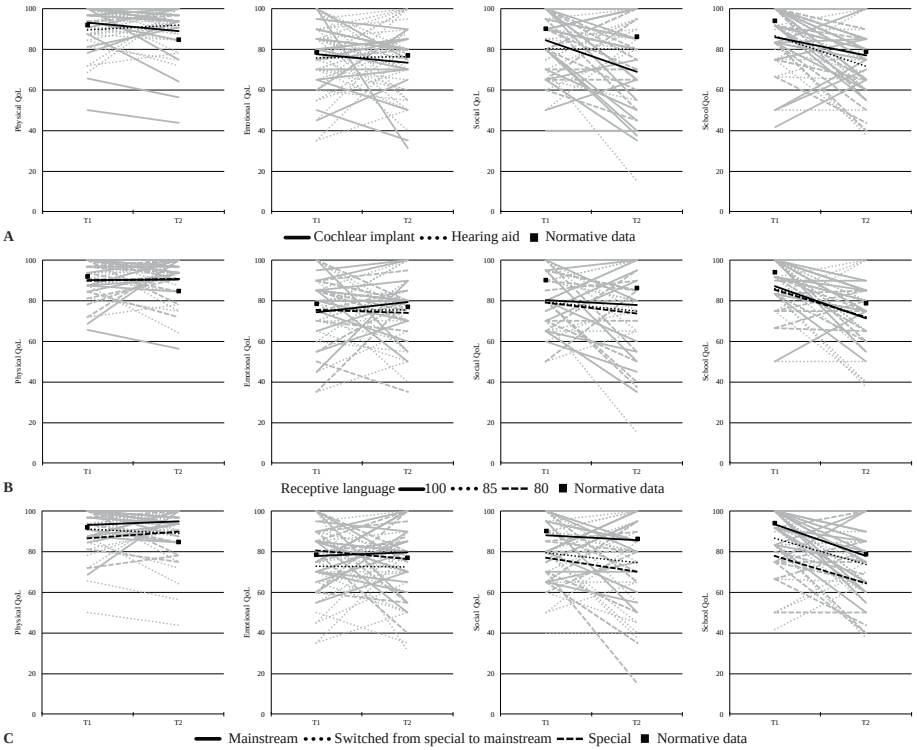

FIGURE 1. Changes in the quality of life (QoL) over time of children with HL as reported by their parents (n = 62). Individual trajectories are in grey and group differences are plotted in black. Note. Normative data = Time 1 parent-reported data and Time 2 self-reported data of Dutch children with normal hearing (Engelen et al., 2009; Schepers et al., 2017). **A.** Children with cochlear implants showed a clinical decrease in their Social QoL at the second time point, while children with hearing aids had similar Social QoL levels at both time points. No significant difference was found between children with cochlear implants and hearing aids in the other subscales of QoL. **B.** Children with HL with adequate receptive language skills (e.g. 100) at age 4 showed a significant increase in their Emotional QoL over time which was not clinically different (>4 points). When receptive language skills were below average (e.g. 80), the Emotional QoL decreased slightly over time. Receptive language skills did not influence the other subscales of QoL. **C.** At both time points, children with HL who attended special education (n = 24.2%) and who switched from special to mainstream education (n = 46.8%) had a clinically lower Social QoL and School QoL than children with HL in mainstream education (n = 29.0%). There were no differences between these educational groups in the Physical and Emotional domain.

DISCUSSION

This longitudinal study examined how type of hearing device and type of education were associated with changes in the QoL of children with HL over a 7-year period. We used the PedsQL™ questionnaire, which includes assessments of Physical, Emotional, Social, and School functioning. The outcomes of this study confirmed that the Emotional QoL and Physical QoL of 4- and 11-year-old children with HL were similar to the QoL of their peers with NH. The Social QoL and School QoL of children with HL in mainstream education were also on par with these measures in children with NH. However, compared to children with and without HL in mainstream education, children with HL who were in special education or who switched from special to mainstream education had lower levels of Social and School QoL. Regarding changes in the QoL, children with HL who had at least average receptive language skills at 4 years of age had statistically but not clinically improved emotional QoL at 11 years of age. In line with findings in children with NH, School QoL decreased between the ages of 4 and 11 years. Social QoL also declined over time, but only for children with CIs; in contrast, the Social QoL of children with HAs did not differ at both assessment times. These findings were all of clinical importance and can be used to modify and improve personalized care for children with HL by creating a focus on their social interactions and school activities.

QoL of children with and without HL

Our findings confirmed those of the meta-analysis by Roland et al. (Roland et al., 2016), in that we found that the Emotional QoL and the Physical QoL of children with HL were similar to those of children with NH at the ages of 4 and 11 years. A novel finding in group differences was the similar level of Social and School QoL of children with HL in mainstream and children with NH at both ages.

Social and School QoL of children with HL in different educational settings

Children in special education and children who switched from special to mainstream education had lower Social QoL and School QoL than children with HL in mainstream education and children with NH at both time points. This is in line with previous studies which found that children with HL in special schools, as opposed to children in mainstream schools, have more problems due to their difficulties with language and communication and presumably some additional non-auditory disabilities, all of which may contribute negatively to their QoL (Hintermair, 2011; Keilmann et al., 2007; Schick et al., 2013; S.C.P.M. Theunissen, Rieffe, Kouwenberg, et al., 2014; Wakil et al., 2014; Zaidman-zait et al., 2017).

Almost half of the children with HL in this study had adequate language skills in the range of children with NH, which enabled them to transfer from special to mainstream education. Therefore, this study is the first to investigate the impact of a school transition on the QoL of children with HL. The Social QoL and School QoL of children who switched from special to mainstream education were lower at both time points compared to children with HL in mainstream education. At the first assessment time point, 4-year-old children with HL were in special education and had to catch up due to language and communication delays (McCreery et al., 2015). It is likely that social interactions and school activities were more challenging at that age (Keilmann et al., 2007; Schick et al., 2013). Seven years later, children with HL who switched to mainstream education may have struggled with the demands of a faster teaching pace and/or with the less favorable acoustics of mainstream classrooms (Hintermair, 2011). Furthermore, due to the level of (extra) noise, children with HL regularly misperceive information in class and social situations, which can lead to feelings of exclusion (A. E. Geers et al., 2013; Mccreery et al., 2015; Nittrouer et al., 2013; Picard & Bradley, 2001; Rieffe et al., 2018; Wolters et al., 2011). These feelings of exclusion might even be enhanced since children with HL in mainstream settings are often the only ones wearing hearing technology in a hearing classroom. This can affect their self-perception, social development, friendships, and eventually their QoL (I. W. Leigh & Leigh, 1999; Rieffe et al., 2018; Xie et al., 2014). Based on the results of this study, it is important to consider specific and long-term guidance regarding school activities and social interactions for children with HL who switch from special to mainstream education.

Changes in QoL over time

According to parents, the School QoL and Social QoL of children with HL changed over time. All children with HL experienced a decline in School QoL after 7 years, which is in line with findings among children with NH (Engelen et al., 2009; Schepers et al., 2017). This decrease may have been related to their developmental stage of adolescence and concomitantly a more demanding educational curriculum for older children, which the children must learn to cope with.

In contrast to our expectations, the receptive and expressive language scores of 4-year-old children with HL did not clinically contribute to the development of QoL. The absence of a clear relation between language skills and QoL in children with HL was also found in other studies on language skills and social emotional functioning (Beitchmen et al., 1986; Constantinescu-Sharpe et al., 2017; Horwitz et al., 2003; Netten et al., 2015, 2018). They found that communication skills and not language skills are more import for social

functioning which in turn can affect the wellbeing of children with HL. Language skills such as vocabulary are learned by professionals in schools and are important to develop communication skills (Moeller et al., 2007; Netten et al., 2015). Yet, the social rules are learned in a more indirect way by observing and communicating with others outside of school or at the playground. Understanding a joke for example requires the understanding behind the vocabulary and relies on the pragmatics within communication. It is therefore more important that children with HL learn to use their language capacities in the right way.

Children with HAs or CIs

Except for Social QoL, changes in the QoL of children with CIs did not differ from changes in children with HAs. The parents of children with HAs reported similar Social QoL when their children were 4 and 11 years old, whereas parents of children with CIs reported a decrease in Social QoL after 7 years. This finding should be interpreted with care due to the difference in group size (the CI group was three times smaller than the HA group) and the difference in degree and etiology of HL between groups. However, three plausible explanations could be suggested for the change in Social QoL over time for children with CIs. First, children with CIs participated in intensive rehabilitation programs in their early years. Such programs gave them access to speech therapists, psychologists, qualified teachers for children with HL, and other professionals. However, for older children with CIs, the frequency of rehabilitation services usually decreases to once a year and children must be more self-reliant which can result in a lower QoL. Second, the decrease in Social QoL could be a consequence of the fact that parents of children with CIs may expect their child to be like children with NH and social problems in their 4-year-old child may go unnoticed (A Zaidman-Zait & Most, 2005). When the children with CIs are 11 years old, they can express themselves concerning their difficulties with social interactions and parents of children with CIs may be, therefore, more aware of the difficulties. Third, regarding the educational settings of these two groups, 75% of children with CIs, but just 37% of children with HAs, switched from special to mainstream education between the two time points. This greater number of children with CIs who switched educational settings may have had more of an impact on their social development than explained previously.

Strengths and limitations

One of the strengths of this study is its longitudinal design. It provides a unique, and valid perspective on QoL changes in children with HL over a period of 7 years, from pre-school to pre-adolescence. It would be informative to follow this cohort into adolescence, when the demands of social interactions and school become even greater. This third time point

would provide more information regarding causal relationships and could further validate our findings. In addition, children in this study were born in the implementation phase of the Newborn Hearing Screening preventing us from drawing conclusions concerning the age at detection or the age at first amplification and QoL. However, factors like audibility, early access to amplification, and family counseling have been proven to influence language skills in children with HL and should therefore be integrated in future studies when studying QoL in this group (J. B. Tomblin et al., 2015). The study had three main limitations. First, the QoL of children with HL was compared to normative QoL data instead of being compared to data from a control group of children with NH. Second, compared to the 4-year-old children who only participated at the first time point, 4-year-old children with HL who participated at both time points had a higher Total QoL as rated by their parents and had mothers with a higher educational degree. These differences together with the response rate of 30.4% may have potentially led to selection bias. From a statistical point of view, the linear mixed models used address this problem if the missing data is missing "at random", i.e. the reason for missing data can be explained by the covariates in the model. As we have included sex and age in the model, we believe that important sources of bias have been considered. This being said, the possibility of bias cannot be eliminated. Third, this study used a generic health-related QoL questionnaire to compare the QoL of children with and without HL and to examine the development of QoL over time for children with HL. Despite the relative positive findings concerning the generic QoL of the children with HL in our study, children with HL could still have hearing-specific problems and consequently a lower hearing-specific QoL (Clark et al., 2012; Rachakonda et al., 2014; Umansky et al., 2011). Future studies should therefore take the development of hearing-specific QoL into account for children with HL.

Conclusion

In this longitudinal study, the Physical and Emotional QoL levels of children with HL were in line with those of children with NH at the ages of 4 and 11 years. Half of the children with HL in this study had appropriate language skills, which allowed them to switch from special to mainstream education. However, for good clinical practice, they should receive extra guidance and long-term support for school activities and social interactions. In particular, school-aged children with CIs may need extra guidance for their social functioning. It is our expectation that these findings can be used to improve personalized guidance for children with HL.

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SUPPLEMENTARY TABLE. Demographic characteristics of the children with hearing loss who participated in the follow-up study and who did not.

	Participants of follow-up study (n=62)	**Drop-outs (n=142)**
Age at first study		
Mean, years; months (SD)	4;5 (0;9)	4;4 (1;0)
Range, years; months	2;6–6;0	2;6–6;2
Sex, *n* (%)		
Male	40 (64.5)	79 (55.6)
Hearing amplification type, *n* (%)		
Hearing aid	50 (80.6)	94 (66.2)
Cochlear implant	11 (17.7)	34 (23.9)
Bone-anchored hearing aid	1 (1.6)	8 (5.6)
Missing	0	
Degree of hearing loss, *n* (%)		
<40 dB (mild)	7 (11.3)	15 (10.6)
41–60 dB (moderate)	28 (45.2)	41 (28.9)
61–80 dB (severe)	14 (22.6)	42 (29.6)
>80 dB (profound)	13 (21.0)	38 (26.8)
Missing	0	6 (4.2)
Mean age at detection, months (SD)	13.40 (16.2)	12.55 (14.4)
Age range at detection, months	0–50	1–60
Mean age at amplification, months (SD)	21.44 (15.0)	18.93 (14.8)
Age range at amplification, months	2–55	1–60
Education, *n* (%)		
Mainstream	20 (32.3)	36 (25.4)
Special	42 (67.7)	56 (39.4)
Missing	0	50 (35.2)
Preferred mode of communication, *n* (%)		
Oral language only	32 (51.5)	48 (33.8)
Spoken and sign-supported	18 (29)	37 (26.1)
Sign and/or sign-supported	5 (8)	23 (16.2)
Spoken, sign, and sign-supported	3 (4.8)	4 (2.8)
Missing	4 (6.5)	20 (14.1)

SUPPLEMENTARY TABLE. Continued

	Participants of follow-up study (n=62)	Drop-outs (n=142)
Maternal education, n (%)*		
Primary/lower general secondary education	4 (6.4)	26 (18.3)
Secondary vocational education	20 (32.3)	49 (34.5)
Higher general secondary education	6 (9.7)	8 (5.6)
College/university	32 (51.6)	55 (38.7)
Missing	0	5 (3.5)
Pediatric Quality of Life Inventory 4.0, Mean (SD)	*n*=61	*n=130*
Total score*	84.4 (10.8)	80.8 (12.2)
Physical*	90.5 (12.9)	85.7 (18.3)
Emotional	76.3 (16.1)	73.9 (14.6)
Social	81.3 (15.6)	79.7 (16.8)
School	86.4 (14.6)	81.9 (17.6)
Language skills, Mean (SD)		
Receptive language	84.70 (19.83)(*n=53)*	82.0 (15.6)(*n=95)*
Expressive language	87.53 (14.05)(*n=50)*	83.8 (15.2)(*n=85)*
Non-verbal intelligence, Mean (SD)	105.21 (13.40)(*n=34)*	101.9 (16.9)(*n=51)*

Bold *p≤0.05, **p≤0.01, ***p≤0.001

The School Career of Children With Hearing Loss in Different Primary Educational Settings: A Large Longitudinal Nationwide Study

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ABSTRACT

Children with hearing loss (HL) are at risk for a lower educational achievement. This longitudinal study compared the school career of a nationwide Dutch cohort with and without HL based on descriptive data of the governmental authority Statistics Netherlands. From 2008 to 2018, 3,367,129 children, of whom 1,193 used cochlear implants (CIs) and 8,874 used hearing aids (HAs), were attending primary and/or secondary education. Sixty-one percent of children with HL attended mainstream and 31% special primary education. Compared to mainstreamed pupils without HL, mainstreamed pupils with HL achieved lower levels for language and mathematics in primary education but eventually attended comparable types of secondary education. Children with HL attending special primary education attained lower types of secondary education compared to mainstreamed peers with and without HL. These findings suggest that future educational (and as a result professional) attainment of a child with HL depends on the type of primary educational setting.

INTRODUCTION

Early detection and hearing rehabilitation with hearing aids (HAs) and/or cochlear implants (CIs), family-centered early intervention, preschool treatment groups, and extra guidance at school have brought great enhancement for the development of children with hearing loss (HL) (Marschark & Spencer, 2011; Moeller et al., 2015; Yoshinaga-Itano, 2004). However, it remains unclear whether children with HL are nowadays able to reach their full potential in education, or that they are still at risk due to their HL (Dammeyer & Marschark, 2016; Illg et al., 2017; Nagle et al., 2016; Rydberg et al., 2009; Winn, 2007). Current knowledge regarding the school career of children with HL is built upon cohort studies that either examined the academic achievements during primary education (Harris et al., 2017; Khairi Md Daud et al., 2010; Qi & Mitchell, 2012; Wauters et al., 2006) or assessed the educational attainment of college students who were able to graduate from secondary or high school (Dammeyer & Marschark, 2016; Illg et al., 2017; Nagle et al., 2016; Rydberg et al., 2009; Winn, 2007). There is a lack of nationwide studies with a long-term follow-up investigating the type of secondary education of a large population with HL. Therefore, the present study examined the type of primary and secondary education in addition to the academic achievements of children with and without HL using a longitudinal design and a nationwide large sample in the Netherlands.

Children with HL in special or mainstream primary education

Previously, in many Western countries children with HL were obliged to attend special schools for the deaf and hard of hearing (Mitchell & Karchmer, 2006). In these schools, children were surrounded by peers with HL and separated from their hearing peers. With current legislation in most Western countries (e.g., Education for all handicapped children act in the United States in 1975; Inclusive education in the Netherlands, August 2014), children with HL are encouraged to attend mainstream schools. As a result, respectively 78% and 85% of the children with HL in the United States and Australia attend mainstream schools (Punch & Hyde, 2010; Shaver et al., 2014). Most of these children are HA-users as their relatively lower degree of HL allows them to attend mainstream schools at an early age (Shaver et al., 2014; Verhaert et al., 2008). Cohort studies examining the educational setting of children with CI reported a wide range of 38 to 64% children who fully or partially attended mainstream classes (Archbold et al., 2002; Christiansen & Leigh, 2002; Punch & Hyde, 2010). To our knowledge, the nationwide percentages of children with CIs or HAs in each educational setting have not yet been identified for other Western countries, such as the Netherlands.

There is still discussion concerning which children with HL will benefit from mainstream or special education (Stinson & Kluwin, 2012). The inclusive education policy of the Netherlands enables children with HL to have access to a mainstream school curriculum at a pace and in the same manner as it is taught to their hearing peers with an option to receive additional support of special education services. Literature has shown that children with HL in primary mainstream schools are likely to have higher academic achievements compared to children with HL in special schools (Marschark et al., 2015; Powers, 1999; Wauters et al., 2006). Still, mainstream education can be challenging for children with HL. Their auditory input is influenced by poor acoustics in large classrooms or background noise due to mumbling classmates which could lead to misunderstanding instructions and explanations of teachers. In other words, instructions in mainstream settings are not always communicatively accessible for children with HL.

Children with HL who have language and/or cognitive delays or special communication needs may lag behind even more in academic achievement due to their inability to keep up with mainstream education. Most of these children are therefore placed in special schools where support is provided in small groups or even on an individual level to allow for intensive guidance on their school performance. Other reasons, such as the severity of HL, additional handicaps, ethnicity, or the reliance on sign language (Israelite et al., 2002; Karchmer et al., 1982; Knoors & Vervloed, 2012; Rydberg et al., 2009; Shaver et al., 2014), may influence the decision whether the child will benefit from special (for the deaf or hard of hearing) or mainstream education. Due to these reasons, children with HL in special education are often supported by sign language and individually evaluated instead of taking standardized academic achievement tests.

Essential subjects in primary education

To continue in secondary education, children need to acquire essential scholastic skills. Among the diverse subjects in school, language and mathematics are two main subjects of standardized achievement tests in primary education. Commencing with a language delay due to a deprived auditory input can continue to affect the development of language and mathematics. Learning to read is one of the biggest challenges children with HL face in school (Geers & Hayes, 2011; Trybus & Karchmer, 1977; Worsfold et al., 2010). Previous studies found that children with HL in general hover between a third or fourth-grade reading level (Qi & Mitchell, 2012) or that roughly 4% of deaf students within special education read on an age-appropriate level (Wauters et al., 2006). Further on in their school career, "learning to read" moves to "reading to learn" which gives children with

reading deficits even more challenges to obtain an educational degree (Walter & Dirmyer, 2013). Factors such as aided audibility, the degree of HL, age at identification and age at cochlear implantation, which influence language development in children with HL have also been found to affect reading development (Archbold et al., 2008; McCann et al., 2009; McCreery et al., 2015; Moeller et al., 2007, 2015).

Children with HL seem to score lower in mathematical assignments (Gottardis et al., 2011; Nagle et al., 2016; Sarant et al., 2015; Swanwick et al., 2005; Traxler, 2000), although this has been less often subject of research compared to language and reading. Difficulties are found in number comparisons, calculation, counting, number facts, numeral language, mathematical concepts, measurement, story problems, multiplication, and fractions (Ansell & Pagliaro, 2006; Frostad & Ahlberg, 1999; Kritzer, 2009; Leybaert & Van Cutsem, 2002). Mathematics often requires reading comprehension and understanding of specific linguistic math terms such as conditionals, comparatives, and inferentials (Traxler, 2000). Hence, mathematical achievement tends to co-vary with reading ability (Edwards et al., 2013; Mukari et al., 2007). Solving mathematical exercises is therefore expected to be extra challenging for children with HL who have reading and language difficulties. Previous studies did not find differences between CI and HA-users in their mathematical achievement (Bull et al., 2018; Marschark et al., 2015), which might result from the heterogeneity within the HL population (Convertino et al., 2009) and the sample sizes (Bull et al., 2018). Current knowledge of the degree of mathematical skills in children with HAs and CIs is still limited and further research is required.

Secondary education and individuals with HL

To the best of our knowledge, no research to date has yet examined which type of secondary education adolescents with HL attend. It is known that college students with HL have a higher risk of obtaining lower educational attainment compared to their hearing peers (Dammeyer & Marschark, 2016; Illg et al., 2017; Nagle et al., 2016; Rydberg et al., 2009; Winn, 2007). It is even estimated that only about 40% of the pupils with HL obtain their secondary school diploma (Idstad & Engdahl, 2019; Powers, 2003; Teasdale & Sorensen, 2007). Lower educational attainment might eventually lead to a higher chance of unemployment later in life (Dammeyer et al., 2019; Winn, 2007).

To date, all available conclusions about the educational achievements of individuals with HL are mostly based on small cross-sectional studies with a high probability of selection bias. Some studies were conducted on larger samples, but these large-scale studies

are weighted heavily toward either profoundly deaf students in special settings (Illg et al., 2017; Qi & Mitchell, 2012) or college students with mild/moderate HL in mainstream settings (Dammeyer et al., 2019; Hendar & O'Neill, 2016; Idstad & Engdahl, 2019; Teasdale & Sorensen, 2007). A longitudinal nationwide large-scale study that covers the whole population of children with HL from primary school years to adolescence is still lacking.

Educational system in the Netherlands

In the Netherlands, education starts at age 4 and includes 8 years in primary education. Schooling is compulsory from the age of 5 to 16 years. Most Dutch children with HL start with specialized preschool treatment groups to support their language and communication skills. Thereafter, parents and professionals decide whether the child will benefit from special education (for the deaf or hard of hearing) or they can keep up with mainstream education. This decision is often based on the level of language, communication, and social skills of the child. There is little empirical evidence which of these two types of educational settings would enable individuals with HL best to reach their full potential (Shaver et al., 2014; Stinson & Kluwin, 2012).

Dutch children are obliged to complete a final test in their last year of primary mainstream education (e.g., Cito, IEP, Route 8) (Lubbe, 2007). It covers compulsory subjects such as language (e.g., reading) and mathematics, but also includes geography, history, and subjects about nature. The standard score of this test estimates the type of secondary education that the child could potentially obtain. Unlike other countries such as the United States and France, secondary education in the Netherlands is uniquely divided into four types from the first year onwards. Based on the outcomes of this final test, Dutch children are divided in either low or intermediate prevocational, general secondary, or preuniversity education (Hakkenes & de Wijs, 2012). This Dutch system (with varied types of secondary education) aims to focus on the potential an individual has for attending and successfully accomplishing secondary education (Dutch Ministry of Education Culture and Science, 2006).

Present study

The main aim of this longitudinal nationwide study was to unravel the school career of children with HL in different educational settings in a large population. First, the distribution of children with HL in either special or mainstream primary education was studied. Second, children with and without HL were compared on their grades for language and mathematics obtained in primary mainstream education based on the outcomes of

a national standardized test (Cito). Moreover, the impact of switching from special to mainstream education on the school career was examined by dividing the mainstream group into children who always attended mainstream education and children who switched to mainstream education. Third, the type of secondary education of adolescents with HL was examined and compared to their typical hearing (TH) peers, taking their primary educational settings (i.e., mainstream, special, and switched from special to mainstream education) into account. Figure 1 illustrates an overview of the research questions. Due to small sample sizes within the HL population, previous research could not identify if the use of either HAs or CIs was related to differences in academic achievement or in the type of education (Bull et al., 2018; Convertino et al., 2009). This study was based on data from the governmental authority Statistics Netherlands which enabled us to design a large-scale longitudinal and nationwide study, and to compare individuals with either HAs or CIs to the Dutch hearing population. A longitudinal follow-up through different school years allowed us to monitor these children with HL from primary to secondary education. An intrinsic limitation of using this kind of nationwide generic collected data is that other HL-related background information is lacking, such as the degree and etiology of HL or the age at detection of HL and intervention.

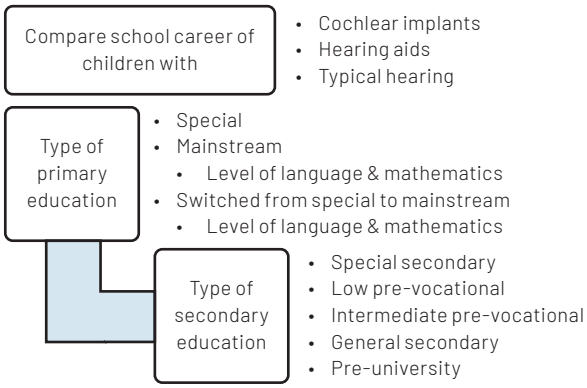

FIGURE 1. Research model of this study

MATERIALS & METHODS

Study design

This longitudinal retrospective study was designed using existing nonpublic microdata from Statistics Netherlands (www.cbs.nl). This third trusted party has nationwide data available with strict regulations on privacy and data anonymity. For example, the day of birth was censored and data that involved 10 individuals or fewer was not to be disclosed. Consequently, some categories needed to be combined to ensure the privacy of small groups. Under these conditions, microdata are accessible for statistical and scientific research without additional approval of an ethics committee. For further information: microdata@cbs.nl. We selected all children who were born between 1995 and 2013. The billed medical care, which is based on the combination of diagnosis and treatment, was available between 2013 and 2017 and was used to define if a child was using a CI or HA. These children received medical care in a hospital at least once during the period from 2013 to 2017. This is because in the Netherlands, children with CIs receive follow-up examinations every 6 months until the age of 11 years, thereafter follow-up changes to once a year. Children with permanent HL and HAs receive follow-up examination every year until the age of 11 years, and afterwards changes to once every 2 years. By using these data, we recognized that we were unable to identify the children with a mild or profound HL who did not use HAs or CIs (estimation of .7% missing that has sensorineural HL). Each child living in the Netherlands is obliged to follow education from the age of 5 to 16 years. Every Dutch child should therefore be enrolled in one of the Dutch schools. Data on the type of primary education that each child was attending were used to track whether they attended mainstream or special schools during the school years 2008 till 2018. Standardized academic achievement scores were available for the school years 2006 to 2018. Type of secondary education was available for the school years 2007 till 2018. Demographic data on every child, including sex, ethnicity, parental educational attainment, and household income were also collected.

Study population

The study population consisted of 4,087,877 children, of which 1,283 used CIs and 9,677 used HAs. These numbers corresponded with the national registry of pediatric cochlear implantations in the Netherlands (https://www.opciweb.nl/ci-centra/ci-centra-in-nederland/aantal-implantaties-in-nederland-t-m-2017/). Children with HL were separated into three groups by type of educational setting during their primary school years: children who only attended mainstream schools, children who (eventually) attended special schools, and children who had switched from special to mainstream schools (characteristics in Table 1). Some children were not registered in public primary schools which resulted in 710,681 (17%) children having missing data regarding their educational settings (due to their age, additional disabilities, emigration, or other unknown reasons). There were significantly more girls in the group with HL who continually attended mainstream schools compared to the group of children with HL in special schools. This female preponderance was also apparent when we compared children with typical hearing (TH) in mainstream and special education.

Additional nonauditory disabilities were most prevalent within children with HAs who attended special education compared to all other groups (most of them had down syndrome [8%] and/or behavioral problems [3.6%]). Children who (eventually) attended mainstream schools were more often autochthonous (native Dutch), had parents with significantly higher educational attainment and a higher household income compared to children who attended special schools. This difference was found in both groups with and without HL.

TABLE 1. Characteristics of children with typical hearing (TH), with cochlear implants (CIs), and with hearing aids (HAs)

		TH	CI			HA		
			Mainstream	Switched	Special	Mainstream	Switched	Special
		(a)	(b)	(c)	(d)	(e)	(f)	(g)
		n=3,165,074	*n*=259	*n*=242	*n*=692	*n*=5,830	*n*=316	*n*=2,728
Sex *n* %	Boys	**1,585,542** **50,1%**[e]	114 44,0%	127 52,5%	**366** **52,9%**[b,e]	2,748 47,1%	**183** **57,9%**[a,b,e]	**1,565** **57,4%**[a,b,d,e]
	Girls	**1,579,532** **49,9%**[f,g]	**145** **56,0%**[d,f,g]	**115** **47,5%**[d,f,g]	**326** **47,1%**[g]	**3,082** **52,9%**[a,d,f,g]	133 42,1%	1,163 42,6%
Additional nonauditory disability *n* %	No	**3,105,633** **98,1%**[d,e,f,g]	**249** **96,1%**[g]	**232** **95,9%**[g]	**664** **96,0%**[g]	**5,626** **96,5%**[g]	**300** **94,9%**[g]	2,378 87,2%
	Yes	59,441 1,9%	10 3,9%	10 4,1%	**28** **4,0%**[a]	**204** **3,5%**[a]	**16** **5,1%**[a]	**350** **12,8%**[a,b,c,d,e,f]
Highest education of mother *n* %	Missing	**979,809** **31,0%**[c]	**85** **32,8%**[c]	50 20,7%	**211** **30,5%**[c]	**1,788** **30,7%**[c]	**97** **30,7%**[c]	**844** **30,9%**[c]
	Primary or secondary education	**487,106** **15,4%**[b,c,e]	26 10,0%	23 9,5%	**201** **29,1%**[a,b,c,e,f,g]	784 13,4%	**52** **16,5%**[b,c]	**677** **24,8%**[a,b,c,e,f]
	Vocational education	**672,424** **21,2%**[e]	54 20,9%	56 23,1%	142 20,5%	1,123 19,3%	62 19,6%	**675** **24,7%**[a,d,e,f]
	Polytechnics or University	**1,025,735** **32,4%**[d,g]	**94** **36,3%**[d,g]	**113** **46,7%**[a,b,d,e,f,g]	138 19,9%	**2,135** **36,6%**[a,d,g]	**105** **33,2%**[d,g]	532 19,5%

TABLE 1. Continued

		TH	CI			HA		
			Mainstream	Switched	Special	Mainstream	Switched	Special
		(a)	(b)	(c)	(d)	(e)	(f)	(g)
		n=3,165,074	*n*=259	*n*=242	*n*=692	*n*=5,830	*n*=316	*n*=2,728
Gross income of household *n* %	Unknown	3,175 0,1%	1 0,4%	1 0,4%	2 0,3%	7 0,1%	1 0,3%	3 0,1%
	€0-15,000	**726,471** **23,0%** [e,f]	53 20,5%	44 18,2%	**199** **28,8%** [a,b,c,e,f,g]	1,166 20,0%	54 17,1%	**681** **25,0%** [a,c,e,f]
	€15-45,000	**317,828** **10,0%** [b,e]	15 5,8%	**30** **12,4%** [b]	**119** **17,2%** [a,b,e,f]	519 8,9%	**35** **11,1%** [b]	**426** **15,6%** [a,b,e,f]
	€45-60,000	280,305 8,9%	20 7,7%	28 11,6%	**78** **11,3%** [a,e]	512 8,8%	35 11,1%	**309** **11,3%** [a,e]
	>€60,000	**1,837,295** **58,0%** [d,g]	**170** **65,6%** [a,d,g]	**139** **57,4%** [d,g]	294 42,5%	**3,626** **62,2%** [a,d,g]	**191** **60,4%** [d,g]	**1,309** **48,0%** [d]
Ethnic origin *n* %	non-native	**626,474** **19,8%** [b,c,e]	29 11,2%	26 10,7%	**236** **34,1%** [a,b,c,e,f,g]	778 13,3%	**57** **18,0%** [b,c,e]	**713** **26,1%** [a,b,c,e,f]
	Native Dutch	**2,538,600** **80,2%** [d,g]	**230** **88,8%** [a,d,f,g]	**216** **89,3%** [a,d,f,g]	456 65,9%	**5,052** **86,7%** [a,d,f,g]	**259** **82,0%** [d,g]	**2,015** **73,9%** [d]

Bold=significantly larger proportion than the category mentioned in superscript (p<.001).

Standardized academic achievement test at the end of primary mainstream education

The national standardized academic achievement test (developed by the National Board of Tests and Examinations and the Central Institute for Test Development [Cito]) was used to indicate the performance level of children at the end of primary education (https://www.cito.com/) (Lubbe, 2007). This exam, often referred to as Cito or the central final test, is conducted by roughly two-thirds of the schools in the Netherlands (schools decide which test they use [e.g., Cito, IEP, route 8]). It indirectly indicates intelligence, motivation, concentration, and drives to learn and has a well-documented reliability (Hakkenes & de Wijs, 2012; Lek, 2020). The test consists of multiple-choice questions covering the obligatory subjects language and mathematics (world orientation such as geography, history, and nature are not mandatory). Questions covering language involve reading comprehension, summarizing, writing skills, and language cultivation (spelling, grammar, and vocabulary). Mathematical tasks cover measurements, geometry, time, money, fractions, and ratios. These are regular questions that require reading comprehension. Dutch schools are obliged to provide test accommodations (e.g., extra examination time, extra support with sign language, pictures, or assistive listening devices) for students with extra needs such as dyslexia or hearing problems. The raw scores were converted to percentile scores for further analyses.

Types of secondary education

Secondary education in the Netherlands starts after primary education at around the age of 12 years and is compulsory until the age of 16. It ranges from 4 to 6 years depending on the type of education. When entering secondary education, pupils are divided into one of the four different types of education: low prevocational (basic and general occupation-oriented education or in Dutch VMBO-basis/praktijk), intermediate prevocational (combination of general and theoretical occupation-oriented education or VMBO-gemengd/theoretisch), general secondary (HAVO), and preuniversity (VWO). Each stream demands increasing intellectual and scholastic abilities (Hakkenes & de Wijs, 2012). Pupils can switch upward or downward between the types of secondary education depending on their academic achievement. After secondary education, pupils can attend further optional higher education: vocational education for graduates of low or intermediate prevocational education, polytechnics for graduates of general secondary education, or university for graduates of preuniversity education (www.epnuffic.nl). Special secondary education in the Netherlands provides education that is mainly focused on acquiring skills for the labor market or finding daytime activities with the opportunity (not mandatory) for acquiring

an educational degree (often low prevocational education) (Dutch Ministry of Education Culture and Science, 2014). The schools thus can provide adjustments to the curriculum based on the developmental capabilities and educational needs of each pupil.

Statistical analysis

We used all nonpublic microdata from Statistics Netherlands that was available in February 2020. The date of birth was censored, and therefore age was calculated based on the year of birth. Children with HL were divided into three groups based on their type of educational setting during primary school years (2008–2018): children who only attended mainstream schools, children who (eventually) attended special schools, and children who had switched from special to mainstream schools. To examine a potential increase in the proportion of children with HL in mainstream schools, the different school years (2008–2018) were compared with dependent samples t-tests. Descriptive statistics were used for the baseline characteristics between groups. To compare the standardized test scores of children with CIs and HAs to children with TH in mainstream education, one-way ANOVA and independent samples t-tests were performed for the percentile scores of language and mathematics. Furthermore, a chi-square ($\chi 2$) test was carried out to examine the proportion of pupils attending each type of secondary education and whether the distributions differed between the different groups. Due to the privacy regulations of Statistics Netherlands some categories have been combined to ensure the privacy of small groups (e.g., merging general secondary and preuniversity education). Statistical analyses were performed with IBM SPSS Statistics 23.0 software package.

RESULTS

Distribution in primary education

The type of education during primary school years is shown in Figure 2 for children with TH (n=3,165,074), CIs (n=1,193), and HAs (n=8,874). Considering all primary school years together, 61% of children with HL (eventually) attended mainstream schools and 31% special schools. Sixty-four percent and 28% of the HA-users and 39% and 54% of the CI-users attended mainstream and special schools, respectively. The type of primary educational setting of the remaining 8% (CI n=90 and HA n=803) of children with HL was unknown or they were already in secondary education. Compared to the TH population, fewer children with CIs and HAs attended mainstream schools (p<.05). Within the HL population, children with HAs attended mainstream education more frequently compared to children with CIs p<.05).

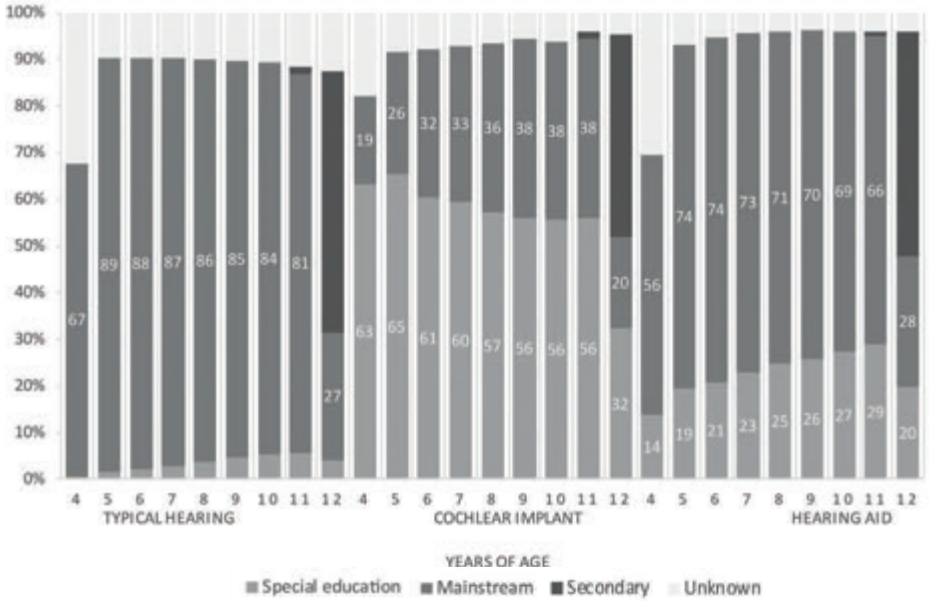

FIGURE 2. Distribution of children within primary educational settings in percentages

At age 5, 65% of the children with CIs were in special schools and 26% in mainstream schools. Eventually at age 11, respectively 56% and 38% of the children with CIs attended special and mainstream schools. This means that the proportion of children with CIs in mainstream schools significantly increased from 5 to 11 years of age (p<.05). Within the total group of CI-users, 20% of the children stayed in mainstream education from the start, 19% switched from special to mainstream education, 50% stayed in special education, and 4% switched from mainstream to special education.

The number of children with HAs in mainstream schools significantly decreased from 5 to 11 years of age (p<.05). At age 5, 19% of the children with HAs were in special schools and 74% were in mainstream schools. At age 11, respectively 29% and 66% of the children with HAs attended special and mainstream schools. Within the total group of HA-users, 8% of the children switched from mainstream to special education, 20% stayed in special education, 3% of the children with HAs were able to switch from special to mainstream education, and 60% stayed in mainstream education from the start.

Subsequently, the proportion of children with HL in mainstream settings per schoolyear was examined over time (2008 to 2018). Only the percentage of children with CIs in mainstream settings considerably increased over time (Figure in supplements). There was a clear distinction, however not statistically different (p>.05), between children with CIs who were born before and in 2005 and onwards: 6-year-old children attended mainstream education more often from the schoolyear 2011 than before and 10-year-old children attended less often mainstream education before the schoolyear 2015 than after 2015 and onwards. Thus, children with CIs born from 2005 and onwards appeared to attend mainstream schools more often compared to the ones born before 2005.

Language and mathematics at the end of primary mainstream education

In line with the fact that two-thirds of the schools use Cito in the Netherlands (Lubbe, 2007), we found that around two-thirds of the mainstreamed children in the data (who were old enough) completed this standardized test (70% [n=1,345,287] of the children with TH, respectively 63% [n=74] and 70% [n=120] of the children with CIs who switched to or continuously attended mainstream education, and respectively 59% [n=110] and 67% [n=2,543] of the children with HAs who switched to or continuously attended mainstream education). Only a negligible number of children in special schools took the standardized test, which impeded us from examining their language and mathematics scores. The average score of language and mathematics was higher for children with TH compared to children with HAs and CIs (Figure 3). Yet, children with CIs who switched from special to mainstream schools had comparable levels of language and mathematics as their hearing peers. Among those who continuously attended mainstream education, children with HAs outperformed children with CIs with respect to language, but not with respect to mathematics.

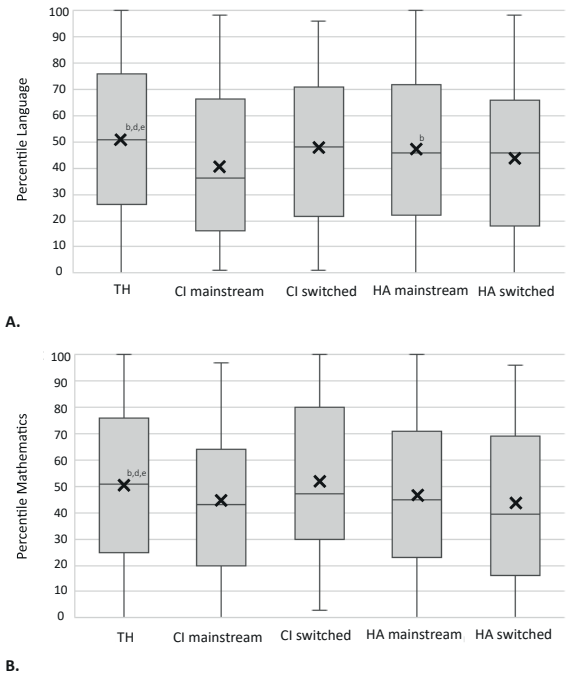

FIGURE 3. Boxplots of the level of language **(A)** and mathematics **(B)** in percentile scores of children with typical hearing (TH), cochlear implants (CI), and hearing aids (HA). The cross represents the mean and the horizontal line the median of each group. For each significant pair, the key of the smaller category appears in the category with a larger mean (p<.05).

In Table 2 the children are divided in quartiles to indicate if their percentile scores fell into a below-average (25th percentile and below), average (25 - 75th percentile), or above average (75th percentile and above) group. Approximately 60 - 70% of the children with HL performed average or above average on language and mathematics. Regarding language specifically, more mainstreamed children with CIs and HAs performed below average compared to children with TH (p<.05). Concerning mathematics, more children with HAs performed below average compared to children with TH and children with CIs who switched to mainstream education (p<.05). Strikingly, more children with CIs who switched to mainstream education performed above average on mathematics compared to their peers with CIs and HAs who always attended mainstream education. In addition, post hoc analysis showed a positive correlation between language and mathematics (r=.501-.682, p<.001) in all groups.

TABLE 2. Percentage of children who performed on average, below, or above average on language and mathematics

	TH	CI		HA	
	(A) *n*=1,345,287	Mainstream (B) *n*=120	Switched (C) *n*=74	Mainstream (D) *n*=2,543	Switched (E) *n*=110
Language					
Below (0-25th)	25,0%	**37,7%[a]**	32,1%	**29,4%[a]**	33,3%
Average (25-75th)	49,3%	43,0%	50,0%	49,0%	51,7%
Above (75-100th)	**25,7%[.d,e]**	19,3%	17,9%	21,6%	14,9%
Mathematics					
Below (0-25th)	25,2%	29,2%	21,6%	**28,9%[a]**	**37,3%[a,c]**
Average (25-75th)	48,9%	53,3%	47,3%	49,9%	41,8%
Above (75-100th)	**25,9%[b,d]**	17,5%	**31,1%[b,d]**	21,2%	20,9%

Bold=significantly larger proportion than the category mentioned in superscript (p<.05). TH=typical hearing; CI=cochlear implants; HA: hearing aids

Type of secondary education

The different types of secondary education was examined between adolescents with TH (n=1,130,777), CIs (mainstream n=83; switched n=89; special n=263), or HAs (mainstream n=2,392; switched n=136; special n=1,092) who were 13 to 18 years of age between the school years 2007-2018 (Figure 4). Adolescents with CIs and HAs, who finished their primary education in mainstream schools, followed roughly the same distribution in secondary education as their hearing peers. However, adolescents with CIs attended low prevocational education (Figure 4.B) and unspecified secondary education (level of secondary education not yet determined)(Figure 4.C) significantly more often compared to TH peers (p<.05). There were also significantly more adolescents with HAs in special, low prevocational, or unspecified secondary education compared to their hearing peers (p<.05; Figure 4E and F). On the contrary, adolescents with TH attended more often general secondary or preuniversity education than adolescents with HL (both children with CIs and HAs; p<.05). Post hoc analysis showed a positive correlation between language scores, mathematical scores and the type of secondary education (language r=.309-.623, p<.021-.001; mathematics r=.309 - .523, p<.07-.001). This indicated that pupils who obtained higher

levels of language and mathematics at the end of mainstream primary education attended types of secondary education with higher intellectual challenge.

After primary education in special schools, adolescents with CIs and HAs attended special secondary education significantly more often and intermediate prevocational, general secondary, or preuniversity significantly less often compared to all other groups (Figure 4D and G). Regarding low prevocational education, more HA-users attended this educational level compared to hearing pupils, CI-users who switched to mainstream education and who stayed in special education, and HA-users who stayed in mainstream education (Figure 4G).

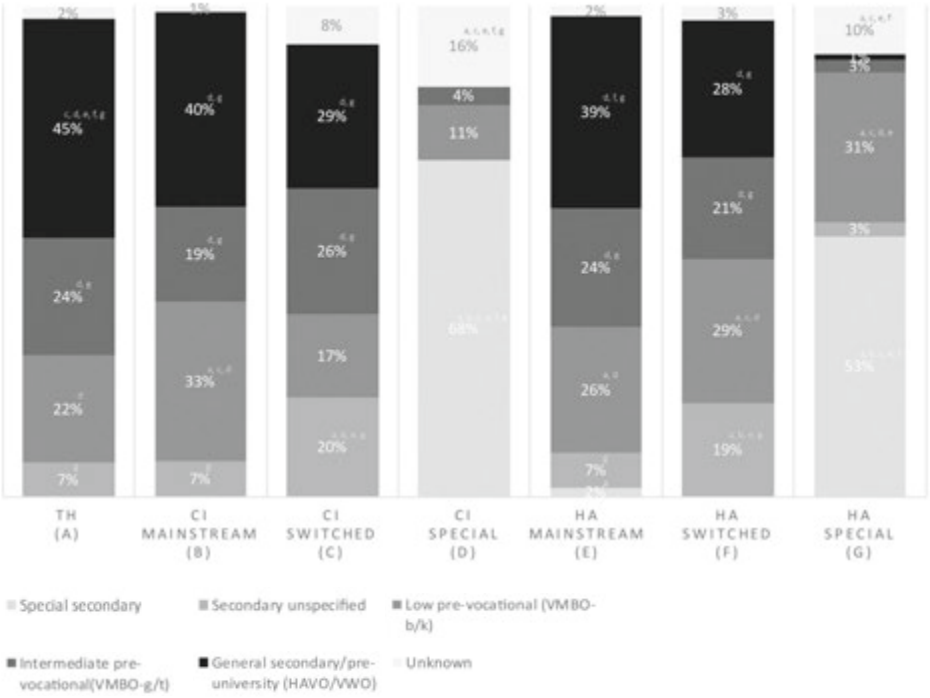

FIGURE 4. Distribution of the type of secondary education within adolescents (age 13 to 18 years) with typical hearing (TH; n=1,130,777), cochlear implants (CI; respectively n=83, 89, 263), or with hearing aids (HA; respectively n=2,392, 136, 1,092) who attended mainstream primary education only, who switched from special to mainstream primary education, and who attended special primary education. For each significant pair, the key of the category with the smaller column proportion appears in the category with the larger column proportion ($p<.05$).

DISCUSSION

This longitudinal retrospective study is to our knowledge the first to compare the school career of a nationwide cohort with HL and their hearing peers from school-age to adolescence. Overall, 61% of the children with HL attended mainstream and 31% special primary education. However, we found that more HA-users attended mainstream education compared to CI-users (64% versus 39%). The majority of children with HL who attended mainstream education reached average or above average levels of language and mathematics similar to their hearing peers. After continuously attending mainstream education, children with HAs outperformed children with CIs regarding language. Yet, children with CIs who switched from special to mainstream primary education achieved comparable levels of language and mathematics as their hearing peers, though children with HAs were unable to reach that score.

This difference between children with CIs and HAs disappeared during secondary education. Adolescents with HL who (eventually) attended mainstream primary education went to similar types of secondary education compared to their hearing peers. Only low prevocational, unspecified, or special secondary education were more often attended, and general secondary or preuniversity education were less often attended by adolescents with HL. Most notably, individuals with HL who have been only in special primary education attended lower levels of secondary education than their mainstreamed peers with and without HL. Many of the adolescents from special primary schools continued their school careers in specialized education. This study, therefore, revealed that not all children with HL, but mainly the children who finished their primary education in special settings are expected to obtain a lower educational achievement after graduating from their secondary school.

Mainstream primary education and standardized achievement outcomes

In line with the literature where children with HL tend to underachieve on scholastic examinations (Edwards et al., 2013; Geers & Hayes, 2011; Marschark et al., 2015; Mukari et al., 2007; Pagliaro & Kritzer, 2013; Spencer PE, 2010; Trybus & Karchmer, 1977), this study also found lower mean levels of language and mathematics in children with HAs and CIs compared to hearing children. However, a majority of children with HL performed above or on average when we evaluated their language and mathematical scores based on the percentile quartile they were in. The reason for this inconsistency might be twofold. First, this study was able to omit selection bias by using a standardized test in

a nationwide population with HL who attended mainstream and not special education. Second, the division of below-, on-, and above-average might be better for evaluating this heterogeneous group of mainstreamed children with HL whose scores covered a broad spectrum. It is likely that the children within the below-average group compromised the average score of the complete group with HL. Moreover, post hoc analyses showed a positive correlation between language and mathematics. This indicates that children who had lower levels of language also underperformed on mathematical tasks and vice versa. This might support the fact that mathematical tasks require an understanding of specific linguistic terms or reading comprehension (Edwards et al., 2013; Mukari et al., 2007).

Thus, this study found that a majority of children with HL in mainstream primary education could keep up with their hearing peers as they showed comparable academic achievements. These are promising results for the growing population of children with HL in mainstream settings. The increase of children with CIs in mainstream settings over time might be a result of early detection of congenital HL, which enabled early awareness and rehabilitation through family-centered early intervention. The newborn hearing screening was completely implemented in the Netherlands in 2005 (Korver et al., 2013), which ensures early development of language and communication (Yoshinaga-Itano, 2004). It is expected that early detection of HL and intervention will continue to enable children with HL to transfer to mainstream education and obtain average educational achievements.

Furthermore, this study corroborated that the educational chances in secondary education of children with an auditory disability are good as long as they can attend mainstream education, even with the different conditions of a secondary school in mind. Acoustics, listening effort, social-emotional inclusion, or the time-frame at which their (HL) identity is developed are key-factors that children with HL have to deal with besides attaining adequate educational achievements in secondary education (Brice & Strauss, 2016; Israelite et al., 2002; Kent & Smith, 2006; Rich et al., 2013; Spencer et al., 2012).

Switching from special to mainstream primary education

After switching from special to mainstream primary education, children with CIs were able to achieve similar levels of language and mathematics as their hearing peers. Possibly, by attending special education in the early years prepared these children to keep up with the curriculum in mainstream schools. Alternatively, these CI-users might have been assigned to special education while in hindsight they would have had even better opportunities to reach their full potential if their initial placement would have been in mainstream education. However, this did not apply to children with HAs. Instead, children with HAs who made the step from special to mainstream primary education lagged behind on language and mathematics compared to their peers without HL. This could be related to the fact that children with HAs usually switch at an earlier age compared to children with CIs. Children with HAs tend to speak relatively well and often have successful interactions with others which might leave their difficulties unnoticed (Tomblin et al., 2015). Yet, the discrepancy between children with CIs and HAs was not maintained in secondary education. This finding may encourage parents and teachers of children with CIs to consider the transition to mainstream education despite their slight delay in language and mathematics as they will eventually attend similar types of secondary education. Thus, not the level of academic performance but the primary educational setting of children with HL was related to the educational achievement later in life.

Special primary education

In total, 54% of all children with CIs and 28% of the children with HAs attended special primary education and did not switch to mainstream education. A subset of these children could have attended some hours or days in mainstream education which is common practice in the Netherlands. However, we were not able to distinguish the amount of participation in our dataset, but only identified in which educational setting each child was registered. This might explain our lower epidemiological numbers compared to the United States and Australia (of which 78% and 85% of the children with HL attend mainstream schools) (Punch & Hyde, 2010; Shaver et al., 2014). It would be interesting to further investigate the reasons for such a large group of children with HL (more specifically children with CIs) staying in special education, including the reasons other than their level of (spoken) language and communication skills. Multiple factors, such as sign language, the severity of HL, additional handicaps (despite low percentages), ethnicity, and so on, have possibly contributed to the fact that these children with HL were assigned to specialized education with more individual support (Israelite et al., 2002; Karchmer et al., 1982; Knoors & Vervloed, 2012; Rydberg et al., 2009; Shaver et al., 2014). It is also possible that a switch to

mainstream education was withheld by parents and/or teachers. As a result, these children with HL continued in specialized settings during their adolescence and attained a lower educational achievement.

Strengths and limitations

No studies to date have examined the level of mathematical skills in such a large group of children with HL. Besides, this study is the first to examine the educational attainment during adolescence as most previous studies were conducted in college years (Dammeyer & Marschark, 2016; Marschark et al., 2015). The longitudinal format made it also possible to delve into the impact of primary educational settings on the educational achievement of adolescents with HL. With the available data of Statistics Netherlands, we could examine a large sample of children with CIs and HAs in the Netherlands without selection biases. However, a limitation of using the national data of Statistics Netherlands was a lack of additional background information, such as the type and degree of HL, the age at detection of HL and at intervention, the reason for educational placement, and the level of support children with HL receive at their school. The standardized achievement test was used in two-third of the mainstream schools, implying that we had missing data of one-third of the total mainstreamed population with HL in the Netherlands. This was however not a consequence of a selection bias, but a decision made within the mainstream schools whether or not they used Cito or other standardized tests. Additionally, a lack of standardized achievement tests in special education prevented us from investigating the levels of language and mathematics of children with HL in specialized settings. This will change as the government of the Netherlands has recently made standardized tests in special primary education obligatory. Adding the grades of these children in special education to the mainstreamed population with HL will most likely decrease the overall mean levels of language and mathematics of the group with HL. Furthermore, the choice for educational setting does not only depend on the academic performance. Some adolescents might lack great interest in obtaining high educational achievements, but would rather have peers that are similar to them. Future studies should therefore consider the social perspectives of each educational setting and include children from more recent cohorts with CIs and HAs that have benefitted from the ongoing innovations in the field of hearing technology and interventions.

Conclusions

The majority of children with HL were able to keep up with mainstream education and attended similar types of secondary education as their hearing peers. In these mainstream settings, children with CIs did not differ or performed better on academic achievements compared to children with HAs. After finishing primary education in special settings, children with HL attended more often special secondary education than their mainstreamed peers with and without HL. On the basis of these findings, extra guidance and precautions should be made in special education to inform caregivers and teachers about future perspectives. This enables shared decision making regarding the best educational setting for children with HL in order to reach their full potential. Moreover, mainstream schools, with the additional support from the Dutch government, need to be more inclusive for children with HL, especially for the ones with CIs.

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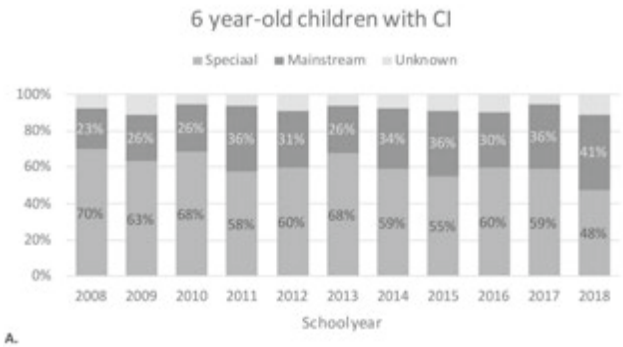

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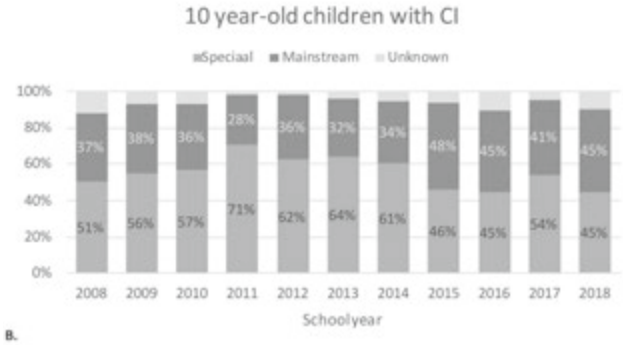

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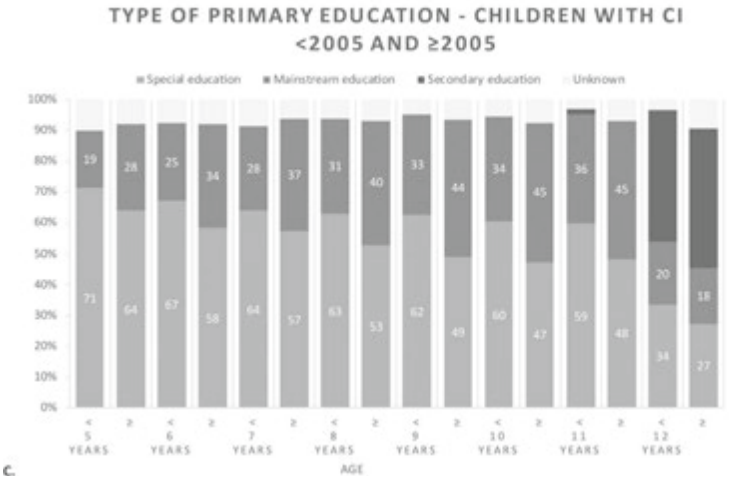

C.

SUPPLEMENTARY FIGURE. Type of primary education of children with cochlear implants (CI) examined over time (2008 to 2018). The distribution is shown at the age of 6 **(A)** and 10 **(B)** years. There was a distinct shift in children with CI attending mainstream settings who were born before (<) and in 2005 and onwards (≥) **(C)**. The number of children with CIs ranged from 147 to 646 per age category as it depended on the year of birth and the available school years.

Main Outcomes and General Discussion

The aim of this thesis was to examine the impact and the growing expectations of audiological rehabilitation for patients with hearing loss (HL). Currently, clinicians are confronted with patients' great expectations for the technically advanced devices on the market. Although individuals with HL are likely to experience benefits after receiving a hearing aid (HA), cochlear implant (CI), or auditory brainstem implant (ABI), challenges with hearing still arise. In this discussion we aim to look beyond the standard of care. We will elaborate on future possibilities and obstacles for children and adults with HL in a world driven by sound and verbal communication. We will provide a critical point of view on the selection criteria for implantation and the increasing overlap between candidates for HAs or CIs. The optimal assessment method is discussed in order to quantify obstacles for patients with HL, especially when in doubt which rehabilitation method to choose. The expectations of social-emotional well-being and educational attainment of children with HL is reviewed in order to debate the areas for improvement in pediatric rehabilitation. These findings will help us answer the questions that adults with HL and parents of children with HL ask when faced with the decision for rehabilitation.

MAIN OUTCOMES OF THIS THESIS

Post-lingually deafened candidates for cochlear implantation

The efficacy of different preoperative measures were compared in **Chapter 2** to define which measure optimally selects post-lingually deafened candidates for CI. This was investigated by using the binary outcome of improvement or no improvement of speech perception with CI and calculating the diagnostic value of each measurement with the sensitivity and inverted specificity. This study showed that the best-aided phoneme score in a quiet free-field setting had the highest diagnostic value to indicate which candidate would improve their speech perception in a quiet setting after implantation. In addition, the best-aided phoneme score in noise was the most accurate in predicting an improvement of speech perception in noise. The latter is particularly useful when deciding upon borderline candidates with high preoperative speech perception in quiet settings, but who function poorly in noise.

Additionally, the collected dataset allowed us to compare various criteria for implantation. More specifically, we investigated the recently implemented criteria of the United Kingdom and Flanders (northern part of Belgium) - **Chapter 3**. In both countries, the criteria of speech understanding became more dominant than the criterion defined by pure tone audiometry. The newly introduced criteria had a sensitivity of 68.4-69.4% (increase of 31%). Broadening the criteria increased the number of candidates by 30%. However, seven out of eight patients that were excluded by the new British and Flemish criteria were able to improve their speech understanding (>0%) if they would have received a CI in our center.

Pre-lingually deafened candidates for auditory brainstem implantation

Many studies have investigated the benefits of CIs and HAs in children with congenital HL. Yet, the alternatives and expectations of deaf children who are not eligible for CI have been less often topic of research. Therefore, the language development of children who received ABIs were evaluated in **Chapter 4** and compared with children who received CIs. Within one year, six out of seven children with ABIs could identify sounds, respond to speech, and use their voice to attract attention. Their development in language skills was slower compared to children with CIs, but on par with children with CIs who also had additional disabilities. A key issue in this comparison is that most children receiving an ABI have multiple disabilities. We concluded that ABIs can provide satisfactory auditory input for deaf children with severe bilateral inner ear malformations. However, having multiple severe additional disabilities negatively influences the ability to understand and

use speech which should be balanced against the surgical risks of brain surgery. With these findings, parents can be optimally informed regarding the expectations of ABIs before implantation.

Impact of childhood hearing loss on the social-emotional and educational development

When studying the effect of rehabilitation, we have the tendency to focus on criteria and function outcomes that can be measured (such as in **chapter 2, 3, and 4**). However, parents of children with HL are also interested in the long-term wellbeing, possible psychological problems that can arise after rehabilitation, or what can be expected of the future level of education of their child. Therefore, the quality of life (QoL) and the level of education of children with HL were studied in two longitudinal studies.

In **chapter 5** we have examined how the QoL of children with CIs and HAs changes over time. We saw that at age 4 and 11 years, children with HL had similar QoL regarding emotional and physical aspects as their hearing peers. However, social and school functioning were lower in children with HL who attended special education and who switched to mainstream education compared to their mainstreamed peers with and without HL. Wellbeing regarding school activities decreased over time, but this was apparent in both children with and without HL. We also found that children with CIs showed a decrease in social functioning when they were 11 compared to 4 years old. Based on these findings, we determined that specific guidance is needed regarding school activities and social functioning within hearing-impaired children who are in special education or who switch to mainstream education.

In **Chapter 6**, we compared school careers of children with CIs and HAs with typical hearing (TH) children in order to examine the extent to which children with HL are at risk of lower levels of education due to their deprived auditory input. We found that 39% of the children with CIs and 64% of the children with HAs were able to eventually attend mainstream education. Despite the fact that the total group with HL, on average, underperformed in language and mathematics of the standardized tests in primary education, more than 60% of these children achieved average or above-average scores. Moreover, the distribution within the level of secondary education after mainstream primary education was similar in the HL as the hearing population. Individuals with HL who attended special primary education obtained significantly lower levels of secondary education than their mainstreamed peers with and without HL. Thus, not all children with HL, but mainly the children who attend special education are expected to obtain lower educational attainment.

GENERAL DISCUSSION

The findings of the five studies included in this thesis show that rehabilitation greatly influences the lives of individuals with HL concerning their speech understanding, language development, social wellbeing, and school career. We would like to further delve into the perspective of patients with HL who are confronted with obstacles and future possibilities in the following sections.

Changing the guidelines in adult rehabilitation

There is a gradual increase of candidates eligible for implantation as discussed in **Chapter 3**. Only severely to profoundly deaf adults with no additional disabilities were eligible for CIs in the early years (Frijns et al., 2002). This expanded quickly to patients who functioned modestly with HAs and had residual hearing or had additional disabilities. The expansion of candidate criteria has been a result of improvements in implant types (Green et al., 2007), new speech coding strategies (de Jong, 2019; Guevara et al., 2016), new surgical techniques (Cullen et al., 2004), improved pre-implant scores (Snel-Bongers et al., 2018), and improved rehabilitation methods (A. Geers et al., 2008; Yoshinaga-Itano, 2004). Nowadays, adults with HAs who are able to function reasonably in quiet but not in noisy situations are also considered as candidates for CI in many places including Leiden, the Netherlands (Snel-Bongers et al., 2018).

The expansion of candidate criteria is well known in ear-, nose-, and throat (ENT) departments of academic centers where implantation take place. Yet, guidelines regarding when to refer patients with HL in peripheral health care centers vary broadly. The guideline for general practitioners (GP) in the Netherlands state that they can consider a referral to an audiological center when adults have severe HL of 70 dB or more (Nederlands Huisartsen Genootschap, 2014).

Currently, commercial hearing aid dispensers, who facilitate HAs for patients with HL in the Netherlands, often use pure tone audiometry to define the amount of HL in dB. Recently, hearing health aid dispensers started to use speech perception measures (in quiet) on a regular basis. They are, however, unaware of the new CI-criteria based on speech perception in quiet and noise. According to the study discussed in **Chapter 2**, speech perception tests in quiet and in noisy environments should be the golden standard to follow when HAs do not satisfy the patient's needs. Thus, peripheral hearing aid dispensers do not use the optimal diagnostic test to measure hearing-related difficulties. In addition, when

Adults with post-lingual HL usually wait 7 years before they start rehabilitation with HAs as they are not willing to acknowledge their HL, wear the visual aids or think that HAs only belong to the elderly (Chan et al., 2017; Rolfe & Gardner, 2016; Wallhagen, 2010).

the product (HA) is not meeting the patient's needs, professionals may first try to solve the issue. This can cause a delay in referring patients with HL to an audiological center and contribute to a longer duration of severe HL before implantation. Therefore, guidelines in the peripheral health care system (including GP's) that assist in decision making regarding the moment of referral or regarding the type of rehabilitation should be updated.

Academic ENT centers that screen candidates for CI may also benefit from a new approach. A new tendency is evolving to change pre- and post-operative screening from only objective measures to a combination of objective and subjective measures. This approach is already operational in England (National Institute for Health and Clinical Excellence, 2019). The needs of individuals with HL can be found by identifying a person's subjective obstacles (Ebrahimi-Madiseh et al., 2020; Pryce et al., 2016). QoL questionnaires, such as hearing handicap inventory for adults or elderly (Lazzarotto et al., 2016) or the impact of HL inventory tool (Stika & Hays, 2016), assist in measuring the subjective point of view of coping with HL. However, it remains difficult to capture the person's own viewpoint in these general questionnaires and the assessment should therefore be personalized towards an open interview (Ebrahimi-Madiseh et al., 2020). By asking open questions one can identify the challenging situations at home, at work, with hobbies, or during sports (Cox et al., 2000; National Acoustic Laboratories: A Division of Australian Hearing, n.d.). Individuals with HL may function poorly due to the fact that they miss crucial information in social situations and that they frequently need to ask to repeat what was said. This can result in loneliness or isolation as they avoid social appointments (Carlson, 2020). One could also encounter problems with hearing when listening to music. The associated degree of impairment depends on the individual's personal or professional interest in playing musical instruments, singing, or dancing. HL can also impair the ability to do sports where individuals are unable to use externally worn assistive tools, are unable to look at a person's face for lip-reading, or are unable to ask for clear pronunciation.

Interviews with patients provide an important contribution to the understanding of people living with HL and allow the screening of all aspects of daily life that may be impacted by HL (Stichting Protocol Hoorhulpmiddelen, 2018). Including a realistic, achievable, and personal goal assures a strong motivation for attaining a successful rehabilitation

Resistance towards CI is known in individuals with HL and peripheral hearing care (Ebrahimi-Madiseh et al., 2020). Examples of patients' concerns for CI are a time investment that is limited by work, social concerns regarding their HL, surgical risks and the sustainability of the implant, or the ability to participate in a hearing society with CI. Only 20-30% of the candidates actually receives a CI in Western countries nowadays (Deborah Vickers et al., 2016b).

(Ebrahimi-Madiseh et al., 2020; Goetz & Schork, 2018). This would imply a more qualitative approach instead of quantitative selection criteria for rehabilitation. Furthermore, it is essential to find a balance between providing enough medical and audiological information, with what one may expect, and being open to a person's value and concern regarding the rehabilitation trajectory (Ebrahimi-Madiseh et al., 2020). Shared decision making where speech perception measures and personalized interviews are combined has a great potential to be the future approach of adult rehabilitation, especially when criteria for CI and HA start to overlap.

This made us think about future studies that should be performed. The implementation of multidisciplinary consultation or usage of one shared database would enhance the collaboration between the periphery and implant centers. Hearing aid dispensers and GP's may easily consult professionals for the next step in rehabilitation when patients encounter HL-related obstacles. Efforts are already being made to start sharing knowledge between the peripheral hearing care system and professionals in academic centers by exchanging hearing questionnaires (Stichting Protocol Hoorhulpmiddelen, 2018). We should start with the collection of nationwide data from the different hearing health care centers and audiologic departments throughout the country. By joining forces, we could collect HL-specific information and refer the patient to the right clinic honoring the current privacy regulations. Big data analysis and the use of artificial intelligence can further examine the area for improvement in rehabilitation.

Selection criteria in pediatric rehabilitation

The question arises if we should apply more lenient criteria within pediatric rehabilitation. We have already seen a shift towards children with additional disabilities who benefitted from implantation (**Chapter 4**, (Anat Zaidman-Zait et al., 2015). Yet, selection criteria for children with HL differ in various ways making a comparison between adult and pediatric selection criteria almost impossible. Firstly, the amount of HL (severe to profound) measured with ABR or pure-tone audiometry is the only objective criterion within young pediatric candidates (Lovett et al., 2015). It is rather difficult to test the speech perception

of young children with residual hearing who are still learning a language. Secondly, children with HL can benefit from their HAs in their early years showing good responses to environmental sounds, but eventually might encounter delays in the development of language or speech. Nowadays, the decision for borderline candidates can be found in bimodal hearing where one would have the best of both worlds: one ear with a CI and one with a HA. In addition, opinions of older children with bimodal hearing who would rather use their CI than HA could be of additional value when deciding on a second CI. Thirdly, choosing between the types of pediatric rehabilitation methods is feasable for parents as they tend to choose the best option for their child. However, parents often have no experience with HL as 92% have normal hearing themselves (Mitchell & Karchmer, 2004). A lack of knowledge about the available rehabilitation trajectories contributes to the concerns of parents. They only pursue the best available rehabilitation method to ensure that their child can optimally participate in a society that is driven by verbal communication.

Tailoring expectations in pediatric rehabilitation

Parents of children with HL have multiple questions at the beginning of rehabilitation. They seek answers regarding the future possibilities and challenges that their child with HL will face in a verbally orientated society. It is important to provide accurate information regarding the consequences of HL. The questions most often asked were studied in **Chapter 4 to 6**.

It is important to sketch an outline of what one may expect concerning the development of language, including to what extent a child will understand its parents' spoken speech is important (**Chapter 4**). In general, children with CI and HA are able to communicate in spoken language with their parents and are able to participate in a society driven by verbal communication. Only children with additional disabilities often retain difficulties with spoken language. Particularly in ABI, parents need to be well informed in order to make a major decision for the future of their child. The advantages and disadvantages of ABI need to be balanced against the option to use only sign language, keeping the severity of additional disabilities, if present, in mind.

For every child with HL, additional communication methods should be discussed with parents. New communication methods include direct translators of spoken language to text, such as Google glasses or mobile applications, thereby keeping in mind that reading is one of the subjects that children with HL find challenging (Qi & Mitchell, 2012; Wauters et al., 2006). Introducing sign supported language in mainstream classrooms for

all children could be a valuable assistance in difficult listening conditions. For example, in the Netherlands the news on the television is also translated in sign language. This would require learning an additional language, a more visual skill instead of a verbal one. Using additional communication methods as a more mainstream instrument in everyday situations will support the concept of individuals with HL being part of a diverse society, where they can participate equally. The social model of disability proposes that what makes someone disabled is not their handicap itself, but the attitudes and structures of society (Goering, 2015). There should be no limits set on what individuals with HL can achieve. The key is to find the support which they need to achieve these things in order to participate fully in a society dominated by sound and verbal communication.

Another frequently asked question at the beginning of pediatric rehabilitation is whether a child with HL will be able to attend mainstream education. We found in **Chapter 6** that thirty-nine percent of children with CIs and sixty-four percent of children with HAs could attend mainstream primary education. These children can reach similar levels of education as their hearing peers. It is important to acknowledge to parents that the type of primary educational setting is related to later educational attainment for their children, as shown by the lower levels of secondary education achieved after special primary education. The demonstrated division in educational outcome shows that children with HL in mainstream education have better chances of achieving an educational degree. However, this does not imply that all children with HL belong in mainstream education as we need to look at the capacities of the child. Our findings could also be interpreted as evidence that the Dutch system of selection is working adequately. The two types of educational systems seem to cooperate and properly investigate which type of education suits best for each child with HL. Figure 1 shows a theoretical model with main determinants of academic achievement. The cognitive and motivational determinants are embedded in a complex system of individual, parental, and school-related determinants and depend on the given social, classroom, and cultural context (Helmke & Schrader, 2001). This scheme illustrates that there might be other problems, that we were unable to measure, that could add to the explanation why children with HL benefitted by each type of education (Shaver et al., 2014).

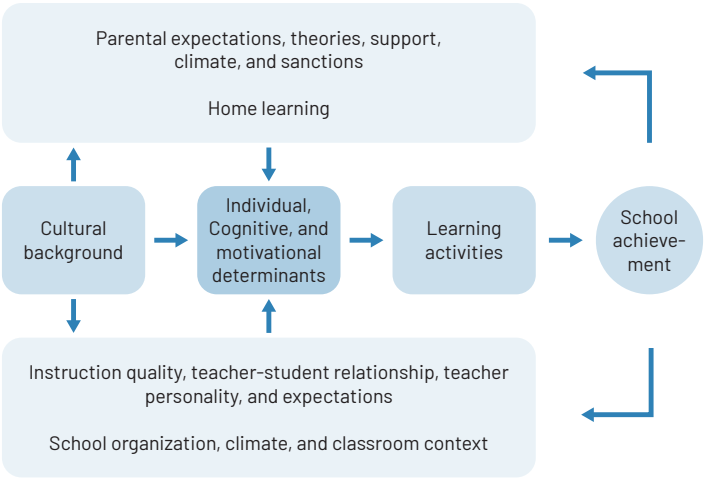

FIGURE 1. Theoretical model of academic achievement (Helmke & Schrader, 2001)

Another explanation of the lower levels of secondary education achieved after special primary education could be related to the reliance on sign language. It is known that children who are able to perform without sign language but are exposed to sign language on a daily basis are in some way reliant on the visual cues (Barca et al., 2013) and would miss important information if not provided via signs. This makes them vulnerable in a society predominantly driven by verbal communication. This situation is to some extent comparable to children following education in a foreign country where they have to learn a second language at a young age. However, sign supported language is less complicated than sign language as Dutch would be the primary language, of which it is supported by visual cues.

In order to catch up on school delays, the ideal solution could be to introduce extra time for children with HL in mainstream settings. This extra time might help children with HL to better adjust to mainstream education in a primary or secondary setting. For example, schools where children with additional disabilities are guided within separate classes for the first (couple of) years of secondary schools. Parents could look for these inclusive schools when changing from primary to secondary education. Just recently, the Dutch government proposed to implement a general secondary education for the first two to three years without the division between the levels of education. This gives children with HL more time to adjust, which eventually could enhance the chance of defining the right educational level for each child (also for adolescents without HL).

Special secondary schools primarily focus on providing practical tools in order to participate in society with a disability and secondarily focus on acquiring a level of education (Dutch Ministry of Education Culture and Science, 2014).

Regarding social functioning, parents are often unaware of the future challenges of their child with HL. They often assume that their child is able to fully participate in society after rehabilitation, including acquiring close relationships with friends, family, or a spouse. The expectation of a child's social functioning right after implantation found in **Chapter 5** underpins the expectation of a 'normal' development. Therefore, providing information regarding the possibility of challenges in their social functioning as early as possible to parents of children with HL is important in order to prevent too great expectations and setbacks, especially in children within special education. By exposing children with HL to hearing peers in mainstream settings, could encourage them to actively participate in social communications and engage in peer relationships, thus also better getting to understand the hearing culture in all its variation. This is known to be difficult for children with HL (Rieffe et al., 2018; Stevenson et al., 2015; S.C.P.M. Theunissen, Rieffe, Kouwenberg, et al., 2014) and therefore social development and well-being need extra focus in primary education for these children.

It would be interesting to study the possibly bidirectional relation between education and social functioning of children with HL in future studies. More communicative difficulties might lead to a lower wellbeing, which in turn can contribute to both failing exams and an increase of psychopathologic symptoms. Also, a lower educational attainment increases the risk of unemployment later in life (Dammeyer et al., 2019; Winn, 2007). Both unemployed and undereducated people are vulnerable to emotional dysfunction which might eventually lead to psychopathologic symptoms such as anxiety and depression (van der Schans et al., 2016). Future longitudinal studies investigating the social-emotional functioning among children with HL should consider the educational setting of the child (S.C.P.M. Theunissen, Rieffe, Netten, et al., 2014). One of the possibilities is to use radio-frequency identification tags to study how and to what extent children with HL interact with their surroundings in order to improve the environment, and subsequently the social interaction between children with and without HL. This is especially interesting in environments that are predominantly designed for children without HL, such as most (if not all) mainstream schools.

Concluding remarks

This thesis showed us that current rehabilitation for patients with HL is very effective, but still has promising capabilities that can be further developed. The population with HL is rather heterogenic which asks for an individual approach. Therefore, subjective screening tools and personal treatment options should be the future method within rehabilitation for patients with HL.

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Nederlandse Samenvatting

Revalidatie voor individuen met gehoorverlies kent in Nederland vele ontwikkelingen. Zo werd cochleaire implantatie (CI) in 1985 in het kader van wetenschappelijk onderzoek geïntroduceerd en uiteindelijk in 2000 geïmplementeerd als standaard zorg voor ernstig slechthorenden. Tot nu toe hebben meer dan 7.500 personen in Nederland een CI gekregen en worden er jaarlijks circa 600 implantaties uitgevoerd (CI-Overleg Nederland, 2019). Recentelijk is het auditieve hersenstam implantaat (auditory brainstem implant; afgekort ABI) beschikbaar gekomen voor kinderen met niet-functionele cochlea's of niet-functionele cochleaire zenuwen (Figuur 1 – **hoofdstuk 1**). De afgelopen paar jaar ondergingen 12 dove kinderen deze nieuwe procedure in het Leids Universitair Medisch Centrum. Deze implantaties en andere ontwikkelingen op het gebied van gehoorrevalidatie hebben het leven en de toekomstperspectieven van patiënten met gehoorverlies in positieve mate veranderd. Toch blijft de werkelijke impact van deze recente implementaties en uitbreidingen in de gehoorrevalidatie onduidelijk. Wat zijn de huidige verwachtingen van patiënten als het gaat om revalidatie? Mag een patiënt met gehoorverlies verwachten dat hij of zij na revalidatie volledig kan deelnemen aan een maatschappij met name gericht op geluid en verbale communicatie, of moet hij of zij de gevolgen van een chronische beperking accepteren? Met andere woorden, wat kunnen we tegenwoordig verwachten van revalidatie voor patiënten met gehoorverlies? Dit zijn voorbeelden van vragen die patiënten en ouders zich momenteel stellen wanneer zij de keuze maken tussen een hoortoestel (HT), CI of ABI. Om deze vragen te kunnen beantwoorden, moet men weten dat de populatie met gehoorverlies zeer heterogeen is. Dit vereist onderzoek naar verschillende uitkomsten, waarbij rekening gehouden wordt met de individuele verschillen binnen de populatie om de impact van revalidatie te onderzoeken. Dit proefschrift is zodoende gericht op het onderzoeken van verschillende aspecten van de huidige revalidatie rondom gehoorverlies. In de verschillende hoofdstukken gaan we in op de selectiecriteria voor volwassen CI-kandidaten (**hoofdstuk 2** en **3)**, de taalontwikkeling bij kinderen met een ABI (**hoofdstuk 4**), en verschillende ontwikkelingsuitkomsten na revalidatie voor kinderen met gehoorverlies, zoals de kwaliteit van leven (**hoofdstuk 5**) en het opleidingsniveau (**hoofdstuk 6**).

Postlinguaal dove en slechthorende kandidaten voor cochleaire implantatie

In **hoofdstuk 2** wordt de doeltreffendheid van verschillende preoperatieve meetinstrumenten vergeleken om te bepalen welke post-linguale dove kandidaat in aanmerking komt voor een CI. Dit werd onderzocht door na te gaan of er wel of geen verbetering van spraakverstaan met CI werd verkregen. Door middel van de sensitiviteit en specificiteit van elk meetinstrument te berekenen werd een ROC-curve (receiver operating characteristic) gegenereerd. Met de oppervlakte onder deze curve kon vervolgens de diagnostische waarde van elk meetinstrument met elkaar vergeleken worden. De resultaten van deze studie toonde aan dat de foneem score met optimaal ingestelde HT in een vrije veldopstelling de hoogste diagnostische waarde had om aan te geven welke kandidaat zijn spraakverstaan in een vrije veldopstelling zou verbeteren na CI. Daarnaast was de foneem score met optimaal ingestelde HT in ruis het meest accuraat in het voorspellen van een verbetering van de spraakperceptie in ruis. Dit laatste is vooral nuttig in de besluitvorming voor kandidaten die een hoge preoperatieve spraakverstaan in stilte hebben, maar moeite hebben om spraak te verstaan als er achtergrondruis aanwezig is.

Het doel van **hoofdstuk 3** was om het effect van nieuwe selectiecriteria voor CI te illustreren, zoals recentelijk is geïmplementeerd in het Verenigd Koninkrijk en Vlaanderen (noordelijk deel van België). In beide landen werd bij de nieuwe selectiecriteria meer waarde gehecht aan het criterium van spraakverstaan dan het criterium van toonaudiometrie. Dit leidde tot een sensitiviteitstoename van 31% ten opzichte van hun oude selectiecriteria (68,4% en 69,4%). De versoepeling van de criteria deed het aantal kandidaten met 30% toenemen. Echter, zeven van de acht kandidaten vielen alsnog buiten de nieuwe Britse en Vlaamse criteria die hun spraakverstaan wel verbeterde na implantatie in Leiden. Deze studie liet zien dat de nieuwe selectiecriteria van het Verenigd Koninkrijk en Vlaanderen hebben geleid tot een groter aantal postlinguaal dove volwassenen dat in aanmerking komt voor een CI. Toch blijkt er ruimte te zijn voor extra versoepelingen van de criteria in de toekomst.

Prelinguaal dove kandidaten voor auditieve hersenstam implantatie

Er zijn veel studies waarin de voordelen van een CI en HT bij kinderen met aangeboren gehoorverlies zijn onderzocht. Echter, de alternatieven voor dove kinderen die niet in aanmerking komen voor een CI of HT zijn in mindere mate onderzocht. Dit was de reden om in **Hoofdstuk 4** de taalontwikkeling van kinderen met een ABI in kaart te brengen en te vergelijken met kinderen die een CI gebruikten. Binnen een jaar konden zes van de zeven kinderen met een ABI geluiden identificeren, konden zij reageren op spraak en gebruikten zij hun stem om aandacht van hun omgeving te trekken. Hun ontwikkeling in taalvaardigheden

verliep echter langzamer dan die van kinderen met een CI. Uiteindelijk bereikten de kinderen met een ABI gemiddeld hetzelfde vaardigheidsniveau als kinderen met een CI die ook andere non-auditieve beperkingen hadden. Hieruit kan er geconcludeerd worden dat ABI een bevredigende auditieve input kan geven aan dove kinderen met bilaterale malformaties van het binnenoor, tenzij zij meerdere ernstige bijkomende beperkingen hebben. Met deze bevindingen kunnen toekomstige ouders van dove kinderen optimaal worden geïnformeerd over de taal en spraakuitkomsten na implantatie met een ABI.

Invloed van gehoorverlies op de sociale en educatieve ontwikkeling van kinderen

Zoals te zien is in voorgaande hoofdstukken, wordt er veel nadruk gelegd op criteria en functie-uitkomsten die makkelijk gemeten kunnen worden binnen de revalidatie voor dove en slechthorenden (**hoofdstuk 2, 3, en 4**). Ouders van kinderen met gehoorverlies zijn echter ook geïnteresseerd in het welzijn op de lange termijn, zoals mogelijke psychische problemen die kunnen ontstaan of het toekomstige opleidingsniveau van hun kind. Dit bracht ons op het idee om de kwaliteit van leven en het opleidingsniveau van kinderen met gehoorverlies te onderzoeken in twee longitudinale studies.

In **hoofdstuk 5** werd er onderzocht hoe de kwaliteit van leven van kinderen met een CI of HT verandert door de jaren heen. We zagen dat op de leeftijd van 4 en 11 jaar, kinderen met een gehoorverlies een vergelijkbare kwaliteit van leven hadden met betrekking tot emotionele en fysieke aspecten als hun horende leeftijdsgenoten. Echter, sociaal en school gerelateerd functioneren was lager bij kinderen met een gehoorverlies die het speciaal onderwijs volgden of hadden gevolgd in vergelijking met hun leeftijdsgenoten met en zonder gehoorverlies die altijd al regulier onderwijs volgden. Welbevinden met betrekking tot schoolactiviteiten nam af in de loop van de tijd, maar dit werd bij zowel kinderen met als zonder gehoorverlies gezien. Ook zagen we dat kinderen met een CI een afname in sociaal functioneren ervaarden op 11 jarige leeftijd in vergelijking met toen zij 4 jaar oud waren. Met deze bevindingen kan er vastgesteld worden dat specifieke begeleiding nodig is met betrekking tot schoolactiviteiten en sociaal functioneren bij slechthorende en dove kinderen die in het speciaal onderwijs zitten of die overstappen naar het reguliere onderwijs.

Door middel van een groot longitudinaal onderzoek, dat mede mogelijk gemaakt werd door het Centraal Bureau van de Statistiek in Nederland, werd in **hoofdstuk 6** de loopbaan in het basis en voortgezet onderwijs vergeleken tussen kinderen met en zonder gehoorverlies.

Hierbij werd gekeken of kinderen met gehoorverlies een risico lopen op een lager onderwijsniveau vanwege hun verminderde auditieve input. Resultaten lieten zien dat uiteindelijk 39% van de kinderen met een CI en 64% van de kinderen met een HT op het reguliere basisonderwijs terecht kwamen. Ook werd er gekeken naar de eindresultaten van de gestandaardiseerde Cito-toets in het basisonderwijs. Op taal en wiskunde presteerde de hele groep kinderen met gehoorverlies gemiddeld ondermaats. Echter, nadat de kinderen werden onderverdeeld in drie groepen (onder-, boven- of gemiddelde score) behaalde meer dan 60% van de kinderen met gehoorverlies een gemiddelde of bovengemiddelde score in deze vakken. Na het regulier basisonderwijs, volgden adolescenten met gehoorverlies een vergelijkbaar niveau binnen het voortgezet onderwijs als de horende populatie. Individuen met een gehoorverlies die speciaal basisonderwijs hadden gevolgd, zaten op een significant lager niveau van voortgezet onderwijs dan hun leeftijdsgenoten met en zonder gehoorverlies die regulier basisonderwijs hadden gevolgd. Dit betekent dat niet alle kinderen met gehoorverlies, maar met name diegenen die speciaal onderwijs volgen grotere kans hebben op een lager opleidingsniveau.

Tot slot

Het onderzoek beschreven in dit proefschrift had als doel de impact en de groeiende verwachtingen van revalidatie voor patiënten met gehoorverlies te onderzoeken. Er zijn veel voordelen van revalidatie met een HT, CI of ABI, maar problemen met het gehoor kunnen zich nog steeds blijven voordoen in een wereld gedreven door verbale communicatie. In **hoofdstuk 7** keken we verder dan de standaardzorg binnen de huidige gehoorrevalidatie. De optimale methode werd besproken om de obstakels van patiënten met gehoorverlies in kaart te brengen, vooral wanneer er twijfel bestond welke revalidatiemethode er gekozen moest worden (waarbij zowel een CI of HT een optie is). Gedeelde besluitvorming, waarbij spraakverstaan testen en gepersonaliseerde interviews worden gecombineerd, hebben grote potentie om de toekomstige voorkeursbenadering binnen de revalidatie voor volwassenen met gehoorverlies te worden. Tevens zouden richtlijnen in de perifere gezondheidszorg (inclusief huisartsen en audiciens) geactualiseerd moeten worden aangezien de selectiecriteria voor implantatie versoepeld zijn en er een toenemend overlappend gebied tussen de typen revalidatie is ontstaan. Hierbij is tijdig verwijzen naar een audiologisch centrum de belangrijkste boodschap.

Op dit moment worden er al pogingen ondernomen om kennis te delen tussen de perifere audiciens en professionals in academische centra door gehoorvragenlijsten uit te wisselen (Stichting Protocol Hoorhulpmiddelen, 2018). De implementatie van multidisciplinair

overleg en het gebruik van één gedeelde database zou de samenwerking kunnen verbeteren waarbij men professionals kan raadplegen voor de volgende stap in revalidatie wanneer patiënten tegen gehoor gerelateerde obstakels aanlopen. We zouden moeten beginnen met het verzamelen van patiëntgegevens door de verschillende audiciens en audiologische centra in het hele land. Door de krachten te bundelen kunnen we specifieke informatie omtrent het gehoorverlies verzamelen en de patiënt doorverwijzen naar de juiste kliniek met inachtneming van de huidige privacyregels. Big data analyse en het gebruik van kunstmatige intelligentie kunnen gebruikt worden om te onderzoeken welk gebied van de revalidatie voor gehoorverlies verbeterd zou kunnen worden.

In **hoofdstuk 7** kwamen tevens de uitkomsten van het sociaal welzijn en het onderwijsniveau van kinderen met aangeboren gehoorverlies aan bod om de verwachtingen binnen de pediatrische gehoorrevalidatie te kunnen schetsen aan ouders. Om de achterstand op school en in het sociaal functioneren in te halen, zou de ideale oplossing kunnen zijn dat er extra tijd voor kinderen met gehoorverlies wordt geïntroduceerd in het regulier onderwijs, zoals een tussenjaar op het basis of voortgezet onderwijs. Extra tijd zou deze kinderen kunnen helpen om zich beter aan te passen aan het regulier onderwijs en aan hun horende leeftijdsgenoten. Dit zou uiteindelijk de kansen kunnen vergroten om voor deze kinderen het juiste onderwijssysteem te bepalen.

In toekomstige studies zou het interessant zijn om de bi-directionele relatie tussen onderwijs en sociaal-emotioneel functioneren van kinderen met gehoorverlies te onderzoeken. Problemen met het gehoor kunnen leiden tot een lager welbevinden, wat zowel kan bijdragen tot het zakken voor examens als tot een toename van psychopathologische symptomen. Ook kan een lager opleidingsniveau leiden tot werkloosheid later in het leven (Dammeyer et al., 2019; Winn, 2007). Zowel werklozen als laagopgeleiden zijn kwetsbaar voor emotioneel disfunctioneren wat uiteindelijk weer tot psychopathologische symptomen kan leiden zoals angst en depressie (van der Schans et al., 2016). Toekomstige longitudinale studies die het sociaal-emotioneel functioneren bij kinderen met een gehoorverlies onderzoeken zouden daarom rekening moeten houden met de onderwijssetting van het kind (Theunissen, Rieffe, Netten, et al., 2014). Een van de mogelijkheden is om met behulp van radiofrequente identificatie tags (RFID) te bestuderen hoe en in welke mate kinderen met gehoorverlies interactie hebben met hun omgeving. Dit is vooral interessant op schoolpleinen en in klaslokalen die zijn ingericht voor horende kinderen (zoals in het regulier onderwijs).

Appendix

QUESTIONNAIRES

PedsQL 4.0 – Oudervragenlijst (2-4 jaar)

In hoeverre heeft uw kind in de AFGELOPEN MAAND problemen gehad met ...

LICHAMELIJK FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Lopen	0	1	2	3	4
2. Rennen	0	1	2	3	4
3. Actief spelen en lichaamsbeweging	0	1	2	3	4
4. Iets zwaars optillen	0	1	2	3	4
5. Baden	0	1	2	3	4
6. Helpen met opruimen van zijn/ haar speelgoed	0	1	2	3	4
7. Pijn hebben	0	1	2	3	4
8. Weinig energie	0	1	2	3	4

EMOTIONEEL FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Zich angstig of bang voelen	0	1	2	3	4
2. Zich verdrietig voelen	0	1	2	3	4
3. Zich boos voelen	0	1	2	3	4
4. Moeite met slapen	0	1	2	3	4
5. Zich zorgen maken	0	1	2	3	4

SOCIAAL FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Spelen met andere kinderen	0	1	2	3	4
2. Andere kinderen willen niet met hem/ haar spelen	0	1	2	3	4
3. Gepest worden door andere kinderen	0	1	2	3	4
4. Bepaalde dingen niet kunnen die andere kinderen van zijn/ haar leeftijd wel kunnen	0	1	2	3	4
5. Mee kunnen blijven doen tijdens het spelen met andere kinderen	0	1	2	3	4

* Vul a.u.b. dit deel in als uw kind naar school of naar een peuterspeelzaal gaat

FUNCTIONEREN OP SCHOOL (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Aan dezelfde schoolactiviteiten deelnemen als andere kinderen van dezelfde leeftijd	0	1	2	3	4
2. Niet naar school/peuterspeelzaal gaan omdat hij/zij zich niet lekker voelt	0	1	2	3	4
3. Niet naar school/peuterspeelzaal gaan omdat hij/zij naar de dokter of het ziekenhuis moet	0	1	2	3	4

PedsQL 4.0 – Oudervragenlijst (5-7 jaar)

In hoeverre heeft uw kind in de AFGELOPEN MAAND problemen gehad met ...

LICHAMELIJK FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Meer dan 100 meter lopen	0	1	2	3	4
2. Rennen	0	1	2	3	4
3. Aan sport of andere lichaamsbewging doen	0	1	2	3	4
4. Iets zwaars optillen	0	1	2	3	4
5. Zelfstandig een bad of douche nemen	0	1	2	3	4
6. Karweitjes doen, zoals het opruimen van zijn/haar speelgoed	0	1	2	3	4
7. Pijn hebben	0	1	2	3	4
8. Zich moe voelen	0	1	2	3	4

EMOTIONEEL FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Zich angstig of bang voelen	0	1	2	3	4
2. Zich verdrietig of somber voelen	0	1	2	3	4
3. Zich boos voelen	0	1	2	3	4
4. Problemen met slapen	0	1	2	3	4
5. Zich zorgen maken over wat hem/haar zal overkomen	0	1	2	3	4

SOCIAAL FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Op kunnen schieten met andere kinderen	0	1	2	3	4
2. Andere kinderen die niet zijn/haar vriend(in) willen zijn	0	1	2	3	4
3. Gepest worden door andere kinderen	0	1	2	3	4
4. Dingen niet kunnen die andere kinderen van zijn/ haar leeftijd wel kunnen	0	1	2	3	4
5. Mee kunnen blijven doen tijdens het spelen met andere kinderen	0	1	2	3	4

FUNCTIONEREN OP SCHOOL (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Opletten tijdens de les	0	1	2	3	4
2. Dingen vergeten	0	1	2	3	4
3. Bijblijven met schoolactiviteiten	0	1	2	3	4
4. Niet naar school gaan omdat hij/zij zich niet lekker voelt	0	1	2	3	4
5. Niet naar school gaan omdat hij/zij naar de dokter of het ziekenhuis moet	0	1	2	3	4

PedsQL 4.0 – Oudervragenlijst (8-12 jaar)

In hoeverre heeft uw kind in de AFGELOPEN MAAND problemen gehad met ...

LICHAMELIJK FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Meer dan 100 meter lopen	0	1	2	3	4
2. Rennen	0	1	2	3	4
3. Aan sport of andere lichaamsbewging doen	0	1	2	3	4
4. Iets zwaars optillen	0	1	2	3	4
5. Zelfstandig een bad of douche nemen	0	1	2	3	4
6. Karweitjes rond het huis doen	0	1	2	3	4
7. Pijn hebben	0	1	2	3	4
8. Zich moe voelen	0	1	2	3	4

EMOTIONEEL FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Zich angstig of bang voelen	0	1	2	3	4
2. Zich verdrietig of somber voelen	0	1	2	3	4
3. Zich boos voelen	0	1	2	3	4
4. Problemen met slapen	0	1	2	3	4
5. Zich zorgen maken over wat hem/haar zal overkomen	0	1	2	3	4

SOCIAAL FUNCTIONEREN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Op kunnen schieten met andere kinderen	0	1	2	3	4
2. Andere kinderen die niet zijn/haar vriend(in) willen zijn	0	1	2	3	4
3. Gepest worden door andere kinderen	0	1	2	3	4
4. Dingen niet kunnen die andere kinderen van zijn/ haar leeftijd wel kunnen	0	1	2	3	4
5. Mee kunnen blijven doen tijdens het spelen met andere kinderen	0	1	2	3	4

FUNCTIONEREN OP SCHOOL (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Opletten tijdens de les	0	1	2	3	4
2. Dingen vergeten	0	1	2	3	4
3. Bijblijven in de klas en met huiswerk	0	1	2	3	4
4. Niet naar school gaan omdat hij/zij zich niet lekker voelt	0	1	2	3	4
5. Niet naar school gaan omdat hij/zij naar de dokter of het ziekenhuis moet	0	1	2	3	4

PedsQL 4.0 – Kindervragenlijst (8-12 jaar)

In hoeverre heb je in de AFGELOPEN MAAND problemen gehad met ...

OVER MIJN GEZONDHEID EN ACTIVITEITEN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Het is voor mij moeilijk om meer dan 100 meter lopen	0	1	2	3	4
2. Het is voor mij moeilijk om te rennen	0	1	2	3	4
3. Het is voor mij moeilijk om te sporten of lichamelijke oefeningen te doen	0	1	2	3	4
4. Het is voor mij moeilijk om iets zwaars op te tillen	0	1	2	3	4
5. Het is voor mij moeilijk om zelfstandig een bad of douche te nemen	0	1	2	3	4
6. Het is voor mij moeilijk om karweitjes rond het huis te doen	0	1	2	3	4
7. Ik heb pijn	0	1	2	3	4
8. Ik heb weinig energie	0	1	2	3	4

OVER MIJN GEVOELENS (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Ik voel me angstig of bang	0	1	2	3	4
2. Ik voel me verdrietig	0	1	2	3	4
3. Ik voel me boos	0	1	2	3	4
4. Ik heb moeite met slapen	0	1	2	3	4
5. Ik maak mij zorgen over wat mij zal overkomen	0	1	2	3	4

HOE IK MET ANDEREN OP KAN SCHIETEN (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Ik heb moeite om met andere kinderen op te schieten	0	1	2	3	4
2. Andere kinderen willen mijn vriend(in) niet zijn	0	1	2	3	4
3. Andere kinderen pesten mij	0	1	2	3	4
4. Ik kan dingen niet die andere kinderen van mijn leeftijd wel kunnen	0	1	2	3	4
5. Het is moeilijk om met andere kinderen mee te kunnen blijven doen als ik met ze speel	0	1	2	3	4

OVER SCHOOL (problemen met...)	Nooit	Bijna nooit	Soms	Vaak	Bijna altijd
1. Het is moeilijk om op te letten tijdens de les	0	1	2	3	4
2. Ik vergeet dingen	0	1	2	3	4
3. Ik heb moeite om bij te blijven met mijn schoolwork (waaronder huiswerk)	0	1	2	3	4
4. Ik ga niet naar school, omdat ik me niet lekker voel	0	1	2	3	4
5. Ik ga niet naar school, omdat ik naar de dokter of het ziekenhuis moet	0	1	2	3	4

ABBREVIATIONS

ABI	Auditory Brainstem Implant
AUC	Area Under the Curve
CAP	Categories of Auditory Performance
CELF	Clinical Evaluation of Language Fundamentals
CI	Cochlear Implant
CITO	Central Institute for Test Development
dB	Decibel
DECIBEL	Developmental Evaluation of Children: Impact and Benefits of Early hearing screening strategies Leiden
DHH	Deaf and Hard of Hearing
HA	Hearing Aid
HL	Hearing Loss
IQ	Intelligent Quotient
IT-MAIS	Infant Toddler Meaningful Auditory Integration Scale
LUMC	Leiden University Medical Center
MCAR	Missing Completely At Random
MUSS	Meaningful Use of Speech Scale
n	Number of participants
NF2	Neurofibromatosis Type 2
NH	Normal Hearing
NHS	Newborn Hearing Screening
PedsQL	Pediatric Quality of Life Inventory
PTA	Pure Tone Audiometry at 0.5, 1, 2, and 4 kHz
QoL	Quality of Life
ROC	Receiver Operating Characteristic
SD	Standard Deviation
SIR	Speech Intelligibility Rate
SPSS	Statistical Package for the Social Sciences
SPT	Speech Perception Test
TH	Typical Hearing
UK	United Kingdom
WISC-III	Wechsler Intelligence Scale for Children-Third Edition

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CURRICULUM VITAE

Tirza van der Straaten was born on 17 September 1990 on Curaçao, the former Dutch Antilles. She finished her secondary education (VWO) at the Peter Stuyvesant College (now Kolegio Alejandro Paula) in 2008 in Willemstad, Curaçao. Hereafter she emigrated to the Netherlands where she started studying Medicine at the University of Leiden. She achieved her Bachelor of Science degree in 2012 and her Master of Science degree in 2015, both at the Leiden University. Her MSc thesis was conducted in Melbourne, Australia where she studied the age-related differences in the quantity of hemostatic proteins. After graduation in 2015, she started working as a medical doctor at the surgery department in Bronovo (MCH) hospital in The Hague. From 2016 till 2020 she was conducting her PhD research at the department of Otorhinolaryngology and Head & Neck Surgery at Leiden University Medical Center resulting in this thesis. In September 2020 she started her residency in general practice (or house doctor) at the Leiden University Medical Center.

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Stellingen behorende bij het proefschrift

OPPORTUNITIES WITHIN AND AFTER REHABILITATION
for patients with hearing loss

1. Bij het selecteren van kandidaten voor cochleaire implantatie bepaalt het beoogde doel in spraakverstaan de keuze voor het meest accurate diagnostische meetinstrument (*dit proefschrift*).

2. Het leren van gesproken taal door kinderen met een auditief hersenstam implantaat vraagt om zowel intensieve begeleiding als doorzettingsvermogen van ouders en leraren (*dit proefschrift*).

3. Een geslaagde overstap van speciaal naar regulier basisonderwijs is mogelijk voor kinderen met gehoorverlies met extra begeleiding voor onderwijs en ondersteuning bij het sociaal functioneren (*dit proefschrift*).

4. De mogelijkheden in het voortgezet onderwijs voor kinderen met gehoorverlies worden voor een belangrijk deel bepaald door het type basisonderwijs dat zij hebben doorlopen (*dit proefschrift*).

5. De sociaal-emotionele ontwikkeling van kinderen met gehoorverlies wordt het beste gestimuleerd op het schoolplein van passend onderwijs.

6. Iemand wordt getypeerd als gehandicapt door de houding en structuur van de maatschappij, niet door zijn/haar beperking.

7. Ouderen met progressief gehoorverlies moeten beter geïnformeerd worden over de mogelijkheden van een cochleaire implantaat zodat zij minder risico lopen op moeizame communicatie en sociale isolatie.

8. Bij CI-kandidaten die Papiaments spreken is het belangrijk om extra aandacht te geven aan klanken omdat een betekenis van een woord daardoor kan veranderen.

9. De planning en hoofdstukken van een proefschrift worden bij eb in het zand geschreven.

10. Promoveren is net als zeilen, ook met tegenwind leer je je doel te bereiken.

11. Een discussie kan het beste volgens de Socratische methode verlopen zodat met een nieuwsgierige blik nieuwe inzichten verkregen worden.

12. Opgroeien op Curaçao geeft een ontwikkelingsvoorsprong op het gebied van multiculturaliteit, wereldburgerschap, muzikaliteit en watersport.