Impact and benefits of early hearing screening

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1 INTRODUCTION

GENERAL INTRODUCTION

This thesis describes the first results of the DECIBEL-study. DECIBEL is an acronym for Developmental Evaluation of Children: Impact and Benefits of Early hearing screening and is a nationwide study initiated in Leiden. The general introduction provides a brief introduction to aetiology, hearing screening programs and developmental outcome of children with permanent childhood hearing impairment (PCHI) and discusses the background and rationale of the study.

Congenital hearing impairment is a serious and relatively common problem. It is widely believed that approximately 1 in a 1000 live newborns has hearing loss at birth.[1;2] This prevalence is expected to double during childhood due to progressive, late-onset and acquired hearing losses.[2-5] Children classify for the diagnosis of PCHI when they suffer from bilateral permanent conductive or sensorineural hearing loss of ≥40 decibels (dB) in the better ear.

A BRIEF INTRODUCTION

AETIOLOGY

It is assumed that the cause of hearing loss in children is hereditary in 50%, of non-genetic origin in 25% and of unknown cause in 25%.[6-8] The cause of PCHI can be present at birth, but hearing loss may not yet be detectable at that early age and gone unnoticed by early hearing screening. The causes of hearing loss which appear later than the post-neonatal period can be classified as delayed-onset, progressive or acquired hearing loss.[9] Congenital infections such as toxoplasmosis, rubella, cytomegalovirus, herpesvirus and syphilis (TORCHES) can produce neurological damage and are important causes of delayed onset and progressive sensorineural hearing loss. The cytomegalovirus (CMV) is the most common congenital infection in the western world.[10] Approximately 85% of infants with congenital CMV infection do not exhibit signs or symptoms at birth, but about 15% of these children will develop permanent sequelae, such as PCHI and general developmental delay, during early childhood.[11-13] Progressive hearing loss is seen in neurodegenerative diseases and syndromes including Pendred and Waardenburg.[6] Acquired hearing loss can be caused by infection, ototoxicity (e.g. medication), trauma and noise-damage. Hearing loss caused by meningitis is the most well known in this category. Since the introduction of a nationwide well-established immunization program for *H. influenzae type B* and *S. pneumoniae* it is clear that meningitis caused by these bacteria has declined in the Netherlands.[14]

In 2007 the Joint Committee on Infant Hearing published a list of risk-indicators for progressive or 'delayed-onset' hearing loss in children with a negative screen for hearing loss at newborn hearing screening (NHS) (Table 1.)[15] The presence of risk-indicators, ranging from parental

concern on children's hearing to congenital infections and neurodegenerative diseases, indicates the need for audiological evaluation in childhood.
Understanding the aetiology of PCHI is important with respect to treatment options, predicting the developmental prognosis and recurrence risk in future siblings and family members.

HEARING SCREENING PROGRAMS

In the Netherlands hearing screening started with distraction methods (DHS): in 1965 the Ewing test and in the 1990s the Compacte Amsterdamse Paedo-Audiometrische Screener (CAPAS).[16] These subjective methods, performed at the age of 9 months, used the orientation reflex of the child to detect hearing loss. This screening method faced three important difficulties caused by the moment and the type of hearing screening. First the inadequate efficiency: at extensive audiological examination a large number of referred children did not suffer from a permanent hearing impairment but from a temporary conductive hearing deficit accompanying an upper respiratory infection, a relatively prevalent condition in children at the age of 9 months. The consequence was a large number of false-positives. This was not only costly but did also decrease the credibility of the test for professionals as well as for parents, with subsequent withdrawal from testing as final result. Secondly, the relatively ''older'' age at detection and start of amplification. Finally, the inability to use the distraction hearing test for children with developmental delay. The aforementioned disadvantages necessitated the introduction of another moment and type of hearing screening. In several countries (but not in the Netherlands) targeted ('high-risk') screening was introduced. In targeted screening children with risk factors for hearing loss were identified in the postnatal period and were taken in audiological follow-up (see Table 1 for risk factors). It was found however, that targeted screening left a large proportion of children with permanent childhood hearing loss unidentified, since many children had no risk factors for hearing loss at all.[17;18] The fact that non-invasive objective screening instruments had just become available coupled with the outcomes of studies showing important developmental consequences should amplification and habilitation be delayed, lead to the initiation of universal NHS programs throughout the world during the last two decades.[19;20] Furthermore, the probable cost savings associated with early identification and intervention reducing the need for further long-term special educational needs contributed to the emergence of NHS programs.[21-23]
The aim of NHS at introduction was (and still is) to identify permanent conductive or sensorineural hearing loss early in life to allow early intervention and counselling. The Joint Committee on Infant Hearing formulated recommendations concerning the time-periods in which the screening process should be accomplished.[15] Those with a positive screen for hearing loss at NHS should have a comprehensive audiological evaluation at no later than 3 months of age and infants with confirmed hearing loss should receive appropriate intervention as soon as possible but no

later than at the age of 6 months.[15] Auditory input during the period of plasticity of the auditory cortex is essential for cortical development.[24;25] Start of habilitation should therefore start before the critical period of 6 months. Habilitation and intervention involve amplification with hearing aids, a cochlear implant or a bone-anchored hearing aid. Amplification is often accompanied by family-support, speech-training and/or the use of sign-language.

Risk indicators that are marked with a "§" are of greater concern for delayed-onset hearing loss.
1. Caregiver concern§ regarding hearing, speech, language, or developmental delay.
2. Family history§ of permanent childhood hearing loss.
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: extracorporeal membrane oxygenation,§ assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop- diuretics (furosemide), and hyperbilirubinemia that requires exchange transfusion.
4. In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,§ such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange- Nielson.
8. Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome.
9. Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.
11. Chemotherapy§

Table 1 Risk indicators associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood.[15]

In the Netherlands the NHS is either performed during home-visits, together with newborn bloodspot screening, or at the well-baby clinic. NHS for well babies is a three stage hearing screening program using transient evoked oto-acoustic emissions (OAE) in the first two stages and automated auditory brain stem response (A-ABR) in the third stage. OAEs are weak sounds produced spontaneously or in response to a specific external stimulus by the outer hair cells of a well functioning ear.[26] The adequacy of the neurons (response-level and latency) is measured with the A-ABR.[27] A uni- or bilateral positive screen for hearing loss at one stage will be followed by a second hearing screening step and a positive screen at the third stage will be followed by referral to an Audiology Center for extensive diagnostic investigation and

confirmation. Newborns admitted to a neonatal intensive care unit (NICU) are at increased risk for developing PCHI and these newborns undergo a 2-stage NICU hearing screening program using A-ABR since 2001.[27] Some children directly undergo extensive diagnostic investigations without completing a hearing screening program because of medical reasons (i.e. external ear anomalies). Nowadays virtually all live newborns participate in hearing screening programs in the Netherlands, being one of the first countries in the world. It is important to realize that the NHS is the only newborn screening program that is not a blood test analysis and for which the treatment is predominantly educational rather than medical.

DEVELOPMENTAL CONSEQUENCES OF EARLY DIAGNOSIS OF HEARING LOSS

The importance of early diagnosis of hearing loss and prompt habilitation received worldwide attention by studies by Yoshinaga-Itano et al.[21;28-30] These studies report that the diagnosis of PCHI should be established within 6 months of age and intervention should be started soon after to optimize development. The degree of hearing loss in early identified children was found to be unrelated to developmental outcome, in contrast to what was found in later identified children.[28] Research by others replicated the finding that children with PCHI identified by NHS had higher (receptive) language scores at school-age compared to children without access to NHS[31] and that early start of intervention contributed to a better language developmental outcome.[32] In recent studies the degree of hearing loss was found to be more predictive of language skills in childhood than the age at diagnosis.[33-35] The evidence, however, for the true effect of early diagnosis and early intervention on developmental outcome in children with PCHI to date, is of poor to fair quality (non-randomized designs, descriptive in nature and based on convenience samples) and other child-relevant outcome parameters (social aspects, quality of life, educational development) were not earlier investigated.[36]

THE CLINICAL PROBLEM

The introduction of the NHS in all of the Netherlands was an enormous effort which increased the awareness of the importance of normal hearing in children among caregivers as well as health care professionals. Several clinical questions, important with respect to the yield and the success of NHS, are however, unanswered to date.

Following a positive screen at hearing screening, can the aetiology be determined? Is a positive screen for hearing loss ('a refer' or 'fail') at hearing screening followed by early intervention? To what extent is a negative screen for hearing loss ('a pass') at hearing screening a guarantee for a normal hearing in childhood? And can we consolidate the evidence for better developmental outcome following early diagnosis of hearing loss?

BACKGROUND AND RATIONALE

To determine the true effect of moment of hearing screening on the developmental outcome in children with PCHI ideally a randomized trial would be preferred. In such a trial live newborns would be randomly assigned to DHS or NHS (personalized or clustered by region of birth) and development would be assessed at a certain age in all children diagnosed with PCHI. The age at developmental evaluation should be carefully chosen, since the assessment alone may influence the habilitation program and thereby the developmental outcome during follow-up. However, apart from practical difficulties (including expense and duration) the initiation of a randomized trial is not considered ethically feasible to date.[21;37]

To overcome these difficulties, we used the regional differences in the assignment of type of hearing screening created by national policy, as an instrumental variable in the present study. In the Netherlands NHS was gradually introduced from 2002 onwards, replacing DHS (at the age of 9 months) region by region (n=65). In June 2006 NHS had replaced DHS in the whole country (Figure 1.) This policy created regional differences in the assignment of type of hearing screening to children based solely on the availability at the place and the date of birth (the 'instrumental variable'). Since the type of hearing screening offered was independent of the developmental prognosis of the individual child, this study, with its 'naturally' randomized design, is expected to be as credible as a randomized trial.[38-41]

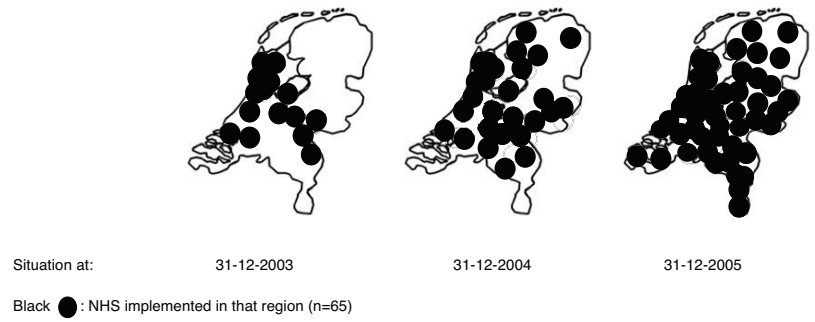

Figure 1 Implementation of newborn hearing screening in the Netherlands

This period of gradual replacement of type of hearing screening was chosen as the period of study (2003-05 cohort) for the present DECIBEL-study. We started the DECIBEL-study with identifying the children of the 2003-05 cohort known to an Audiology Center with PCHI at the age of 3-5 years. This gave us information on the epidemiology of PCHI and the yield of NHS in

our country. Detailed information on the aetiology of hearing loss and developmental outcome of children with PCHI was collected in children participating in the DECIBEL-study.

STUDY AIM

The aim of the DECIBEL-study was to evaluate the impact and benefits of early hearing screening (NHS) in comparison to hearing screening at the age of 9 months (DHS).
The main objectives of this thesis are:

1. Determine the aetiology of permanent childhood hearing impairment in children in the Netherlands.
2. Determine the sensitivity of the newborn hearing screening program and the characteristics of the children unidentified by NHS.
3. Study the effect of newborn hearing screening on developmental outcome in 3-5 year old children with permanent childhood hearing impairment compared to children in distraction hearing screening.

OUTLINE OF THIS THESIS

In CHAPTER 2 the aetiology of permanent childhood hearing impairment is evaluated and compared with the results of a systematic review on this topic. CHAPTER 3 describes the role of congenital cytomegalovirus in children with permanent childhood hearing impairment. CHAPTER 4 discusses what doctors know and should know about prevalence, clinical symptoms and prevention of cytomegalovirus. CHAPTER 5 describes what proportion of children with permanent childhood hearing impairment of congenital cause at the age of 3-5 years was unidentified by NHS. In CHAPTER 6 the periods of time elapsed following a negative screen for hearing loss at hearing screening are analysed. CHAPTER 7 presents the impact of early hearing screening on developmental outcome in childhood as determined in a unique (pseudo-randomised) design. The main findings of the DECIBEL-study in the context of current literature and suggestions for future policy and research are discussed in CHAPTER 8. The final chapter of this thesis is CHAPTER 9. This chapter provides the NEDERLANDSE SAMENVATTING VOOR 'DE NIET MEDISCH ONDERLEGDE LEZER'.