Impact and benefits of early hearing screening

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SUBMITTED

5 NATIONAL STUDY ON NEWBORN HEARING SCREENING: PROGRAM SENSITIVITY AND CHARACTERISTICS OF CHILDREN UNIDENTIFIED

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ABSTRACT

Objective

Determine the sensitivity of the current newborn hearing screening programme (NHS), analyse the characteristics of children unidentified by NHS and explore the possible need for hearing screening later in childhood.

Design

Nationwide, retrospective follow-up.

Setting

Population based study in the Netherlands.

Participants

All children born in 2003-05 and screened by a hearing screening programme (well babies and neonatal intensive care (NICU) graduates).

Main outcome measures:

Sensitivity of the NHS programme (based on the proportion of children known with permanent childhood hearing impairment (PCHI) in 2008 who were identified by NHS). Age at diagnosis, severity and aetiology of PCHI in children unidentified by NHS.

Results

The sensitivity of the current NHS programme was 0.83 (0.79 for well babies and 0.96 for NICU graduates). PCHI was confirmed before 36 months of age in 96% of the study cohort. Of the children unidentified at NHS >50% had moderate hearing loss. In these children no predominant cause of PCHI was found.

Conclusions

With our current NHS programme we are able to identify the majority of children with PCHI of congenital cause. The results of the present study do not support the initiation of audiological screening in childhood (later than NHS). Compared to the previously used distraction hearing screening programme, all programme characteristics are greatly improved.

INTRODUCTION

Permanent Childhood Hearing Impairment (PCHI) is a serious and relatively common condition. Children are classified as having PCHI when they suffer from bilateral permanent conductive or sensorineural hearing loss of ≥40 decibels (dB) in the better ear. If PCHI of congenital cause is identified later than the neonatal period those children are at risk for developmental delay. Early detection and subsequent timely intervention in children with PCHI has been suggested to lead to a better developmental outcome later.[21;31;37;87-91]

When objective hearing screening instruments for newborns became available, Newborn Hearing Screening (NHS) programmes were implemented throughout the world to identify hearing loss of congenital cause early. The NHS programme increased the proportion of cases of PCHI referred early.[1] However, to date it is not clear what proportion of children with PCHI in childhood is unidentified following a universal NHS programme.

Earlier research showed that children with delayed onset and progressive hearing loss present sometime in childhood. Together with acquired hearing loss this results in an increase in the prevalence of PCHI early in life. The prevalence of PCHI in live newborns is reported to range from 0.94 per 1000[1] to 1.18 per 1000 newborns.[2] At the age of 5-10 years the prevalence of PCHI varies between 1.33 (CI95% 1.22-1.45)[54] and 1.44 (1.41-1.48) per 1000 and increases to 1.63 per 1000 (CI95% 1.59-1.67) at the age of 8-13 years.[5]

This increase in prevalence of PCHI during childhood suggests that, with a NHS programme, it is not unlikely that approximately 34% of children with PCHI of congenital cause are unidentified by NHS and need to be diagnosed after the neonatal period.

We set up a nationwide follow-up study to evaluate (1) the sensitivity of the current hearing screening programme, (2) the characteristics of children unidentified by NHS, and (3) the possible need for audiological screening (in addition to NHS) in childhood. The results of this study are expected to contribute to our knowledge concerning the yield of the NHS programme.

PATIENTS AND METHODS

HEARING SCREENING PROGRAMS IN THE NETHERLANDS

The aim of NHS is to identify permanent conductive or sensorineural hearing loss early in life to allow early intervention and counselling and subsequently promote developmental outcome. The current NHS programme in the Netherlands includes the NHS for well babies and the NHS for NICU graduates.

From 2002 onwards the NHS for well babies co-existed with and gradually replaced a distraction behavioural hearing screening programme (DHS, offered at the age of 9 months), which had been carried out in our country since 1965. NHS for well babies has been offered to all well babies born or living in our country before the age of 6 weeks since 2006. The NHS is performed either during home-visits, combined with newborn bloodspot screening, or at a well-baby clinic. It is a three stage hearing screening programme using transient evoked oto-acoustic emissions (OAE) in the first two stages and automated auditory brain stem response (A-ABR) in the third stage. A uni- or bilateral positive screening at one stage will be followed by a second hearing screening step and a positive screen for hearing loss at the third stage will be followed by referral to an Audiology Center (AC) for diagnostic investigations, confirmation and treatment, if necessary. Newborns admitted to a neonatal intensive care unit (NICU) are at an increased risk for developing PCHI and since 2001 these newborns undergo a 2-stage NICU hearing screening programme using A-ABR.[27] Some children directly undergo extensive diagnostic investigations without completing a hearing screening programme because of medical reasons (i.e. external ear anomalies).

STUDY-DESIGN AND DATA-COLLECTION

Data were collected within the framework of the DECIBEL-study. All children born in our country between January 1st 2003 and December 31 2005 who were offered hearing screening (well babies and NICU graduates) were included in the present study. In this 2003-05 study cohort the number of children identified with PCHI following NHS was determined (a positive screening for hearing loss at NHS and PCHI confirmed by diagnostic investigation at an AC). The number of children known with PCHI at the age of 3-5 years (known to an AC in 2008) was also determined.

All data from well babies who were screened with NHS are registered by the Dutch Foundation for the Deaf and Hard of Hearing Child (NSDSK).[92] All data on NICU-graduates are registered by the coordinator of the NICU-hearing screening programme.[93] Following approval by the administrators (with permission from the youth health care organisations for well babies and the NICU privacy committee for NICU graduates) these data (including information on the positive predictive values of the hearing screening programmes) were available for analysis. No registration data were available for children in the DHS programme and, as a consequence, detailed information on the number of children identified with PCHI by DHS is not available.

Follow-up data of the 2003-05 study cohort were collected in 2008. Following ethical approval by the Medical Ethics Committee of the Leiden University Medical Center, all AC's (n=22) in the Netherlands agreed to collaborate in the identification of children from the 2003-05 study cohort, known with PCHI at the age of 3-5 years. In the Netherlands the AC is the designated

organisation for children with PCHI, not only for diagnostic evaluation, but also for amplification. Counselling is organized by family care organisations. Professionals at the ACs conscientiously examined, with assistance of two of the researchers (AK and SK), all records of children from the 2003-05 study cohort to ascertain in- or exclusion for this study in 2008. The inclusion criteria were: born in the Netherlands between January 1st 2003 and December 31st 2005, offered NHS or DHS and documented PCHI at the age of 3, 4 or 5 years. PCHI was classified on the basis of the most recent hearing tests performed by the audiologist.

Minimal background data were collected (initials, sex and date of birth). Duplicate entries of children were minimalised by the use of a database to discover possible matches based on these background variables. Additionally, the age at diagnosis of children in DHS was determined. In children in NHS not only the age at diagnosis, but also the result of hearing screening, the degree of hearing loss, and the cause of PCHI (if known) were noted from medical and audiological records, parental questionnaires and relevant diagnostic tests.

When causes leading to hearing loss were present at the time of NHS screening (present before, at, or shortly after birth) PCHI was classified as congenital in this study. Congenital PCHI may be due to genetic causes, intrauterine infections, congenital anomalies or perinatal risk factors.

In our country virtually all children with PCHI of congenital cause will have presented at an AC for diagnostic investigation by the age of 4. It is unlikely that children with more severe hearing loss of congenital cause will present later than school-age (48 months in the Netherlands) since a certain level of communication skill is required to start school. There is also an extensive network of youth health care organizations monitoring the development of all children during childhood, which should detect (language) developmental delay.

STATISTICAL ANALYSIS

The sensitivity of the current hearing screening programme in our country was calculated using the number of children from the 2003-05 study cohort identified with PCHI by NHS and known with PCHI in 2008. The sensitivity of the individual programmes (DHS and NHS for well-babies and NHS for NICU-graduates) was also determined. To make the effect of the current NHS on age at diagnosis of PCHI clear a comparison was made with the age at diagnosis of children in DHS.

Since NHS was gradually introduced (since 2002, region by region, n=65) not all well-babies born in 2003-05 in our country were offered NHS. However, all NICU-graduates were offered NHS in that period. A weighting factor was used to determine the prevalence of PCHI, as if NHS for well babies would have been offered for the full period 2003-2005 (See Table 1. for the formula).

Since hearing screening programmes are set up to detect children with hearing loss caused by factors present at birth, children with an acquired hearing loss in childhood (e.g. due to meningitis, medication or trauma) were identified and included only in the appropriate analysis.

Descriptive statistics were used to generate results for the type and result of hearing screening, the degree of hearing loss and the cause of PCHI. The interquartile range (IQR) was used to describe the range in age at diagnosis. All statistical tests were carried out using SPSS version 16.0 (SPSS Inc., Chicago, IL, USA).

RESULTS

The 2003-05 study cohort included all 582.214 live births in that period. Of those 338.525 were screened by NHS (326.697 by NHS for well babies and 11.828 by NHS for NICU graduates). PCHI was confirmed following a positive screen for hearing loss at NHS in 403 children in the 2003-05 study cohort (214 well babies and 189 NICU graduates) (Table 1.). In 2008, at follow-up, 448 children were found to have PCHI of congenital, or not confirmed acquired cause, and had been screened by NHS (279 well babies, 169 NICU graduates) (Table 2.). In detail: 65 extra well babies were identified at an AC with documented PCHI (65/326.697 or 0.19/1000 live newborns screened) and 20 NICU graduates who initially had PCHI confirmed following a positive screen for hearing loss at NHS were not known to an AC with PCHI in 2008. In addition, 132 children who had been screened by DHS were identified with PCHI.

	NUMBER OF CHILDREN SCREENED	NUMBER OF CHILDREN WITH CONFIRMED BILATERAL PCHI	PREVALENCE OF CONGENITAL PCHI PER 1000 LIVE NEWBORNS
POPULATION-BASED PERSPECTIVE			N=582.214
Following NHS well babies 2003-05	326.697	214#	0.67
Following NHS NICU graduates 2003-05	11.828	189	16
Total following NHS in 2003-05		403	
Weighted total following NHS 2003-05##		563	0.97

Confirmed hearing loss following NHS for well babies: 0.67 per 1000 or
((326.697 * 0.003 *0.93)* 0.24) /326.697
Confirmed hearing loss following NHS for NICU graduates: 16 per 1000 or 189 /11828
Weighted prevalence if NHS had been offered to all well babies and NICU graduates in 2003-05:
0.97 per 1000 or 563 /582.214

The following figures were used to calculate the number of children with confirmed bilateral PCHI in well babies: Percentage screen not pass 0.29, documented follow-up 93 and confirmation of PCHI in 24.1. 4

Weighting factor= 1.75 or (582.214-11.828) /326.697

Table 1 The number of children with confirmed hearing loss following NHS in the 2003-05 cohort.

Information concerning the hearing screening result at NHS was available for 367 out of the 448 children with PCHI of congenital, or not confirmed acquired cause. A negative screen for hearing loss was documented in 36 of the 65 extra identified NHS well babies and also for 6 NHS NICU graduates (Table 2.). The proportion of NICU graduates unidentified by NHS for NICU graduates was 6 per 11.828 children (0.5/1000 live NICU graduates screened).

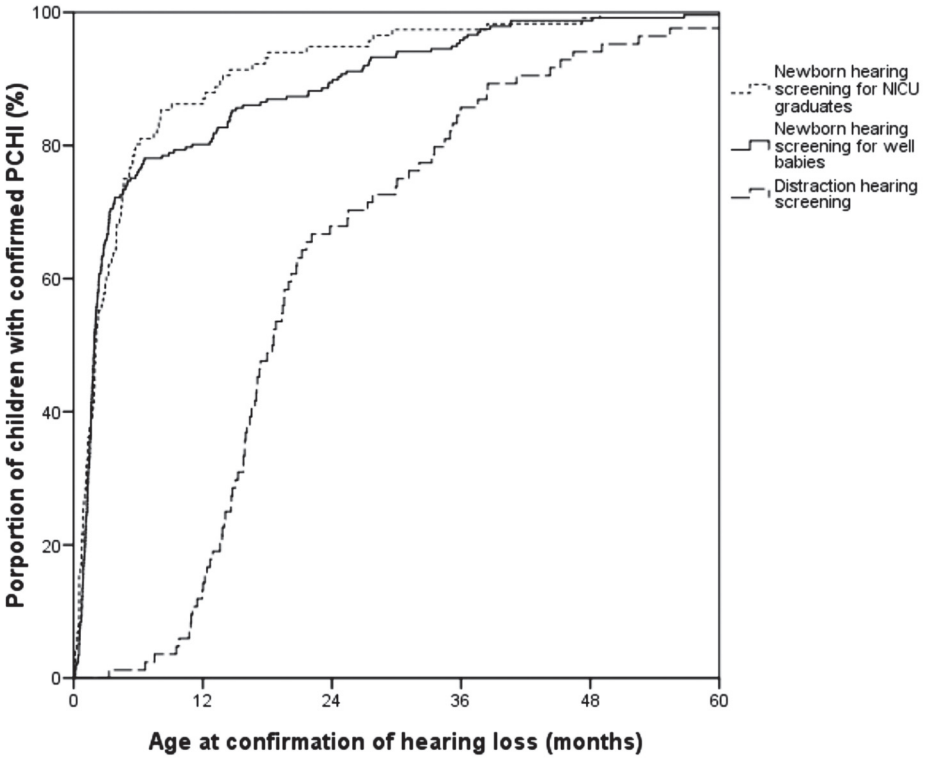
The weighted total prevalence of PCHI following NHS in the Netherlands, based on the 2003-05 study cohort (adjusted for when NHS for well babies would have been offered for the full period) was calculated to be 0.97 per 1000 live newborns (See Table 1.).
The hearing screening programme sensitivity was calculated using the prevalence of PCHI and the proportion of children unidentified by NHS and was based on an annual birth-rate of 195.000. The sensitivity of the current newborn hearing screening programme in our country was 0.83 (187/225) (NHS well babies 0.79 (132/168) and NHS NICU graduates 0.96 (55/57)). All other NHS screening programme characteristics (specificity, positive and negative predictive value) were much better in the current NHS programme than in the formerly used DHS.

CASE-BASED PERSPECTIVE		NUMBER OF CHILDREN WITH CONFIRMED PCHI#	NUMBER OF CHILDREN WITH CONGENITAL, OR NOT CONFIRMED ACQUIRED PCHI##	NUMBER OF CHILDREN WITH DOCUMENTED NEGATIVE SCREEN AT NHS (EXCLUDING ACQUIRED LOSS)##
2003	NHS- well babies	54	53	10
	NHS- NICU babies	55	54	3
2004	NHS- well babies	103	101	13
	NHS- NICU babies	57	57	2
2005	NHS- well babies	129	125	13
	NHS- NICU babies	59	58	1
Total		457	448	42

Confirmed hearing loss at the age of 3-5 years following NHS for well babies: 0.88 per 1000 or 286/326.697
Confirmed hearing loss at the age of 3-5 years following NHS for NICU graduates: 14.5 per 1000 or 171/11.828

The best estimate based on maximum 10 per 1000. ## Based on 367 complete datasets (hearing screening result and aetiology).

Table 2 The number of children with permanent bilateral hearing impairment at the age of 3-5 years in the 2003-05 cohort.



Note: NHS well babies: 100%= 124 children, NHS NICU graduates: 100%= 243 (total complete NHS cases 367 children) with confirmed PCHI in a screened population of 338.525 children (326.697 well babies and11.828 NICU graduates, adjusted for gestational age at delivery). DHS: 100%= 85 children (complete cases) with confirmed PCHI in a screened population of 229.215 children.

Figure 1 Proportion of children with confirmed PCHI by age.

The separate programme characteristics of the hearing screening for NICU graduates, running longer than the NHS for well babies, were better than those for the programme for well babies (Table 3). At any given age until follow-up, the proportion of children with confirmed PCHI was larger for NHS NICU graduates and NHS well babies than for children in DHS (Figure 1.).

The median age at confirmation of PCHI (not corrected for gestational age at delivery) following a negative screen for hearing loss at NHS was 25.7 months for well babies and 17.6 months for NICU graduates. The majority of children unidentified by NHS presented at an Audiology Center for diagnostic evaluation before the age of 3 years (75%) and suffered at that time, from

	PCHI AT 5 YEARS	NO PCHI AT 5 YEARS	TOTAL	PPV/NPV
CURRENT NEWBORN HEARING SCREENING PROGRAM				
Positive screen for hearing loss at NHS and bilateral hearing loss confirmed	187	453	640	0.29
Negative screen for hearing loss at NHS	38	194.322	194.360	0.99
Total	225	194.775	195.000	
Sensitivity/Specificity	0.83	0.998		
NEWBORN HEARING SCREENING WELL BABIES				
Positive screen for hearing loss at NHS and bilateral hearing loss confirmed	132	416	548	0.24*
Negative screen for hearing loss at NHS	36	190.473	190.509	0.99
Total	168	190.889	191.057	
Sensitivity/Specificity	0.79	0.998		
NEWBORN HEARING SCREENING NICU GRADUATES				
Positive screen for hearing loss at NHS and bilateral hearing loss confirmed	55	37	92	0.60**
Negative screen for hearing loss at NHS	2	3849	3851	0.99
Total	57	3886	3943	
Sensitivity/Specificity	0.96	0.990		
DISTRACTION HEARING SCREENING				
Positive screen for hearing loss at DHS and bilateral hearing loss confirmed	154	95966	9.750	0.02***
Negative screen for hearing loss at DHS	41	185.209	182.250	0.99
Total	195	194.805	195.000	
Sensitivity/Specificity	0.79	0.95		

* PPV for confirmed bilateral hearing loss following (uni or bilateral) refer at NHS.[15] ** PPV for confirmed bilateral hearing loss following (uni or bilateral) refer at NICU hearing screening.[16] ***PPV DHS, national figure.

Table 3 Sensitivity and specificity of hearing screening programs

a moderate hearing loss (40-60dB)(>50%). No major cause of PCHI, present in the perinatal period (hereditary, congenitally or perinatally acquired), was identified in children with a negative screen for hearing loss at NHS in this study (Table 4).

To check whether most children with PCHI of congenital or not confirmed acquired cause, would have presented for diagnostic investigation by the age of 4, this study included children with PCHI aged up to 5 years. Of all children initially unidentified (n=45), two children (born in 2003) presented at an Audiology Center for diagnostic investigations because of parental concern when older than 48 months of age.

		NICU BABIES N=6 (%)	WELL BABIES N=36(%)
Diagnosis	Mean age in months	24.3	26.7
	Median age in months (IQR)	17.6 (14.9-35.4)	25.7 (16.6-35.6)
Degree of hearing loss	Moderate (40-60 dB)	4 (66.7)	19 (52.8)*
	Severe (61-90 dB)	2 (33.3)	11 (30.6)
	Profound (>90 dB)	0	6 (15.7)*
Etiology of hearing loss	Hereditary	2 (33.3)	13 (36.1)*
	Congenital infection	-	2 (5.6)
	Perinatal problems	2 (33.3)	1 (2.8)
	Unknown	2 (33.3)	20 (55.6)*

*Child presented later than 48 months for diagnostic evaluation (n=2).

Table 4 Characteristics of children with a documented negative screen for hearing loss at NHS.

COMMENT

The results of the present study show that with our current NHS programme we are able to identify the majority of children with PCHI of congenital cause known to an AC in childhood (total programme sensitivity 0.83). Of all children screened with NHS per year, the proportion of children with PCHI of congenital, or not confirmed acquired cause, unidentified was relatively small (n=38/195.000). Compared to the previously used DHS, the programme characteristics have greatly improved.

To our knowledge, only one earlier study has been published on hearing screening programme sensitivity to date.[94] In the present study we determined the sensitivity of the programme in a much larger and national study.

To put our results in context: the findings are consistent with previous studies that show that the number of children with PCHI increases during childhood and that a small proportion is unidentified by NHS. Unlike earlier studies the present study was a follow-up of a cohort over time and it showed that the increase in prevalence up to the age of 3-5 years was relatively small. This proportion might possibly increase later in childhood.

Our study has certain limitations. First, the identification of children with PCHI of the 2003-05 study cohort at the ACs in 2008. Although the AC personnel and the researchers did their utmost best to identify all children, some could possibly have been lost due to administrative difficulties and recall bias. This might have led to an underestimation of the number of children unidentified by NHS. However, it seems unlikely that an extra 92 children (necessary to increase the prevalence

found following NHS by 34% during the first 5 years of life as reported in some studies [1;2;54]) could have existed, without being identified with PCHI. Important factors contributing to the lower than expected prevalence could be the national immunization programme for children, the screening for irregular erythrocyte antibodies and Human Immunodeficiency Virus in pregnant women and also the low prevalence of congenital CMV infection in our country. Next to health-care differences between countries studied, methodological differences could have contributed to the differences found in the prevalence increment (personal nationwide cross-sectional identification versus questionnaire ascertainment). Secondly, we were only able to quantify acquired hearing loss which was documented. We assume there may be a small proportion of children suffering from an acquired hearing loss which we have not recognized as such. This may have caused an overestimation of children with PCHI of congenital cause. Thirdly, in this study 2 children were identified later than the age of 48 months. It is not impossible that after closure of data-collection very late onset hearing loss remained undetected or appeared (especially in a small number of children born in 2004 and 2005). It is not likely that the proportion of children with PCHI of congenital cause who were left unidentified by NHS and presented later than 48 months of age, is large. The extensive network of youth health care organisations monitoring development would probably have detected and referred these children. Finally, the data suggest that children with confirmed PCHI following a positive screen for hearing loss at NHS may not always be known to an AC in childhood (n=20 in NHS NICU graduates). There are several possible explanations for this: the severity of hearing loss may have decreased over time (maturation), the degree of hearing loss did not necessitate amplification, the medical condition of the child led to the judgment (parental or professional) that PHCI was relatively less important (e.g. in children with multiple disabilities), the family moved out of the country or the child died before extensive diagnostic investigations could be performed (in NICU-graduates the late mortality is approximately 5%).[95]

A negative screen at NHS does not guarantee normal hearing in childhood. We found however, that this problem was not as large as might have been feared. It is unknown if all the children initially unidentified suffered from late onset or progressive hearing losses or that there was a false negative screening result (i.e. the child had a negative screen for hearing loss, even though hearing loss was present at the time).[96] This study showed that the severity of hearing loss at diagnostic evaluation in these children was most often of moderate degree, suggesting that the hearing loss at the time of NHS might have been just about, or just under, the detection threshold used in NHS (40dB). Even though later than the aim of NHS programmes, amplification and habilitation before school-age were possible in the majority of these children.

If the aim is to identify all children with PCHI by NHS, the detection level for hearing loss should be decreased. This would however result in an increased workload and financial costs and

would increase the proportion of children with false positive screening results. The results of the present study (the programme sensitivity and the characteristics of children unidentified with PCHI after NHS), do not support the initiation of audiological screening in childhood (later than NHS).

CONCLUSION

In conclusion, this nationwide follow-up study shows that the sensitivity of the current NHS programme in detecting children with PCHI is high and difficult to improve even further. The findings of the present study, in which data were collected during the implementation period of NHS, emphasize the need that careful attention should be paid, even following a negative screen at NHS, to the child's auditory capacities. It is important to be aware that PCHI may become manifest later than the neonatal period in some children and that some children acquire hearing impairment in childhood.

ACKNOWLEDGEMENTS

We thank all children and parents for participation in the DECIBEL-study.