Reliability and effectiveness of screening for hearing loss in high risk neonates


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Reliability and effectiveness of screening for hearing loss in high risk neonates

R J McClelland, D R Watson, V Lawless, H G Houston, D Adams

Abstract
Objective—To establish the reliability and effectiveness of screening for hearing loss by brainstem auditory evoked potential testing in high risk neonates.

Design—Seven year investigation of newborn babies admitted to a special care baby unit and monitored through a regional children’s audiology unit.

Setting—Special care baby unit and children’s audiology department, Belfast.

Subjects—405 neonates admitted to the baby unit, during 1 October 1982 to 31 March 1987.

Main outcome measures—Presence of hearing impairment, type and severity of hearing impairment, mortality.

Results—85 children failed the screening test, 62 of whom were followed up. Five children had severe bilateral sensorineural impairment and 12 had conductive impairment requiring surgical intervention. A further 18 had severe neurological disorder detected. The sensitivity of screening was 100% and specificity was 88%. If the procedure was introduced into routine clinical practice the mean age at diagnosis for all children with severe perinatal hearing impairment would be 11 (median 1) months. The mean age at diagnosis with the health visitor screening service was 23 (19) months (difference 10 months, 95% confidence interval 6 to 16 months; p<0.0001).

Conclusion—Screening for hearing loss in high risk neonates is highly reliable and cost effective. It also provides valuable neurophysiological information. Routine testing of these infants would result in over half of all children with severe bilateral perinatal sensorineural hearing impairment being identified by 2 months of age. This would make an important contribution to the habilitation of this socially, emotionally, and educationally vulnerable group.

Introduction
Severe prelingual hearing impairment has important consequences for language acquisition, communication, and cognitive, social, and emotional development. Indeed evidence is increasing that even moderate hearing loss in very young children can be detrimental. It is now generally accepted that appropriate remedial measures should be implemented at the earliest possible age and to this end screening for hearing loss in preschool children has been implemented in most health districts in the United Kingdom. The mainstay of current programmes is distraction testing carried out at 7-9 months of age. However, serious doubts have been raised regarding the efficacy and cost effectiveness of such programmes, and health visitor screening has been suggested to be inefficient and costly and to result in considerable delays in diagnosis. Attention has increasingly turned towards developing methods for the early and accurate evaluation of high risk infants.

In view of the uncertainties regarding the relative merits of these two approaches we began a seven year study in 1982. The study had two related aims. The first was to assess the validity and reliability of a screening programme for high risk neonates. The second was to compare the relative success of this programme with existing services and to assess the potential for improving the service by screening children at risk. The long term follow up in our study was made possible by the presence of a single regional children’s audiology unit, to which all children with suspected hearing impairment were routinely referred (fig 1). Identification of false negative test results was also facilitated by the presence of two community screening tests for all children, one at 7 months and one at 3 years of age, and by routine pure tone audiometry at entry to school.

Subjects and methods
The evaluation was based on 405 neonates admitted to the special care baby unit of the Royal Maternity Hospital, Belfast, during 1 October 1982 to 31 March 1987. A careful assessment was made of the following risk factors: familial deafness, rubella infection during pregnancy, birth weight <1500 g, congenital malformations of the ear, apnoea, asphyxia (Apgar score <7 at 5 minutes), respiratory difficulty (requiring respiratory support or more than respiratory oxygen), sepsis, hyperbilirubinaemia (requiring phototherapy or exchange transfusion), and exchange transfusion. The auditory brainstem evoked potential testing was usually carried out after any acute conditions had settled; the modal gestational age was 37 (range 28-42) weeks. Neonates were investigated in their bassinet or incubator. Surface electrodes (silver/silver chloride) were attached to the vertex and both mastoid processes (impedance <5 kΩ). The ear canals were inspected to ensure they were clear. Sound stimuli were generated by a Disa audiometer (Disa Elektronik, Denmark) using 80 μs pulses of alternating polarity delivered monaurally at a rate of 16 Hz through a TDH 39
Sensitivity and specificity of evoked potential screening for serious neurological or hearing impairment

<table>
<thead>
<tr>
<th>Clinical status</th>
<th>Normal or Screen screening Severe minor result impairment impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fail</td>
<td>36 44</td>
</tr>
<tr>
<td>Pass</td>
<td>0 319*</td>
</tr>
<tr>
<td>Sensitivity= 100%, specificity= 88%.</td>
<td></td>
</tr>
</tbody>
</table>

*One child who died of systemic causes was excluded.

earphone (Instrument Systems, New York) at two test levels, 30 dB normal hearing level (nHL) and 65 dBnHL.

Electrical activity recorded from vertex and ipsilateral mastoid was amplified ×50,000 and analogue filtered with a band width of 30-3000 Hz by using a Grass Model 12 amplifier (Grass Instrument Company, Quincy, Massachusetts). Averages were formed from 2048 individual responses with a purpose built portable microcomputer. The criterion for passing the test was a definite response at 30 dBnHL in both ears.

Any child who failed the initial screening test was referred to the regional audiology unit, where all who survived were independently assessed and followed up for a minimum of five years (mean follow up seven years). Follow up investigation included detailed otoscopic examination, behavioural audiometry for younger children and pure tone audiometry for older children, acoustic impedance testing, and evoked potential audiometry, as necessary. Based on the results of follow up children were assigned to one of the following five categories: normal hearing, conductive loss, sensorineural loss (including mixed conductive and sensorineural loss), did not survive, and lost to follow up.

In addition most children who passed the initial screening were reviewed at the end of the study through the clinical medical officer register. This provided a record of the results of health visitor screening at 9 months, screening at 3 years, and pure tone audiometry during the first year of school.

We also analysed data on all children born during the study period and referred to the regional audiology unit. These included any child who had passed the screening test and who was later suspected of having hearing impairment. In addition to audiological assessment, details of perinatal risk factors were sought.

Analysis of statistical significance was based on either 2 or Mann Whitney U tests. The 95% confidence interval was calculated in all cases. The study was approved by the ethics committee of the faculty of medicine, Queen’s University.

Results

INITIAL SCREENING

A total of 405 neonates were screened. Of these, 85 failed the test and 30 had no responses at 65 dBnHL in at least one ear (major impairment).

All perinatal risk factors for deafness were positively associated with a raised auditory threshold, largest correlations being obtained with Apgar score and estimated maturity at birth. Fifty two neonates had Apgar scores of 6 or less at 5 minutes after birth. Twelve (23%) of the 52 had major impairment compared with only 16 (5%) of the 342 neonates with Apgar scores greater than 6 (difference in means 18%, 95% confidence interval 7% to 30%; p<0.001). Apgar scores were not available for 11 infants. Eighty five neonates had gestational ages less than 31 weeks. Of these, 15 (18%) had major impairment in one or both ears compared with 15 (5%) of the 320 neonates with gestational ages of 31 weeks or greater (difference in means 13%, 9% to 17%; p<0.001).

FOLLOW UP EVALUATION

None of the children who passed the screening test had been referred to the regional audiology unit with suspected congenital hearing impairment at the end of study. Two hundred and thirty three (73%) children were systematically evaluated through the clinical medical officer service. All of these were confirmed as having no perinatal hearing impairment.

Five children who failed the screening test were lost to follow up due to change of address. Of 19 neonates who died (15 before discharge from hospital), 18 failed the screening test. The child who passed the screening test subsequently died from systemic causes and without any evidence of neurological or audiological impairment. The principal aetiological factors in the other children were severe birth asphyxia with central nervous system signs (eight children), major congenital abnormalities (seven), and very low birth weight (<1500 g) (three).

Sixty two children who failed the screening test received follow up audiological evaluation. Hearing impairment was confirmed in 32 (8% of 405 screened, 5% to 11%). Twenty five children had conductive impairment, 12 of whom required surgical intervention. The remaining children had sensorineural impairment, including one child with a unilateral non-functioning ear. Five children required hearing aids (best ear threshold: mean 76 (range 60-90 dB). One child failed to attend after the initial follow up.

Twenty seven children passed the initial review and three children were considered to have normal hearing at second review. The sensitivity of the test procedure (number of children with impairment correctly identified) for the 30 dB stimulus was 100%.

The sensitivity for serious impairment was also calculated. The test correctly identified 18 children who had severe neurological and possibly also audiological disorder. Together with the 18 children with sensorineural or conductive deafness requiring intervention described above, a total of 36 (9% of 405) children had severe neurological or audiological impairment (6% to 12%). The corresponding sensitivity and specificity respectively were 100% and 88% (table).

Response to the high level stimulus (65 dB) alone gave a sensitivity of 61% and a specificity of 98% for severe neurological or audiological impairment. Of 14 neonates who had definite responses at screening, four subsequently died and 10 had severe sensorineural or conductive losses requiring intervention.

COMPARISON WITH EXISTING SCREENING METHODS

From the register of all children with suspected hearing impairment in the regional audiology unit we identified a total of 119 children with severe congenital bilateral sensorineural impairment born during the study period. From the registrar general (Northern Ireland) reports on live births, the estimated incidence of severe perinatal hearing impairment for the period was 0-96/1000 live births. Figure 2 compares the frequency distribution of age at diagnosis in children in the screening study, in children at risk outside the study group who were referred to the regional audiology unit, and in those referred by conventional

![Image](https://example.com/image.png)

**FIG 2—Frequency distributions of age at diagnosis of sensorineural impairment for four methods of detection: neonatal screening in special care baby unit, screening by health visitor, referral of children at risk to regional centre for auditory brainstem evoked potential testing, and other (children not detected or not screened by health visitor). Mean age at diagnosis indicated by ▫**
routes, usually after health visitor screening. Progressive
delays in age at diagnosis were observed. The
largest delays were experienced by those children who
did not receive any screening and whose referral was
initiated by parental suspicion, the mean age at diag-
nosis being 24 (median 19) months. However, even
with health visitor screening mean age at diagnosis was
21 (13) months. Children known to be at risk at the
time of birth but who were admitted to special care
units not participating in the study experienced some
delay due to the usual process of referral to the regional
centre for audiological assessment. Their mean age at
diagnosis was 5 (5) months compared with 1 (1) month
in the study group.

![Chart](https://example.com/chart.jpg)

FIG 3—Predicted age at diagnosis in 119 children with sensorineural
impairment if those at risk had been routinely screened at birth and
actual age at diagnosis with current procedures. Mean age at diagnosis
indicated by ▼

From the histories of the 119 neonates we deter-
mained that definite risk factors were known at birth in
64 (54%) infants. But only 21 neonates, including the
five subjects in our neonatal study, were actually
identified at birth. The remaining 43 were referred by the
more conventional routes. Figure 3 shows what the
substitution of age at diagnosis in the 119 children
would have been if all 64 children at risk had been
given early evoked potential audiology testing (optimal
screening) and compares this with the actual distri-
bution for those children who did not receive auditory
brainstem evoked potential testing. The mean age at
diagnosis by conventional procedures was 23 (19)
months. The corresponding mean age at diagnosis for a
service which included electrophysiologic investiga-
tion of all children at risk would be 11 (1) months
(mean difference 10 months 95% confidence interval 6
to 16 months; p<0.0001).

Discussion

The aim of our study was to assess the reliability
and cost effectiveness of auditory brainstem evoked
potential testing in a high risk group of neonates.
The prognostic validity of auditory brainstem evoked
potential testing can be assessed only through careful
and lengthy follow up, but in most studies the follow
up rates are relatively low. In the studies reviewed by
Murray et al follow up data were available on only
13%; and in those reviewed by Jacobson and Hyde
the average retest rate for children who failed the test
was only 60%. In our study outcome information was
obtained on all but five of the children who failed the
initial screening test (94%). In addition, a careful
record has been kept of all children born during the
study period who required audiological assessment.
This provided a unique opportunity to assess the
sensitivity of the method. At the end of 1990, when
the youngest child was 4 years old, none who passed
the initial screening test had been referred with
suspected or confirmed perinatal hearing impairment,
giving a sensitivity of 100%.

Cost Effectiveness

A second issue is cost effectiveness. Several studies
have shown that the positive predictive value of
auditory screening increases if the test procedure is
delayed until infants are 6 months old. This has
implications for the efficiency and cost effectiveness of
early screening. We found a positive predictive value of
29-4%—that is, only one child in four who failed the
screening test had confirmed evidence of hearing
impairment requiring intervention. From the narrow
perspective of audiological screening the testing of
children who subsequently died could have been
avoided by delayed testing. On the other hand, the test
correctly identified all children at serious risk of either
neurological or audiological impairment giving a posi-
tive predictive value of 45%. The highest proportions
of children failing the test were observed among very
low birth weight neonates and those whose Apagar score
at 5 minutes was less than 6. Though targeting such
very high risk groups would improve the positive
predictive value, this would be at the cost of failing to
identify several children with hearing impairment.

The issue of cost effectiveness requires considera-
tion of factors other than the test procedure. There are
organisational costs associated with bringing infants
back to the clinic for testing. We had considerable
difficulty in bringing 12 children (15% of those who
failed the test) back for follow up investigation. The
reasons for these difficulties were lack of parental
understanding, transport costs, inability to get babys-
sitters, and ill health among other members of the
family. These difficulties were confined to families
from the lower socioeconomic groups, among whom a
low uptake of health care services is well recognised.
Even greater difficulties and costs probably would have
occurred with many more children had the initial
screening been delayed until 6 months of age.

The cost to the child and the family of delay in
diagnosis must also be considered. The neonatal
nursery provides a unique opportunity to assess all
children at risk. Once newborn babies leave hospital
considerable delays generally occur before hearing
impairment is finally diagnosed. The efficiency of high risk hearing screening in
neonates can be defined as the number of children
correctly diagnosed as a proportion of the total tested.
In the present study the efficiency for severe sensori-
neural impairment was 1 3%. For all forms of hearing
impairment requiring intervention this rises to 4-5%
The cost of such a service can also be estimated. From
our experience a skilled technician can investigate and
report on five children a day or 1100 a year (assuming
44 working weeks). With an efficiency of 4-5%,
50 children requiring remedial treatment would be
correctly identified. Our estimated cost of technician
time and equipment is £15 000 a year. This gives an
estimated cost of £300 for each child who has hearing
impairment correctly diagnosed. The efficiency of
auditory brainstem evoked potential screening can be
further improved by using automated methods. In any
cost-benefit analysis the reassurance given to
parents of babies at risk who pass the test must also be
considered.

Other Screening Methods

Three objective methods for audiological screening are
currently available: auditory brainstem evoked
potential, the auditory response, and oto-
acoustic emissions. Although we cannot comment on
the relative merits of these three procedures, it should
be noted that auditory brainstem evoked potential is
the only test that provides information on the integrity
of much of the central auditory pathway. This informa-
tion is of interest for audiological and neurological
evaluation. Many children in special care baby units
require auditory brainstem evoked potential testing for neurological investigation in itself. If auditory brainstem evoked potential testing is done then two test intensities should be used, 30 dB for auditory screening and a high level stimulus to evaluate the morphology and latencies of the evoked potential components.

CONCLUSIONS
We recommend that all infants in special care baby units are systematically screened. From our observations of the regional register, children with a family history of deafness, congenital and perinatal infection, or malformations of the head or neck should also be screened at birth. If all such children were screened routinely at birth our findings suggest that over half the children with severe congenital sensorineural impairment could be identified at birth. In addition more children with conductive impairment who require treatment would also be detected. Of course a screening service in isolation is not sufficient. It must be linked to the full continuum of services embracing diagnostic evaluation, habilitation, family counselling, and follow up.

This study was supported by grants from the Department of Health and Social Security (Northern Ireland), Birthright, and Peel Medical Trust. Professor Mark Haggard, University of Nottingham, provided valuable criticism of an earlier draft of this paper. We thank the nursing and medical staff of the Royal Maternity Hospital and the Provinces clinical medical officers.

Comparison of female to male and male to female transmission of HIV in 563 stable couples

European Study Group on Heterosexual Transmission of HIV

Abstract

Objective—To identify risk factors for heterosexual transmission of HIV and to compare the efficiency of male to female and female to male transmission.

Design—Cohort study of heterosexual couples. Regular partners of HIV infected subjects were tested and both members of the couples interviewed every six months. HIV prevalence in partners was analysed according to the characteristics of the couples.

Setting—Nine European countries.

Subjects—563 couples comprising 156 female index patients with their 159 male partners and 400 male index patients with their 404 female partners. Partners reporting risk factors other than sexual contacts with the index patient were excluded.

Main outcome measures—HIV infection in partners and high risk sexual behaviour.

Results—Overall, 19 (12%) male partners and 82 (20%) female partners were infected with HIV, suggesting that male to female transmission is 1.9 (95% confidence interval 1.1 to 3.3) times more effective than female to male transmission. An advanced stage of HIV infection in the index patient (odds ratio 17.6; 4.9 to 62.7) and sexual contacts during menses (3.4; 1.0 to 11.1) increased the risk of female to male transmission and stage of infection (2.7; 1.5 to 4.9) and an oral sex (5.1; 2.9 to 8.9), and age of the female partner (3.9; 1.2 to 13.0) for age >45 years) increased the risk of male to female transmission. None of the 24 partners who had used condoms systematically since the first sexual contact was infected.

Conclusions—Several factors which potentiate the risk of transmission through unprotected vaginal intercourse have been identified. Knowledge of these factors could be helpful for counselling partners infected with HIV and their sexual partners.

Introduction
Several studies have examined the risk of sexual transmission of HIV from infected men to their female partners.1,2 HIV prevalence among female partners of infected men ranges from 15% to 30% in most studies from Europe and the United States. In addition to unprotected vaginal intercourse anal sex and advanced clinical or immunological stage of HIV infection in men have been shown to significantly increase the risk of transmission.

Since many more men are infected with HIV than women in most developed countries, transmission from infected women to their male partners has been poorly studied. Even in regions where HIV is predominantly acquired through heterosexual contact few data are available. Only one large study on clients of prostitutes has been published.3

We present the results of a European multicentre study, the aims of which are to measure the risk of and identify the risk factors for heterosexual transmission; to compare the relative efficiency of male to female and female to male transmission; and to assess the effectiveness of counselling safer sex through the prospective follow up of couples. The preliminary results on male to female transmission of HIV have been published.4

This paper focuses on the analysis of the risk of female transmission.

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