Six year effectiveness of a population based two tier infant hearing screening programme

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Aims: To determine whether a two tier universal infant hearing screening programme (population based risk factor ascertainment and universal distraction testing) lowered median age of diagnosis of bilateral congenital hearing impairment (CHI) >40 dB HL in Victoria, Australia.

Methods: Comparison of whole population birth cohorts pre and post introduction of the Victorian Infant Hearing Screening Program (VIHSP). All babies surviving the neonatal period born in Victoria in 1989 (pre-VIHSP) and 1993 (post-VIHSP) were studied. (1) Pre-1992: distraction test at 7–9 months. (2) Post-1992: infants with risk factors for CHI referred for auditory brain stem evoked response (ABR) assessment; all others screened by modified distraction test at 7–9 months.

Results: Of the 1989 cohort (n = 63 454), 1.65/1000 were fitted with hearing aids for CHI by end 1995, compared with 2.09/1000 of the 1993 cohort (n = 64 116) by end 1999. Of these, 79 cases from the 1989 cohort (1.24/1000) and 72 cases from the 1993 cohort (1.12/1000) had CHI >40 dB HL. Median age at diagnosis of CHI >40 dB HL for the 1989 birth cohort was 20.3 months, and for the 1993 cohort was 14.2 months. Median age at diagnosis fell significantly for severe CHI but not for moderate or profound CHI. Significantly more babies with CHI >40 dB HL were diagnosed by 6 months of age in 1993 than in 1989 (21.7% v 6.3%). Compared to the six years pre-VIHSP, numbers aided by six months were consistently higher in the six years post-VIHSP (1.05 per 100 000 births versus 13.4 per 100 000 births per year).

Conclusions: VIHSP resulted in very early diagnosis for more infants and lowered median age of diagnosis of severe CHI. However, overall results were disappointing.

The late diagnosis of bilateral congenital hearing impairment (CHI) sufficient to require hearing aid fitting (approximately 40 dB HL) remains an important public health issue. CHI has a relatively high incidence of approximately 1–2/1000 live births, and early detection and remediation prior to 6 months of age appears to substantially decrease the resulting speech and language impairments.

Recent advances in techniques suitable for neonatal screening have resulted in the implementation of universal neonatal screening (UNHS) programmes. The Wessex Study, the only existing controlled trial comparing detection rates in hospitals with and without screening, showed that high coverage can be achieved within institutions, resulting in much earlier detection of many neonates with CHI. False positive rates are widely judged to have fallen to acceptable levels, and UNHS appears to be very acceptable to parents and professionals. However, overall success of UNHS programmes will ultimately be judged on population detection rates and long term outcomes, not detection rates in neonatal institutions. Such studies are not yet available.

Some continue to argue that, until such evidence is provided, hearing screening should be limited to those with defined risk factors. There is general agreement that it is worthwhile to at least screen newborns with risk factors for CHI, who include up to approximately 60% of the children with CHI. However, screening only children with risk factors could by definition identify no more than 60% of CHI, and in fact has not approached this level because accurate identification has in practice proved to be a challenging task.

In yet another approach, some countries with strong community nursing services, such as the UK and Australia, have for many years used universal distraction testing programmes at the age of 7–9 months. Although good results are possible with this test in ideal circumstances, no group has reported programme success on a population basis.

A combination of the latter two approaches (neonatal screening for infants with a risk factor, followed by behavioural screening for all other infants later in the first year of life) has been suggested as one possible system to detect the majority of children by the age of 12 months. Based on the findings of a state government review committee before UNHS gained its present popularity, this approach was adopted in 1992 for trial in the state of Victoria, Australia (population 4.5 million). The Victorian Infant Hearing Screening Program (VIHSP) aimed to reduce median age at diagnosis, aid fitting, and early intervention entry for children with CHI throughout the state, its target being to diagnose and fit with aids all infants with bilateral CHI >40 dB HL by the age of 12 months.

We report outcomes for two complete birth cohorts born before and after the implementation of VIHSP, each followed to the age of 6 years. To our knowledge, this is the first population study to report long term follow up of two different strategies for screening for bilateral CHI >40 dB HL.

METHODS

Screening methods

Pre-VIHSP

Prior to VIHSP, risk factors were not routinely ascertained and babies with known risk factors were not routinely referred for audiological assessment, although some auditory brain stem evoked response (ABR) testing facilities did exist in the state. Maternal and child health nurses screened most infants with

Abbreviations: ABR, auditory brain stem evoked response; CHI, congenital hearing impairment; UNHS, universal neonatal hearing screening; VIHSP, Victorian Infant Hearing Screening Program
the distraction test at 7–9 months. However, the screen used did not have strict pass/fail criteria and referral mechanisms for screen failures varied considerably across the state.

Post-VIHSP
Paediatricians, neonatal intensive care unit staff, and maternal and child health nurses were responsible for referral of babies with one or more of eight risk factors of CHI (table 1). Babies identified as “at risk” below the age of 3 months were referred to an outpatient appointment with their regional audiologist for ABR testing, since at that time automated ABR was not widely available. A “pass” was defined as a reliable response at 40 dB HL (average hearing across 500, 1000, and 2000 Hz) in either ear. Milder impairments were not sought. While some audiologists used ABR only to “screen” referred babies to rule out a hearing loss of >40 dB HL, others carried out full ABR testing on all referred babies. Babies with a risk factor identified beyond this age were assessed using the method judged most appropriate for each baby by the audiologist.

All other babies underwent a revised distraction test in maternal and child health centres at 7–9 months of age. Based on the Ewing Distraction Test, this behavioural screen involved presentation of a low frequency voice sound and a high frequency sound using a Manchester rattle (Ewing Foundation, UK) at an intensity less than 40 dB (A). The new test had more stringent pass/fail criteria than previously and a standardised protocol for delivery. Some of the key points of the new protocol were that only the distractor (maternal and child health nurse) decided whether the infant had responded. A “pass” response was defined as a direct localising response to the noisemaker. A “fail” response was defined as no response within 3–4 seconds of stimulus onset, movement to the wrong side, or any other movement. Presentation of stimuli was limited to 12 per test. Eight pass responses consisting of correct responses to the rattle and the voice at least twice on each side, with no more than four fails, constituted a pass on the screen. Five fail responses or failure to give correct responses to the rattle and the voice twice on each side constituted a fail on the screen. The distraction test screen was repeated 4–6 weeks after the first screen if pass criteria were not met, to reduce numbers of false positives caused by transient conductive losses.

Training and supervision
The State Government endorsed VIHSP prior to implementation, as did the professional groups representing all 575 maternal and child health nurses, 120 paediatricians, and 50 audiology assessment centres across Victoria. Vigilance about early signs of CHI and rapid response to any parental concerns were emphasised. All parents received literature at the time of screening, supplemented by information in the parent held Victorian Child Health Record. Programme information was disseminated through direct mailings, regional workshops, professional journals, and articles in the lay press. A training video demonstrating the modified distraction test was distributed to all maternal and child health nurses, all of whom underwent training in its administration. Two senior audiologists were employed to visit maternal and child health nurses for further training and supervision, and a quality assurance programme to monitor performance of the distraction test was implemented. At a workshop for audiologists, over 90% correctly interpreted a variety of standard ABR traces and inter-rater reliability was over 90%. Regular feedback about VIHSP performance was given to all participating professionals by means of a newsletter, maternal and child health nurse regional workshops and local paediatric conferences.

Referral and data collection
Referral forms were developed for infants at risk and for those failing two distraction tests. These forms also served as data collection forms, recording the infant’s sex, gestation, birth date, referral date and source, and reason for referral. Parent consent and audiology assessment results from the first appointment were also recorded.

Additional information was collected from parents of children with CHI who were fitted with hearing aids from both the 1989 (pre-VIHSP) and 1993 (post-VIHSP) birth cohorts. The 1989 cohort was chosen as this was the most recent year in which all children would have turned 3 years by the time of implementation of VIHSP, and hence would be minimally affected by its activities which are largely directed at infants in the first year of life.

Australian Hearing provides and fits all hearing aids for Australian children and has for many years maintained a population database recording age of first appointment (used in the present study as a proxy measure of age at diagnosis) and age at hearing aid fitting for these children. Australian Hearing provided non-identifying information for children with CHI from the two birth cohorts (dates of birth, hearing aid fitting, aetiology, degree of loss), enabling assessment of the impact of VIHSP at population level rather than analysing such milestones only for those children successfully screened. The state’s Department of Human Services provided information on numbers of cases screened by distraction test and referrals made by maternal and child health nurses to audiologists.

Use of the Australian Hearing database meant that only those children with bilateral significant hearing impairment requiring the fitting of hearing aids were regarded as “true positives” from the hearing screening programme. Acquired conductive hearing impairments, unilateral sensorineural losses, and very mild bilateral losses which did not require aiding but were identified as a result of screening were not included in the assessment of outcomes.

Statistical methods
Analyses were performed using the Statistical Package for the Social Sciences (SPSS). Descriptive statistics were generated for each birth cohort. Medians were compared using the Mann–Whitney U test, proportions using χ² tests or Fisher’s exact test as appropriate, and differences in means using unpaired t tests. Positive predictive values were calculated for risk factor and distraction screening respectively for the 1993 birth cohort.

RESULTS
Of the 1989 birth cohort, 63 454 babies survived the neonatal period; 105 (1.65/1000) were diagnosed with CHI and had
hearing aids fitted by 31 December 1995, for whom 74 (70%) parent questionnaires were returned. In 1993, 64 116 infants survived the neonatal period; 134 (2.09/1000) were diagnosed with CHI and had hearing aids fitted by 31 December 1999, for whom 82 (61%) parent questionnaires were returned. The difference in overall prevalence between the two cohorts approached significance (χ² = 3.22, p = 0.07) because of an increase in the numbers with mild CHI fitted with aids by 6 years, but prevalence of CHI >40 dB HL was similar (1.24/1000 v 1.12/1000, χ² = 0.40, p = 0.53). These figures exclude all cases of hearing impairment where the loss was known to have been acquired, for example, meningitis, head injury, chemotherapy (seven cases in the 1989 and seven in the 1993 birth cohort). Three cases included in the 1993 cohort had evidence of some progressive loss but were retained for analysis as comparable data were not available for the 1989 cohort.

Referral patterns from the 1993 birth cohort

VIHSP referral forms for infants with a risk factor for CHI were received for 2827 infants (4.4% of the birth cohort). Maternal and child health nurses recorded that 49 358 infants (77% of those eligible) had a distraction test at 7–9 months. Department of Human Services figures indicate that 5262 (8% of the birth cohort) were referred to an audiologist after the 7–9 month screen, but VIHSP data were received for only 3299 infants who failed two distraction tests (5% of the 1993 cohort).

Results of screening

Table 2 shows reasons for referral for referred infants born in 1993 for whom full documentation was returned to VIHSP, including 49 of the 134 ultimately diagnosed with CHI. It was known that some cases ultimately aided were identified via VIHSP pathways even though no forms were received, so these numbers are almost certainly an under-representation. Congenital abnormalities of the head and neck and birth asphyxia were the strongest predictors of CHI, whereas the risk factor category generating the largest number of referrals (family history) was a much weaker predictor. As a screening tool, the distraction test performed poorly, with a very high false positive rate. Only 18 of 3299 (0.5%) children failing two distraction tests were subsequently fitted with aids, compared with 31 of 2827 (1.1%) referred for an identified risk factor.

Age at diagnosis, aid fitting, and early intervention entry

Australian Hearing data show that implementation of VIHSP was followed by an immediate and sustained rise in mean number of aids fitted by 6 months of age per year, from 0.01 per 1000 births in the six years prior to VIHSP to 0.13 per 1000 births (p < 0.001) in the six years after VIHSP implementation (fig 1).

Proportions with CHI >40 dB HL fitted with hearing aids by 12 months differed little between the 1989 and 1993 cohorts (24% v 31%, χ² = 1.03, p = 0.31). Infants with a hearing impairment with a known risk factor (identified before or after diagnosis) were 23 times more likely to be diagnosed by 6 months of age than infants without a known risk factor (relative risk = 23.2, 95% CI 3.2 to 169.1, p < 0.001). This pattern was observed across all severity groups. Less than half of the infants with profound losses who did not have an identified risk factor were diagnosed by 12 months of age (table 3).

Figure 2 shows cumulative ages of diagnosis for the 1989 and 1993 birth cohorts by severity of hearing impairment; table 4 shows changes in median age of diagnosis and in proportions diagnosed by age 6 months. As expected, since VIHSP targeted only CHI >40 dB HL, the proportion of mild CHI (21–40 dB HL) diagnosed by 6 months of age changed little (3.8% pre-VIHSP v 4.1% post-VIHSP, p = 1.0, Fisher’s exact test). Overall the median age at which CHI >40 dB HL was diagnosed tended to be lower in the 1993 than the 1989 cohort, though this fall was statistically significant only for severe (not moderate or profound) hearing loss. Significantly more babies with CHI >40 dB HL were diagnosed by 6 months of age in 1993 than in 1989 (21.7% v 6.3%, χ² = 7.48, p < 0.01), with the biggest difference again occurring for the severe group (35.0% v 0%, p < 0.01, Fisher’s exact test). In 1993 only 11 children were born with profound CHI, fewer than in 1989 (n = 26) or any other year in the past two decades, which may have weakened our ability to detect true changes in this group.

![Figure 1](http://adc.bmj.com/) Number of infants fitted with hearing aids by 6 months of age pre- and post-VIHSP.
Table 3  Proportions diagnosed with bilateral hearing loss by 6 and/or 12 months from the 1993 cohort by presence of risk factor

<table>
<thead>
<tr>
<th>Babies with known risk factors (n=49)</th>
<th>Babies without known risk factors* (n=71)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosed by 6 mth</td>
<td>Diagnosed by 6 mth</td>
</tr>
<tr>
<td>Mild</td>
<td>1/18 (5.6%)</td>
</tr>
<tr>
<td>Moderate</td>
<td>4/16 (25.0%)</td>
</tr>
<tr>
<td>Severe</td>
<td>7/9 (77.8%)</td>
</tr>
<tr>
<td>Profound</td>
<td>4/6 (66.7%)</td>
</tr>
<tr>
<td>Total</td>
<td>16/49 (32.7%)</td>
</tr>
</tbody>
</table>

*Includes children for whom a risk factor was not present, as well as children for whom no data about risk factors were available.
†Includes two children for whom severity of hearing loss was not known.

Table 5 compares median ages at milestones in the pathway to treatment of moderate or greater CHI between the two cohorts for children whose parents returned detailed questionnaires to VIHSP after diagnosis. There was a general tendency for events to occur at a younger age in the 1993 cohort; however, no median value for any milestone on the pathway to intervention was significantly lower in 1993 than in 1989.

Although not shown in table 5, the median time from first referral for audiological assessment to fitting of hearing aids was 5.8 months for the 1989 cohort and 4.1 months for 1993 cohort (Mann–Whitney U, p = 0.50).

DISCUSSION

Principal findings

This two tiered infant hearing screening programme achieved a modest increase in the number of infants with bilateral CHI >40 dB HL fitted with aids by 6 months and a decrease in median age at diagnosis for children with severe CHI.

Table 4  Median age at diagnosis of CHI and percentage diagnosed by six months with bilateral hearing loss at different severities of impairment; pre-VIHSP (1989) and post-VIHSP (1993) birth cohorts

<table>
<thead>
<tr>
<th>Severity of HI</th>
<th>n</th>
<th>Median age at diagnosis (mth)</th>
<th>% diagnosed by 6 months of age</th>
<th>p value*</th>
<th>Pre-VIHSP</th>
<th>Post-VIHSP</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>26</td>
<td>39.6</td>
<td>3.8</td>
<td>0.07</td>
<td>4.1</td>
<td>0.10†</td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>31</td>
<td>36.4</td>
<td>3.2</td>
<td>0.55</td>
<td>10.5</td>
<td>0.25†</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>22</td>
<td>22.4</td>
<td>0</td>
<td>0.003</td>
<td>30.0</td>
<td>&lt;0.01†</td>
<td></td>
</tr>
<tr>
<td>Profound</td>
<td>26</td>
<td>10.3</td>
<td>15.4</td>
<td>0.29</td>
<td>36.4</td>
<td>0.16†</td>
<td></td>
</tr>
<tr>
<td>Moderate + severe + profound†</td>
<td>79</td>
<td>20.3</td>
<td>6.3</td>
<td>0.18</td>
<td>21.7</td>
<td>&lt;0.01§</td>
<td></td>
</tr>
</tbody>
</table>

*Mann–Whitney U test.
†Mild not included, as not the target of VIHSP.
‡Fisher’s exact test, §Fish’s exact test.
Mild = 21–40 dB HL; moderate = 41–60 dB HL; severe = 61–90 dB HL; profound = >90 dB HL.
original goal of diagnosis and intervention (including hearing aid fitting) by the age of 12 months of all infants with moderate or greater CHI was not met. Even in cases where the screening test gave a true positive result, aids were sometimes not fitted until many months later.

The prevalence of risk factors for hearing impairment quoted in the literature varies between 3.9% and 12% depending on precise definitions and whether risk factors were ascertained before or after diagnosis.** Our yield from risk factor referrals (1.9%) was very similar to the yield for high risk infants screened in the Wessex study (1.86),1 but only 4.3% of our 1993 birth cohort was documented as being referred with a risk factor compared to the 8.1% of screened babies that had a documented risk factor in the Wessex study. Thus under ascertainment of risk factors probably contributed significantly to the low rates of early diagnosis. We do not know how much of this occurred as a result of not asking the questions, not interpreting the answers correctly, or parents being unaware of the risk factor at the time of questioning.

Despite this, ascertainment of risk factors and subsequent referral of these infants for outpatient audiology led to more and earlier diagnoses of CHI than did the modified distraction test. Infants with a risk factor were more likely to be referred for audiology, and earlier diagnoses of CHI than did the modified distraction test. Infants with a risk factor compared to the 8.1% of screened babies that had a documented risk factor in the Wessex study. Thus under ascertainment of risk factors probably contributed significantly to the low rates of early diagnosis. We do not know how much of this occurred as a result of not asking the questions, not interpreting the answers correctly, or parents being unaware of the risk factor at the time of questioning.

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**Strengths and weaknesses of the study**

This study has several important strengths. The study was population based, covering two entire birth cohorts within a large geographically defined community. Reliable, population based information on hearing aid fitting was available, allowing confident estimates to be made of the numbers of children with CHI >40 dB HL who were not detected through the screening system. Many cases could be followed to the point of entry to early intervention, enabling identification of where the successive delays in the pathway to intervention occurred. Finally, outcomes were evaluated to six years, whereas most studies have reported results only for those successfully screened and with follow up limited to at most one to two years. While short follow up is sufficient to calculate the incremental yield of one programme over another, longer follow up allows inclusion in the denominator of the many children not detected by the screening programme (whether false negatives of testing or children who were not screened) and not diagnosed until the preschool years.

Weaknesses of the study included the lack of information about children who passed their screens, and missing data for children who were referred but for whom VIHSP did not receive data. Sophisticated electronic data collection systems to track children through all stages of the screening, referral, and diagnostic processes may now hold solutions to this problem, though this will depend on ease of use, resourcing, and training. All cases of known acquired losses were excluded from all analyses. However, in many studies, some late diagnosed children could have had an acquired hearing loss but, in the absence of earlier hearing tests or clear precipitants, been inadvertently misclassified by Australian Hearing as having a congenital loss. Our prevalence of known acquired and progressive losses in the 1993 cohort (seven acquired and three progressive—that is, 0.16/1000) is similar to that reported elsewhere,7 giving us confidence that any misclassification would be minimal (and should in any case affect both cohorts equally). Finally, over the past two decades age of diagnosis has slowly but steadily fallen even in the absence of new programmes, and this may have accounted for some of the improvements seen between 1989 and 1993.

**Implications and future research**

This two tier hearing screening programme improved the early diagnosis of children with severe hearing impairment and increased aid fitting in early infancy, but overall achieved only...
modest success. Based on current evidence, universal neonatal hearing screening appears likely to be much more successful. However, a population perspective such as that provided in this study is needed to fully show the successes and failures of any programme, incorporating long term follow up of entire birth cohorts and methods for ascertaining cases missed by the screening programme as well as why they were missed. Such reports will also help quantify the true benefits of universal neonatal hearing screening programmes, which currently appear to offer the best hope of early detection and lasting benefit for hearing impaired children.

ACKNOWLEDGEMENTS

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REFERENCES


ARCHIVIST

More on erythromycin and pyloric stenosis

A paper in the *Lancet* in 1999 (354:2101–5) described an increase in infantile hypertrophic pyloric stenosis in babies given prophylactic erythromycin after exposure to whooping cough. Now more data have been reported from a hospital in Indianapolis.

The retrospective cohort study (Barbara E Mahon and colleagues. *Journal of Pediatrics* 2001;139:380–4) included 14 876 infants born between June 1993 and December 1999. Forty-three (0.3%) of these infants developed pyloric stenosis, most of them (29) between 3 and 6 weeks of age. Thirty-six of the infants with pyloric stenosis were boys but erythromycin had been prescribed equally for boys and girls (3.2% and 3.1%). No infant given erythromycin after 2 weeks of age developed pyloric stenosis but 6 of 226 (2.7%) prescribed erythromycin in the first 2 weeks did so. Five of 182 (2.7%) infants prescribed erythromycin in the first week and one of 44 (2.3%) prescribed erythromycin in the second week developed pyloric stenosis (relative risk 10.5 for erythromycin in the first 2 weeks). All of the infants who developed pyloric stenosis had been given erythromycin for 14 days or longer. None of 1243 infants treated with erythromycin eye ointment developed pyloric stenosis. There was a suggestion (not statistically significant) of an increased risk of pyloric stenosis after treatment of the mother with a macrolide antibiotic in the last 10 weeks of pregnancy, especially if the infant was also treated.

Erythromycin given for 14 days or longer to infants up to 2 weeks old increases the risk of pyloric stenosis tenfold.