Risk Factors for Hearing Disorders: 
Epidemiologic Evidence of Change over Time in the UK

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Abstract

The prevalence** of all bilateral sensorineural hearing impairments of at least 40 dB HL was assessed by a retrospective ascertainment study to be about 1.2 children per 1000 births per annum, over the period 1983–1988. This prevalence was highly variable over year and region of the study. For all profound (at least 95 dB HL) impairments, the prevalence was 1 in 2703 children per birth cohort. The contribution of acquired impairments and inter-region transfers after birth was not trivial, being over 25 percent for the profound impairments. The proportion of the bilateral moderately profoundly hearing impaired who might have been in the neonatal intensive care unit, have a family history of childhood deafness, or craniofacial anomaly recognizable prior to maternity discharge was 64 percent. Changes in the relative distribution of the etiology of hearing impairment seem to have taken place since the EC 1969 birth cohort study. These changes should have considerable impact on the planning of services for hearing-impaired children and child health surveillance in general.

Key Words: Children, epidemiology, hearing disorders, prevalence

Several studies have investigated the epidemiology of childhood hearing impairments in different parts of the world. In the UK (Martin et al., 1981; Martin, 1982; Newton, 1985; Peckham, 1986; Davis and Wood, 1992), Europe (Martin et al., 1981; Feinmesser et al., 1982; Parving, 1983, 1984; Kankkunen and Liden, 1982; Dias, 1990), US and Canada (Schein and Delk, 1974; Schildroth, 1986; Davidson et al., 1989), and Australia (Uptold, 1988), different protocols have been used to assess the prevalence and etiology of different aspects of childhood hearing impairments. These data are extremely useful within their own context. The limitations occur mainly when trying to generalize the results obtained in one country or district to another, or when comparing the data over time. One of the main restrictions on the data is that they are usually collected as part of the audit for the service given to the hearing impaired, and are not necessarily designed and collected independently of that service. This is because the

**In the nomenclature of epidemiology, the term “incidence” refers to the number of new cases in the population over a given unit of time. The term “prevalence” refers to the number of cases in the study population who have a particular characteristic at a particular time. The present study population covered a number of birth cohorts, and comprised roughly three incident populations: (1) the congenital hearing impaired who were ascertained by the cutoff for the study; (2) the children who acquired hearing impairment; and (3) the children with progressive impairments that we had also ascertained. Thus, we use the term “prevalence” to indicate how many children in the birth cohorts were hearing impaired by the time the children were 5 years old. This is standardized as the prevalence for a 1-year cohort. It is, in effect, the incidence in a 1-year cohort by the age of 5. Thus, for children with congenital hearing impairment, who are identified in the first year of life, the apparent incidence is the same as the prevalence at 1 year of age.
need to be able to compare data using standard protocols for data collection are not always priorities for the routine service providers. However, with the availability of better information technology such as the Pediatric Audiology Record System (PARS) (Marshall et al, 1993), it is now possible to register uniform data with agreed protocols, at a cost and effort that are acceptable to the user. In addition, an emphasis by purchasers on testing the quality of the services provided means that good information, provided through such databases, has to be available for routine service audit; thus, the real cost of providing uniform data for epidemiologic purposes is falling substantially. The benefits of the good use of database systems are now becoming more readily accessible to routine clinical users (e.g., over 30 percent of health districts in England are using such databases in spring 1995).

The aim of this paper is to assess what, if any, changes there are over birth cohorts (approximately 15 years apart) in the epidemiology of childhood hearing impairments. The three aspects of epidemiology that we will attempt to cover are the prevalence of permanent childhood hearing impairment (Davis and Parving, 1994), the risk groups for childhood hearing impairment, and the presumed etiology clinically attributed to the impairments. The data are restricted to areas of the UK that have collaborated with this project and collected data concerning hearing-impaired children on birth cohorts, rather than by year of referral. The first problem in examining change over time is to have a baseline for the comparisons. This is a very difficult matter, as the protocol and aims of previous cross-sectional studies may not be entirely relevant to our present aims. The major study that comparison is made with, at least for the purpose of illustration of substantial changes, is the UK sample from the European Community (EC) study (Martin et al, 1981) of children born in 1969 and assessed at age 8 years. No comparable cross-sectional data exist for all of the UK in the 1980s, and we are restricted to the areas chosen in this present study. The temporal comparison we wish to make is between children born in 1969 and those born around the mid-1980s.

Even without any comparison with the previous study, the data presented below concerning the demography and etiology of hearing impairment help to inform the assessment of (1) the audiologic health care needs of hearing-impaired children, (2) the relevant audiologic services that should be demanded on behalf of the children, and (3) the proposed quality standards for future services. The main aims are (1) to quantify the size of the hearing-impaired population and characterize it along relevant audiologic and time domains, (2) to try to quantify the risk groups relevant to strategic planning issues such as neonatal screening, (3) to attempt to attribute etiology to a subgroup of the hearing-impaired children, and (4) to document the distribution of referral patterns in terms of age of intervention.

METHOD

Areas of Study

Data were available from three geographically defined administrative health areas in the UK: Nottingham District Health Authority (DHA), Oxford Regional Health Authority (RHA), and Sheffield District Health Authority (DHA). The data from Nottingham and Sheffield were collected by the respective pediatric audiology departments as ongoing projects evaluating neonatal and other pediatric screening procedures. The Oxford data were collected from clinical case notes, as part of the Oxford Regional Register of Early Childhood Impairments project (Johnson and King, 1989). These data, with respect to hearing threshold levels, were checked by the pediatric audiology department at Reading. All three centers used age-appropriate audiometric procedures.

Birth Cohorts

The data reported here are a subset of the data collected, and have been selected according to audiologic and statistical selection criteria described below from the total number of children that we know have been supplied with hearing aids, whether or not they currently

<table>
<thead>
<tr>
<th>Area</th>
<th>Birth Cohorts</th>
<th>Total Births</th>
<th>Total Aided</th>
<th>Number Meeting Study Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nottingham DHA</td>
<td>1983–1988</td>
<td>47,036</td>
<td>142</td>
<td>68</td>
</tr>
<tr>
<td>Oxford RHA</td>
<td>1984–1986</td>
<td>98,082</td>
<td>146</td>
<td>95</td>
</tr>
<tr>
<td>Sheffield DHA</td>
<td>1986–1988</td>
<td>19,811</td>
<td>48</td>
<td>37</td>
</tr>
<tr>
<td>Overall</td>
<td>1983–1988</td>
<td>164,929</td>
<td>336</td>
<td>200</td>
</tr>
</tbody>
</table>

*Excluding perinatal and neonatal mortality.
use them. Table 1 shows the number of children from each district, the birth cohorts considered, and the relevant number of births in each area.

Criteria for Inclusion in the Study

The criteria for inclusion in the ascertainment study were that the hearing impairments were derived from tests using age-appropriate testing procedures (the youngest child at ascertainment was 3 years of age; the median age was 5 years). Only children with bilateral hearing impairment of a sensorineural or mixed type of at least 40 dB HL, averaged over the mid-frequencies 0.5, 1, 2, 4 kHz, were included in the study. Obviously, those children who had not been detected in the areas at the time when the study was done were not included. In the UK, most children with these degrees of hearing impairment should be detected by the public health system by about 5 years of age. The sensitivity of the screens and surveillance procedures are not perfect (i.e., they are less than 100%), and we presume that they vary across the districts (Davis and Wood, 1992). Notification of hearing impairments, particularly in Oxford RHA, where several health districts combined to notify a central register, will also reflect this variability. The prevalence estimates are highly likely, therefore, to be underestimates of true prevalence. To minimize this problem, a statistical analysis was undertaken to examine, for each area, whether there was a significantly different profile of severity of hearing impairment over time. Most recent birth cohort years were excluded until there was no difference in severity profile over birth cohort (p < .05), minimizing the risk of underascertainment as a function of severity. It is usually the mild and moderate hearing impairments that are not discovered until much later than the profound impairment, and this procedure minimizes the chances of that bias.

Table 1 shows that there were 200 children with hearing impairments that met the criteria. Of these children, 21 (10.5%) were not born in the district of residence and transferred into that district at some stage during the following years. An assumption might be that there are an equal number of transfers out balancing the transfers in, unless the reason for transfer in is to get nearer to regional centers of excellence and thus better hearing services! This has been known to happen recently in the provision of cochlear implants, but, over the period of this study, no definite case of the latter was identified.

To put the inclusion criteria into perspective, we note that Davis and Wood (1992) have shown that, for Nottingham DHA, the prevalence of children issued with a hearing aid was about 2 per 1000. About one third (35%) of this total hearing aid group (n = 142) from Nottingham DHA in this study have moderate (40–69 dB HL), 16 percent severe (70–94 dB HL), and 16 percent profound (95–130 dB HL) impairments. Thus, two thirds of hearing aid users might normally be included by the severity and statistically derived cohort criteria.

The data were analyzed and confidence intervals (CIs) obtained using the appropriate logistic models (with binomial errors and logit link function) through the GLIM4 statistical software for generalized linear interactive statistical modelling (Francis et al, 1993).

RESULTS

Prevalence of Hearing Impairment

Table 2 shows the prevalence of hearing impairment averaged over the birth cohorts for each area and for the severity bands defined above. As expected, from the presumed poisson distribution (which is the appropriate statistical distribution for estimating most rare events), there was considerable variation in the number of hearing-impaired children per cohort, making

| Table 2 Prevalence (95% CI) per 1000 Births per Annum of Bilateral Hearing Impairment Averaged over 0.5, 1, 2, and 4 kHz as a Function of Area and Severity |
|-----------------|-----------------|-----------------|-----------------|
| Severity of Hearing Impairments (dB HL) | 40–130 | 70–130 | 95–130 |
| Nottingham DHA | 1.45 (1.14–1.83) | 0.70 (0.50–0.99) | 0.38 (0.24–0.61) |
| Oxford RHA | 0.97 (0.79–1.18) | 0.54 (0.41–0.71) | 0.32 (0.22–0.45) |
| Sheffield DHA | 1.87 (1.35–2.58) | 0.96 (0.61–1.50) | 0.61 (0.34–1.07) |
| Overall | 1.21 (1.06–1.39) | 0.64 (0.53–0.77) | 0.37 (0.29–0.48) |
Table 3 Prevalence (95% CI) per 1000 Births per Annum of Bilateral Hearing Impairment Averaged over 0.5, 1, 2, and 4 kHz as a Function of Severity and Acquisition

<table>
<thead>
<tr>
<th>Acquisation of Hearing Impairment</th>
<th>40-130</th>
<th>95-130</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital resident at birth</td>
<td>0.98 (0.84-1.14)</td>
<td>0.27 (0.20-0.36)</td>
</tr>
<tr>
<td>Transfers included</td>
<td>1.10 (0.95-1.27)</td>
<td>0.30 (0.23-0.40)</td>
</tr>
<tr>
<td>Transfer and acquired impairments included</td>
<td>1.21 (1.06-1.39)</td>
<td>0.37 (0.29-0.48)</td>
</tr>
</tbody>
</table>

yearly comparison meaningless for a birth cohort of a typical health district (3-6000). Taking the data for moderate hearing impairments or worse, there is a significant difference (p < .01) in this prevalence between Oxford RHA and Sheffield DHA and also (p < .05) between Oxford RHA and Nottingham DHA. The overall estimate of 1.21 per 1000 per annum is weighted more towards the Oxford data due to the larger number of children in the birth cohort. A less biased estimate may be to take the Nottingham estimate of prevalence at 1.45 per 1000 per annum, as it is suspected that the Oxford data may have deficiencies in the ascertainment of mild-moderate hearing impairments due to the different nature of the data collection. At least severe and profound hearing impairment severity categories do not differ significantly over area, and the joint estimate of prevalence at 0.64 per 1000 per annum and 0.37 per 1000 per annum, respectively, are probably good first estimates. The estimates given in Table 2 include all children with hearing impairment satisfying the criteria set out above, including all children who have transferred into the district and all acquired and progressive hearing impairments. Table 3 shows the prevalence in terms of congenital hearing impairments for children born in a district, and then including the transfers in and the possibly acquired hearing impairments.

The acquired impairments were 9.5 percent of all the moderate to profound bilateral hearing impairments. Meningitis was the main etiology, accounting for 6.5 percent (CI 4-12%) overall, but almost 16 percent (CI 9-27%) of the profound impairments. For profound hearing impairments, the risk is of the order of 1 in 3748 for congenital impairments who are still resident in their area of birth. Including all impairments now resident in the area, this risk is of the order of 1 in 2703.

For congenital impairments overall, the profound are 27 percent of the selected study group, with the severe being 23 percent and the moderate 50 percent, and this is relatively constant over the areas of the study. However, Oxford has 48 percent moderate, with Nottingham and Sheffield having 54 percent. For all impairments, the profound plus severe increase to 52.5 percent, 56 percent for Oxford, and 49.5 percent for Nottingham and Sheffield. The differences between Oxford and Sheffield are statistically significant.

Characteristics of Hearing-impaired Children

Table 4 shows the distribution of children with congenital hearing impairment with respect to whether they were resident in the neonatal intensive care unit (NICU) and also whether they had a family history of childhood deafness. Overall, 43 percent of the children did not fall into one of these two categories. A further 7 percent of these 43 percent were classified as having a craniofacial abnormality (CFA) noticeable at birth. Thus, a three-category risk factor incorporating NICU status, family history, and CFA might account for up to 64 percent (CI 57-72%) of the hearing-impaired children in this study.

Table 4 Percentage of Children in Three Groups: NICU, Family History, and Other for Each Severity of the Hearing Impairment

<table>
<thead>
<tr>
<th>Severity of Hearing Impairment</th>
<th>Family History (%)</th>
<th>NICU History (%)</th>
<th>Other (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moderate (40-69 dB HL)</td>
<td>22</td>
<td>38</td>
<td>40</td>
</tr>
<tr>
<td>Severe (70-94 dB HL)</td>
<td>28</td>
<td>24</td>
<td>48</td>
</tr>
<tr>
<td>Profound (95-130 dB HL)</td>
<td>27</td>
<td>30</td>
<td>43</td>
</tr>
<tr>
<td>Overall</td>
<td>25</td>
<td>32</td>
<td>43</td>
</tr>
<tr>
<td>(CI 21-34)</td>
<td>(CI 26-40)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
There is no significant variation in the proportion of children with a family history of hearing impairment over severity; however, there is some evidence that the proportion of NICU-history children is greater in the mild and moderately hearing impaired. The NICU children in this study comprised about 7 percent of all births over the three areas.

For the Nottingham and Sheffield data alone it is possible to examine the NICU data in more detail to examine how many of the NICU children with no risk factor on the Joint Committee Risk Register had a material hearing impairment. In this study, 14 percent (CI 6–29%) of the NICU hearing-impaired children had no risk on that register, compared with Mauk et al (1991) with 23 percent (12–39%). The majority of these 14 percent were in the NICU for prematurity-related reasons but did not have any specific high-risk factor.

Davis and Wood (1992) had a risk of hearing impairment in the NICU of about 1 in 150. For this study, the estimate of this risk was about 1 in 222 NICU children having at least a moderate hearing impairment. This compares with 1 in 1407 of all other children or 1 in 2223 for those remaining children who do not have a positive family history of childhood deafness, maintaining a tenfold increased risk for the NICU children over not-at-risk non-NICU children.

**Age of Referral**

Table 5 shows the age of referral for diagnostic audiology for the congenitally hearing-impaired children who were born in the district in which they are now resident. The median age of referral overall was 12 months, with an interquartile range of about 13 months.

The NICU status makes an impact on the age of referral, as it should, because, during this time (1983–1988), targeted neonatal screening was being conducted on a pilot basis in some of the districts. The effect of severity of impairment has an effect on the upper quartile but not very much at the median or lower quartile.

**Etiology**

It was not possible to look at etiology in great detail over the three areas of the study. Several children had more than one possible etiology and it was very difficult for the clinician to put one primary etiology in each case. However, given those limitations, it was possible to obtain an etiology for 77 children with moderate to profound hearing impairments in Nottingham between 1983–1989. These were divided into 4 percent each for rubella, meningitis, and syndromal deafness. Of the 18 percent who had a family history of childhood deafness, there were 5 percent who were classified as autosomal recessive hearing impairments. A further 25 percent were classified as presumed autosomal recessive and 22 percent as unknown. For the remaining 24 percent, 20 percent were classified as having perinatal complications (e.g., birth asphyxia, fits or seizures, and usually gentamycin), with 4 percent having no complications other than prematurity.

**DISCUSSION**

Apart from looking at the implications of the cross-sectional data presented above, a prime consideration here is to compare the data with that from the UK 1969 cohort of Martin (1982). The data presented here are about 15 years later. The progress in determining uniform protocols for assessing degree of hearing impairment has enabled us to rely on data obtained from children younger than those in the EC study; however, the extent to which the data are open to absolute comparison is open to argument. Notwithstanding the differences in definitions and protocols, there is probably a 20 percent increase in the prevalence of hearing impairment. The main reason for this may be an artifact due to poorer ascertainment in the mild-moderate severity categories in the earlier study. There are probably about 1 in 800 bilaterally at least moderately hearing-impaired children in the UK, in the cohort of children presently (1992) aged 5, with about 1 in 2700 being profoundly impaired. The proportion of children from NICU with hearing impairments appears to have
increased fourfold (odds ratio increase 4.1, CI 2.7-6.0); the proportion with a family history of childhood deafness has also increased about twofold (odds ratio 2.4, CI 1.6-3.6). These increases are balanced by a decrease in rubella to one fifth of its 1969 proportion (odds ratio 0.18, CI 0.05-0.60). There appears to be no change at all in the proportion of hearing impairments due to meningitis at 6.5 percent in both studies, accounting for about 16–20 percent of all profound impairments.

The age of referral for a hearing aid was just below 3 years on average in the EC study, and this has decreased in the areas studied here to about 12 months on the median for age of referral to the diagnostic audiology service. Thus, there has been a substantial reduction in this critical age; however, for many hearing-impaired children in this study, it is still well into the second year before referral is effected.

The public health implications of these data are (1) that the NICU graduates should be a major target for neonatal screening using ABR or click evoked otoacoustic emissions methods, (2) that better understanding of the etiologies of the NICU hearing-impaired children should eventually lead to a reduction in complication rates for premature babies and those ill at birth, and (3) that the family history group should be effectively targeted on maternity wards and offered neonatal screening as a priority. Finally, all children with meningitis should have a diagnostic assessment of hearing at a reasonable interval post-discharge.

These data have given a glimpse of some of the present epidemiologic characteristics of hearing-impaired children in some areas of the UK. They may generalize to other areas of the UK and other countries with similar birth rates, prematurity rates, rubella vaccination programmes, and childhood surveillance programmes (Hall, 1992). However, there is still a tremendous amount of work that needs to be done to achieve a uniform set of assessment protocols and data collection so that a proper benchmark can be put down to assess future changes in prevalence, risk factors, etiologies, and age of referral.

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REFERENCES


