ABSTRACTS
Presentations at the BACDA Study Day
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Identification of Hearing Impairment: A Multi-Centre Study

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A collaborative nine-centre study was designed to follow the routes to identification of all chil;dren up to the age of seven years, newly diagnosed with permanent hearing impairment (>50 dBHL) during the period 1993-94. Ages of identification were compared with the standards set by the National Deaf Children's Society (NDCS), ascertaining whether these targets could be achieved with current service provision. Of the 126 children identified, 104 had congenital sensori-neural hearing loss: 19% were identified by the age of 6 months and 39% by their first year. These results compare unfavourably with the NDCS targets of 40% and 80% respectively and point to the need for modifications of current practice.

Hyperacusis

Josephine Marriage PhD.

"Hyperacusis" is used to describe an intolerance or aversion to sounds that would be tolerated by most normal listeners. Although the term "hyperacusis" can be applied to peripheral and central causes of over-sensitivity to sound, this presentation will focus on the central auditory processing disorders.

Hyperacusis is one of many physical and behavioural features of the Williams syndrome phenotype. The author's initial research work in this area examined auditory over-sensitivity in subjects with Williams Syndrome (WS). These Ws hyperacusics represent a clinical group with a shared genetic lesion. However, over the course of the project it became apparent that there are a number of other clinical groups for whom oversensitive hearing is a major problem.

A synopsis of the work on hyperacusis in Williams Syndrome will be presented, followed by some data from hyperacusics in the general population. Thirdly, the role of sensory sensitivity in children with communication disorders and other learning disabilities will be discussed. Strategies for

A Register (or shared list) of Hearing Impaired Children - Benefits, Costs and Implementation

Adrian Davies

The Department of Health funded review of the role of neonatal screening in the identification of children with congenital permanent hearing impairment collected information on the present screening and audiological services available in the UK. As part of this study five focus groups were held around the country. One of the important aims of these groups was to take soundings about what different professionals felt about hearing screening in the UK, and what aspects of the services they thought were good and which needed the most improvement.

Contrary to my initial belief that everyone would want better neonatal screening, the major concern was with the information systems that were available and the provision of a seamless service to the children and their parents. Furthermore, the research we have undertaken in the Trent region concerning building a database, for research, on all the children in the cohort born 1985-93 underlined the fact that there were inconsistencies in the data held by the different services (acute, community and education). We recommended in our report to the Trent NHS executive that at the district level there ought to be a shared list of children with a hearing impairment and that this ought to facilitate a seamless service. In my opinion this would also help in making the Children Act requirement on the local authority more explicit. In addition to the needs of those responsible for the health-care needs of hearing impaired children the professional and representative bodies involved in childhood deafness have also expressed a considerable interest in a database of children with permanent childhood hearing impairment (PCHI).

However, several questions have to be asked about such an undertaking. What exactly are the benefits of a shared list of children with PHCI? These might include benefits for the services provided such as: — shared knowledge, more

efficient access to information concerning the action of different professionals by parents and other professionals; better ability to audit the services provided; understanding of what are the necessary and major items of information that need to be shared; as well as giving considerable potential for health service research and the possibility of answering other basic research questions. The nature of these benefits will be elaborated in more detail.

There are several difficulties in the implementation of shared lists! Firstly, all the providers have to agree to collaborate in building up the list. Secondly, a decision has to be taken whether the list is local or wider in nature. Thirdly, the information on the list has to be agreed and uniformly implemented. Fourth, the ongoing nature of the list has to be made clear. Is it something that should be continuously updated or something that is periodically updated. Finally, the criteria for who should be on the list will need to be made.

Although many of these issues have been aired previously, the argument will be made that now is an ideal time to consider earnestly seeking the funds to undertake such a task so that the services provided to hearing impaired children and their families can benefit from the uniform application of demonstrable improvements in service provision for hearing impaired children.

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NATIONAL CENTRE FOR CLINICAL AUDIT

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What can the NCCA do for you?
What can you do for the NCCA?

The NCCA - a brief history

- Contract between BMA/RCN and Dept. of Health
- Started 1995
- Funded for 3 years
- self financing from June 1998

Purpose

- To facilitate best clinical picture in healthcare by promoting clinical audit through a multidisciplinary context.
- To improve patient care through promoting real change toeards best practice. To achieve this,NCCA will evaluate and disseminate information on audit activities, especially development of methodologies and clinical practice guidelines, and establish criteria for assessing audits.

Main functions of the NCCA

- gather and disseminate audit information.
- network for lead clinicians and organisations involved in audit.
- clinical audit database.
- enquiry service for clinicians, audit support staff and others.
- develop and disseminate quality improvement principles to ensure high quality clinical audit.
- support the above with a library function.

The NCCA - a partnership, e.g.

- BMA
- RCN
- Royal Colleges
- Patient organisations etc. etc.

Current work

- developing consensus criteria for audit.
- commissioning monograph on audit.
- assembling bibliographic database.
- creating database of contacts.
- inviting submissions to national audit database.
- distributing Audit Action Pack.
- running workshops.
- enquiry service.

Future direction

- work with partners and others to create combined information service.
- draw together existing data and make it accessible as a single entity.
- get overview of enquirers' needs.
- get overview of providers' activities.
- improving cost effectiveness of audit initiatives.
- high standards of presentation.
- moving towards comparable information.

Early: A National Audit of Hearing Loss in Children. A Report and Findings

Tim Williamson. BACDA Audit Subcommittee

It is the goal of children's audiology services to diagnose deafness as early as possible, and to provide appropriate amplification quickly. However, there is considerable doubt as to how well the services achieve this. This makes it a good topic for audit.

In 1993 BACDA set up an audit committee, consisting of Janet Lowe (Chair), Doreen Roberts, Peter Watkin, and Tim Williamson. They proposed a national audit to look at the early diagnosis and management of permanent hearing loss in children - called **Ear**ly. An application for DoH Central Audit Funding was successful, and by January 1995 data collection had started.

Early was designed to provide the most useful information required to improve the service at "district", Health authority, and Regional level. Central to this was the need to identify the key people in each "district" who took the clinical lead for children's audiology services, to be co-ordinators for **Ear**ly. BACDA members were well placed to take this on, and they account for most of the "district co-ordinators". They were asked to complete forms for each newly diagnosed deaf child - with a PTA of >40dB. The data on the forms included the hearing thresholds, the ages of first suspicion, of confirmation, and of aiding, together with reasons for delay at any stage. These data sets were entered onto a computerised National Register by Audit Staff in Manchester.

The data from the first 18 months are available for analysis. The completeness and quality of the data will be discussed. They can be analysed in any number of ways. For this presentation, most of the analyses will be national, but indications will be given of how use can be made of analyses by Region or even by "district".

The ages of first suspicion, confirmation and of aiding are usually considered to key performance indicators. The **Ear**ly national results are as follows:

Median age of suspicion	9 months
Median age of confirmation	15 months
Median age of aiding	18 months

The times between these stages will be reported, together with the reasons for delays. Also, the results from **Ear**ly will be discussed in relation to those from the last national audit (Martin 1982), and the NDCS Quality Standards.

Finally, the weaknesses and strengths of **Ear**ly will be discussed. The main tasks ahead for **Ear**ly will be firstly, to provide feedback "packs" for each Region, so that the analyses at Regional and even "district" level can be discussed and improvements made, secondly, to prepare a full report for the DoH, and thirdly, to address the issue of further funding, and completing the audit cycle.

Obscure Auditory Dysfunction (OAD) - an overview

Josie Higson and Mark Haggard, MRC Institute of Hearing Research, Nottingham

Obscure Auditory Dysfunction (OAD) is one of the terms used to describe the condition in which an individual complains of difficulties understanding speech in background noise but has normal results on pure-tone audiometry. It is a descriptive label for a reported problem, allowing the complaint to be recognised and further explanatory tests to be carried out. It is not a diagnostic label for a person meeting specified criteria on certain tests. For short, the term "OAD" can also be applied to the person.

The need for research on OAD was twofold: a) to provide a set of tests which could offer explanations for the difficulties in OAD and, if possible, more precise diagnostic labels, and b) to provide a management strategy. An emerging literature in the late 1980's suggested that various psychoacoustic deficits "underlay" OAD. However, in a large-sample case-control study, Saunders and Haggard (1989,1992) showed this definition to be inadequate by defining a multi-factor model of performance, cognitive/linguistic, psychological and psychoacoustic factors. A clinical test package (Saunders, Field and Haggard, 1992) is now available which has been purchased by over 60 audiology departments throughout the UK. The management strategy includes patients acquiring a better understanding of their problem along with practical advice on hearing tactics. This work has since been validated in application by two further studies (Higson et al., 1994,1996).

The package is restricted to adults only. At present a similar package suitable for children does not exist, although children fitting the same general description are occasionally referred. It is not clear that obtaining norms for two or three batteries appropriate to different age ranges would repay the research investment, given the limited numbers, and limited courses of action. Where there are strong reasons to suspect a rare specific neuropathology, an electrophysiological or imaging approach may be more effective. The success of the behavioural approach in adults is based on the assumption, shown in follow-ups to be correct, that a neuropathological approach is not justified in the vast majority of these adults.

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Professional Concerns in OME - the Symptom Concern (SYMCON) study

Josie Higson, Joan Birkin, Mark Haggard, MRC Institute of Hearing Research, Nottingham

Since Glover's report in 1938 of a 13-fold variation of tonsillectomy rates between areas in England, the literature has continued to report examples of medical practice variation. There has been public health concern at the 2-fold variation in treatment for Otitis Media across healthcare regions in England (Effective Healthcare Bulletin: 4, 1992), and larger variation (over 4-fold) among Scottish health boards (Russell and Bissett, 1994). Professional uncertainty has been proposed as one of the major factors explaining medical practice variation. Wennberg et al., 1982, commented that procedures associated with low variation are those where the diagnosis can be achieved reliably and for which a consensus concerning the value of the procedure exists within the medical professions. Otitis media with effusion is not easy to assess as regards severity and persistence, nor does it enjoy a consensus of opinion regarding its treatment, Consequently we might expect research into professional uncertainty surrounding OME to be ultimately fruitful in reducing the overall variation in its management.

The Symptom Concern Study aims to understand the differences within and across professions in judged frequency of OME symptoms and the level of importance/ concern each professional group associates with the symptoms when reported. Discrepancies with the parental view are being examined also. Only the results from questionnaires sent to Community Doctors, GPs and ENT Consultants are reported here.

The first aim of the analysis was to simplify the 38 questions about judged frequency and the 38 questions about concern into a meaningful subset of factors which could be used to examine across-group differences. Factor analysis revealed 7 factors for the "frequency" domain and 8 for the "concern" domain, explaining a 55% and 67% of the variance respectively. The rotated factors correspond well with the 7 *a priori* sections into which the questionnaire was structured. The few main discrepancies made sense, e.g. the "presentation" section simplifies down into "acute symptoms" only, and "hearing problems in presentation" combine with the general hearing section. The factor structure for the "concern" domain further differentiated the hearing section into 2 factors, one of which comprised the symptoms of difficulty hearing/listening in a one to one conversation and in a group, leaving the more general indication of an impairment (turning up TV/radio and talking louder than necessary) to enter another factor. Correlations between corresponding factors are sufficiently low to indicate that the two aspects of questioning, about frequency of report and about associated concern, are indeed sufficiently different to justify addressing separately.

The extracted factor scores were used to examine differences between the groups with one-way analysis of variance and paired t-tests with Bonferroni correction for the number of paired comparisons. These showed several differences among the three professions in the judged frequency of symptoms. Community Doctors come out more strongly as allocating both to behaviour and to speech/language problems a frequent place in OME presentation than is the case with the other two groups. ENT Consultants report concentration problems as a less frequent sign than do the other two groups. GPs attribute balance and hearing symptoms less frequently. However acute symptoms figure more frequently for GPs than for the other two groups. The latter difference may reflect possible confusion in the diagnosis of OME in general practice where access to appropriate diagnostic tests is not always available.

Similar analyses in the "concern" domain showed Community Doctors to be significantly more concerned about acute symptoms, which is perhaps related to less frequent observation of these symptoms in a possibly more purely defined caseload. They are also significantly more concerned about lack of concentration than ENT Consultants and about speech/language problems than both of the other two groups.

In summary, a stable pattern of variation between professional groups in concern and frequency for the reported symptoms of OME has been identified. Some of the differences relate to differences in probable caseload specification, but others are less easy to explain and may reflect inter-professional differences in awareness of the importance of these symptoms.

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BACDA PRIZE 1996

The 1996 BACDA Prize was awarded to Dr. Kate Hooper for her paper describing the problems of distraction testing in her area. Her entry is summarised here.

An Audit of the Health Visitor Distraction Test.

Dr. Kate Hooper

Introduction

It is accepted that the early diagnosis of hearing impairment in children is beneficial and that early habilitation is associated with a more favourable outcome[1]. The methods of screening for hearing loss in infancy are currently the subject of much discussion and the development of B.S.E.R. audiometry and O.A.E. has raised the possibility of screening all babies within the first few weeks of life. At the present time many areas operate a targeted screen at this stage, focused on the group of infants with known risk factors for hearing loss and this can be expected to identify 50-60% of hearing impaired infants.

However, in most areas the detection of unsuspected hearing impaired infants remains largely the responsibility of the health visitor. Traditionally, all infants have been screened using the distraction hearing test, generally carried out at about seven months, although in some areas this has now been replaced by a programme of surveillance and a parental questionnaire completed with the health visitor, again when the baby is about seven months old[2].

In Doncaster a targeted neonatal screen has been in place since 1992 and, as expected, is identifying about half the cases of congenital hearing loss. Health visitors carry out a distraction test at seven months and in recent years achieved a coverage rate of 96% (rising to 98% in 1995) with a referral rate to second tier services of just under 8%. However, a more detailed examination of referral patterns showed a range of referral rates of 0 - 25% between different health visitors, a pattern which was repeated almost identically over two consecutive years. Such a variance prompted local concern and this, together with national doubts over the value of the distraction test[3], led to the decision to carry out an audit of the test.

Following this a working party was formed to consider the best format for the audit and it was decided to carry out an operational audit and to combine this with a retrospective study of the route of identification of sensori-neural hearing loss over the years 1989-1993. Two experienced paediatric audiologists were employed on a part time basis for six months and, together with the author and an audit officer, designed a tick box data collection form. This was first piloted in a second tier clinic to check for consistency between the two auditors. Before the audit started a series of meetings were held so that the audit could be discussed with health visitors. All were assured that the results would be anonymous and they were encouraged to raise any concerns. In fact, nearly all were very positive towards the audit. Health visitors were contacted directly by the audiologists (who had no previous knowledge of their work patterns or referral rates) and arrangements were made to observe them testing in their normal working environment. In all it was possible, within the time constraints of the study, to observe a total of 154 tests and 54 (74%) health visitors. All the results were recorded and subsequently used for analysis.

The areas looked at in the operational audit can be divided into three main categories and these will be discussed under the following headings:

1 Test environment
2 Conduct of the tests
3 Quality issues.

Test Environment

The choice of test situation was made by the health visitor concerned and was governed by local factors such as client compliance and availability of facilities. The percentage usage of each test situation was:

Home	51%
Clinic	33%
G.P. Surgery	16%

The test environments were assessed looking at ambient background noise, number and loudness of interruptions during testing, and overall suitability, e.g. size, furnishings, windows and reflective surfaces. Overall, the home came out as the most favourable situation with the local clinic the worst.

Conduct of test

In this section the auditors looked at both the distractor and the person producing the test stimuli (tester). At the time of the audit only health visitors had been trained to take the latter role. The distractors were from the following groups:

H.V.	32%
N.N.	57%
Other	11%

Distractor

The auditors found that this part of the test was being performed to a high standard in all cases, e.g.

Technique correct	95.5%
Timing Correct	99.4%

There was no difference in performance between the different groups of workers.

Tester

Although only health visitors were supposed to take the role of the tester, it was found that in 5% of tests other workers were being used. In some areas the test was being carried out to a high standard but in other cases significant problems were highlighted. Despite all staff having been trained to use and having access to warblers, 21% of health visitors were not using them and, of those who did, 27% were not using them in the correct position. Of great concern was the finding that only 7% of tests included a no-sound trial and in 20% the baby was not given any reward for turning round. In over 90% of tests the timing of the sound presentation was judged to be correct with randomised sides and frequencies being used.

The auditors asked the health visitors to produce 'conventional' sounds at the end of the test and their loudness was measured on a sound level meter with the following results:

	<40dBA	>40dBA
Rattle	89%	11%
'ss'	81%	19%
Hum	73%	27%

Interestingly, when these measurements were correlated with the test venue it was found that there was a higher percentage of over-loud stimuli from tests conducted in the noisier settings, perhaps testers subconsciously allowing for background noise.

It would be hoped that the use of warblers, instead of conventional stimuli would result in a more accurate test. However, when the warblers were checked a disturbingly high number were found to be out of calibration.

PERCENTAGE OF WARBLERS RECORDING >40dBA

	Calibrated 30dBA	Calibrated 35dBA
500Hz	0%	0%
2kHz	0%	27%
4kHz	12.5%	0%

Quality Issues

In this section the audit looked at a variety of issues which would affect the total quality of the test, for instance communication with parents and recording of results. Again a variable picture was demonstrated. In 98% of tests the equipment was ready and in place before the test started but only 50% of testers checked that their warblers were working satisfactorily before using them. The results of the hearing tests were fully explained to the parents in 94% of cases. However, parental concern about the child's hearing was only sought in 52% of tests.

It was shown earlier that there were two main pairs of professionals working together; H.V/H.V or H.V./N.N. Traditionally, the test was always done with two health visitors working together and so it was decided to look at how the two different groups performed. There was no difference between the actual standard of distraction but in several other ways the H.V/N.N. pairing outperformed the traditional pair.

	H.V/H.V.	H.V./N.N.
Warbler Used	65%	85% (x^2 0.01>p<0.05)
Reward Never Used	40.5%	12.7% (x^2 P<0.01)
Test explained	54.8%	75.7% (x^2 P<0.05)

The reason for these discrepancies is not clear although it may be that when two health visitors work together neither one adopts the lead role. Additionally N.N. have only been trained over the last few years.

Finally, the author carried out a brief retrospective audit of the route of detection of S.N.H.L. over a five year period between 1989 and 1993. Looking at bilateral losses greater than 40dBHL, 30 children had been identified of which 40% had come from the H.V. screen. 23% were considered to have incorrectly passed the screen and, as expected, the introduction of a high risk neonatal screen in 1992 reduced the number 'available' for detection.

Conclusions

In conclusion, it was felt that the site chosen by the health visitor for the test was often out of their control but that in many cases relatively cheap alterations to clinics could improve the testing conditions. The role of the distractor was being carried out to a high standard but there were problems with the delivery of test stimuli, which could result in a wrong test result. In particular, re-training needed to be directed at the consistent and proper use of regularly calibrated warblers and the importance of using rewards and no-sound trials. Health visitors should always ask about parental concern before starting the test and explain the results after the test is completed.

The results of the audit were presented to all health visitors and a number of recommendations were made to improve the screen. These included a worksheet to be used at all tests, the H.F.R. and warbler being the onl;y stimuli permitted to decide whwther a baby should pass or fail and a regular calibration programme for warblers. A detailed test protocol is now issued to all staff at their initial training and a copy is available at all base points. The particular issues highlighted by the audit were noted for inclusion in the regular refresher courses and were also discussed at H.V. meetings. Finally, it was decided that N.N.s would be offered training so that they could take either test position, although a fully qualified health visitor must be present at all tests.

The screen is currently being re-audited as part of an audiologist's M Sc. dissertation and we await her findings with interest.

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NATIONAL EVALUATION OF OUTCOMES FROM PAEDIATRIC COCHLEAR IMPLANTATION

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This article describes a collaborative project between the Medical Research Council's Institute of Hearing Research (IHR) and researchers at the Universities of Manchester and York. The project will be the first national (i.e. very wide scale) study of the effectiveness of cochlear implantation for children conducted anywhere in the world. The initial phases of the study are supported by the Hearing Research Trust with funding from the National Lotteries Charities Board. The success of the study will depend upon the co-operation of professionals involved in the audiological care of deaf and hearing-impaired children, and in their education. During the next few months, we shall be writing directly to many members of BACDA to ask for a small amount of specific help. This article explains the background to the study.

Cochlear implantation for children has developed rapidly in the United Kingdom over the last seven years from a low base of activity during the 1980s. By the end of 1996, 600 children had received implants and the rate of provision had risen to nearly 200 new cases each year. The resulting annual cost to the NHS is about twelve million pounds. The growth in activity has been spurred by evidence that implantation can lead to achievements in the use of spoken language that are not attained by equally-impaired children who use hearing aids. Nonetheless, the provision of implants to children remains contentious, in part because of four uncertainties: Are the average benefits large enough to make a material difference to the lives of the children and their families in the longer term — e.g. in enhanced academic achievements and increased quality of life? Do all implanted children benefit, or only a subset? Are equally good results achieved by all sizes and styles of hospital programme? Overall, is implantation of children a cost-effective use of NHS resources?

Our first goal is to paint a representative picture of outcomes from paediatric implantation. To do that, we shall aggregate and compare outcomes for all children who have received implants in the UK, building on a productive relationship between IHR and providers of implantation services which developed during the evaluation of the (mainly adult) National Cochlear Implant Programme.

The second goal is to make controlled comparisons between implanted and un-implanted children. We shall take careful account of the different styles of habilitation, communication, and education for which members of each group have opted. We start from the point of informed detachment: we do not presume that any option will prove to be more effective than any other in permitting success in education and fulfilment in other aspects of life. To enable such comparisons to be made, a large comparison group must be identified. Accordingly, the first phase of the study will be a national ascertainment of all children with hearing levels 40dB in the birth cohorts 1980-1995 living in the UK. This is an important goal in its own right. Children with implants will be identified in collaboration with providers of implantation services. We shall seek the collaboration of health carers, principally Senior Clinical Medical Officers, and educators, principally Heads of Services and Schools for Hearing-impaired Children, in identifying the other children with hearing levels 40dB.

In the second phase of the study, parents, teachers and health carers will be asked to complete questionnaires. They will cover all implanted children, all other deaf and profoundly-impaired children, and a random sample of moderately and severely impaired children. Questions will concentrate on (i) the natural history of the impairment, (ii) use/non-use of cochlear implants, hearing aids and other assistive devices, (iii) skills in the perception and production of signed and spoken languages, (iv) attainments in reading, writing, and arithmetic, (v) socialisation and engagement in the process of education, (vi) quality of life of children and their families, and (vii) the quality of care that it has been possible to provide in health and education. Groups of children with and without implants will be carefully matched for the purpose of comparing outcomes in order to control for variables known to influence outcomes.

In the third phase, a stratified sample of approximately 200 children will be invited to participate in face-to-face testing to corroborate responses gathered in Phase II. In parallel with these activities, we shall measure the costs associated with different choices in health care and in educational care, including costs born by families themselves. These data will permit comparisons of the cost-benefit and cost-utility of implantation in relation to other options for deaf and hearing-impaired children.

In summary, the study has several purposes: to provide evidence on which children and their parents can base choices; to help purchasers of health care to target resources effectively; to allow providers to steer programmes in the direction of greatest cost-effectiveness; and to inform judgements about the optimal concentration of services within the NHS. Overall, therefore, the results should inform the (re)habilitation of all deaf children in the UK. We hope that members of BACDA will be able to help us to achieve this goal.

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VISUAL IMPAIRMENT IN ASSOCIATION WITH HEARING IMPAIRMENT (INCLUDING USHER'S SYNDROME)

Daniel V. Lang

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Mrs. Angela Alderman the Head of the County (educational) Audiology Service in Cornwall requested my view on visual screening for hearing impaired children with special reference to Ushers Syndrome. I reviewed the recent literature and prepared a response based on the services and needs in Cornwall. Any views on this would be welcome either directly to myself or through the newsletter

If an individual has significant hearing impairment then vision is extremely important for safety and development. Wager and Whale (1988) screened a total of 120 children, aged between 3-16, in schools for the deaf and partially hearing units in Birmingham. They used Snellen, Sheridan Gardner or Kay tests (with sign language communication if required). The total number of children with an ocular defect was 59 (49%), 39 of these were receiving appropriate treatment; nevertheless, 13 warranted further refractive examination. The major categories of defect were acuity problems and acuity problems associated with squint. The study did not comment on specific aetiology causing the visual impairments.

The paper did not state whether universal pre-school orthoptic screening was in place as it is in Cornwall (at age 3. 5). The authors concluded that it was important to consider the effect an ocular problem may have on a child with hearing impairment and emphasised the need for these children to have full orthoptic, refractive, and ophthalmological examination where appropriate. The incidence of Congenital Rubella Syndrome has dropped through immunization and this will cause a significant reduction in children with combined visual and hearing problems.

Ushers Syndrome

This is a very rare genetic condition characterised by sensorineural hearing loss with retinitis pigmentosa. There is a progressive deterioration of the retina which can cause night blindness, tunnel vision and finally severely reduced central vision.

Dr. Valerie Newton referred to a prevalence of 3 : 100.000; estimating that only 3-6% of children with congenital hearing impairment will have Ushers Syndrome. Davis (1993) provided prevalence figures showing that 1: 2000 children at age 5 have sensorineural hearing impairment $\geq$ 80 dBHL increasing to a prevalence of 1:770 for hearing impairment $\geq$ 50 dBHL.

Should all cases of sensorineural hearing loss be screened for Retinitis Pigmentosa?
If so how and at what age?

Newton (1988) addressed the issue in a paper on the aetiological evaluation of the hearing impaired child.
" Ushers disease cannot be diagnosed by retinal inspection in the first few years of life and in the young child the diagnosis would depend upon electroretinography. Very young children would need sedation or anaesthetic for the test and then the problem arises of parent counselling if the test indicates the disease. Establishing the diagnosis confirms the genetic nature of the condition in families in which the child is an isolated case, but presents the problem of telling the parents who have just discovered that their child is hearing impaired that they may also have a progressive loss of visual acuity. As the course of the visual defect is variable, the progression in an individual child would not be easily predicted. The disease does not usually

manifest its visual aspects before adolescence, so some clinicians may prefer to wait until the child was old enough for the diagnosis to be made by inspection of the retina. The decision could have implications for families contemplating having another child".

The gene for Ushers syndrome has recently been mapped and pre-natal diagnosis may be available in the future. This may add to the complexity of early management of such cases and the advice we give to parents.

Should all hearing impaired children in Cornwall have their vision screened ?

Vision should be considered at all opportunities for preschool paediatric surveillance: neonatal check, 6 week check, 8 month check, 18 month check, 3 year check. Vision is formally screened at the preschool orthoptic check (3.5 years) where the commonest findings will be acuity problems (refractive errors) and squint. Visual acuity is then monitored through school health. All hearing impaired children protected by the 1993 Education Act should have their vision checked annually by the school health service. We should ensure all hearing impaired children receive these checks.

Should all children with sensorineural losses requiring hearing aids be referred routinely to an ophthalmologist ? If so when ?

Newton (1988) stated that the eye is implicated in many conditions associated with significant childhood hearing loss and ophthalmological examination should be routine. Wager and Whale (1988) reinforced this view. We should bear in mind that the population of hearing impaired children in Cornwall will not be the same as the population referred to a specialist centre such as Manchester. Some children are routinely referred in Cornwall.

If we are aware of any child with severe to profound hearing impairment who develops balance problems, night vision problems, clumsiness, lack of attention, inexplicable accidents, discomfort to bright lights, sun, white objects we should discuss referral to ophthalmology. Bear in mind that the above symptoms usually present between 10 and 30 years of age.
Sense recommend (1994) that peripheral vision and night vision should be screened in school. We must consider this carefully. If we introduce a specific screen we must tell the parents why we are doing it. Only about 3-6 % of the congenitally hearing impaired will have Ushers syndrome. So for 97-94% of cases of congenitally hearing impairment introduction of a formal screen may burden the family with a serious worry about a condition that will not affect their child. On the other hand early diagnosis of Ushers Syndrome is important as referral for cochlear implant could be considered when vision is as good as possible to facilitate rehabilitation post implant.

Conclusion

This brief review considers some of the issues relating to coexisting visual and hearing impairment. On balance it would seem wise to obtain a full ophthalmological opinion at least on children with severe and profound sensorineural loss. If we formally screen in early infancy using electroretinography and in school for peripheral vision and night vision problems then it raise the issue of informing the parents about the purpose of the screen. This may cause significant anxiety.

Paediatric audiologists, community paediatricians and teachers of the deaf should be aware of the visual signs and symptoms of Ushers Syndrome as well as the much more common problems of visual acuity which may affect any hearing impaired child and request ophthalmological assessment if concerned.

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Unilateral Hearing Loss in Children

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(God would not " have simply hung a second ear on our heads purely as a mechanical safety factor in a chancy world") (Markides, 1977)

Editor's Note: I would like to apologise for the omission of the references when this article was printed previously. It is now included complete.

Introduction.

Unilateral hearing loss is often diagnosed and managed in community audiology clinics. This varies from the advice "your child has one 'dead' ear, this does not matter" to prescription of sophisticated hearing aid systems. In this article I have looked very briefly at the underlying physics and physiology of unilateral hearing loss and then reviewed the literature for some guidance on the best management.

The advantages of Binaural hearing

Most animals have two ears so that if one is damaged basic hearing is preserved. Two ears ensure full binaural hearing, which gives localisation of sound and better speech discrimination in noise. A child with hearing in one ear only is at a disadvantage as he/she has partial loss. Often the child himself, the parents and teachers do not fully understand the level and nature of the disadvantage of only hearing with one ear. Binaural hearing is more sophisticated. It gives the advantages of localisation of sound and better speech discrimination in noise.

Localisation of sound depends on three factors: firstly the difference of time, intensity and phase of arrival of sounds at the two ears. Stevens and Newman (1934) confirmed that low tones are localised on the basis of phase difference, and high tones are localised on the basis of intensity difference. Secondly central (neural) processing in the nuclei of the superior olivary complex codes the direction of sounds, the auditory cortex controls the ability to attend to sounds. (Moore, 1991). Lastly a role is played by the pinna. Its complex structure with many facets reflects sound meaning that some sound localisation occurs even when only one ear is functioning.

Two ears give an advantage in discrimination of speech in noise. There are three factors involved in this, firstly a simple summation - 3dB advantage of two ears over one. This may not seem much, but at threshold speech material may have a 6% per dB rise in discrimination score, so 3dB = 18% improvement in discrimination of connected speech. There is the squelch effect - the facilitation of speech in noise, meaning the binaural listener can ' tune' into wanted sound and ignore background noise depending on orientation of wanted sounds and noises, adding another 3dB. Finally the head shadow effect - if noise is one side and wanted sound the other the noise will be suppressed by the head. So a listener who can move his head gains considerably from the possession of two ears.

Unilateral (Monaural) Hearing loss in children may be a partial or total loss, and may be congenital or acquired. Bess 1986 (and other papers e.g. Bovo et al, 1988 and Culbertson 1986) showed audiological problems of speech discrimination and attention difficulties in children with unilateral hearing loss in noise which lead to academic delay and behavioural problems. The severity of the problems appear to relate to the severity of hearing loss, the age of onset and right ear impairment.

Suggestions to habilitate children with unilateral hearing loss

Carefully prescribed amplification for the poorer ear might be expected to help. In practice the children will not tolerate hearing aids, even if carefully calculated CROS (microphone at deaf ear, sounds transmitted to good ear) systems are used. Radio aids appear to improve speech discrimination in noise, but are not commonly available in the ordinary schools these children usually attend. (Updike, 1994)

The following are the main factors to consider to help children with monaural hearing loss. Some are applicable to all hearing impaired children, some to unilateral loss only. (Bess,1988 Brookhouser et al 1991 ,Northern and Downs 1991)

At Clinic:

1 Identify a.s.a.p. - especially if attentional or behavioural problems. In the UK these children are frequently picked up by the 5 year school entry audiometry screen. Once identified these children should have a full investigation to establish, where possible, the cause of the unilateral hearing loss.

2 Audiological supervision. There should be an annual review to check there is no change in the hearing level of the poor ear, and to promote the preservation of hearing in the good ear. The opportunity can also be taken to discuss the implications of the unilateral hearing loss with the child and the parents.

3 Counsel re preservation of good ear:
- early and efficient treatment of ear infections.
- ear protectors in noise.
- protective helmets when cycling or skate boarding.
- advise re scuba diving

4 Counsel child and parents re likelihood of directional difficulties, e.g. in traffic the child may not be aware of direction of unseen oncoming traffic.

5 Alert carers and teachers: advise re classroom placement. i.e. seat with good ear towards sound sources, and bad ear towards unwanted noise.

6 Involve teacher of the deaf, and if appropriate speech therapist.

At home and in School:

1 Gain child's attention before speaking, if necessary by non auditory means, e.g. touching, and ensure attention is maintained.

2 Use familiar vocabulary, and simple sentence structure, if necessary rephrasing, - these children are likely to be language delayed.

3 Use visual supplement to communication.

4 Minimise background noise.

With these factors in mind the community audiologist and school doctor should be able to help the teaching staff in ordinary schools to prevent secondary handicap resulting from the disability of unilateral hearing loss in children.

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