ABSTRACTS
Presentations at the BACDA Study Day

Tinnitus in Children
Jonathan W.P. Hazel

Over the last 7 years we have had the opportunity to apply retraining techniques developed in the adult tinnitus clinic to children and teenagers. An understanding of the neurophysiological mechanisms of tinnitus based on the Jastreboff model are extremely appropriate for tinnitus in the paediatric audiology clinic, and this model, together with illustrative case presentations will be presented.

The importance of tinnitus and hyperacusis in children, is that it can be extremely intrusive and disturbing and affect all aspects of the child's life, particularly schooling, where tinnitus interferes with concentration and particularly during homework and sleep. Hyperacusis can actually make school attendance, with its many loud sounds, impossible for the child.

All of these problems can be addressed by a retraining approach. In adults this customarily takes 18 months to be effective, but in children, where there is a much higher degree of plasticity in central auditory pathways concerned with the processing of tinnitus related neuronal activity, the results can be very much faster. In all cases the retraining approach must involve the parents as well as the child.

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A Review of Imaging in Audiology
Dr. David Baguley

This lecture will review the current status of imaging from a clinical audiology perspective. The physical basis and clinical benefit of the traditional imaging techniques of CT and MRI will be reviewed. The fascinating new developments in functional imaging shall be described, namely the use of SPECT PET and fMRI, the clinical application of these techniques to cochlear implants and neuro-otology will be considered.

Dr. David Baguley, Consultant Audiological Scientist

The Educational Implications of Minimal Hearing Loss - John Briggs

The difficulties faced by children with minimal hearing loss often appear to be out of proportion to the extent of their hearing loss.
A division needs to be drawn between those children who have a hearing loss sufficient to impede their development of language and those for whom environmental factors, especially in education, have a significant negative impact on their access to the spoken word.
Some of these children will have normal or near-normal peripheral hearing, but may have other auditory difficulties, or learning difficulties.
The importance of a broad based approach to the identification of such children and the use of classroom amplification systems will be discussed.

John Briggs, Educational Audiologist, Specialist Advisory Service

Child Protection in Audiology
Mary O'Sullivan

Perusal of the literature on child protection and communication reveals that

- Abused and neglected children's language is delayed
- Where there are problems with communication, there are difficulties with discipline
- Where there are problems with discipline, abuse is more likely and
- Children with otitis media with effusion tend to have linguistic difficulties

In a small study looking at the hearing of a group of children who attended for a Child Protection medical examination, it was found that the prevalence of hearing problems was 48%, as opposed to 8% in the general population at age 6 (as established by the School Sweep Screen). Most of these were mild. Whilst numbers in this study were small, nevertheless this is a striking finding, and merits further investigation. In the context of the related factors above, there may be more significance in the relationship between hearing problems, communication, child behaviour and the child-parent interaction where there are child protection concerns than has been recognised previously.

Reports from the United States suggest that deaf children are more likely to be abused. Children with disabilities in general are more likely to be abused, and this may be related

to difficulties with their care, behaviours which may be challenging, difficulties with attachment and difficulties with communication. Deaf children also frequently have communication difficulties with their parents, and they may be targeted by perpetrators but not have the communication skills to disclose abuse, or, as suggested by Ridgeway, may even 'believe that abuse is part of being deaf".

Finally, children who are abused and/or neglected can present to a community paediatrician, an audiologist or an E.N.T. surgeon, and these children deserve that their problems be recognised and addressed. Traumatic eardrum perforation due to a slap on the ear with an open hand, haematomas on the external ear, recurrent bleeding from both ears and non-organic hearing loss have all been described in association with child abuse. Abusing parents do not seek help, and it is up to us professionals to recognise that something is wrong, and know how to take appropriate action, or at least have access to senior colleagues for advice when these issues arise.

Mary O'Sullivan,
Consultant Community Paediatrician (Audiology)

BACDA members involved in training Specialist Registrars in Audiological Medicine and wishing to have guidance from the Royal College of Physicians, please contact Joanna Gurr on tel: 0171- 935 1174, ext 395, or fax: 0171- 486 4160 or e-mail: joanna.gurr@rcplondon.ac.uk. She will send you a copy of the logbook and training syllabus on request.

RECENT ADVANCES IN THE GENETICS OF NON-SYNDROMAL SENSORINEURAL HEARING IMPAIRMENT/DEAFNESS

Professor R F Mueller

INTRODUCTION

Since the mapping of the first gene for non-syndromal sensorineural hearing impairment/deafness (NSSNHI/D) in 1992, there has been rapid progress with the mapping of a large number of further genes. In the last 2 years mutations have been identified in the first 3 genes responsible for NSSNHI/D. Over the next couple years, these developments will lead to molecular diagnostic tests becoming routine making it possible to determine the cause of the hearing impairment/deafness in a significant proportion of children sporadically affected with NSSNHI/D. This will allow precise Mendelian recurrence risks rather than the empiric recurrence risks used in genetic counselling at present.

EPIDEMIOLOGY OF NSSNHI/D

Studies over the last 4 decades have shown that congenital or childhood-onset sensorineural deafness affects approximately one in 1000 children. The majority, about 70%, are non-syndromal, i.e. occur in isolation, in the absence of associated features. Of these, approximately two-thirds are due to genetic causes. Various studies have estimated that autosomal recessive genes account for two-thirds to three-quarters, one-sixth to one-third are due to autosomal dominant genes, the remainder (0-5%) being due to X-linked or mitochondrial inheritance.

GENETIC COUNSELLING/RECURRENCE RISKS

For the single gene syndromal causes associated with hearing impairment and in families in which hearing impairment/deafness has an obvious single gene pattern of inheritance, it is possible to offer a precise Mendelian chance of recurrence. NSSNHI/D, in contrast, most commonly occurs sporadically, i.e. as an isolated case within a family. It is not possible to reliably discriminate between acquired and genetic causes of hearing impairment purely on the basis of clinical and/or audiological criteria, e.g. by the age of onset, the severity of hearing impairment or the shape of the audiogram. The chance of recurrence of hearing impairment/deafness in subsequent offspring is based on empiric data, i.e. observed recurrences of hearing impairment/deafness in such families (Table 1). Depending on the study cited, the recurrence risk for a couple who have a sporadically affected child with NSSNHI/D, ranges from 1 in 10 to 1 in 5. In one study, the chance of recurrence approaches 1 in 4 if the hearing impairment was greater than 80 dB, consistent with the majority of sporadic NSSNHI/D being due to an autosomal recessive single gene cause.

NSSNHI/D LOCUS HETEROGENEITY

Segregation studies of the hearing status of offspring of different mating types, e.g. parents with normal hearing and parents with hearing impairment/deafness, have led to estimates that the number of genes responsible for NSSNHI/D ranges from 5 to 36 or more loci. The demonstration of linkage, cloning and identification of mutations in the genes responsible for NSSNHI/D will allow precise Mendelian rather than empiric recurrence risk figures to be given when counselling families who have a child sporadically affected with NSSNHI/D.

MAPPING OF GENES FOR NSSNHI/D

Since the mapping 6 years ago of the first autosomal dominant gene, there has been rapid progress in the mapping of a further 13 dominant, 18 recessive, 6 X-linked loci and 2 mitochondrial mutations responsible for NSSNHI/D[1-3] (Figure 1, Table 2).
Even with the known genetic heterogeneity of NSSNHI/D, mapping dominant and X-linked loci of genes is relatively straightforward provided that individual families with sufficiently large enough numbers of affected individuals are collected. Mapping of autosomal recessive loci for NSSNHI/D is potentially more problematic, requiring a different approach. Collecting families with two or more offspring affected with NSSNHI/D with normal hearing parents would require a large number of families to have sufficient mathematical power to be able to map the locus of a single autosomal recessive gene. Since there is no clinical or audiological means at present to distinguish between families where different autosomal recessive genes are responsible for NSSNHI/D, the known genetic heterogeneity means that it would be virtually impossible to demonstrate linkage for even a single gene. Identification of families in a variety of different populations in which consanguinity is common and is especially complex, i.e. there are multiple affected individuals in different branches of the family tree all of whom are related to a common ancestor, allowed this problem to be overcome. This approach, known as autozygosity mapping, has resulted in the mapping of 18 loci responsible for autosomal recessive non- syndromal sensorineural hearing impairment/deafness.

CLONED NSSNHI/D GENES

Recessive

DFNB1

The first locus responsible for NSSNHI/D was mapped in complex consanguineous Tunisian families[4] and confirmed in a complex consanguineous British Pakistani family[5] and a highly consanguineous Bedouin family[6]. A study of the segregation of polymorphic markers linked to DFNB1 in 19 families with NSSNHI/D from Australia and New Zealand were compatible with the majority being due to a gene at that locus[7]. A similar study of 30 Italian and 18 Spanish families with NSSNHI/D showed that the majority, approximately 80%**,** were likely to be due to a gene at the DFNB1 locus.[8]

Identification of the gene responsible for DFNB1 was due, in part, to a serendipitous observation. A dominant locus, DFNA3, responsible for dominant NSSNHI/D had also been mapped to the same region of the long arm of chromosome 13 in which DFNB1 had was located[9]. A family was studied in which an inherited skin disorder (palmoplantar keratoderma) and sensorineural hearing impairment were segregating as an autosomal dominant. The pattern of inheritance of polymorphic markers linked to that locus on the long arm of chromosome 13 suggested that genes mapping to that region known to be expressed in the skin were potential candidate genes. One such gene was the connexin 26 (Cx26) or gap junction protein beta-2 (GJB2) gene. Gap junctions (or connexons) are plasma membrane channels formed by 6 individual subunits called connexins. There are a number of different connexins, each of which has 4 transmembrane domains and are characterised by different molecular weights. Connexons make up intercellular channels between adjacent cells facilitating the exchange of ions and small molecules up to 1 kiloDalton in size. Immunohistochemical studies showed that GJB2 is widely expressed in a variety of different tissues and within the inner ear at high levels in the stria vascularis, basement membrane, limbus and spiral prominence of the cochlea.

Identification of a T to C point mutation in the Cx26 gene of the persons with sensorineural hearing impairment/deafness in the family resulting in a substitution of methionine to threonine (M34T) in the first transmembrane portion of the GJB2 protein suggested it was likely that Cx26 was responsible for the hearing impairment/deafness. The identification of 2 different substitutions in 3 complex consanguineous British Pakistani families with autosomal recessive NSSNHI/D known to be linked to the DFNB1 locus on chromosome 13 which generate a premature stop codon in the first and second transmembrane domain leading to a severely truncated GJB2 protein, confirmed Cx26 as being the gene responsible for NSSNHI/D at DFNB1[10].

A single base pair deletion of a guanine (G) in a stretch of 6 G's, variably known as 30 or 35del G, accounts for the majority of mutations (> 60%) in the families from Mediterranean area which had been shown to be linked to DFNB1[11]. This single base pair deletion results in an alteration of the reading frame resulting in the generation of a downstream stop codon which leads to truncation of the gap-junction protein in the first transmembrane domain. This single mutation was also found to be responsible for the majority, approximately 70%, of mutations in Cx26 in families linked to chromosome 13 from Tunisia, France, New Zealand and the UK[12]. Further studies have shown that this mutation is responsible for approximately 50% of sib-pairs and between 1 in 10 to 1 in 3 individuals sporadically affected with NSSNHI/D[13,14]. The hearing impairment in the majority of individuals with mutations in Cx26 is usually bilateral and symmetrical, with the degree of hearing impairment/deafness usually being severe or profound, although occasionally moderate.

In individuals with NSSNHI/D who have been found to be heterozygous for mutations in the Cx26 gene, studies are under way to determine whether there is a second mutation in the 5' promoter region of the gene. Studies have estimated that the frequency of the 35delG mutation could be as high as approximately 1 in 20 in the general population[14]. Although a number of other less common mutations have been reported in the Cx26 gene which are responsible for causing NSSNHI/D"[11,15,16] polymorphic variants which are unlikely to have functional consequence has also been identified[11]. Of further interest is the presence in persons with normal hearing of

the M34T Cx26 variant reported in the original family which lead to the suggestion of Cx26 as being a candidate gene for NSSNHI/D[17], raising the possibility that this sequence difference could also be a polymorphic variant and therefore not be of any functional consequence.

DFNB2

The mapping of the second autosomal recessive locus responsible for NSSNHI/D to the same region of the long arm of chromosome 11, 11q35[18] which contained one of the loci for a syndromal cause of sensorineural hearing impairment/deafness known as Usher's syndrome, Usher's type 1b, suggested that the same gene could cause both non-syndromal and syndromal hearing impairment/deafness. Identification of one of the unconventional myosins, myosin VIIA, as being responsible for the deafness and vestibular problems in an inbred strain of mouse known as shaker-1[19], led to the identification of the human homologue also being responsible for one of the forms of Usher's syndrome (congenital sensorineural deafness, retinitis pigmentosa and vestibular dysfunction) known as type 1b[20,21]. This meant that by virtue of its location, myosin VIIA was also an obvious candidate gene for DFNB2. Screening of the myosin VIIA gene in 8 Chinese individuals with NSSNHI/D revealed 2 persons with mutations in the gene.[20] A complex consanguineous family with multiple individuals affected with profound deafness which mapped to region of DFNB2 has also been identified as being homozygous for a mutation in the myosin VIIA gene[22]. In addition a locus for dominant NSSNHD/I, DFNA11, has also been mapped to the same region of the long arm of chromosome11 through a family with dominant late-onset progressive hearing loss. The individuals with hearing loss in the family have been shown to have a mutation in the myosin VIIA gene[23].

The unconventional myosins share structurally conserved head domains or regions which move along actin filaments by means of actin-activated ATPase activity and have divergent tails which are thought to move different macromolecular structures relative to actin filaments. Expression of the myosin VIIA gene is limited in the inner ear to the inner and outer hair cells of the cochlea and sensory hair cells of the vestibule.

At present it is unclear how frequently mutations in myosin VIIA are responsible for autosomal recessive or sporadic NSSNHI/D. Population estimates of this are likely to be difficult to obtain because of the large size of the coding sequence for the myosin VIIA gene and consequent difficulty in offering mutation screening routinely.

DFNB4

The locus on the long arm of chromosome 7, DFNB4, was originally reported to be associated with NSSNHI/D[24]. The mapping of the gene for the syndromal cause of sensorineural hearing impairment/deafness known as Pendred's syndrome (congenital sensorineural hearing impairment/deafness and goitre) to the same region[25,26] suggested that the situation could be the same as with Usher's 1b and DFNB2. The family which led to the mapping of the DFNB4 locus has been re-investigated and have findings consistent with Pendred's syndrome. This led to the identification of the PDS gene responsible for Pendred's syndrome.[27] Perhaps not surprisingly, a large consanguineous family from India with profound non-syndromal sensorineural autosomal recessive deafness which mapped to the region of the long of the chromosome 7 containing DFNB4 have been shown to homozygous for mutations in the PDS gene[28]. It is not clear at present how frequently mutations in the PDS gene might lead be responsible for autosomal recessive and sporadic NSSNHI/D.

Dominant

DFNA1

A family first reported over 25 years ago with late onset progressive hearing impairment/deafness[29] led to the mapping of the first locus for NSSNHI/D, DFNA1[30]. Identification of the gene responsible, HDIA1[31], reveals it codes for a protein which is homologous to members of the formin gene family which is involved in cytokinesis and establishing cell polarity. The frequency with which it leads to autosomal dominant NSSNHI/D is not known at present.

X-linked

DFN1

The first X-linked locus for NSSNHI/D was originally reported as being associated with isolated early onset progressive sensorineural deafness[32]. Subsequent review of the family revealed additional clinical features including visual disability, dystonia and mental retardation, i.e. it was syndromic[33].

DFN3

The gene responsible for DFN3, POU, causes mixed hearing impairment/deafness, i.e. both conductive and sensorineural[34]. This, along with the rarity of X-linked causes for inherited deafness means that mutations in the POU gene are unlikely to be a common cause of sporadic NSSNHI/D.

Mitochondrial

Mitochondrial mutations are often associated with syndromal disorders of which hearing impairment/deafness is a feature[35], but can occur as maternally inherited NSSNHI/D[36-39].

REDUCTION OF HETEROGENEITY

It is not yet possible to determine which estimate of the possible number of loci responsible for NSSNHI/D is likely, in fact, to be correct. The realisation that different mutations in the same gene can cause both syndromal and non-

syndromal hearing impairment/deafness means that the co-localisation to the short arm of chromosome 11p14-15 of Usher's syndrome type 1c and DFNB18 and Usher's syndrome type 1d and DFNB12 and to the long arm of chromosome 10 could represent the same phenomenon. The observation of mutations in the same gene resulting in both dominant and recessive NSSNHI/D means that the co-localisation of further loci causing NSSNHI/D may allow further reduction of the total number of loci involved. It is important to note that some of the existing loci for NSSNHI/D map to the same region of the chromosomes and may, in time, be shown to be due to mutations in the same gene, e.g. DFNB8 and DFNB10 on the long arm of chromosome 21q22 and DFNB7 and DFNB11 on the long arm of chromosome 9. One report of linkage, DFNB15, using autozygosity mapping in inbred families has revealed two regions of autozygosity on the long arm of chromosome 3 and the short arm of chromosome 19[40].

FUTURE DEVELOPMENTS

The identification of regions of homology in humans for mouse models for inherited deafness, along with the isolation of cochlea specific expressed sequences and cDNA which map to those regions should allow the identification of further genes responsible for inherited NSSNHI/D in the near future[13,41].

CONCLUSION

In the short term, the identification of a common mutation in Cx26 which can be easily screened for, means that in a significant proportion of families where the hearing impairment/deafness is known to be recessively inherited, the gene responsible can be identified. Screening for the 35delG mutation in Cx26 is likely to be the first routine molecular genetic test for individuals sporadically affected with NSSNHI/D. This will provide accurate Mendelian recurrence figures for families where mutations in the Cx26 gene are identified. If blood is taken from a sporadically affected child with NSSNHI/D to test for acquired causes of hearing impairment/deafness, such as TORCH titres for congenital infections, it is likely in the near future to become routine practice for an aliquot to be saved/sent for molecular genetic diagnostic testing. It is clear that the diagnostic rate for molecular genetic testing even at this early stage of development far surpasses anything else presently available.

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TABLE 1

CHANCE OF RECURRENCE OF HEARING IMPAIRMENT IN A RELATIVE OF PERSON WITH HEARING IMPAIRMENT

Relative	**Risk (%)**
Sibling	1/10-1/5 (10-20%)*
Offspring	1/1 6 (6.25%)
Nephew/Niece	1/130 (0.77%)
Offspring of hearing sibling	1/250 (0.4%)
Offspring of affected parent and child	2/5 (40%)**
Offspring of affected parent with affected sibling	1/100 (1%)

* if the parents are consanguineous, the risk is 1/4 (25%)

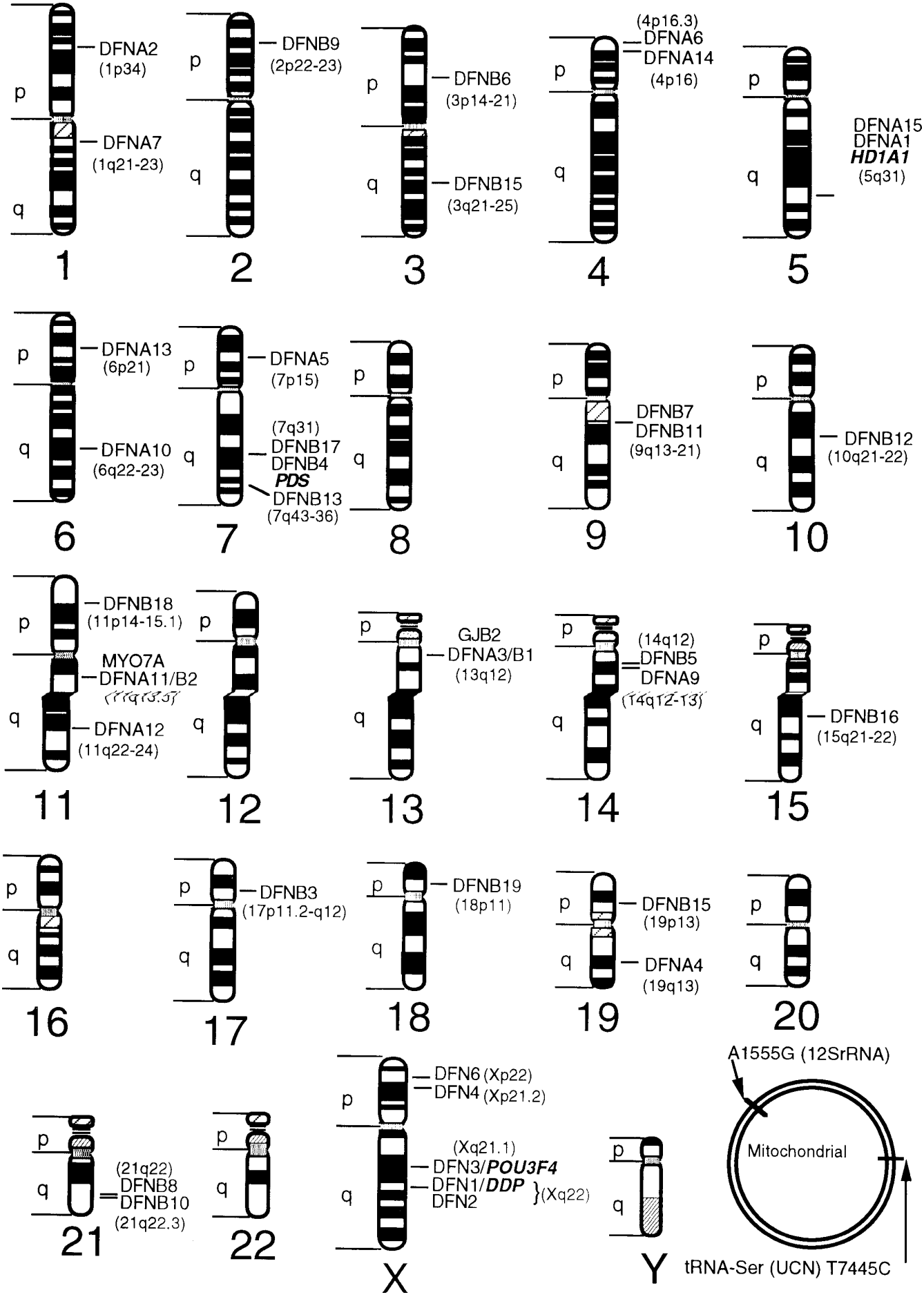
** although the chance of inheriting the gene is 1/2 (50%), the lower chance of having hearing impairment/ deafness is due to incomplete penetrance

TABLE 2

LOCI AND GENES RESPONSIBLE FOR NSSNHI/D[3]

LOCUS NAME	**CHROMOSOMAL LOCATION**	**GENE**
Dominant		
DFNA1	Sq31	HDIA1
DFNA2	1p34	
DFNA3	13q12	GJB2
DFNA4	19q13	
DFNA5	7pl5	
DFNA6	4p16.3	
DFNA7	1q21-23	
DFNAS	11q22-24*	
DFNA9	14q12-13	
DFNA10	6q22-23	
DFNA11	11q12/3-21	MYOVIIA
DFNA12	11q22-24	
DFNA13	6p21	
DFNA14	4p16	
DFNA15	Sq31	
Recessive		
LOCUS NAME	**CHROMOSOMAL LOCATION**	**GENE**
DFNB1	13q12	GJB2
DFNB2	11q13.5	MYOVIIA
DFNB3	17p11.2	
DFNB4	7q31	
DFNB5	14q12	
DFNB6	3p14-21	
DFNB7	9q13-21	
DFNB8	21q22	
DFNB9	2p22-23	
DFNB10	21q22.3	
DFNB11	9q13-21	
DFNB12	10q21-22	
DFNB13	7q34	
DFNB14	reserved	
DFNB15	3 q21-25/19p13	
DFNB16	15q21-22	
DFNB17	7q31	
DFNB18	11p14-15. 1	
DFNB19	18p11	
DFNB20	reserved	
LOCUS NAME	**CHROMOSOMAL LOCATION**	**GENE**
X-linked		
DFN1	Xq22	DDP
DFN2	Xq22	
DFN3	Xq21.1	POU3F4
DFN4	Xp21.2	
DFN5	reserved	
DFN6	Xp22	

FIGURE 1 - IDIOGRAM OF THE HUMAN CHROMOSOMES WITH LOCATION OF THE VARIOUS LOCI AND GENES FOR NSSNHI/D



BACDA PRIZE 1997

The 1997 BACDA Prize was awarded to Dr. Margaret Miles. Her paper on the speech perception test for the diagnosis and assessment of young deaf children is reproduced here.

The Smile Test: A speech perception test for the diagnosis and continuing assessment of young deaf children
Dr. Margaret Miles, MB.BS., MSc.

Introduction

There has always been a need for speech perception tests that are reliable, repeatable, and suitable for very young children to assess deaf children's auditory performance as part of their ongoing management. This need has been enhanced by the advent of cochlear implants. In the assessment of severely and profoundly deaf children a full battery of audiological tests is usually undertaken, and may need to be frequently repeated:

- pure tone audiometry
- objective tests
- speech perception tests.

The design of speech perception tests

Speech perception is a pattern recognition process where the listener hears (perceives) certain acoustic cues and selects an appropriate category into which the item fits. Speech perception tests use speech material but aim to assess the level of hearing. The response to the speech material may depend more on the experience and expectations of speech than on the level of hearing (Martin, 1987).

There are many speech perception tests that have been designed by various workers over the last century. Work started with adults, and was only later modified to be suitable for children. In the design of speech perception tests for adults the tests are graded by auditory challenge. In work with young children the developmental aspects of the test have to be considered. A test that is audiologically sound may be unusable for a young child who cannot understand or respond to the test material. Apart from all the known problems of designing speech perception tests young children have a low threshold of boredom.

In the English speaking world speech perception tests have been developed independently in the UK, the US and Australia. A full review of these tests showed that there is little material suitable for young hearing impaired children. In the UK there are good speech based tests (Distraction, Kendall and McCormick toy tests) for screening populations to identify children with hearing loss. These tests have been expanded for general use in the assessment of mild to moderate hearing losses but are not satisfactory for use with children with severe or profound losses. A nationally accepted battery of tests for the assessment of speech perception in children does not exist. There are as many tests and test batteries as there are centres. Simple, well researched and trialled speech perception tests for the continuing assessment of severely and profoundly deaf young children are needed. In the UK the most recent (but unpublished) test battery is the Tests of Auditory Perception of Speech for Children, TAPS test, devised by Reid in 1992. The TAPS test evaluates a hierarchy of auditory skills needed for the perception of speech and is aimed at children between the ages of 2 and 15 years.

Factors in the construction of speech perception tests

Hearing level and auditory ability:

- level of hearing loss, mild to profound
- pre-lingual or post lingual onset of deafness
- age of diagnosis
- efficacy of amplification (Boothroyd, 1995).

Content of the speech used:
Speech should be the test stimulus and should reflect everyday speech. Subjects are more likely to identify familiar, or favourite words so it is important to check the words are within the vocabulary of the child and that there is equal familiarity within the lists (Martin 1987). Some words, e.g. food, toys, may be more intrinsically attractive than others. Using tests from other languages or cultures is fraught with difficulties as usage changes (e.g. UK crisp / US chip). Translating material to and from English into foreign languages (as is suggested in the TAPS test) will also mean unpredictable changes in the phonemic content of the test material.

The child's age and developmental level:
The development of a child's vocabulary and use of speech as symbols should be considered. The younger the child the

more realistic the representation of the object needs to be (Sheridan, 1960). The test words used should be known by the majority of the hearing impaired children of the age to be tested. There is a recognised developmental path of attention control in normal children (Cooper et al. 1978). The test needs to be short, some hearing impaired children have very short attention span for auditory material. The tester has to be prepared to expend considerable effort on gaining and maintaining the child's attention on task.

Test design features:

♦ *Test material*. This must be suitable for a small child to handle. It is difficult to decide whether to use toys or pictures. Pictures need to be large, robust and resistant to creasing, chewing etc. and must be clear, colourful and attractive representations of the objects, not cartoons or caricatures. Toys are difficult to duplicate and transport and are breakable and expensive. The items need to be adequately separated when presented to the child, so that the tester is clear which response is being indicated by the child.

♦ *Recorded / live voice / monitored live voice.* Recorded live voice is the preferred method of delivery for work with adults. The advantage of recorded material is that the test items are delivered in a standard fashion. Young children find recorded material rather intimidating and can have unrealistically low scores. The advantage of live voice is the rapport with the child. Visual clues can be used to indicate the onset of the test stimulus. Monitored live voice can be the best solution, as the voice can be accurately measured, while maintaining the rapport with the child (Tyler 1993).

♦ *Carrier Phrase*. The words may be presented with a carrier phrase, or in isolation. It is important with children to use a natural style; the use of a carrier phrase presents more complicated speech to the child but is closer to the everyday experience of the child. It also enables the tester to maintain the child's concentration throughout the test (Olsen and Matkin, 1979).

♦ *Auditory / Visual.* Most tests are delivered in an auditory alone paradigm. All visual clues have to be eliminated to assess the child's use of audition. Lip-reading is prevented by the tester covering his mouth with his hand or a screen. This immediately presents a problem with deaf children, who are taught to watch mouths and make use of lip-reading, facial and visual clues to help them understand speech.

♦ *Open set / closed set*. In open set tests there is no limit to the responses that can be given, in closed set the alternatives are presented to the subject. Closed set tests are better for speech discrimination tests in young children as they are able to identify the test items, even if they are unable to name them. The limitations of the subject's speech production, and the tester's subjective interpretation of the response are therefore eliminated. Closed set tests can be repeated over time.

♦ Training / learning of test materials. The results depend to some extent on the familiarity of the test materials. Some tests are strongly influenced by learning factors. In closed set word discrimination tests learning factors do not affect the outcome. In some situations, e.g. children with cochlear implants who need repeated testing, the learning factor can be helpful. The knowledge of the vocabulary will make the test easier for the child.

♦ *Test format*. This needs to be simple and focused. Small children are unable to cope with many variables and are easily distracted or confused during a test if there is superfluous material. Some test paradigms are unsuitable for the younger age group, e.g. the oddity paradigm - identifying the odd one out - is not within their ability at early developmental stages.

♦ *Test instructions.* Thought has to be given as to how the instructions are going to be conveyed to the profoundly deaf child. Their understanding of speech may be limited, and the test material may need to be introduced using sign, gesture and improvisation. It may also be difficult to be sure that the child has understood the test instructions.

♦ *Standardisation*. The test needs to be presented in (as far as possible) a standardised way each time, to prevent differences in response related to the mode of presentation of the test. This can present problems when live voice tests are used as inevitably different testers vary in the test procedure. The instructions need to be detailed and precise. Either live voice or recorded speech must be standardised. However work with very small children can have so many other limitations that the design of a standardised test may be unachievable for deaf children with a limited linguistic repertoire.

♦ *Test / retest reliability.* Trials of the same material with the same subjects, repeated over time, or of the same material with different subjects will give an idea of reliability of the test. Equivalent lists may contain the same items in different order.

♦ *Test construction.* Consideration must be given to the number of items, the child's attention span, knowledge of the test items. Statistical methods, e.g. the binomial theorem, can be used to demonstrate that the results obtained have some validity.

Design of Smile test

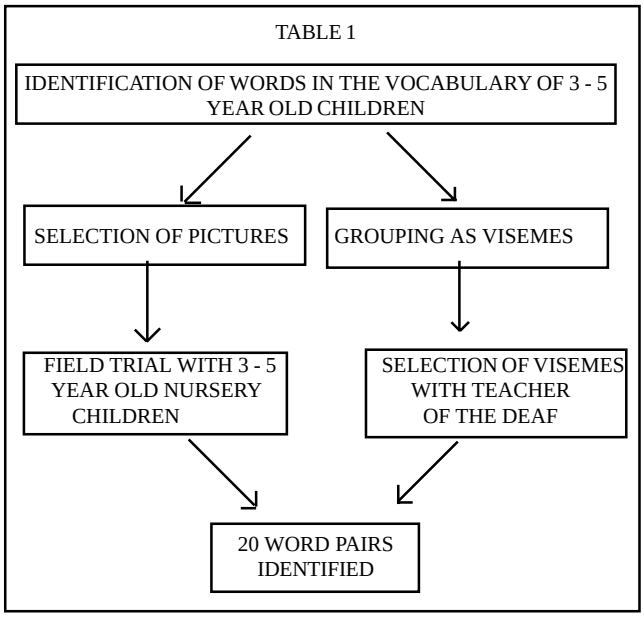
The Smile test consists of 20 pairs of pictures, which are visemes i.e. "visually homophenous (visually similar)" pairs. The picture pairs are shown to the subject, who is asked to identify one item out of each pair, using measured live voice, with the mouth uncovered. A raw score is obtained, and from this a measure of the level of likelihood that the child is using audition to identify the pictures can be obtained.

It was decided to call the new test the 'The Smile Test' because it reminds the tester that children will respond better to a

friendly atmosphere and a positive approach. A smile will blur any remaining lip-reading clues. It is also an anagram of the devisor's name. The Smile test is based on a subtest of the TAPS test. The test should determine the extent to which the child is relying on visual clues to recognise words, and the extent to which he/she is able to use his hearing to differentiate visually identical words. It is not a test of lip-reading, but rather an attempt to exclude lip-reading as a source of information to identify the words. This test is designed to demonstrate the child's ability to integrate visual and auditory information.

Development of test:

Table 1 shows a flow chart of the development of the test.



Pairs of words were sought that were visemes and in the vocabulary of 3 -5 year old children.

♦ *Identification of words* in the vocabulary of 3 - 5 year old children. 179 words were identified from a variety of sources.

♦ *Selection of pictures***.** The pictures were taken from Ladybird books, speech therapy material and amateur photographs. A number of words had to be rejected immediately as it was not possible to provide a simple picture in which it was clear which item needed to be identified. Problems were met with for some nouns, e.g. representations of paint, spot, pin. Numbers, prepositions (one, up) were also unpictureable in a way that was meaningful to children as young as three years. It was decided not to add the written name of the pictures as it is important in this "user friendly" test that children struggling with literacy skills are not discouraged by thinking their reading is being tested.

♦ *Field trial* with 3 - 5 year old Nursery Children. 64 words, 71 pictures, were collected, and shown to a group of 24 normal hearing children aged 3-5 years old, who were considered to have normal speech and language development (attending North Kensington Nurseries) to check the words were in their vocabulary. During this field trial it became clear which words were familiar to the children, and which pictures were presenting difficulties.

♦ *Groupings as visemes*. The words were grouped into approximate viseme groups. The first plan was to identify 5 groups of a minimum of 4 visemes (e.g. man / fan / bag / pan). However it became clear that it would not be possible to find sets of 4 (or even 3) true visemes that would be in the vocabulary of 3 -5 year olds. It was finally decided to look for pairs of words (diads) that were visemes that could be used for the test.

♦ *Selection of visemes* with teacher of the deaf. A strict definition of visemes was adopted. The words needed to have

- ❖ rhyming central vowel
- ❖ whole vocal tract in a similar position for the vowel
- ❖ consonants needed to be identical on the lips and the other articulators, especially the tongue.

They were tried before an experienced lip-reader (who is considered to be in the top 10% of lip-reading ability using the CUNY/UCL Videolaserdisc lip-reading test, 1996), and the pairs were only accepted if she was unable to discriminate the words without the use of her hearing aids.

20 word pairs identified. 20 Diads were needed for statistical validity to form the SMILE test. The ones chosen were those that best fitted the criteria of

- ❖ phonemic balance
- ❖ familiar pictures
- ❖ not lip-readable

cat / hat	**hat / hand**	**pan / man**	**bag / back**
bath / path			
bear / pear			
tea /sea	**meat / peas**	**tree / green**	**pea / bee**
pin / bin	**chip / ship**		
dog /doll			
toes / nose	**goat /coat**	**boat / bone**	**rose / road**
moon / boot	**shoes / juice**	**nuts / sun**	

Test Procedure:

The test material consists of a ring binder with the pictures of the 20 word pairs mounted on A4 sheets, so that each pair could be shown to the children, who are invited to chose one of the two pictures. The test is first explained verbally to mother or carer and child. The child is seated at a small table about 1 metre opposite the tester in a sound proofed room (ambient noise less that 35 dBA). Each picture pair is shown to the child, and he is asked by speech, sign and/or gesture to identify them. During testing the pictures should be held below the tester's face so that child can see lips and pictures simultaneously.

The tester says: **"What is this?"** pointing to the salient part of the picture. If the child names the item the tester repeats it. If the tester is satisfied that the child knows both the items of the pair the test proceeds; if child does not know one of the items the tester discards that pair. Once established that the child knows the items the tester asks the child to identify one of them saying:

"Show me the....."

with the mouth uncovered, using a natural unforced smile to blur any residual lip-reading clues, at a speech intensity range of 55-65 dBA (a sound meter should be placed or held by the child's ear to check this sound level). If the child says

"what ?"

or otherwise asks for a repeat the tester may repeat the request once, using the same level of vocal intensity. After that the tester should smile, and say something like

"You show me....." or **"That's what the game is all about!"**

The tester accepts the child's responses, being careful not to indicate by attitude or facial clues which are correct or incorrect. It must be remembered hearing impaired children are very aware of non-verbal clues. The tester has to use some effort to maintain the child's concentration.

The child is first shown plate / plane, gate / cake, bed / pen to familiarise him with the test procedure. Once the tester is satisfied the child is able to co-operate the test can proceed. All the test 20 picture pairs should be shown to the child. During the test the tester records the results on the score sheet, ticking ⌣⌣ the correctly named test item, putting '\' through an incorrectly named item, and striking through the pair if they were not correctly identified, and therefore not used in the test. A comment is made on the child's reaction to the test in terms of mental state, level of cooperation.

Once the test is completed a note is made of how many pairs were used, and how many pairs the child correctly identified. Both a raw score and an estimate of the likelihood that the child is using audition are obtained.

Statistical analysis:

The results can be analysed using the binomial probability distribution (Olsen and Matkin, 1979). Conditions required for use of binomial probability distribution are:

- A sample of n experimental units are chosen without replacement (i.e. once an experimental unit has been chosen it cannot be used again).
- Each experimental unit possesses two mutually exclusive characteristics - in this case the choice between one of two pictures. The response can be called correct or incorrect, or success or failure.
- The probability that a single experimental unit possesses the success characteristic is the same for all experimental units.
- The outcome for any one experimental unit is independent of the outcome for any other experimental unit.

The random variable k counts the number of successes in the trials, here presentations of the test items. Table 2 shows the exact probability from the binomial distribution of a child answering k or more questions out of n presentations, under the assumption that the answers are made randomly (i.e. probability that an answer is correct is 0.5). For example if 19 visemes are used the probability of getting 14 or more correct is 0.032. A simplified table (Table 3) was devised to put on the individual child's test results sheet. This table shows the four groups of degrees of evidence (from 0 to 3) that the subject is using hearing to discriminate the visemes. Using this table the tester can quickly check the degree of likelihood that the child is using audition by checking the test result (number of correct answers and number of test pairs presented).

Pilot of Smile Test:

The test was piloted to check that it could be used with the target group, and to highlight any test design problems.

- *Subjects*. The SMILE test was used with 15 children aged 3.8 to 11.7 years, who were attending the Nuffield OP department, or Community Paediatric audiology clinics in N. Kensington. All the children used speech as their only or principal means of communication, with English as a first language.

- *The results* of the test comprise:

i. Raw score: the number of items identified correctly out of up to 20 pairs.

ii Using the table (Table 3) the tester can see

- ❖if the test is valid (enough pairs used to give a result)
- ❖the level of evidence of audition.

Table 4 shows the results. The children are listed in order of the severity of their hearing losses, from normal to profound.

- *Children's response to the test*. All the children (whatever their hearing level) completed the test with little or no difficulty and appeared to enjoy the test. It was relaxed and easy to do the test without covering the mouth to hide lip-reading clues. The children felt they were playing a game and had a sense of success even when failing (this should be treated with caution; deaf adults report disappointment when they realise what a disability they have after years of being praised for their efforts in responding to paediatric audiological tests. "Well done!" means to the tester "Good effort" and to the subject indicates that his hearing level is improving, which may not be the case). The children with the more severe losses show less evidence of audition. The results show that the 4 children with 4 frequency average hearing levels (aided or unaided) of <20 dBHL scored 19 or 20 items out of 20 pairs, giving a top score of 3, very strong evidence the child is using audition.

The children whose 4 frequency average hearing losses (unaided) were profound, >70 dBHL (Northern and Downs, 1991) had lower scores, for example 10 /17, 9 /16, giving a score of 0 - 1, no or some evidence of using audition. The children with hearing levels between these two extremes gave a scatter of results.

♦ *False Negative results.* If a child is not using audition to identify the test items he would be expected to achieve a score of 50% due to chance. If a child fails most of the test items it is likely that he is deliberately choosing the wrong items, and one could surmise that he is using audition.

Discussion and further development

The test could be further developed and consideration should be given to the following factors:

Test construction

♦ *Quality and clarity of the pictures.* A more professional test pack could be devised. The use of a digital camera that enables the images to be put onto computer and modified, would give better pictures. A toy based test could be developed for younger children, who would relate better to toys than to pictures.

♦ *The language content* could be further analysed. The words could be analysed and weighted for auditory challenge. The frequency balance should reflect spoken English so that the test will reflect the subject's hearing for everyday speech. There are differences in voicing and voice onset times. Deaf children use different clues to pick up the differences between words. Children favour some items in the test pairs;. food, familiar items or colourful or attractive pictures may be preferred. At present there is no weighting in the test material for these factors.

♦ *Test vocabulary*. A larger trial of the vocabulary of deaf children needs to be undertaken. The field trial of the words to check their familiarity was carried out with hearing North West London children and gives an indication of the suitability of the vocabulary. It is acknowledged that the vocabulary content of hearing and deaf children can be different (Bishop et al, 1988).

♦ *Live voice / monitored live voice / recorded voice.* The test is delivered live voice. The test material could be presented as a computer test; either monitored live voice, to maintain the rapport with the younger children, or using recorded voice. This would improve the standardisation of the test.

♦ *The lip readability.* One teacher of the deaf assessed the word pairs for lip-readability. The lip-readability of the viseme pairs should be assessed more extensively.

Standardisation and Validation:

♦ *Subjects.* The subjects are varied in chronological age, developmental age, hearing loss and effectiveness of amplification. A full field trial controlling for these factors would be needed to validate the test.

♦ *Randomisation*. A random choice of test item was made by the tester for each test pair, trying to keep an equal number of requests for left and right hand pictures to avoid bias. The test protocol could include a specification of which picture out of each pair is requested. Several lists would be needed so that if the test is used repeatedly children would not learn which items are requested.

♦ *Test / retest measures* have not been done as part of the development. If there is high test / retest variability the number of test items may need to be increased to maintain validity.

♦ *Comparison with existing tests.* There is a comparison with the child's hearing level as assessed by the audiogram, aided and unaided. The relation between pure tone audiometry and speech discrimination tests is considered poor. The Smile Test needs to be compared to an existing standardised test. The Subtest 4 of the TAPS Test is comparable, but has not been standardised. The HAVE Test (Osberger et al, 1991) is very similar to the TAPS Test.

Dr. Margaret Miles, MB.BS, MSc

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Table 2 The probability of k or more correct answers out of n questions

	k											
n	20	19	18	17	16	15	14	13	12	11	10	9
20	3 <0.0001	3 <0.0001	3 0.0002	2 0.0013	2 0.006	1 0.021	1 0.058	0 0.132	0 0.25	0 0.41		
19		3 <0.0001	3 <0.0001	3 0.0004	2 0.002	2 0.0096	1 0.032	1 0.084	0 0.18	0 0.32		
18			3 <0.0001	3 0.0001	3 0.0007	2 0.004	1 0.015	1 0.048	0 0.12	0 0.24		
17				3 <0.0001	3 0.0001	2 0.0012	2 0.006	1 0.025	1 0.072	0 0.16	0 0.31	
16					3 <0.0001	3 0.0003	2 0.0021	1 0.011	1 0.038	1 0.058	0 0.23	
15						3 <0.0001	3 0.0005	2 0.004	1 0.018	1 0.059	0 0.15	
14							3 0.0001	3 0.0009	2 0.0065	1 0.029	1 0.090	0 0.21
13								3 0.0001	2 0.0017	1 0.011	1 0.011	0 0.13
12									3 0.0002	2 0.003	1 0.019	1 0.073

Grouping according to degree of evidence:-

0	-	p>0.1	No evidence
1	-	0.01 < p <0.1	Some evidence
2	-	0.001 < p < 0.01	Strong evidence
3	-	p < 0.001	Very strong evidence

Table 3 Degrees of evidence that the child is using audition

Number of Presentations	Number of correct responses: 9	10	11	12	13	14	15	16	17	18	19	20
12	1	1	2	3								
13	0	1	1	2	3							
14	0	1	1	2	3	3						
15	0	0	0	1	1	2	3					
16	0	0	0	1	1	2	2	3				
17	0	0	0	1	1	2	2	3	3			
18	0	0	0	0	1	1	2	3	3	3		
19	0	0	0	0	1	1	2	2	3	3	3	
20	0	0	0	0	0	1	1	2	2	3	3	3

Using Number of Correct Responses and Number of Presentations Find Number and Ring Box

0 Not using Audition p>0.0: no evidence

1 May be using Audition 0.01<p≤ 0.1: some evidence

2 Probably using Audition 0.001<p≤ 0.01: strong evidence

3 Using Audition p≤ 0.001: very strong evidence

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Table 4 Results of the Pilot Test

SUBJECT	AGE	HEARING LOSS (in dB HL)		SCORE	RATING				COMMENT
		Unaided	Aided		0	1	2	3	
1	5.6	<20	-	20/20				☐	previous glue ear
2	7.5	<20	-	20/20				☐	
3	5.3	<20	-	19/20				☐	speech delay
4	9.8	46	15	20/20				☐	
5	11.3	60	20	20/20				☐	
6	9.3	48	28	17/19				☐	tested without hearing aids
7	3.8	55	28	15/17			☐		
8	11.7	78	30	20/20				☐	
9	7.5	75	34	12/16		☐			mild dev. delay
10	7.4	62	36	14/17			☐		mild dev. delay
11	8.5	85	40	13/20	☐				
12	6.6	98	41	16/19			☐		one aid only working
13	7.0	>100	54	8/16	☐				
14	10.2	>100	64	10/17	☐				mainly signing
15	5.3	85	n.a.	9/16	☐				one aid only working

Audit of Children under review in Community Paediatric Audiology Clinics in the Mid Sussex NHS Trust 1995/96

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SUBJECT OF AUDIT

The main aim of any Community Paediatric Audiology service is to provide prompt and accurate assessment of children with suspected hearing loss and then to provide appropriate advice, monitoring, referral onto and liaison with other services including the Primary Health Care Team, Tertiary Audiology clinics, ENT, Education and Social Services. This audit was designed to look at several aspects of this service within the Mid Sussex NHS Trust.

The **challenge** for service provision is that **Otitis Media with Effusion (OME)** is very common in early childhood (point prevalence of 15-25%, with cumulative incidence at over 80%)[1]. As most cases will resolve spontaneously, a period of "watchful waiting" is frequently advocated, whereas **Severe Congenital Hearing Loss** is rare (1-2 per 1000) and difficult to recognize in young children. However, early diagnosis is of great importance to maximize linguistic, social and cognitive skills. **The two conditions may coexist**.

BACKGROUND

The Community Paediatric Audiology Service in Mid Sussex

This is administrated by the Child Health Bureau (CHB) in Chichester. A well established computer system serves the West Sussex Child Health Service. This system includes a Special Conditions File which records data on children with significant medical conditions - including those with permanent hearing loss. However there is no database for Community Paediatric Audiology - clinics are organized manually and very little detailed information is available on the service. The total population served (0-18yrs) is approximately 33,000 (Sept. 1996: Source: CHB). Community Paediatric Audiology is a very small speciality. In the year to 31.3.96, 1630 children were seen in 207 sessions across the Mid Sussex NHS Trust by WTE 0.45 medical staff and approximately 10 hours per week nursing time. All clinics rely on help from the Teachers of the Deaf who assist with at least one clinic per month in each locality. Full details of the service are available elsewhere[2].

Standards

In 1993 the National Deaf Children's Society (NDCS) consultation document[3] listed targets for quality standards in Paediatric Audiology. In particular a target was set to detect **80%** of bilateral congenital hearing impairment in excess of 50 dBHL (averaged across the frequencies 500Hz, 1 kHz, 2kHz, and 4 kHz) within the first year of life and 40% by the age of 6 months.

In 1994 Clare Haddad's extensive Regional Audit of Infant Hearing[4] identified that these targets were not being met and that actual or suspected OME was the most common delaying factor in the diagnosis of permanent hearing impairment in the South West Thames Region. "Watchful waiting" (repeated reviewing by Community Audiologists) was identified as the main cause in delay in detecting some cases. Recommendations were made to prevent this delay and as a direct result of this in 1995 a SWTRAAG (South West Thames Regional Audiology Audit Group) audit[5] looked at multiple reviews This resulted in the regional recommendation that all trusts/districts were to establish a protocol for onward referral to tertiary clinics of children whose threshold hearing levels had not been established within 12 weeks or a maximum of 3 visits. This regional audit also produced data to support the need for local guidelines for various aspects of Community Paediatric Audiology and highlighted the need for a future re-audit.

In 1994 SWTRAAG set standards on waiting times for appointments in Community Paediatric Audiology Clinics. Staff in the Mid Sussex NHS Trust were concerned that these standards were not being met locally whereas in other trusts across the region they were.

Thus there were 2 main issues to be addressed:

1. Were children with a significant hearing loss being inappropriately reviewed in Community Clinics resulting in their diagnosis and subsequent management being delayed?

2. Were children with mild or fluctuating losses, who could have been discharged back to the Primary Health Care Team, being reviewed unnecessarily thus resulting in long waiting times for clinic appointments?

OBJECTIVES

- To establish whether children are being reviewed appropriately in Community Paediatric Audiology Clinics.
- To assess the performance of the service in respect of SWTRAAG (waiting times and onward referral) and NDCS (age at diagnosis) standards.
- To draw up guidelines on the management of children seen in clinics and to instruct staff (medical and nursing) in their use.
- To re-audit following the introduction of guidelines.

METHOD

- Tool, time and financial resources were identified, a laptop computer was borrowed for the duration of the audit and computer training was acquired.
- Quality indicators were identified.
- A database and a data collection form were designed and piloted.
- Data on children with a congenital hearing bilateral loss > 50 dBHL was retrieved from children's notes and collated. *(Because of the small number involved, children from the Crawley/Horsham Trust for which SR also has responsibility were included in this part of the audit).*
- Management guidelines were produced after discussion with Audiological, Community Paediatric, and ENT colleagues.

In May 1996, the guidelines were introduced to all staff working in Community Paediatric Audiology clinics within Mid Sussex and Crawley/Horsham. The notes of all children seen in Mid Sussex clinics in June 1995 were audited and compared to those seen in June 1996. It was possible to obtain the notes for **every child** seen within the two audit periods. This reflects the efficiency and cooperation of the administrative staff for audiology at the Child Health Bureau in Chichester. The results were analysed using Access (2.0) and Excel (5.0)

SUMMARY OF RESULTS

1. Number of children seen

The total number of children seen was very similar in both June 1995 and June 1996 although the numbers seen in each locality varied.

2. Ages of Children seen

In both years the majority of the children seen were in the preschool age group. It is this age range that requires specialist Paediatric skills.

3. Referrer

Children were referred by many different professionals but the majority in both years were referred by health visitors.

4. Main Reason for Referral

Failed screening and parental concern were the most common reasons for referral in both years.

5. Visit Number

The vast majority of children (87% in 1995 and 80% in 1996) were being seen for the first or second time. Multiple reviews (i.e. children being seen for the 4th or more occasion) made up between 7% and 11% of clinic work load. The great majority of these children had a clearly justifiable reason for multiple review e.g. known permanent loss/Down's Syndrome.

6. Thresholds

At the audit visit threshold hearing levels were established in 85% of cases.

7. Degree of Hearing Loss

Some degree of hearing loss was confirmed at the audit visit in 44% of children in 1995 and 35% in 1996

8. Overall Outcome

	1995 (%)	1996 (%)
Discharged, no action	41 (42)	44 (45)
Review, no action	47 (49)	44 (44)
Discharged/reviewed + referred ENT	9 (9)	11 (11)
Discharged/reviewed + referred tertiary	0 (0)	0 (0)
TOTAL	97 (100)	99 (100)

Quality Indicators and Data Collection Procedures

AUDIT TOPIC: Community Paediatric Audiology Clinics

Objective 1: To ensure that children are being reviewed and referred on appropriately - SWTRAAG guideline.

2: To compare waiting times for initial clinic appointments with SWTRAAG standards #.

Time Period: June 1995 Number of Children: 97

June 1996 Number of Children: 99

ASPECT OF SERVICE	STANDARD	ACHIEVED	EXCEPTIONS	DEFINITION AND INSTRUCTIONS
Establishment of hearing thresholds within 12 weeks of initial assessment or referral to tertiary clinic	**100%**	1995 1996 **89% 93%**	Parental refusal	Using database, identify all children whose hearing thresholds had not been established at audit visit if 12 weeks after initial visit and check for referral
Reason for multiple review to be clearly stated in notes	**100%**	1995 1996 **80% 100%**	Nil	Using database, identify multiple review children, where multiple review + > 3 visits including initial, check for details
#Waiting time for initial appointments <18mths of age : within 6 weeks 18mths - 3 yrs : within 8 weeks >3yrs <5yrs : within 12 weeks >5yrs : within 17 weeks Overall* : within 9 weeks (*Patients' Charter)	**100%** **100%** **100%** **100%** **100%**	1995 1996 **31% 12.5%** **20% 50%** **74% 55%** **87% 94%** **63% 53%**	Referral date unknown Appointment delayed at request of Audiological Physician or referrer	Using databases, calculate waiting time for each child according to age at date of referral

Objective 3: To compare age at diagnosiswith NDCS standard for permanent losses.

Time Period: March 93 - March 1996 Number of Children: 15

ASPECT OF SERVICE	STANDARD	ACHIEVED	EXCEPTIONS	DEFINITION AND INSTRUCTIONS
Age at detection of congenital bilateral hearing loss >50dBHL	**40%** by 6mths **80%** by 1 yr	**40%** by 6mths **60%** by 1 yr		Review notes of all aided children, using Special Conditions file and information from Sensory Support Team

9. Waiting times for initial clinic appointment

% of initial visits which met SWTRAAG Standards

	1995	1996
<18mths	31%	12.5%
18mths - 3yrs	20%	50%
>3 yrs < 5yrs	74%	55%
>5yrs	87%	94%
Overall	63%	53%

In no age groups in either year were these standards met in 100% of cases. **The situation was much worse in the younger age group where early identification is vital.** There had been a deterioration between 1995 and 1996.

10. Age at detection of assumed congenital bilateral hearing loss >50dBHL in better ear

- Over the 3 year period 1993 – 96, 15 children were identified to be in this category.

- Of these 6 (40%) were diagnosed by 6 months of age (i.e. met NDCS standard).

  A further 3 (20%) were diagnosed by 1 year (total 60% – standard 80%). For the remaining children waiting times for clinics was a contributory factor to delay in some cases

There is no directly comparable data from previous years but prior to 1993 no child had been diagnosed under 6 months in what was then the Mid Downs Health Authority (now Mid Sussex and Crawley/ Horsham Trusts). Clare Haddad's audit[4] revealed that across the region only 8% of children were diagnosed by 6 months and only 23% by 1 year. Thus the age at identification of congenital hearing loss appears to be improving, largely as a result of at risk neonatal screening.

CONCLUSIONS

This was the first time detailed information had been obtained on many aspects of the Community Paediatric Audiology Service in the Mid Sussex NHS Trust. From the data analysed our conclusions are:

- Children are not being reviewed unnecessarily
  - Thresholds are quickly established
  - Children are referred on promptly when indicated
- Note keeping has improved
- Waiting times for initial appointments are well below regional standards
- Age at diagnosis of congenital loss appears to be improving

RECOMMENDATIONS

l. A confidential secure database which makes re audit and administration of this service easier should be incorporated into the new community system which at present is being planned.

2. The waiting list problem needs to be seriously and urgently addressed by managers and purchasers as this audit clearly demonstrates that the present resources are being used efficiently.

Dr. Maggie Bruce and Dr. Susan Rose

References

[1] "Screening Children's Hearing – a review of the literature and the impact of Otitis Media" M.P.Haggard & E.G.Hughes. Pub.HMSO Dec 1991

[2] "The Community Paediatric Audiology Service (Mid Sussex A, Crawley/Horsham Trusts)" S. Rose. 1996

[3] "Quality Standards in Paediatric Audiology Part I. Guidelines on Standards for Early Identification of Hearing Impairment in Children" National Deaf Children's Society Consultation Document. June 1993

[4] "The Screening and Surveillance of Infant Hearing in the South West Thames Region." Clare Haddad. Nov 1994

[5] "Regional audit of Children attending 2nd Tier (Community) Paediatric Audiology Clinics for Multiple Review: March/April 1995". M.P.Bruce