ABSTRACTS
Presentations at the BACDA Study Day: "Eyes and Ears"

Usher syndrome: Diagnosis, disclosure and information needs for young people and their families

Mary Guest

Deaf Genetic advances in locating and identifying genes for Usher Syndrome have raised the profile of this major cause of deafblindness in adults. However, there is no medical treatment either to arrest or reverse this condition yet. Disclosure and long-term informational needs for young people and their families are discussed in light of this scenario.

Clinical and genetic types of Usher syndrome (after W.J. Kimberling 1998)

Clinical 1 Type	Sub type	Chrom.	Freq. %	Audiologic	Vestibular	Retinal
1	1a 1b 1c 1d 1e 1f	14q 11q 11p 10q 21q 10q	10 81 6 <1 <1 <1	Congenital and profound	Congenital and severe	Pre-puberty
11	11a 11b 11c	1q41 5q ?	83 5 5	Congenital, stable, sloping mild to profound		Early teens Late teens Pre-puberty
111	111	3q	(7)	Progressive	Progressive	Variable

Usher Syndrome: disclosure and information needs

Pre-diagnosis child/young person presents as:

1. Profoundly deaf
- ½ late motor development
- ½ extinguised vestibular
- ½ gait ungainly

2. Partially hearing
- ½ normal milestones
- ½ normal vestibular response
- ½ normal gait

<———— ½ poor night vision ————————>
<———— ½ in daylight stumbles over low objects ———>
<———— ½ central visual acuity normal ————————>

Generally perceived as a hearing impaired child with no severe visual impairments

DIAGNOSTIC PROCESS need to understand:

- ½ impact of diagnostic tests e.g. dark adaptation, dilation of pupils on deaf child
- ½ how diagnosis given to family members
- ½ what information given influences future attitude
- ½ leads to blindness+ blind registration+ no cure+ very rare+ learn braille+ blind at 25 = no future
- ½ family members require a construct which helps them see a future
- ½ child/young person's perception? What do they understand?

USHER SYNDROME: DISCLOSURE AND INFORMATION NEEDS (cont.)

Post diagnosis

- **child/young person needs age/language appropriate information not secrecy**
- **understanding of how their vision is functioning for child/young person/family**
- **suggestions to overcome practical problems e.g. improved lighting, marking hazards**
- **compensatory strategies: crossing roads, making yourself visible, scanning**
- **need contact with well informed professional e.g. teacher, audiologist**
- **all need somewhere to shed their feelings**

Central Visual Processing

Gary Green
Reader in Neurophysiology
Newcastle Medical School

This talk will review current thinking about the structure and function of the central visual system. The division of the central system into basic Parvo and Magnocellular systems will be described and the cortical analysis of images will be briefly outlined.

The development of the central visual pathway and how this may be disrupted will be discussed. The implications for assessment and therapy will be described.

The effects of damage on the adult visual pathway will be briefly described and again the implications for assessment and management will be reviewed.

A particularly recent improvement in our understanding of how problems in central visual processing can affect language development will be described. Links with problems in hearing are particularly evident in the situation where individuals are characterised as dyslexic. The implications for our understanding of central deafness will be explored in the context of our understanding of vision.

Ophthalmological Investigations in the Diagnosis of Sensorineural Deafness

David Calver

The ophthalmological investigations of all children with sensorineural deafness serves two purposes. Firstly to define in some cases the underlying cause of the deafness and possible ocular disease associated with this and this may involve specialised electro-diagnostic examinations. Secondly to ensure that there are no underlying refractive or motility disorders.

CHARGE

Dr Jeremy Kirk
Consultant Paediatric Endocrinologist
Birmingham Children's Hospital

HISTORICAL PERSPECTIVE

1979

- Hall described 17 children with multiple congenital anomalies with associated choanal atresia.
- Hittner et al. described a similar group of 10 children with coloboma, congenital heart disease & hearing loss.

1981

- Pagon coined acronym.

CHARGE

C oloboma
H eart Defect
A tresia Choanae
R etarded Growth & Development
G enital Hypoplasia
E ar Anomalies

- Diagnosis based on minimum of four out of six characteristics present.

MAJOR CRITERIA – reflect those findings that occur commonly in CHARGE but rarely in other conditions

Criterion	Includes	Frequency (%)
Coloboma	Iris, retina, choroid, disc Microphthalmia	80-90%
Choanal atresia	Uni/bilateral, stenosis/atresia	50-60%
Ear abnormalities (Characteristic)	External ear (lop or cupshape) Middle ear (CSOM, ossicular malformations) Mixed deafness, cochlear defects	90%
CNS abnormalities	I, VII, VIII, IX &/or X	70-90%

MINOR CRITERIA — occur less frequently, or are less specific to charge

Criterion	Includes	Frequency (%)
Genital hypoplasia	UDT, micropenis, Delayed/absent puberty	70-80%
Developmental delay		100%
Heart defects	All types	75-85%
Short stature		70%
Orofacial clefting	Cleft lip &/or palate	15-20%
Tracheo-oesophageal fistula		15-20%
Distinctive face	Broad forehead, facial asymmetry, full nasal tip	70-80%

OCCASIONAL FINDINGS

Finding	Includes	Frequency (%)
Thymic/parathyroid hypoplasia	DiGeorge (22q+ve)	Rare
Renal anomalies	Dysgenesis, horseshoe	15-25%
Hand anomalies	Poly/ectrodactyly	Rare
	Altered palmar creases	50%
General appearance	Webbed neck	Rare
	Sloping shoulders	Occasional
Abdominal defects	Omphalocoele	Rare
	Umbilical hernia	15%
Spinal anomalies	Scoliosis, hemivertebra	Rare

OVERLAP SYNDROMES

Some characteristics of CHARGE overlap with other syndromes:
- VACTERL association.
- DiGeorge sequence.
- Velocardiofacial syndrome.
- Retinoic acid embryopathy (Accutane).
- PAX2 abnormalities.

CHARGE

- Individuals with all four major characteristics or three major and four minor characteristics unquestionably have CHARGE.
- A diagnosis of CHARGE should be considered in any neonate with coloboma, choanal atresia, asymmetric facial palsy, or classical CHARGE ear in combination with other congenital anomalies.

AETIOLOGY OF CHARGE

Associated with one or more features of CHARGE
CHROMOSOMAL:
- Deletion 4q,9p,13q, 22q11.2
- Trisomy 18,21, Partial trisomy 1q,6p,7q,8q
- Tetrasomy 12p

SINGLE GENE: At least 10 disorders described plus some families with familial choanal atresia plus at least 1 additional feature of CHARGE: Burn syndrome, Bamforth syndrome, Lammer syndrome, acro-renal-ocular syndrome.

AETIOLOGY OF CHARGE (3)
TERATOGENS
- Thalidomide, infants of diabetic mothers, fetal valproic acid syndrome, fetal methimazole syndrome and ? environmental agents.

DEVELOPMENTAL
- Intrauterine accident, compression, vascular compromise.

AETIOLOGY OF CHARGE (4)

- Most cases are sporadic, although there is evidence of parent to child transmission, and recurrences amongst siblings born to normal parents suggesting possible germ line mosaicism.
- There is statistically advanced paternal age, with fathers >34 years in 43% of cases.
- None of 38 cases of CHARGE had detectable abnormalities of 22q (DGS/VFCS), 7q36 (holoprosencephaly), or 10q25 (PAX2).

MOTOR DEVELOPMENT

- Mobility at 20-25 months (”five-point crawl” or ”back-shufflers”). Upper body hypotonia may be marked.
- Normal crawlers walk at 35 months (back-shufflers at 57 months).
- Older children often have problems maintaining balance (? vestibular dysfunction).
- Marked delays in motor function likely to be multifactorial: visual field defects, decreased visual acuity, hearing impairment, vestibular disturbance, truncal hypotonia &. chronic fatigue all play a role.

DEVELOPMENTAL & EDUCATIONAL OUTCOME

- Much of the earlier literature suggested that mental retardation was almost inevitable for CHARGE patients.
- Although true for many patients, other types of learning difficulties such as communicative disorder, attention deficit disorder, pervasive developmental disorder & obsessive compulsive disorder have also been noted.
- The degree of early developmental delay may not be a good predictor of ultimate educational capabilities.

DEAFNESS IN CHARGE

- 90% of children with CHARGE have hearing impairment,
- Audiological assessment may be difficult in children with multiple sensory deficiencies.
- Hearing loss may be progressive.
- Thelin has described a characteristic wedge-shaped audiometric pattern which may be unique to the condition.
- Chronic serous otitis media is common, especially in patients with choanal atresia or cleft palate.

SURVEY ON HEARING LOSS IN CHARGE (n=72)

Ear & hearing anomalies	
• Unusual shaped ears	87%
• Small ear canals	71%
• History of otitis media	98%
• Grommets	74%
• Hearing loss	89%

SURVEY ON MANAGEMENT OF HEARING LOSS IN CHARGE (2)

Amplification worn in 83**%**	
• Behind the ear hearing aids	41%
• Bone conduction hearing aids	5%
• In the ear hearing aids	3%
• FM/auditory trainers	46%
• Cochlear implants	0%

”WHO’S IN CHARGE”

- These patients are likely to need multidisciplinary care.
- As with other similar conditions, it is important this care is appropriately coordinated.
- The following are likely to be responsible for coordination of this care:
  - ENT (paediatric) & Audiology
  - General Paediatrician
  - Community Paediatrician
  - Support services such as SENSE.

Jeremy Kirk would be interested in receiving details and DNA on any patients with CHARGE, as they are looking for putative genes.

Family Support Group
Mrs. Sheila Draper, 115 Boundary Road, Colliers Wood, London SW19 2DE. Tel. 0181 540 2142

The Education Assessment Process Throughout The Pre-School Years

Jo Franklin, Advisory Teacher

SENSE Family Centre, Ealing

This presentation will be based on my work as the Advisory Teacher for the Deaf within a multi-disciplinary team at the Sense Family Centre. I shall look at the approach to developmental assessment taken by this team and the key issues behind this method, including the paramount importance of parental involvement and a child-centred outlook. I will discuss the huge range of children we see, and will consider the significance of combined sensory impairments and multi-sensory impairment (including vestibular and proprioceptive disturbance). I shall look at "formal" educational assessment tools and integration of information from "clinical" assessments and how this can best be disseminated. I shall then look at the effects that this information has on the management of such children, looking at amplification issues, the child's environment and the creation of "reactive environments"

Educational Assessment and Management of Children with Multisensory Impairment in the Pre-School Years

The types of children seen at the Family Centre.
- Huge spectrum, not a homogeneous group. Definitions!
- Usually very little known about hearing, sometimes not investigated (N.B. B.A.T.O.D. guidelines)

The way the Centre works. Open referrals.
My role as the Teacher of the Deaf on the team

Assessment
- Needs to be child - centred, multi disciplinary and involve those who really know the child.
- Working with families.
- NB. Hearing loss may very well not be the most important problem child has.
- Can't be prescriptive, can offer guidelines and suggestions based on observations.

A sympathetic environment .
- Place,
- People,
- Time,
- Child's state,
- Distraction-free .....etc.

cf. *Murdoch, H. (1994) "He Can Hear When He Wants To!, Assessment of Hearing Function for People with Leaming Difficulties".*
British Journal of Learning Disabilities. Vol. 22

Pre-assessment observations.
- Summary of child's needs. Initial "Profiles"
- Response repertoires.
- Neurological probabilities and specific medical conditions.
- Awareness levels.

cf. *Mc Cracken, W (1994) "Deaf Children With Complex Needs: A Piece in the Puzzle " Journal of the British Association of Teachers of the Deaf. (18), 2.*

The importance of Sensory Integration.

Some Published Assessment Materials,
- *Callier Azusa Scale (Robert Stillman, 1978. Callier Hearing and Speech Centre, Dallas.)*
- *McInnes & Treffry ("Deafblind Infants and Children: A Developmental Guide", 1982.)*
- *Affective Communication Assessment (Coupe, Barton, Barber, Collins, Levy, & Murphy, 1985, Manchester Education Committee)*
- *Vision for Doing (Aitken & Buultens, 1992, Moray House, Edinburgh.)*
- *Pre-Verbal Communication Schedule (Kiernan & Ried, 1987 NFER-Nelson.)*

The Family Centre Developmental Assessment approach.
Functional Assessment.
So many different reasons for assessment.
What happens to the information? The report.
cf. Etheridge, D (Ed) (1995) "The Education of Dual Sensory Impaired Children" David Fulton: London. Especially Chapter 1, Educational Assessment of Deafblind Learners, Aitken, S.

Management.
- Issues surrounding amplification. Persuading people it's worth it! (professionals and families)
- Management of the environment e.g.. acoustic
- Management of resources, need for 1:1 work, use of intervenors.

Reactive Environments e.g .
- Resonance board
- Little room cf. work of Lilli Nielson
- Sound box

The Most Important Aspects of The Family Centre Approach;

- **Child at the centre.**

Then give that child.....
- **Clarity**
- **Simplicity**
- **Structure**
- **Consistency**
- **and TIME**

Education Approaches to Multisensory Deafness

Norman Brown,School of Education, Birmingham

Having given a brief overview of the diversity of current educational provision and support for individuals with multi-sensory impairment, I shall turn attention to some of the educational strategies used and the theories behind them, together with thoughts on the part played by hearing within the complexity, hopefully leading to discussion of how those in the audience could best contribute to the advancement of knowledge and services for such individuals and their families.

Targeting Greenwich Babies

Ceridwen Deacon, CMO Audiology
Greenwich Healthcare Trust

The Audiology service in Greenwich has had a rather chequered career. In the 1980s and early 1990s it was run as an independent unit within the Child Health Service but the audiologist in charge was not really answerable to anyone and the only medical contact she had was with a consultant ENT surgeon who held a weekly clinic in the community audiology base. In 1993 audiology was drawn more into the fold of community paediatrics and two junior doctors of whom I was one were involved in setting up a working party to look at ways of improving the service. By this time there were long waiting lists due staff shortages, withdrawal of funds when posts were not filled and morale was at an all time low. A new audiologist was appointed who severely criticized our report for not including any mention of neonatal screening. This was taken to heart and a second working party with more senior personnel was convened and looked closely at this question.

The report published in 1996 recommended universal neonatal screening as the most desired option and failing that a targeted neonatal screen. In 1994/5 it had been possible for me to have full time study leave (from a **50%** contract) to go to the Institute of Laryngology and Otology of UCL at Grays Inn Rd. in London and complete the MSc in Audiological Medicine. On my return to clinical work most of my time was allocated to audiology and I became the lead clinician.

At this time we did not have any funds to buy equipment for neonatal screening and so we did the next best thing which was to meet with the acute paediatricians, hospital staff in SCBU and midwives and ask them to refer all babies who fulfilled the at risk criteria. The criteria we adopted were much the same as several other centres were using and can be summarized as any babies spending more than 48 hours in SCBU, those with a family history of sensorineural hearing loss or who had abnormalities of the head and neck and consanguinity. These referrals were sent to audiology where I vetted them and arranged to see them for an assessment at 4 months. If there was a family history of sensorineural hearing loss I contacted the health visitor and asked her to find out more details so that I could decide whether or not the baby should be referred immediately to Guys with whom Greenwich has a contract for a third tier service.

When the babies were seen by me in audiology at 4 months I took a detailed history which included the baby's auditory behaviour and listened to the parents opinion of their baby's hearing. Most babies showed a clear response to sounds and were reviewed again at 7 months when we completed the relevant page in their patient held record.

As a result of all the liaison with the hospital staff both in paediatrics and in maternity it became clear that there was a great deal of support for neonatal screening. So much so that the acute paediatricians encouraged me to put in a bid for equipment through the hospital equipment budget and supported me by saying that they considered it to be essential hospital equipment. With support from my community clinical services manager we put in a bid which was agreed in full.

Within 6 months of starting our "second best" targeted neonatal screening we had an automatic ABR screener from Nottingham, 2 trained health care assistants from SCBU and were beginning a new screening programme in the hospital. This a joint acute and community initiative and to date nearly 70 babies have been tested in the first 4 months. The HCAs work 12 hour shifts and are able to test the babies during the less busy periods. Night duty has been an ideal time as everything is so much more quiet, both in terms of work and acoustics!

The equipment is kept in an office next to SCBU so that babies from the maternity ward and the occasional outpatient can be tested. (These babies would not be allowed into SCBU because of the risk of infection). The programme seems to be running well and in no time it seems I will be carrying out a 6 month audit.

The thing that has been most gratifying about this has been the level of cooperation we have had from everyone in the trust who has been involved, from the director of finance to the maintenance man who secured all the redundant loops of wires and bolted the computer onto the trolley. The staff on SCBU, the paediatricians and the midwives have been wholehearted in their support.

So take heart all you out there who want to start up a new service but have no funds. It is amazing what can be done with existing facilities and what happens when the audiology profile is raised! Our next goal is universal neonatal screening.

Watch this space!

SUMMARY REPORT
FOCUS GROUP FOR PARENTS OF PROFOUNDLY DEAF CHILDREN
A Collaborative Project: Cheshire Community Healthcare Trust East Cheshire NHS Trust Mid Cheshire Hospitals Trust
Commissioned by the District Audiology Group for South Cheshire

1. BACKGROUND

1.1 This focus group was commissioned by the District Audiology Group for South Cheshire with particular support from Dr. Lesley Batchelor, Consultant Community Paediatrician (Audiology), East Cheshire (NHS) Trust (ECT) and Dr. Wendy Floate, SCMO – Audiology, Mid Cheshire Hospitals Trust (MCHT). The organisation of the focus group was undertaken by Carolyn Kural, Clinical Audit Facilitator, at Cheshire Community Healthcare -Trust (CCHT) and Diane Barwick, Clinical Audit Analyst, East Cheshire (NHS) Trust.

1.2 The aims of the focus group were:
a) To identify information needs from the parents' perspective at the time of initial diagnosis of profound deafness and to determine whether these had been met.
b) To identify possible areas for improvement.

1.3 35 invitations were sent out with only four parents accepting the invitation to attend the focus group. One parent who was herself deaf requested a lip speaker and this was arranged through the Cheshire Deaf Society. The meeting was held on 18 March 1998 in Holmes Chapel.

1.4 The following is a summary of the main issues raised by those who attended the meeting as well as background information provided by the paediatric audiologists, teachers of the hearing impaired and speech and language therapists on what actually happens at present.

2. ISSUES RAISED

2.1 Delay from Initial Suspicions of Deafness to Confirmed Diagnosis

2.1.1 The parents were concerned about the length of time it took for their child to be given a confirmed diagnosis of profound deafness. Although there had often been indicators of possible hearing impairment, it was felt that these had not always been picked up or acted upon. In particular, the parents felt that they had not always been kept informed of the professional's suspicions regarding their child's hearing loss.

2.1.2 Children with a profound hearing loss are often diagnosed as a direct result of parental concern. Health visitors have a key role in ensuring that concerns about hearing are referred directly to the paediatric audiologist for early testing. However, the first point of suspicion may arise as a direct result of the health visitor distraction test at eight months.

2.1.3 In young infants who invariably have middle ear effusions, test interpretation can be very difficult. Co-operation between ENT consultants and paediatric audiologists facilitates appropriate management with insertion of grommets where indicated so that hearing losses can be further evaluated by the audiologist.

2.2 Diagnosis

2.2.1 The way in which the diagnosis of a significant hearing loss was given had caused much emotional turmoil for two of the parents. Whilst they accepted that there was no easy way in which to break the news, it was felt that those giving the diagnosis should be properly trained and should be sympathetic to the parent's feelings. More information about why the diagnostic auditory brain stem test (ABR) was necessary would enable parents to decide whether they wished to bring a partner or friend with them.

2.2.2 In practice this may be difficult because only one parent is usually present at the time of the first diagnostic appointment. However, when auditory brain stem testing is carried out, both parents or another close relative/friend should be invited so that if a significant hearing loss is diagnosed, they can be informed together.

2.2.3 Ideally the paediatric audiologist should be on site to offer support, counselling and discussion on management of the child's hearing loss. Technicians performing diagnostic testing should be trained to give appropriate basic information.

2.2.4 A trained paediatric audiologist responsible for the overall management of the child will base the final diagnosis on the results of the behavioural and objective tests.

2.2.5 At the time that three of the children had their auditory brain stem tests, it was necessary to go to another centre, however the test can now be carried out at local hospitals so this is no longer necessary.

2.2.6 Issues about audiological waiting times were raised by the parents and it is acknowledged that clinics are frequently congested and it is felt that this is, in part, due to a high number of referrals for children with mild hearing losses.

2.2.7 The paediatric audiologists consider that there is a need

for a two tier sifting system with more trained paediatricians. Having a universal neonatal screening system and a targeted health visitor distraction test would also help to ensure that children with possible hearing problems are seen earlier by the audiologist and would enable more time to be given to those children for whom there are parental or professional concerns.

2.3 Universal Neonatal Testing

2.3.1 If universal neonatal testing is to be initiated, it was considered important there should be an agreed protocol about who gives the actual diagnosis and how it is given. Training for screeners will be vital to ensure that set protocols are adhered to and that the parents are referred on immediately to the paediatric audiologist if there are any queries.

2.3.2 If a decision is made to introduce universal neonatal hearing screening in this area, it will result in earlier diagnosis and management of profoundly deaf children and some of the problems highlighted by the focus group will become things of the past.

2.3.3 In the meantime, earlier diagnosis and habilitation is facilitated through a locally sensitive service and co-ordinated teamwork.

2.4 Information

2.4.1 Information should be readily available at the point of diagnosis and subsequently as needed around:
• What the diagnosis means for the child and the family
• What support is available, how it is accessed and how it will help the child
• What options are available for the child's education
• Which benefits the family may be eligible to claim
• Support groups both nationally and locally

2.4.2 It was suggested that much of this information could be contained in a ”Who’s Who” guide.

2.4.3 Involvement of the teacher of the hearing impaired as soon as possible after diagnosis is imperative. They will be able to work with the parents and the audiologist and support the parents through the early days after diagnosis.

2.4.4 Some districts also involve the social worker for the hearing impaired. Information about the National Deaf Children’s Society is also given to parents at this time.

2.5 Speech and Language Therapy

2.5.1 There was great concern among the parents about the limited amount of speech and language therapy that was offered to these children both at the pre-school level and within the school setting and the difficulty of getting the need for therapy written into the child’s statement.

2.5.2 There were also concerns about the perceived conflict between professionals about whether speech and language therapy was appropriate for a very young child with a hearing impairment. This conflict is acknowledged by the speech and language therapists who feel that there is a divided opinion amongst educational professionals as to the role of the speech and language therapist with hearing impaired children

2.5.3 The speech and language therapists consider that the most effective way forward is close collaborative team working between the speech and language therapist and the teacher of the hearing impaired to produce a worthwhile package of care for these children and that more research is needed within this area of work. Both ECT and CCHT/ MCHT have recently produced guidelines for their local services to enable closer joint working between speech and language therapists and teachers of hearing impaired children.

2.5.4 It is acknowledged that there is a lack of speech and language therapy provision for this group of children. Ideally, an assessment of the child’s speech and language should be made by a speech and language therapist at the early pre-school stage as part of the inter-disciplinary team approach.

2.5.6 At the school stage speech and language therapists consider that therapy for children with a hearing impairment should be an integral part of their educational provision.

2.5.7 Historically there has been different Speech and Language Therapy Service provision in ECT and CCHT. ECT has a limited service but does have a specialist speech and language therapist working within the partially hearing unit who undertakes some pre-school work. CCHT has no specialist speech and language therapy service and only a service to cochlear implanted children within two out of the three partially hearing units within its boundaries. General advice only is offered to pre-school children with a hearing impairment who are referred to local clinics as there is no specialist pre-school domiciliary service available for this client group.

2.6 Education

2.6.1 It was felt by the group that if parents wished their child to go into main stream education, then adequate support from a teacher of the hearing impaired should be readily available.

2.6.2 In schools where there was a separate unit for children with special needs, it was important that teachers address the problem of integration between hearing children and those who were hearing impaired. This did not happen naturally and structures needed to be in place to ensure that integration was encouraged.

2.7 Attitudes of Professionals

2.7.1 Although in the main there were good reports of the

patients' dealings with health professionals, there were instances when this was not the case. The parents felt that the professionals should be trained to break bad news to parents in a sympathetic manner and that there was never any excuse for being abrupt or rude.

3. RECOMMENDATIONS

3.1 At all times professionals should introduce themselves and state their professional group.

3.2 Written information to be given to the parent regarding the diagnostic ABR prior to the appointment for the test.

3.3 Office staff to be thoroughly briefed about the content of the written information so that they can give appropriate advice over the telephone.

3.4 Parents should be invited to bring a friend/relative with them when they attend for the diagnostic ABR.

3.5 A senior member of the Paediatric Audiology staff should be immediately available at the time of the diagnostic ABR to give the diagnosis and first line support.

3.6 At the time of diagnosis parents to be given a contact telephone number. In reality this may be the home telephone number of a senior medical person or the office number of a teacher of the hearing impaired and also the Deafness Support Network.

3.7 Teacher of the hearing impaired to be informed by phone/fax immediately following diagnosis with a view to contacting the family within 24 hours and visiting them within 48 hours.

3.8 "Who's Who" to be compiled and given to all parents at the time of diagnosis.

3.9 In order to maintain a good local service, provision should be made for the updating of equipment.

3.10 Staff performing diagnostic testing and neonatal screeners should receive appropriate training, preferably through attending a "Breaking Bad News" course organised by SCOPE.

3.11 There should be discussion between Health Purchasers, Education and Provider Trusts in relation to pre-school provision and the integration of speech and language therapy into the child's educational provision.

3.12 Main stream schools should foster social and emotional integration between hearing impaired and hearing children.

3.13 Discussions with the South Cheshire Health Authority to continue regarding implementation of universal neonatal screening and targeted health visitor distraction test.

3.14 There should be good liaison regarding early referral and management of children with mixed hearing loss (i.e. development of middle-ear problems in children with sensorineural hearing loss) between general practitioner, medical paediatric audiologist and ENT consultant.

Carolyn Kural/Diane Barwick

Audit of Children Who Failed the Sweep Test for Hearing at School Screening at Primary 1, 4, and 7

Dr. A Joshi
Dumfries & Galloway Child Health Directorate

Introduction

School provides a good opportunity for population screening. Most districts in the UK continue to screen children's hearing when they enter school. Programmes for detection of significant sensorineural hearing impairment in the pre-school years should pick up the majority of such cases long before school entry. Mild sensorineural losses may escape pre-school screens. Otitis media with effusion (OME) will be the commonest cause of hearing loss detected by school hearing screening. A number of professional bodies and working parties on child health have made recommendations on screening protocols.

The Advisory Committee on Services for Hearing Impaired People Report (1981)[1] recommended one universal screen during the first year of school between age 5 and 6, and recommend no subsequent screening except in special circumstances, e.g. no previous test result, children with special needs and children with language delay.

The first edition of 'Health for all Children' edited by *Hall (1989)*[2] recommended that the universal school entry screen should be continued with no further routine test of hearing. The third edition of 'Health for all Children' edited by *Hall (1996)*[3] has recommended the continuation of school entry sweep test with evaluation of the diagnostic and referral pathway and of the sweep test itself with no further routine screening test of hearing.

Professors *Haggard and Hughes (1991)*[4] recommend abandoning any screens at later ages than school entry as the subsequent yield of treatable new OME cases and undetected sensorineural cases is small.

'The Health Needs of School Age Children' edited by *Polnay (1995)*[5] recommended that the current sweep test of hearing on school entry at age five should be retained. Pupils new to a school should be tested if previous results are unknown, as well as children for whom there is parental or professional concern. Children with special educational needs should have access to hearing test and specialist audiology services. There should be a well defined policy on hearing screening and referral with methods of monitoring the results.

The British Association of Community Doctors in Audiology (1994)[6] in its document on Paediatric Audiology Services, recommended keeping the universal pure tone sweep at school entry. The document recognised the variation in screening protocols used by different authorities.

The Scottish Office, *Department of Health (1996)*[7] recommended continuation of sweep testing at school entry. At present in Dumfries and Galloway school hearing screening is carried out in Primary 1, 4 and 7. It is evident from the above mentioned professional bodies that further screening after Primary 1 is not recommended. However due to a high local failure rate in Primary 4 and Primary 7 in the academic year 1996 – 97, it was considered essential to carry out an audit before making recommendations for future screening programmes.

In response to the first edition of 'Health for all Children' edited by *Hall (1989)*[2] The *Faculty of Community Medicine of the Royal College of Physicians (1989b)*[8] recommended indicators for monitoring screen as below. These indicators were adopted for the purpose of this study to evaluate the yield of the screening.

- The number referred to specialists
- The number and percentage of these actually seen by a relevant specialist
- The number and percentage of those seen, for whom treatment was started

Aim

To provide information on which to base future hearing screening programmes for Primary 4 and 7.

Objectives

Identify:-

1. Those children who failed hearing tests and the number who failed initially, at retest and at subsequent screening at each stage
2. The number of children seen by an Ear, Nose and Throat (ENT) specialist
3. The number of children who received treatment e.g. grommets, hearing aids, follow-up
4. Reasons for their failure
5. Children with significant hearing problems first diagnosed in Primary 4 & 7

Method

The study was based on children from a sample of 29 schools, 16 primary schools in Nithsdale and Annandale and Eskdale localities and 13 primary schools in Wigtown locality. Children and schools were a mixture of rural and urban with children from all socio-economic backgrounds. A total of 1368 children in Primary 1, 4 and 7 in Dumfries and Galloway region were involved. This constituted 25% of the Primary 1, 4 and 7 population.

The hearing screening of this sample of children was carried out between September 1997 and January 1998. Screening takes place in two stages with those failing initial screening receiving a repeat test 4-6 weeks later. Names of children failing an initial hearing screening test were collected from test result lists routinely provided to the Child Health Department by the Audiology Department. Data as per the enclosed data form (Appendix I) were collected from case records and the findings were collated and analysed.

Findings

A total of 1368 children were identified and 1358 (99%) screened, the remainder having left the region or been absent at all screening stages. A total of 159 children failed at initial screening.

While all initial failures are reported, the main focus for presentation of findings is on failures in Primary 4 and 7 in accordance with the audit aim.

Initial Failures

Table 1 provides a breakdown of initial failures in each group.

Table 1 Children Failing Initial Hearing Screening

	Total	Screened	Failed	Percentage
Primary 1	494	491	61	**12%**
Primary 4	425	423	61	**14%**
Primary 7	449	444	37	**8%**

Failures At All Stages

Only one child failed screening in Primary 1, 4 and 7 (fluctuating hearing loss due to otitis media with effusion (OME))

Primary 4 Failures

Only 3 of the 61 had failed in Primary 1 also, clinical details for whom are given below:-

1 normal at retest and attending ENT due to recurrent discharge from right ear
2 mild unilateral loss (21-22 dB) due to eustachian tube dysfunction (ETD)

Table 2 - Screening Test Results for Primary 4 Failures

Total No. of children failed the initial screening	61 (14%)
Number of children passed the retest	43 (70%)
Absent or left school	3
Children failed retest	15

Table 3 - Audiometric Evaluation of retest failures (n= 15)

Children with **Bilateral Moderate** Hearing Loss (41 – 70 dB)	0
Children with **Unilateral Moderate** Loss (41 - 70 dB)	1
Children with Bilateral Mild loss (21 - 40 dB)	11
Children with **Unilateral mild** loss (21 - 40 dB)	3

Based on an **agreed local definition of significant hearing loss** warranting referral to the ENT Department, only 5 of the 15 children, i.e. 5 of those with **bilateral hearing loss of 25 dB or more**, met the criteria:

- Hearing loss due to otitis media with effusion (OME) 2 28dB
- Hearing loss due to Eustachian Tube Dysfunction (ETD) 1 30dB
- Awaits appointment at ENT (Family History of Deafness) 1 27dB
- No record of ENT attendance 1 37dB

Table 4 Outcome of all Primary 4 Failures

Referred to ENT	5	
Attended ENT		4
Failed to attend ENT		1
Already attending ENT	2	
No record of attendance at ENT	33	
Seen by Community Paediatrician at Pilot Community Hearing Assessment Clinic	21	10 discharged 11 under review

As children could be referred by their GP following failure at initial testing, the findings in Table 4 represent the outcome for all failures at any stage (n = 61)

28 out of 33 children with no record of attendance at ENT, had normal hearing at retest. 5 children had bilateral mild loss.

The 4 who attended ENT were managed as follows:–
- Already attending ENT and had grommet inserted 1
- Advised grommet insertion 1
- Advised olive oil and was to be reviewed but failed to attend 1
- Awaits ENT Appointment (Family History of Deafness) 1

Primary 7 Failures

8 children (22%) had failed in Primary 4 also but passed Primary I screening. Their clinical details are given below:-

- 2 with hearing difficulties due to ETD, 1 normal at retest, and the other 25 dB bilateral loss
- 2 already attending ENT, 1 child has had a hearing aid since P.4 and 1 was on annual review due to high frequency loss
- 4 with no record of attending ENT, 2 with 21-22 dB and 2 with 30-40 dB hearing loss

Table 5 – Screening Test Results for Primary 7 Failures

Total number of children failed the initial screening	37 (8%)
Number of children passed the retest	15 (41%)
Absent	1
Children failed retest	21

Table 6 – Audiometric Evaluation of Retest Failures (n = 21)

Children with Bilateral Moderate Loss	1
Children with Unilateral Moderate Loss	1
Children with Bilateral Mild Loss	13
Children with Unilateral Mild Loss	6

Based again on the agreed local definition of significant hearing loss warranting referral to the ENT Department, 10 of the 21 children, i.e. with bilateral hearing loss of 25 dB or more, met the criteria.

• Hearing Aid since P4	1	29dB
• Hearing loss due to ETD	1	25dB
• Hearing loss due to OME	1	33dB
• Uncertain diagnosis	1	45dB
• Spurious school audiogram, normal hearing at ENT	1	31dB
• No record of attendance at ENT	5	30-40dB

(See page17 for details of the children with 30 – 40 dB loss)

Table 7 - Outcome of All Primary 7 Failures

Referred and attended ENT	8	
No record of attendance at ENT	15	9 normal at retest 1 mild loss of 22 dB 5 as above, details page 9
Seen by GP	3	2 Said to be normal 1 Normal at testing at DGRI
Seen by Comm. Paediatrician at Pilot Community Hearing Assessment Clinic	9	Discharged - 6 Under Review – 3
Already attending ENT	2	

As children could be referred by their GP following failure at initial testing, the findings in Table 7 represent the outcome for all failures at any stage (n = 37).

The 8 who attended ENT were diagnosed or managed as follows:–

• Grommets inserted	2
• Eustachian tube dysfunction	2
• Low frequency Sensorineural hearing loss	1
• Under review (uncertain diagnosis)	1
• Already attending ENT with OME	1
• Spurious school audiogram, normal at ENT	1

Discussion

The aim of Primary 4 and 7 audiological screening at school is to identify children with significant hearing loss which may have developed since Primary 1 testing. School age screening may have wider benefits apart from detecting new cases of otitis media with effusion and undetected sensorineural cases including unilateral losses which have eluded pre-school detection. The screen can help maintain awareness of hearing problems in school either informally, or by using the screener in a structured staff training programme. The school screen result can be a useful baseline when hearing loss occurs later due to OME. Non-organic hearing loss may be detected by school screen and the school health services are well placed to consider the physical, psychological and social aspects of such cases. Child Protection issues may exist for these children.

Disadvantages of school screening should also be considered. Seasonal variation of catarrhal symptoms may alter the pass / fail pattern. The screen will miss significant OME cases which arise after the screen. Cost of screen and follow-up must be considered. Disruption of class activity may be a problem. Parental consent and confidentiality of test results can raise difficulties.

In many areas of England and Scotland, Primary 4 and 7 screening have been discontinued in accordance with the recommendation of Professors Haggard and Hughes and Professor Hall as the yield of treatable new OME cases and undetected sensorineural cases is small.

In the initial audit proposal, children with non-organic hearing loss (NOHL) were cited as a possible cause for concern should Primary 7 screening be discontinued. This condition has also been referred to as 'functional hearing loss or pseudohypacusis'. This is a condition in which there is an apparent hearing loss in the absence of clinical or audiological evidence: indeed, audiological assessment for inconsistencies is the mainstay of diagnosis. Several studies, *Brooks and Greoghegan (1992)*[9] suggest that NOHL is a manifestation of, or reaction to, stress which could be social, psychological and child abuse meriting detailed investigation and treatment. It was hoped to identify from the audit the number of children with this diagnosis and hence the extent of potential difficulties should screening cease. However, lack of confirmed diagnosis for suspected

NOHL and non-attendance at ENT made it impossible to reach any conclusions.

The screening frequencies and intensity level criteria by which children are deemed to have failed a test vary throughout the country *Lang (1996)*[10] The criteria for failure vary from 20 dB through 25 – 30 dB at one or more frequencies (Appendix II). The Polnay Report (1995)[5] which is generally viewed as a definitive guide to the health needs of school age children suggests the criteria for failure of 30 dB at 500 Hz and 25 dB at 1-4 KHz. The Scottish Office, *Department of Health (1996)*[7] supports the Polnay Report recommendations.

Within Dumfries and Galloway, the screen intensity level currently used is 20 dB at all frequencies of 250 Hz, 500 Hz. 1, 2, 4 and 8 KHz. This may have influenced what appear to be high failure rates in the 1996-97 figures.

Primary 4 Screening

In Dumfries and Galloway, out of 423 children screened, 5 children (1.18%) were found to have significant hearing loss. One child was already attending ENT and had grommet insertion. One was advised grommet insertion and one awaits an ENT appointment in view of family history of deafness. From these findings, it is evident that Primary 4 screening produces very small numbers of children who go on to have active treatment at ENT.

Primary 7 Screening

Out of 444 children screened, 10 children (2.25%) were found to have significant hearing loss. Five out of these (50%) had no records of attendance at ENT after failure in Primary 7 screening. One has had a hearing aid for a while, only one child had grommet insertion due to OME and one had ETD. One child had fluctuating hearing loss (uncertain diagnosis).

The five children with 30 - 40 dB hearing loss who have no record of ENT attendance have an interesting background. 3 of them have failed hearing tests repeatedly since 1994. One of these has major behaviour problems and has attended a Child Psychiatrist. One has social and emotional problems confirmed by the GP. Two had attended ENT in the past and had grommet insertion, one in 1992 and the other child in 1994. Neither has attended ENT since. The fifth child had also attended a Child Psychiatrist due to emotional problems. Although audiometric evaluation of such children is fraught with difficulties it is still necessary for them to have regular follow-up.

The audit revealed that a very low number of children with significant hearing loss requiring further intervention are detected by Primary 7 screening but did highlight the need for improved follow-up procedures for children such as those above.

Conclusion and Recommendations

1. The benefits of school screening are well recognised but because of the low yield of treatable new OME cases and undetected significant sensorineural cases, it is felt that Primary 4 and 7 screening should cease, bringing the service into line with national recommendations and practice elsewhere in Scotland.

2. Existing arrangements whereby health professionals and education staff and parents are free to refer at any time children with suspected hearing loss and 'at risk' of hearing impairment should be reinforced.

3. To minimise children lost to follow-up, an improved system for immediate follow- up of screening failures should be put in place, e.g. Locality based Children's Hearing Assessment Clinics (CHAC) run by Community Paediatricians and an Audiologist.

4. Improved arrangements for accurate diagnosis and follow up of children with suspected non-organic hearing loss should be developed e.g. through a multiprofessional group.

5. Any saving of resources made from retracting Primary 4 and 7 screening should be invested towards the development of CHAC throughout the region.

6. Screening frequencies and intensity level criteria should be reappraised

Acknowledgement

The Author would like to express sincere appreciation and thanks to the audiology staff for providing information, audit facilitator and administrative staff

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APPENDIX I

CHILD'S DETAILS

Name	
Date of Birth	
School	
Class	

Date of First Test -

		500	**1**	**2**		**4**	**Average**
Audiogram	Rt.						
	Lt.						

Type of Loss	Mild	Mod	Severe	Profound	Unilat	Bilat	Only Low/High Freq.

Date of 2nd Test -

		500	**1**	**2**		**4**	**Average**
Audiogram	**Rt.**						
	Lt.						

Type of Loss	Mild	Mod	Severe	Profound	Unilat	Bilat	OnlyLow /High Freq.

Previously failed in - Primary 1 Primary 4 Primary 7 YES NO

		500	**1**	**2**		**4**	**Average**
Audiogram	**Rt.**						
	Lt.						

Type of Loss	Mild	Mod	Severe	Profound	Unilat	Bilat	OnlyLow /High Freq.

Attended ENT - YES / NO

Diagnosis - at ENT

Eustachian Tube Dysfunction
Otitis Media with Effusion
Sensory Neural Hearing Loss
Non-Organic Hearing Loss
Indiopathic
Normal

Management

Discharged
Under Review
Grommet Insertion
- Proposed
– Operated
Hearing Aid
Annual Review
Other

Appendix II

School Hearing Screening in South West England

TABLE 1 (a-d)

a. At what age was hearing screened? (n = 10)

Age	No. of Districts
5	7
5 & 8	l
5 & 11	2

b. Who carried out screening? (n = l0)

Category	No. of Districts
School nurse	5
Screener (not MTO)	4
MTO-Audiology	1

c. Methods of screening (n = IO)

Method	No. of Districts
Pure tone sweep	10
Speech disc. test	I
Tympanometry	0

What is the screen intensity level? (n = IO)

Level in dBHL	No. of Districts
20	5
25	5
30	0

AUDIOLOGY FOLLOW-UP POST MENINGITIS IN CHILDREN UNDER 15 YEARS

Dr. W. Alkass

Lead Clinician: Dr. J Richards – Consultant in Public Health Medicine, West Kent Health Authority

Aim:
To ensure that all children under fifteen years of age, whose usual place of residence was West Kent Health Authority, receive a hearing test within two months of being diagnosed with Meningitis.

Source of data:
Cases of Meningitis reported to Public Health Department at West Kent Health Authority. These cases were checked against individual hospital information systems where possible.

Method:
Specifically designed data collection form was used to collect age; sex; Meningitis strain; date of inpatient episode and discharge; where subsequently referred for hearing assessment and date of outpatient appointment. We also noted whether the patient DNA for hearing assessment.

Time scale:
All children diagnosed during 1994.

Results:
Seventeen cases of meningitis were reported across the four districts. Ten of these children were boys.

The following table identifies cases by districts:

District	Frequency	Percentage
District ”1”	5	29.4%
District ”2”	3	17.6%
District ”3”	7	41.2%
District ”4”	2	11.8%
Total	17	100%

Fourteen (82.4%) cases were diagnosed with meningococcal; two (11.8%) with pneumococcal, and one (5.9%) case of E.Coli strain.

There was no one method of referral for hearing assessment following discharge from hospital.

• Districts 1 and 2 refer to Paediatric Audiology Consultants based at Cobtree Ward, Preston Hall.

• District 3 refers to hospitals and clinics within their area.

• District 4 refers to ENT department within their area.

All but two children were referred for hearing assessment following discharge from hospital. One child was discharged outside the WKHA area and discharge letter was forward to GP in receiving area. One child who was an inpatient on an adult ward received no follow-up.

Time from receipt of referral to appointment date ranged from 16 to 137 days (one child no follow-up; one child discharged out of area. There were no records available for three children) [see appendix i].

Time from Discharge to Follow-up Appointment ranged from 15 to 199 days. Again there is no information on the child referred out of area and for the one for whom no follow-up was made. [see appendix ii].

Ten (58.8%) of the fifteen children attended the follow-up appointment. There was no data available for the remaining five children, however, no child was positively identified as DNA for first appointment offered.

The outcome of the follow-up assessment was as follows:

Outcome	Frequency	Percentage
No follow-up	2	11.8%
Passed hearing test	9	52.9%
One-sided hearing loss	1	5.9%
For retest at later date	1	5.9%
Discharged, hearing expected to improve with age	1	5 9%
Further referred to Paed. Audiology, Cobtree	3	17.6%
Total	17	100%

The three children who were re-referred to Paediatric Audiology were initially followed up by an ENT department

Degree of Deafness Detected

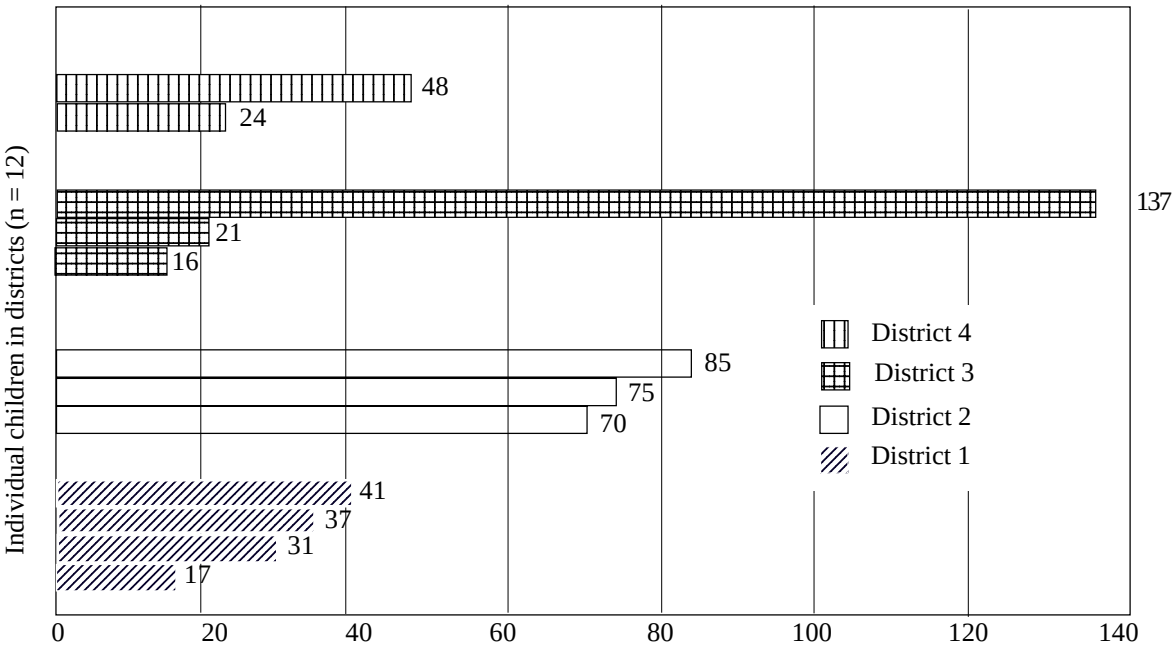
One bilateral sensory neural hearing loss
One left sided hearing loss for annual checks at school
One inconclusive result – for retest at 3 months
One otitis media which is expected to improve with age
One child with some middle ear dysfunction

Recommendations:

That all consultant paediatricians/physicians consider the implementation of a uniform method and mode of referral for hearing assessment for survivors of meningitis pan West Kent area. Whilst the ideal time for testing by audiology department would be prior to discharge this may not be practical in real terms. Dr. Alkass, Consultant Paediatric Audiologist has prepared guidelines for preferred follow-up and these are included for consideration. [appendix iii] Repeat for one year from implementation of Guidelines.

Appendix i

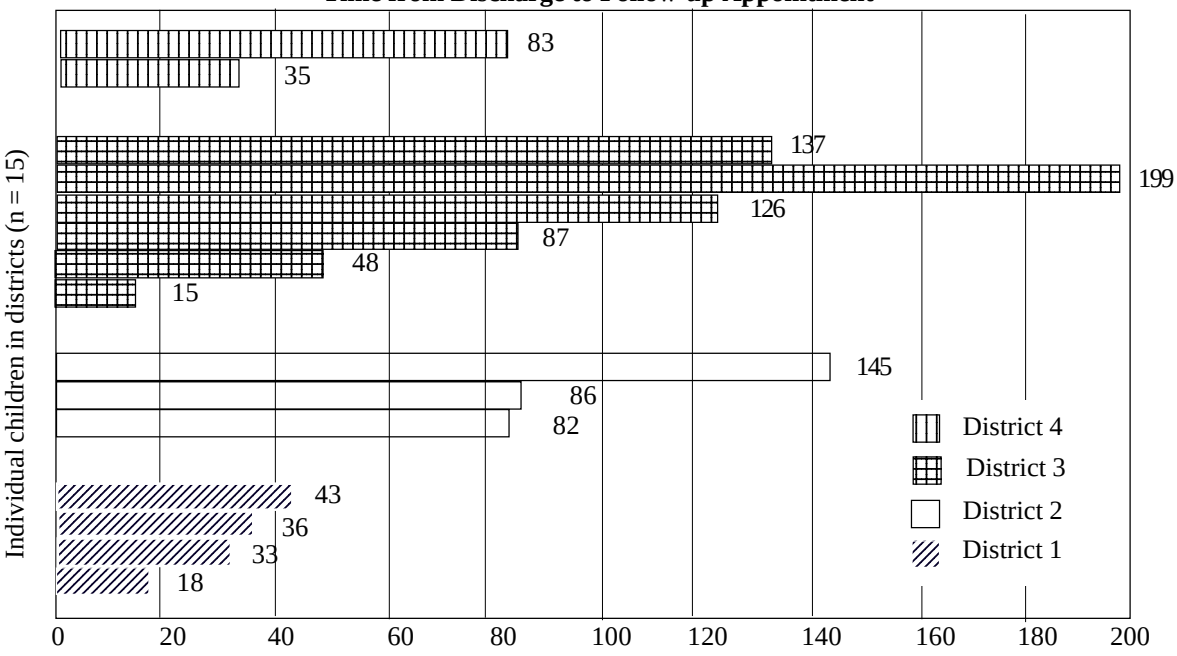
Time from Referral to Appointment



1 child out of area (dist 2)
1 child no f/up (dist 2)
3 children no data available (Dist. 3)

Appendix ii

Time from Discharge to Follow-up Appointment



1 child out of area (dist 2)
1 child no f/up (dist 1)

Appendix iii

REFERRAL TO PAEDIATRIC AUDIOLOGY DEPARTMENT FOLLOWING MENINGITIS

The following child has recently suffered .....meningitis.
Please send an appointment for a hearing test.

Name _____
Address: _____
_____
_____
Hosp No _____
Date of Birth_____ /_____ /_____

Admitted _____ /_____ /_____ Discharged _____ /_____ /_____

Consultant: _____

Health Visitor: _____

Drugs Administered: _____
_____
_____
_____

Neurological Complications: _____
_____
_____

C.S.F. Findings (Especially C.S.E sugar): _____
_____

Signed: _____ Date_____ /_____ /_____

Designation:_____

Please forward to: Audiology Department
Cobtree Ward
Preston Hall
Aylesford
Kent
ME20 7NJ
Telephone number 01-622 790696

Appendix iii

GUIDELINES FOR AUDIOLOGY FOLLOW UP FOR POST-MENINGITIC CHILDREN

Dr. W. Alkass MB, ChB, DCH, MSc Kent Paediatric Audiology Service

The Problem

Sensorineural hearing loss is the most common serious complication of meningitis occurring in about 10% of survivors (Fortnum & Davis 1993). It is also the single most important cause of acquired permanent sensorineural hearing loss in children (Davis & Wood 1992). Children under 5 years of age are particularly at risk. All types of bacterial meningitis can lead to this complication but pneumococcal and Haemophilus Influenzae meningitis pose special risk. Hearing losses can be of any degree or configuration (mild to profound, unilateral or bilateral, symmetrical or asymmetrical) (Fortnum & Davis 1993).

Current Practices

The study carried out by Dr. J. Richards and Mrs. A. Brindle has highlighted several interesting aspects of the current situation in the West Kent Health Authority namely:

1. There is no agreed protocol to follow up by all Districts.

2. Different routes of referrals.

3. Three children out of seventeen were not referred for an Audiological Assessment.

4. Time from referral to actual assessment and further follow up vary widely although there is a trend in the time interval per District, it is difficult to draw any conclusions because of the small numbers involved.

Further similar but larger studies from Liverpool and Nottingham show similar results. The Nottingham Study (Fortnum 8 Davis 1993) covered the period between January 1980 and December 1989 and included a total of 265 survivors, 18.4% were not referred for Audiological Assessment. Of the total under study only 77.4% had their formal hearing assessment (but not necessarily following a formal referral route).

The Liverpool study (Riordan et al. 1993) covered the period of 7 years and 181 survivors. Of those, only 78% had formal referral and of the total only 75.8% had formal hearing assessment.

The authors of both studies conclude that in order to increase the number of post-meningitic children who have formal Audiological Assessment, routine referral while the child is still in hospital or on discharge reinforced by an explanation of the importance of the assessment and re-referral, if necessary, at the Outpatient clinic should be recommended.

The importance of written guidelines which should be brought to the attention of junior Paediatric Staff cannot be over-emphasised.

Suggested Guidelines

1 All children with proved or suspected bacterial meningitis should be referred by the attending physician (Consultant/Registrar/SHO) during the child's stay in hospital or on discharge.

2. Referrals should be made to Kent Paediatric Audiology Service for the following reasons:

a. Urgent appointment can be arranged within weeks.

b. Clinic venues can be chosen to be near the child's home thus minimising the DNA rate.

c. Staff are geared to dealing with children of all ages and their parents.

d. Cliinics are usually less frightening to children who have been through the traumatic hospital admission experience.

e. Close and immediate contact with Educational and Social Services if needled.

f. It will be easier to collect epidemiological data using the Paediatric Audiology Records System (PARS).

3. A special pro-forma will be used for the referral. This has all the necessary demographic and clinical information.

4. Referrals can also be made by telephone to the Paediatric Audiology Service, especially when hearing loss is suspected (in one exemplary case, a 10 year old boy was seen and diagnosed within a week of discharge followed immediately by the rehabilitation process including hearing aid fitting). The form shows the telephone number to ring.

5. The pro-forma and these guidelines should be brought to the attention of the junior hospital staff joining the Paediatric Team on their training.

6. When the child attends for the hospital follow up a check should be made to ensure that the Audiology Clinic has been attended. Otherwise the reasons for non-attendance or non-test should be explored and the appropriate course of action taken (e.g. re-referral, reinforcement, chasing up appointment, etc.).

7. Re-appraisal of the performance of the new system to be carried out periodically.

8. So far cases of viral meningitis have also been referred. However, participating Paediatricians may want to discuss and debate this point.

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Protocol for aetiological investigation for children with sensorineural hearing loss

At a recent committee meeting use of protocols for investigation of aetiology was discussed. I present 2 protocols which are in use. I felt that these particular protocols are both fairly practical, but would welcome comments. The first protocol comes from the European Concerted Action on Hearing Impairment and has been adapted by Professor Bob Mueller, Consultant Clinical Geneticist from Leeds.

History	**Family History**	**Examination**	**Investigation**	**Specialist referral**
Developmental progress Syncopal episodes	Clefting Pigmentary abnormalities	Pigmentary abnormalities Goitre Dysmorphic features Vestibular screening Fundoscopy	Serology <1 year Thyroid function Urinalysis ECG	Audiological ENT Ophthalmological CT/MRI

Because this protocol came from a genetics department it omits to mention referral to genetics.

Professor Mueller said that the most useful investigation for children with non-syndromal sensorineural hearing loss is to test for Cx26 (Connexin 26). 25-50% of profoundly deaf children will have Cx26, and 10-25% of children with severe losses.

We had some discussion at the last Yorkshire Regional Meeting about whether or not CT or MRI scan is essential. The pick-up of inner ear CT abnormalities is probably high enough to warrant it.

The second protocol was developed for children living in the Optimum Health Services area of Lewisham and Southwark and was supplied by Philip Evans, Consultant Audiologioal Scientist.

PAEDIATRIC AUDIOLOGY SERVICE OPTIMUM HEALTH SERVICES

Unless they are known to be in the care of a paediatrician in an acute unit, children found to have sensorineural hearing loss will be referred to a local consultant community paediatrician, or another designated person (with a copy of the protocol), to arrange the investigations required by the protocol (except for the hearing tests on siblings and parents). The person to whom the child is referred will be asked to notify the children's parents of the results directly and also to inform the Consultant Audiological Scientist.

If parents decline a referral to a geneticist, the clinician should recommend that the hearing- impaired child should be referred at about 15 years of age.

AETIOLOGICAL INVESTIGATIONS AND REFERRALS FOR CHILDREN WITH SENSORINEURAL HEARING LOSS

ALL CHILDREN

Investigation	**Target Condition**	**Refer to**
Urine dip-stick test for haematuria	Alport's syndrome	Hospital paediatrician, local CCP or other designated medical personnel
Ophthalmic examination	Reduced visual acuity Usher's syndrome Retinopathy due to foetal infection	
Developmental assessment	Developmental delay	
ECG	Jervell-Lange-Neilson syndrome	
Sibling hearing tests	Familial hearing loss	Tertiary paediatric audiology service or refer to local audiology service

CONDITIONAL INVESTIGATIONS/REFERRALS

Criterion	**Investigation**	**Target condition**	**Refer to**
Aged 6 months or less	Rubella- specific IgM (serum)	Congenital rubella	Hospital paediatrician, local CCP or other designated medical personnel
	CMV-specific IgM (serum/urine)	Congenital CMV	
	Syphilis (FTA)	Congenital syphilis	
Progressive hearing loss	MRI scan	Space-occupying cranial lesions	
Indications of possible thyroid disorder	Thyroid function test	Pendred's syndrome	
Indications of general developmental delay	Amino acids	General retardation	
Signs of branchial arch abnormalities	Renal ultrasound	Oto-renal syndrome	
One or more of: - relevant family history - possible syndrome - parents wanting more children – parental request	Genetics referral	Genetic or chromosomal abnormality	
Only if genetics referral made	Parental audiograms	Familial hearing loss	Tertiary paediatric audiology service or refer to local audiology service