

PAEDIATRIC COCHLEAR IMPLANTATION - The role of the community Doctor Presentations at the BACDA Study Day

A Brief Overview of Paediatric Cochlear Implantation in the UK

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Who could have imagined ten years ago that within the next decade we would be witnessing one of the greatest innovations ever in the history of audiology and otology. The teaching had always been that there was no surgical treatment for sensori-neural hearing loss and yet here we are now witnessing children with cochlear implants performing far better than many children with hearing aids. As our experience grows we are seeing some implanted children performing far better than hearing aid users with severe hearing losses and far better than hearing aid users with profound hearing losses. The full impact of cochlear implantation on services for hearing-impaired children has yet to be realised but it is a technique that we must all think about and offer to appropriate cases.

As the age of implantation declines and as babies/children with complex needs in addition to hearing disorders are considered for implantation there will be specific demands on the part of clinicians to assess candidature and to provide specialist support over and above that normally available for hearing aid users (which, regrettably, are not always satisfactory).

The early pioneers of the technique in the UK were accused of undertaking research on children and of threatening the very existence and survival of a Deaf culture by reducing their numbers. It is very gratifying to see that the procedure is now recognised within a routine service context and to have won the praise and admiration of some (not all) members of the deaf community for our work in this area. Congenitally deaf children are now being implanted alongside deafened children and the progress of both groups is, surprisingly, indistinguishable if the implant is provided within the first few years of life. This fact poses a challenge to some of our traditional ideas about neural plasticity but it does not weaken the argument for early hearing aid fitting. What it does demonstrate is the remarkable degree of information provided by cochlear implant over and above that provided by a hearing aid, particularly in the high frequency region, for individuals with severe/profound hearing-impairments.

As technology advances further and new generations of cochlear implants become available within the next year or so we must be prepared to raise our ceiling of expectation to new heights.

REFERRAL AND ASSESSMENT PROCEDURES FOR PAEDIATRIC COCHLEAR IMPLANTATION

Sue Archbold, Nottingham Paediatric Cochlear Implant Programme

Paediatric cochlear implantation began in the UK amidst much controversy in 1989, and since then over 300 children have been implanted. As the long-term benefits of implantation for appropriate children have become apparent, the numbers of centres implanting children has grown, as has the demand upon them. It is important that there is some consistency of practice in accepted candidature and that referrers in the UK are aware of these, so that there is equity of provision. However, these guidelines for referral have changed over the years, and continue to be looked at critically, in the light of outcome measures being obtained. The guidelines and procedures outlined here are those of the Nottingham programme, but are generally recognised elsewhere. The schedule of a paediatric cochlear implant programme demonstrating the long term commitment needed from the outset is appended.

REFERRAL GUIDELINES

The selection criteria of Staller, Beiter and Brimacombe (1991) are still accepted:

- bilateral profound deafness
- age 2-7 years
- no radiological contraindications
- no medical contraindications
- little or no benefit from conventional amplification
- a strong auditory/oral component in the educational setting
- psychological suitability
- appropriate family and educational expectations and support.

The audiological criteria have relaxed over the years; at present children with aided responses of 60dB across the frequency range will be considered for implantation. This must be assessed following a full trial of appropriate hearing aids, without the presence of a conductive overlay. Implant centres are now considering carefully children previously considered borderline, looking in particular at their high frequency responses, given the high frequency benefits given by the implant system.

The age at which implantation is considered has reduced considerably over recent years, with increased benefits being seen in those children implanted young. However, caution is still expressed about those under the age of two; the full assessment procedure must be carefully carried out, and a full hearing aid trial have been possible.

The assessment protocol of the Nottingham programme is appended; the initial referral is usually accompanied by the form PU1 from the referring Consultant to enable funding to be obtained from the child's health authority. It is vital that the long-term commitment to implantation, should it prove appropriate, is given before the assessment procedures begin.

Information about implantation is sent out to the parents of the child, and the referrer, parents and the child's teacher are asked for further information about the child's functioning before the child is called for assessment. At the Nottingham programme, the visits are spaced out to allow the parents time to consider fully the decision they are taking on behalf of their child, and for the necessary relationship to develop between child, family, local professionals and the implant team. The first visit is usually for the audiological evaluation, and for initial discussions about the procedure, and its implications. If the child has been deafened by meningitis, then the radiological evaluation will take place first to establish whether ossification is taking place in the cochlea.

Standard practice has been to use computed tomography (CT) to assess the status of the cochlea, and to look at the auditory canal and the eighth nerve. Magnetic Resonance Imaging (MRI) is now increasingly being used, as it gives further information for the surgeon. While the child is in hospital the surgeon will carry out a full otolaryngological assessment, as well as looking at the child's general medical condition. The surgeon will particularly look for evidence of active middle ear disease. Children who present as complex cases, with other problems, will involve the surgeon assessing the child in conjunction with an appropriate specialist. Counselling of the parents of children who are candidates for implantation is essential, and the implant surgeon must ensure that parents are fully informed of the risks of implantation, and also are aware of the commitment that they are making. Electrophysiology follows the radiology; conventional ERA using auditory brainstem response and occasionally electrocochleography is essential for the objective confirmation of the hearing loss.

While the clinic-based assessments are being carried out, it is important that visits are made by implant staff to home and to school to assess the child's functioning there. Teachers of the deaf and speech and language therapists will establish baselines of language and communication skills, and the reports from an educational psychologist will be

requested. It is vital to establish whether the child has any other difficulties which may preclude successful use of the implant system, and also to establish close cooperation should implantation take place. Much of the responsibility for the long-term use of the implant system lies with the child's own teacher of the deaf, and this support must be sought and sensitively evaluated. The Nottingham programme uses its own version of the Children's Implant Profile (Hellman et al. 1991) to assess the issues of candidacy in these children and finally the decision whether to implant can be made with the child's parents.

OUTCOMES

The assessments used by the Nottingham programme are given in McCormick et al. (1994); it is essential that all children are not only evaluated fully prior to implantation, but at regular intervals thereafter. The appended outcomes sheet gives some of the assessments which are found useful in demonstrating progress. These evaluations are essential to demonstrate benefit to potential purchasers of cochlear implantation, but also to ensure that each child's device is monitored, and to promote optimum use of the system at all

times. Each implant programme has a responsibility to evaluate the progress of all children; cochlear implantation is an expensive and invasive procedure, and it is vital that the candidature of children is based upon facts as far as possible. It is also vital that those referring children ensure that they keep abreast of the current guidelines for referrals, the potential benefits and the potential problems surrounding the process of paediatric cochlear implantation.

References:

Hellman S A, Chute P M, Kretchmer R E, Nevins M E, Parisier S C and Thurston L C (1991). The development of a children's implant profile. American Annals of the Deaf 136, (2) 77-81.

McCormick B, Archbold S, Sheppard S (eds) (1994) Cochlear Implants for Young Children. London: Whurr.

Staller S J, Beiter A L, and brimacombe J A (1991). Children and multichannel cochlear implants. In Cooper H, (Ed) Cochlear Implants: A Practical Guide, p 283-231. London: Whurr.

NOTTINGHAM PAEDIATRIC COCHLEAR IMPLANT PROGRAMME

Ropewalk House, 113 The Ropewalk, Nottingham NG1 6HA Tel: 0115 948 5549 Fax: 0115 948 5560

GUIDELINES for the referral of children for cochlear implantation

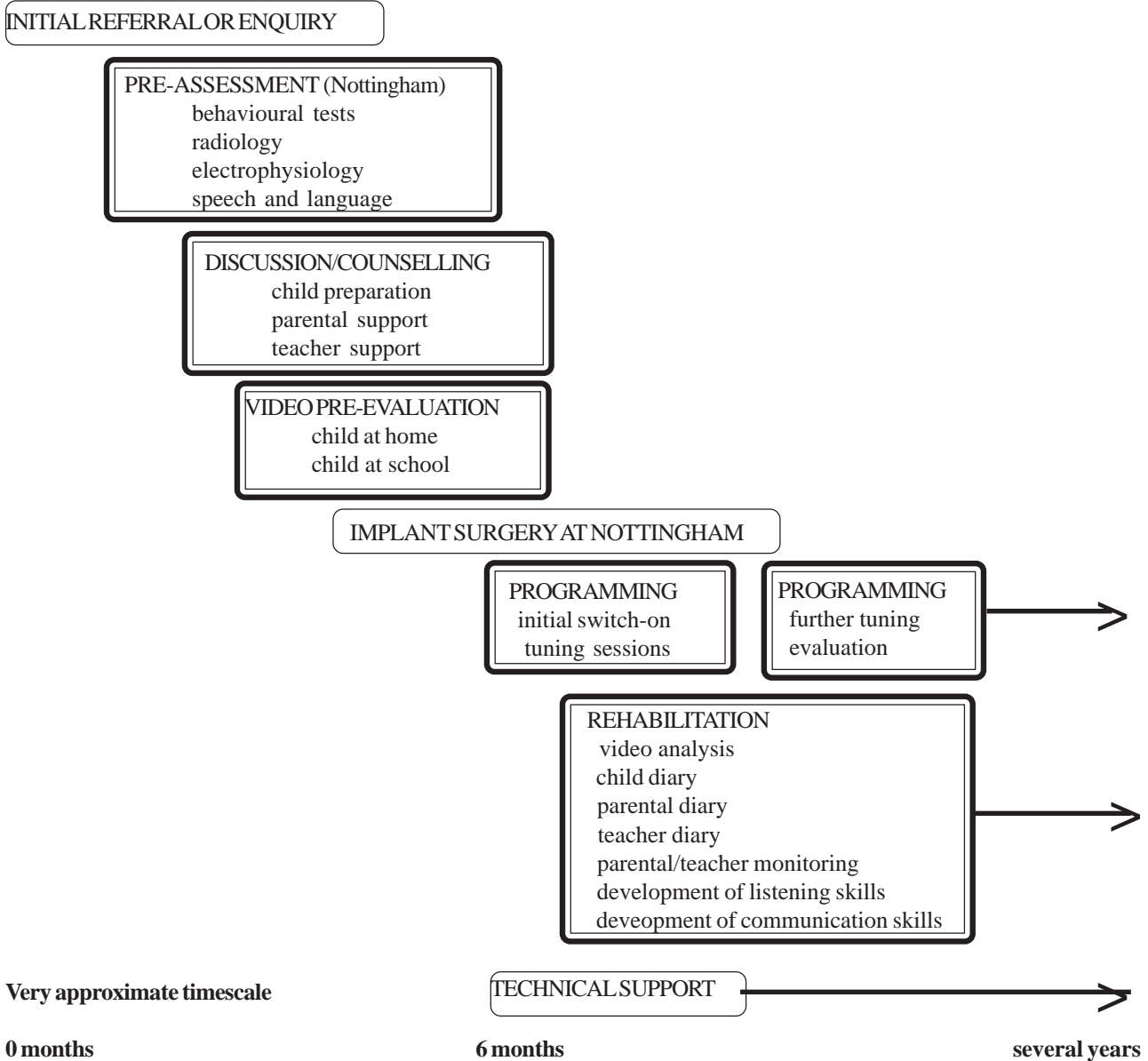
- 1 Referrals are only accepted from consultants in otolaryngology and audiological medicine.
- 2 In general priority is given to children between the ages of two and five, and referrals are accepted for those with either congenital or acquired losses.
- 3 The hearing loss should be profound or total and there must be a clear lack of hearing aid benefit over a period of many months. Measures should include sound-field warble-tone threshold determination obtained in the absence of any conductive overlay with the most powerful hearing aids and well fitting moulds. These *aided* responses should be greater than 60dB(A) across the frequency range from 500Hz up to and including 4kHz. Reliability of conditioning can be gauged by obtaining vibrotactile bone-conduction responses. The team prefers to carry out the necessary electrophysiological assessments and radiology at the University Hospital in Nottingham.
- 4 As children have to participate in the rehabilitation programme, they should have no other significant learning difficulties and be considered psychologically stable.
- 5 There should be strong parental support with realistic expectations. The parents must accept that their child will always have a hearing impairment and that the implant will not be a cure for deafness but rather a sensory aid. The ultimate benefit achievable after several years cannot be predicted prior to the operation and it will not be known where along the scale of benefit the child will be placed; from the extremes of only environmental sound appreciation through to open-set discrimination of speech and ability to use the telephone. Parents must have given careful consideration to other forms of rehabilitation, including manual systems or communication. They must also be prepared to act as an integral part of the rehabilitation programme.
- 6 There must be strong local support and a commitment on the part of relevant local services to work with the implant team. The child's teacher of the deaf will be particularly involved in developing and monitoring the child's use of the implant and a strong oral/aural commitment is necessary, whatever the educational setting.
- 7 The Nottingham group provides a total implant programme of assessment, counselling, implantation, tuning and rehabilitation. We see the programme in its entirety and will not consider offering a part of the programme separately.
- 8 The full costs will be requested from the child's health authority. Full details of current costs are available.
- 9 The Nottingham implant team provides assessment, information and support prior to the operation, the resource and expertise to implant and "tune" the device, follow-up rehabilitation and ongoing support and information. There is an understanding that the team will arrange for long term monitoring and maintenance of equipment, the cost of which will be charged to the referring health authority.

Appendix A

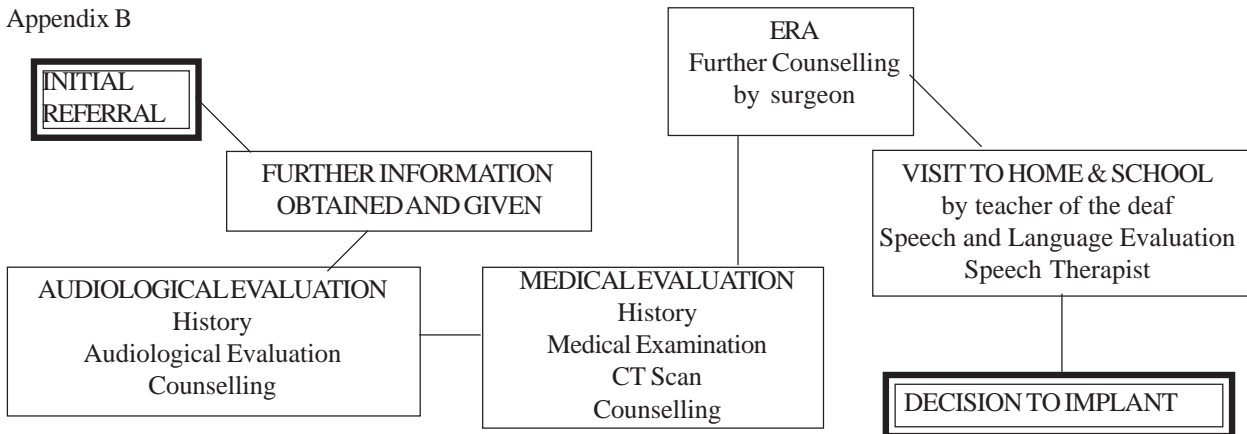
CHILDREN'S IMPLANT PROFILE (CHiP)

Team impressions of factors important to implant use and success		No Concern	Mild to Moderate Concern	Great Concern
1	Chronological age			
2	Duration of Deafness			
3	Medical/Radiological			
4	Audiological Assessment			
5	Speech & Language Abilities			
6	Multiple handicap			
7	Family Structure & Support			
8	Educational Environment			
9	Availability of Support Services			
10	Expectations of Family/Child			
11	Cognitive Ability			
12	Learning Style			

SCHEDULE OF PAEDIATRIC COCHLEAR IMPLANT PROGRAMME



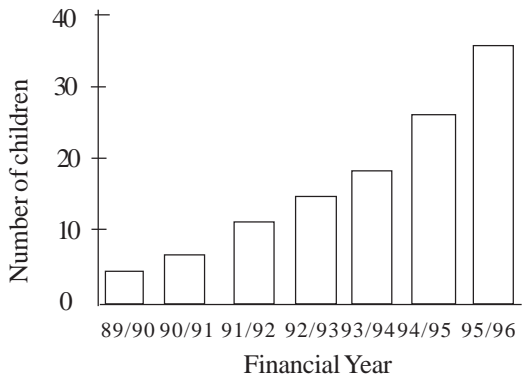
Appendix B



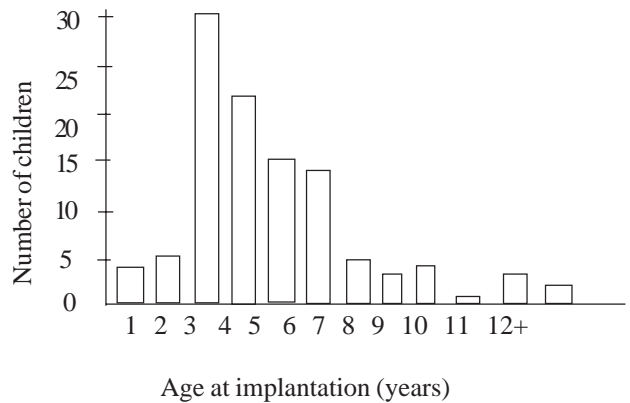
CURRENT OUTCOMES - April 1996

The Nottingham Paediatric Cochlear Implant Programme, established in 1989, specialises in the implantation of young deaf children, and to date 113 children have been implanted. All children are currently wearers of their implant systems. The following information summarises the present achievements of the children and the programme.

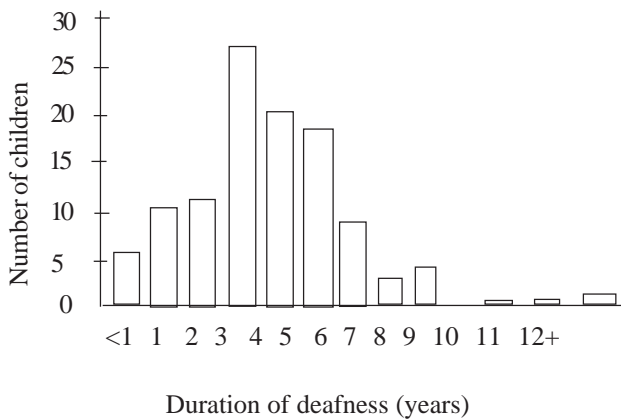
Numbers of children in the financial years 1989/90 to 1995/96



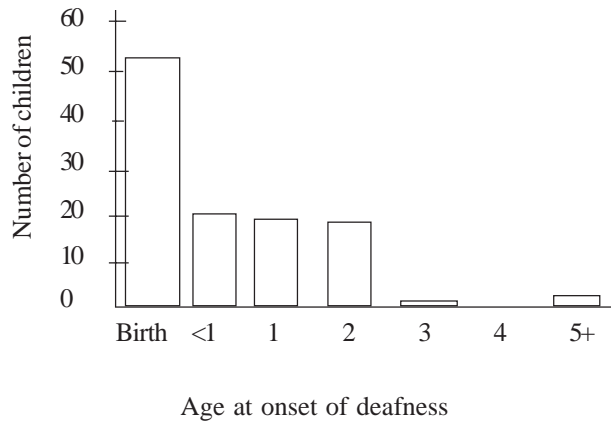
Distribution of deafness at time of implantation



Distribution of deafness at time of implantation



Distribution of ages at onset of deafness

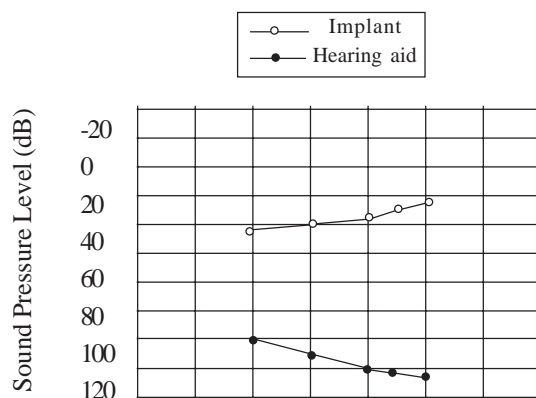


Surgical Summary

113 Children implanted
 2 explanted: no use if auditory input
 5 device failures; 4 successfully reimplanted. 1 considering reimplantation

Audiological summary

Mean sound field thresholds using implant measured six months after implantation compared with mean thresholds using hearing aid before implantation.



Functional changes

Categories of Auditory performance: Assesses children's functional abilities over time

Category of Auditory Performance	Before implant	Time after implant (months)						
		0	6	12	24	36	48	60
Use telephone with known speaker			1	2	2	2	2	2
Understand conversation without lipreading			2	4	7	12	12	6
Understand common phrases without lipreading			8	9	21	14	4	
Discriminate some speech sounds without lipreading		9	51	54	15	3	1	1
Identify environmental sounds	1		15	2				
Respond to some speech sounds	4	41	9					
Aware of environmental sounds	9	24						
Not aware of environmental sounds	87	3						
Total numbers of children	101	77	86	71	45	31	19	9

Speech Intelligibility Rating: Categorises the change in speech intelligibility of the children over time. Categorises range from (1) no recognisable words in spoken language to (6) intelligible to all listeners.

Speech Intelligibility Rating	Before implant	Time after implant (months)						
		0	6	12	24	36	48	60
Intelligible to all	1	1	2	3	2	3	3	4
Intelligible to listener with little experience	1	1			2	6	1	
Intelligible to experienced listener	5	2	1	10	15	19	8	
Unintelligible	8	12	17	30	24	6		
Sign language	53	52	53	26	3	2	1	
Preverbal	42	28	14	5	1			
Total numbers of children	93	96	88	74	45	32	18	5

Summary

We predict that, of children implanted below the age of five, over 90% will be using spoken language as their primary means of communication five years after implantation. Our results confirm that a major determinant of success following implantation is duration of deafness. This implies the need for early diagnosis, and prompt referral for evaluation for implantation if appropriate.

For the future...

Our implant programme is continuing to look at long-term outcome measures for cochlear implantation, including educational achievements and future employment.

COSTS 1996/97

YEAR 1: A minimum of eight full-day visits to Nottingham £28,000

- * Audiological assessment/ERA
- * Medical assessment/CT scan
- * Speech and language assessment involving visits to home and school.
- * Preparation of the child, family and local professionals by the implant team.
- * Surgical implantation
- * Setting up and tuning of the device
- * Medical checks
- * Rehabilitation and monitoring of child's progress and functioning of the device.
- * Monthly visits to home and school by members of the rehabilitation team.
- * An emergency service (spares/supplies/reasonable maintenance costs)

YEARS 2 & 3: A minimum of three full-day visits to Nottingham £4,250

- * Medical checks
- * Tuning and monitoring of the device
- * Rehabilitation and monitoring of child's progress and functioning of the device.
- * Bi-monthly visits to home and school by members of the rehabilitation team.
- * An emergency service (spares/supplies/reasonable maintenance costs)

ANNUAL MAINTENANCE: A minimum of one full-day visit to Nottingham £2,300

- * Audiological and medical checks and rehabilitation
- * Annual visit to home and school by members of the rehabilitation team.
- * A replacement speech processor every 6 years as recommended by the manufacturer.
- * An emergency service (spares/supplies/reasonable maintenance costs)

NOTTINGHAM PAEDIATRIC COCHLEAR IMPLANT PROGRAMME INFORMATION DAY: 30 NOVEMBER 1996

We are holding an information day for parents and professionals on Saturday 30 November 1996 at the Post graduate Medical Centre, City Hospital, Nottingham. This day will again be aimed at giving an overview of assessment, implantation, fitting and tuning of the device and rehabilitation for appropriate deaf children. The price, including coffee and lunch will be £25.00. No crèche facilities are available. Details from Maureen Ross, Paediatric Cochlear Implant Programme, Ropewalk House, 113 The Ropewalk, Nottingham, NG1 6HA. Tel. 0115 9485549. Fax: 0115 9485560

NOTTINGHAM PAEDIATRIC COCHLEAR IMPLANT PROGRAMME ADVANCED WORKSHOP — 17/19 APRIL 1997

The Nottingham Paediatric Cochlear Implant Programme will be running an Advanced Workshop from April 17/19 1997 for professionals with interests in children with cochlear implants. Numbers attending will be strictly limited to allow full participation. Below is a brief resumé of course content - for further information please contact Conference Nottingham, Regent House, Clinton Avenue, Nottingham. Tel. 0115 985 6545. Fax 0115 985 6533:

- audiological assessment - particularly of the very young, and deaf children with additional needs
- the tuning process - particularly of complex cases, and the very young
- electrophysiology and objective measures
- device failures and problems
- ways of workup with very young, profoundly deaf children, including children with visual or nonsensory impairment
- speech and language assessments
- monitoring progress: video analysis
- outcome measures: long term results based on up-to-date NPCIP data
- workshop sessions: individual case studies and problem solving opportunities

Cost: £250 (Residential) inc. full board at University Hall of Residence
 £150 (Non-residential) inc. meals and refreshments throughout the course.

**COCHLEAR IMPLANTS
THE COMMUNITY DOCTOR'S PERSPECTIVE.
Jane Lyons, SCMO, Bury**

As community doctors we work with many different professionals in order to co-ordinate and deliver the full range of audiology services, from screening and training to diagnosis and management.

This work requires a team approach, and flexibility in order to effectively manage and co-ordinate the needs of the hearing impaired child. There may be input from many different professional groups, e.g. teacher of the deaf, audiology doctor and technicians, paediatrician, speech therapy, educational psychologist, ENT surgeon, geneticist.

A cochlear implant adds another dimension to the hearing aid provision and habilitation of the hearing impaired child.

This requires close liaison with the teacher of the hearing impaired and the parents, but also careful co-ordination with other professionals such as paediatrics and ENT, before considering referral for assessment for a cochlear implant

There are many issues surrounding the needs of a hearing impaired child, and in my view referral to a cochlear implant

centre means involving another professional in the multidisciplinary team which manages the hearing impaired child.

So I was curious as to how cochlear implant centres were perceived as fitting into this multidisciplinary team. As it is a relatively new development, with various centres in different areas of the country with varying amounts of funding, there are likely to be teething problems.

Rather than present simply my personal view, I felt it would be more helpful to draw on the experience of colleagues and invite comments. I circulated all 247 BACDA members with a questionnaire. 63 (26%) replies were received.

I did not intend this to be a comprehensive survey, but to look at the interaction between cochlear implant centres and local professionals, and as a starting point to prompt further questions. I do not think there are any really serious problems, but there are undoubtedly some gaps, so that I am anxious for us not to get complacent but to look critically at our work and always strive for improvement.

QUESTIONNAIRE REPLIES

No. of respondents 63.

Some of the respondents had referrals to more than one centre so that some, but not necessarily all, answers on one questionnaire sheet were answered for more than one centre.

*Total = number of replies to this question.

1	Are there any children in your Trust who have been referred for a cochlear implant?	Total 63
	Yes 56 (14 not sure of nos), (89%) No 2, (3%) Don't know 5, (8%)	
	If "Yes," how many? 131	
2	Are there clear written referral criteria?	Total 66
	Yes 16, (24%) No 28? (42%) Don't know 21? (32%) Not answered 1, (2%)	
3a	Are you involved in the referral process?	Total 63
	Directly responsible 10, (16%) Advisory capacity 19, (30%) No involvement 32, (51%)	
	Not answered 2, (3%)	
b	Who makes the referral?	Total 63
	ENT surgeon 34, (54%) Audiological physician 6, (10%) Audiological scientist 1, (1%)	
	SCMO 8, (13%) Not specified, not sure (or arose from referral for 2nd opinion) 14, (22%)	
4a	Do you know who comprises the cochlear implant team and their individual roles?	Total 64
	Yes 38, (59%) No 14, (22%) Don't know 10, (16%) Not answered 2, (3%)	
b	Do you know who to contact for specific questions?	Total 64
	Yes 46, (72%) No 8, (13%) Don't know 6, (9%) Not answered 4, (6%)	

5a Is there a written assessment and habilitation programme, that both the family and professionals can understand?				Total 63
Yes 22, (35%)	No 8, (13%)	Don't know 31, (49%)	Not answered 2, (3%)	
b Is the programme?				Total 63
Provided by the team 26, (41%)		Provided locally 6, (10%)		
Mixture 18, (29%)		Not answered 13, (20%)		
6a Is there adequate notification of appointments?				Total 63
Yes 21, (33%)	No 1, (2%)	Don't know 38, (60%)	Not answered 3, (5%)	
b Is the reason for the appointment made clear?				Total 63
Yes 17, (27%)	No 0	Don't know 41, (65%)	Not answered 5, (8%)	
7 Is a professional from the local multidisciplinary team invited to contribute to or attend each visit?				Total 63
Yes 17, (27%)	No 19, (30%)	Don't know 22, (35%)	Sometimes 4, (6%)	
Not answered 1, (2%)				
8 Are there provisions for bilingual families?				Total 63
Yes 6, (10%)	No 8, (13%)	Don't know 48, (76%)	Not answered 1, (1%)	
9 Do you have other comments on how you would like to see cochlear implant programmes develop?				
Comments from 25, (40%)				
What is your post?				
Consultant community paediatrician 5 (8%)				
SCMO/Head of Paediatric Audiology 51 (81%)				
CMO/Staff grade/Associate specialist 7 (11%)				Total 63
Are you the lead clinician?				Total 63
Yes 35, (56%)	No 21, (33%)	Not answered 7, (11%)		

Comments

Below are the comments written on the questionnaire replies, from the 25 (40%) respondents who provided comments.

1 Communication: Liaison.

Involvement of local professionals, i. e. community child health and audiology, but also social services, because of need to give consideration to ongoing social and family problems.

Liaison between education, health and implant team needs to be clear and precise.

Easier access to team members.

Better feedback from team prior to decision to operate.

Information.

More written information.

Written criteria.

Wider distribution of information.

Guidance for non-attenders or poor compliers who see an implant as an easy option.

More emphasis during the assessment process on the amount of work that will be expected from parents.

More public awareness through primary health care team.

2 Funding.

There is insufficient funding in some regions.

Some Purchasers/Trusts feel cochlear implants in children are still experimental, and it may be useful for implant teams to talk to them directly.

N. B. "Cochlear implantation in the U. K 1990-1994 report by the MRC Institute of Hearing Research on the evaluation of the National Cochlear Implant Programme" distributed from the NHS Executive to all Health Authority and Trust Chief Executives.

3 Annual report, perhaps from the British Cochlear

Implant Group to record progress, a report has been published but it referred to only one implant centre.

Set national standards and guidelines with defined areas of responsibility.

One person with experience of more than one implant centre indicated there are varying levels of communication and degree of rehabilitative support.

4 Each implant team should include a teacher of the hearing impaired.

5 There should be a few specialised centres providing out-reach support. — but conversely:-
There needs to be a local implant service.

In Summary

National standards with an annual report

Specific contact person and updated list of team

members.

Interpreters readily available for bilingual families.

Communication and information, including feedback, e.g. by telephone after initial appointment.

Plan of management available to parents and local professionals.

Specific slots for involvement of local professionals.

Reason for each appointment clearly specified.

Adequate notice of appointments to local professionals as well as parents.

Clear written habilitation programme, with centre and local roles clearly defined and agreed in advance.

Jane Lyons
SCMO - Audiology, Bury

BIRMINGHAM PAEDIATRIC COCHLEAR IMPLANT PROGRAMME

Working with the Child, Family, Professionals and Voluntary Groups
Helen Griffiths

The Paediatric Cochlear Implant Programme at Birmingham Children's Hospital NHS Trust currently has fifty profoundly deaf children with cochlear implants. These fifty children come from a variety of social and cultural backgrounds, range from toddlers to teenagers, some have additional difficulties; what they have in common is deafness.

The Birmingham Paediatric Cochlear Implant Programme works with children throughout England and Wales; the local services vary. It is valid to describe the collaborative working relationship with each child and their family as unique.

The roles of the child, parents, professionals and voluntary agencies are considered. Recent legislation and reports have stressed the importance of inter-agency working and working with parents as partners. Collaborative work has benefits and implications for families during the assessment stage, and during the rehabilitation programme.

The Birmingham Children's Hospital NHS Trust is a major UK centre for paediatric cochlear implants. Since 1993 fifty profoundly deaf children have had cochlear implant surgery. The children range from toddlers to teenagers; some have an acquired hearing-loss, others are congenitally deaf; they come from a variety of religious, cultural and ethnic backgrounds; their communication modes vary from oral English to British Sign Language (BSL); some children have additional difficulties (additional sensory impairment, physical and learning difficulties).

The adult cochlear implant programme, based at the Queen Elizabeth Hospital, Birmingham, has, since 1990, provided cochlear implants for over a hundred profoundly deaf people. Once children who receive cochlear implants on the paediatric cochlear implant programme end their education, they transfer to the adult programme.

Whilst the teams are separately organised, close working links exist and some key personnel are shared. The benefits of shared experience and expertise, to the professionals in

both terms, cannot be undervalued.

The Birmingham Paediatric Cochlear Implant Team includes qualified teachers of the deaf. A significant amount of outreach work is undertaken by the teachers of the deaf and speech and language therapists during the pre-operative rehabilitation. The children continue to be seen regularly for rehabilitation for three years after receiving their implant.

Mission statement/aims

The Birmingham Paediatric Cochlear Implant Programme aims to

- provide information and advice; and multi-disciplinary assessment to families who seek to improve the auditory status of their deaf child;
- provide surgery and rehabilitation; supply and maintain equipment for deaf children for whom a cochlear implant is considered suitable.

To sustain effective provision, the implant centre actively develops liaison with the families and local professionals.

Referral criteria

Age: Congenitally deaf children up to the age of 10 are accepted for assessment for a cochlear implant. Above this age it is believed that the benefits for a child are very restricted.

Children with an acquired or progressive hearing loss are accepted for assessment up to the age of 17. Teenagers older than this are considered by the adult programme based at Queen Elizabeth Hospital. The care of children given implants by the paediatric programme continues to be provided by the implant centre team as long as they are in full time education, or it is felt that they will benefit from the input of the implant centre teachers of the deaf.

Hearing: In order to gain benefit from a cochlear implant, children must have a bilateral sensorineural hearing loss such that they are unable to gain any useful benefit (in terms of hearing for speech) from appropriately fitted hearing aids.

Health: Children need to be in reasonable physical health and fit enough to undergo the operation with general anaesthetic. Active middle ear disease must be treated and cleared before the implant operation takes place.

Additional handicaps: Children with additional disabilities are considered for an implant. Each child is assessed on his or her own merits. Additional disabilities only deter the team from cochlear implantation if, after careful assessment, it is decided that the child's other difficulties prevent him or her from gaining useful benefit from an implant.

Parental involvement: The decision to proceed for assessment rests with the parents on behalf of their child. It is therefore important for them to be well-informed, understanding both the potential benefits and drawbacks of the implant. Following implantation they must be committed to the programme of habilitation established by the implant centre team.

Educational support: All the local professionals closely involved with the child with an implant, must be committed to providing the optimum input and environment in order to maximise the child's benefit from the implant. This includes a commitment to and understanding of the importance of spoken language, whatever the communication method used in the school or nursery, and the individual programme established by the implant centre team.

Rehabilitation

The high level of professional support to children, and local professionals, facilitates/ensures the children's use of their cochlear implants to gain optimal benefits. The aim of the rehabilitation programme is to establish and sustain each child's use of the device, and to increase the potential of each profoundly deaf child to use the auditory information:

- to process information (i.e. understand environmental sounds)

- to process linguistic information (i.e. understand speech sounds linked with visual clues)
- to use the auditory channel preferentially (i.e. to process linguistic information to acquire spoken language)

Learning to listen and understand the sensations of sound from the cochlear implant is achieved by listening throughout the day, at home and at school. The local professionals working with a child with an implant (teachers and speech and language therapists) along with the child's family need to work closely with the implant centre team to provide the optimum conditions for a child to recognise, identify and discriminate sounds.

Individual rehabilitation programmes are devised by the implant centre rehabilitation team, working with the local professionals, to meet the developmental, cognitive, linguistic, social and emotional needs of the child. Each programme builds upon the child's existing knowledge, understanding and skills, and incorporates activities appropriate to the child's age and developmental level. Elements of rehabilitation programmes include:

- recognising, identifying and discriminating environmental sounds
- emphasis on oral / aural communication
- speech therapy
- facilitating the transfer of auditory information to all modes of languages (speaking, listening, reading and writing)
- re-tuning the device, and providing maintenance backup
- supporting the child, his/her family and local professionals

The role of the rehabilitation team at the implant centre is to monitor, assess and advise so that the child can benefit from the cochlear implant. Formal assessments are carried out at 3, 6, 12, 24 and 36 months post-implant. The rehabilitation team sees the child regularly at home, school and at the Implant Centre.

Most difficult words and phrases for parents

Individual words and phrases in order of misunderstandings	Total no.of times misunderstood	Total no.of times used	Per cent of times misunderstood
peers/peer group	17	22	77
self-image concept	13	16	81
description of child's functioning/ description of functioning	11	39	28
year level expressed as 3 year 8 mth. level, etc	10	17	59
visual (acuity, cues, defects, discrimination, memory, sequential memory)/ visual motor	10	18	56
cognitive (development, functioning, skills)	8	9	89
social (skills, interaction, reasoning)	8	15	53
gross motor (coordination, development, skills)	8	22	36
fine motor (control, function, skills)/fine movement	7	10	70
expression/expressive language, vocabulary)	7	16	44
standard deviation written as -0.2 S.D., etc.	6	6	100
3-4 word level (etc.)	6	8	75
auditory (association, memory reception, skills, sequential memory, work)	6	7	86
Derbyshire Language Scheme	5	7	71
mean scale score	5	6	83

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INFORMATION TO FAMILIES AND PROFESSIONALS

Elizabeth Tyskiewisz

“Information” is not a subject that is generally analysed in detail, although it plays a vital role in the Cochlear Implant process. Three groups are identified who have slightly different information needs: families, referrers, and support teams. For each group, informational needs, sources of information and practice on the Manchester Programme are outlined.

The need for accurate, up to date information to all parties is discussed.

Manchester Paediatric Cochlear Implant Programme

— A specialist clinical resource, providing:

Assessment and evaluation to establish appropriateness of cochlear implant to child’s/family’s needs

Implant fitting, maintenance and monitoring

Habilitation and support for implant user, family, teachers, speech & language therapists and others through:

- A programme of Centre/home/school based sessions in accordance with family’s needs and preference
- In-service training and collaboration with support team
- Troubleshooting and response to technical queries

Information to Families and Professionals

What information is needed?

Where can it be obtained?

How is the need addressed in Manchester?

A. INFORMATION TO FAMILIES

When making the decision

Benefits and limitations

Outcomes, in general, and predicted for their child

Risks and possible complications

Implications for family life, work, finances

Results of reporting and research

Updates on technology, expected developments

Alternatives available

Arguments against

Expectations of them and of their child

What they can expect from the service

When using the implant system

Technical information, checking and maintenance

Medical information (restrictions on treatment)

Clear goals and methods of achieving them

Expectations of their child, feedback on progress

Whom to contact with queries & concerns

Contact with user groups, access to newsletters & publications

Sources of information

Implant centres

Implant companies

Voluntary organisations & user groups

Other parents, older children & adult users

Publications (textbooks, newsletters, journals)

International associations

Internet

Before the implant

Parents’ information seminar (centre)

Printed material

Loan of textbooks

Family to family contact

Assistance from adult users

Observation of other children

Attendance at “ diagnostic habilitation “ sessions

Suggested contacts

Reading list

Family information session (home)

Discussion & response to queries

After the implant

User manual and technical information

Demonstration of care routines

Telephone troubleshooting

Immediate medical or technical check on request

Regular attendance at habilitation sessions, with

individual discussion and goal setting
"Goals" booklet, video, contact book
Copies of all correspondence and reporting
Distribution of information e.g. re events (Centa
Parks meeting) technical information (problems with
electrostatic discharge) copies of articles ("
Understanding the Map")

B. INFORMATION TO SUPPORT TEAMS

Team Members

Teacher of the Deaf
Mainstream teacher
Non teaching assistant
Speech-language Therapist
Audiology Department staff
Health Visitor

Information Needed

How do the child's needs now differ?
What are the expectations for the child?
Goals and techniques for auditory learning
When should we be concerned? (health & safety,
equipment malfunction, child's responses)
Where to obtain information and assistance
What observations and recordings are helpful in
monitoring progress & information management
What can I expect from the implant centre?
What can I not expect from the implant centre?

Sources of information

Technical manuals
Implant Centre documentation
Implant companies
Textbooks
CI centre staff
Training courses
Internet
Professional networks
Parents, adult users

In Manchester

Pre-implant school information visit
Full inservice training package included
Printed information, reference list
Video of sessions, contact book "Goals"
School visits
Attendance at centre
Telephone contact
Copies of reports & correspondence

C. INFORMATION TO REFERRERS

Where are the programmes?
What is the funding policy of the relevant
purchasers?
What are the referral criteria for the programme?
What information needs to be included with the
referral?
How long is the wait to be seen, to undergo
assessment, to receive an implant?
What aftercare/follow-up will be required from the
local service?
What items will need to be provided? (e.g. batteries)
What training & information are available?
Who should be contacted about queries?
What is the content of the follow-up programme?
How is liason maintained, and how are outcomes
reported?
What support is available for families whose child
cannot have an implant?

Sources of information

Implant centres
Training courses and conferences
Implant companies
Health authority & DOH policy documents
Journals and other publications
Textbooks (if recently published)
Voluntary organisations
Experienced parents, adult users
The professional network

In Manchester

Never refuse an invitation to speak at a gathering
of relevant professionals
Correspondence and telephone contact
Updates on arrangements sent to all involved
professionals once or twice per year
Training and information meetings offered locally,
when first implant is fitted in the area

Elizabeth Tyskiewicz
Manchester

1995 BACDA PRIZE AWARD

The 1995 BACDA prize was awarded to Dr. Gill Parry for her work looking at aetiology and prevalence of childhood hearing loss in Bradford in Asian and non-Asian children. This was part of the work she carried out for her dissertation for the MSc in Audiological Medicine at Manchester University. Her entry is summarised here.

Aetiology and prevalence of childhood hearing impairment in Bradford

Gill Parry
Consultant Community Paediatrician (Audiology)
Bradford Royal Infirmary

Abstract

The study was carried out in 1990 on children resident in Bradford, born between 1974 and 1987.

210 children were identified as having bilateral sensorineural or permanent congenital conductive hearing loss equal to or greater than 30dBHL.

For the children born in Bradford the prevalence of hearing loss was 4.69 per 1000 Asian births (2.68 for severe/profound loss) and 1.38 per 1000 non-Asian births (0.63 for severe/profound loss).

The increased prevalence amongst the Asian children was due to genetic hearing loss or hearing loss of unknown aetiology. For the Asian children born in Bradford, 51% had genetic hearing loss, mainly thought to be due to non-syndromal autosomal recessive hearing loss, compared to 26% of non-Asian children.

The increased numbers of hearing-impaired children place stresses on both health and education services in Bradford.

Key words : sensorineural hearing loss, prevalence, aetiology, genetic

Introduction

The school for the deaf in Bradford has mainly Asian children, although only 30% of the births in Bradford are Asian.

I wanted to see whether there were more deaf Asian children in Bradford than expected, and to look at possible causes.

80% of the Bradford Asians are Pakistani Muslims, from the Mirpur district of the Azad Kashmir in Northern Pakistan.

Congenital abnormality is a serious problem in the Asian population in Bradford. All types of congenital abnormality are more common, and multiple congenital abnormality is much more frequent, often not recognisable as a particular syndrome. (Gillies *et al* 1984)

The generally quoted prevalence of hearing loss is 1/1000 live births. The EEC study of childhood deafness in the European Community found an overall prevalence of hearing loss greater than or equal to 50 dBHL of 0.9 per thousand live births (Commission of the European Communities 1979). From that study the prevalence for Britain was 1.0 per thousand live births. Newton (1985), studying losses of 25dBHL or more in the better ear, found a prevalence of 0.8 per thousand live births. Das (1988), studying losses of 30dBHL or more in the better ear, found 1.078 per thousand births.

Criteria

All children living in Bradford with bilateral sensorineural or congenital conductive hearing loss averaging 30dBHL or greater in the better ear over the frequency range 250Hz to 4KHz, born between 1974 and 1987 were included.

Children with conductive hearing losses were only included if the hearing loss was permanent e.g. congenital atresia. Children with average losses less than 30dB were not included, nor were hearing-impaired children attending Bradford schools but not resident in Bradford.

Method

The health authority handicap register was examined, and also the records held by the peripatetic service for the hearing impaired, to identify the group of children to be studied.

The children's school medical record (10M), health visiting record and clinic card were studied as available, including copies of hospital letters and perinatal records.

Hearing loss was classified as slight, moderate, severe or profound, using the BATOD definitions.

For details of classification of aetiology see Appendix 1.

Results

210 children resident in Bradford were identified with a hearing loss of 30dBHL or greater, and their ethnic groups are shown in Table 1.

For the purposes of analysis the children were divided into two groups:-

(1) Asian (Pakistani, Indian, Asian from Africa and Bangladeshi)

(2) Non-Asian (Afro-Caribbean, African and European).

182 of the children were born in Bradford and these were analysed further.

The aetiology of the hearing loss fell into five main groups as shown in Table 2 and Figure 1. The majority of the children had hearing loss due to either genetic or unknown aetiology. 51.4% of the Asian children had hearing loss of genetic origin, compared to 26.0% of the non-Asian children. The aetiology was unknown in 32.4% of the Asian children and 39.0% of the non-Asian children.

During the period 1974-1987 there were 77,993 births in Bradford. (22,377 Asian births and 55,616 non-Asian births). The prevalence of hearing loss in this population was 2.3 per 1000 births (1 in 429 births).

If this is divided into ethnic groups we find that the prevalence rate for Asian children was 4.69 per 1000 births and for non-Asian children 1.38 per thousand births (Table 3). For Asian children with severe/profound loss the prevalence was 2.68 per 1000 births and for non-Asian children 0.63 per 1000 births. The prevalence of hearing loss amongst *all* children resident in Bradford is 5.99 per 1000 Asian children and 1.66 per 1000 non-Asian children.(Table 4)

For those children born in Bradford the prevalence rate is compared for different aetiologies in Table 5.

The differing prevalence for genetic and unknown losses between Asian and non-Asian children is significant at the 1% level.

Genetic Group. This group contained 74 children who were born in Bradford. The increase in deafness amongst the Asian group was mainly due to non-syndromal autosomal recessive deafness. (Table 6)

Unknown. For 64 children (34 Asian, 30 non-Asian) no cause could be determined for the hearing loss. Children who only had one first cousin with a hearing loss and no other affected relatives were placed in the unknown group.

Consanguinity First cousin marriages were recorded for 40 Pakistani sets of parents. Fifteen sets of notes recorded that parents were unrelated. Information about parental consanguinity was unavailable for 155 children.

Discussion.

Design of study Retrospective analysis as used in the

present study is always open to criticism. In the present study a great effort was made to identify all hearing impaired children.

Diagnosis of aetiology of hearing loss is to some extent dependent on what investigations were performed either in the neonatal period or later at the time of diagnosis of the hearing loss. Presumed aetiology may change as further information becomes available or there are new signs or symptoms.

It would have been useful to test the hearing of all parents and siblings of hearing-impaired children.

Prevalence of hearing loss The prevalence of hearing loss in non-Asian children is very similar to that recorded in other studies, but it is substantially higher in Asian children.

The prevalence rate in the present study is an underestimate as no allowance is made for children who moved out of the area. If we include children who have moved into Bradford the prevalence rises to 5.99/1000 Asian children and 1.66/1000 non-Asian children. This represents the number of hearing-impaired children that are cared for in Bradford, for whom resources must be made available by both the health and educational services. It emphasises the problems which may arise if local planning is based on national statistics, and the easily memorised figure of 1/1000 quoted.

As the prevalence is increased even when we only consider children born in Bradford it is not due to selective immigration of deaf children.

Ethnic groups In this study all the deaf Asian children were included in one group. Whilst I am not concluding that the different ethnic groups included here as Asian (Pakistani, Indian and Asian from Africa) form a homogeneous group, the numbers of hearing-impaired Indian children and Asian children from Africa living in Bradford were too small for meaningful analysis separately. It might have been better to analyse the Pakistani Muslim population separately however as it has been suggested that the term 'Asian' is too broad - masking important variations in country of origin, religion, language, diet (Senior and Bhopal 1994).

Consanguinity It was unfortunate that insufficient information was available about consanguinity in the present study to allow further analysis or to draw any conclusions. Further studies of some of the families have shown first cousin marriages for the last three generations.

In 1981 midwives in Bradford analysed marriages for all patients attending antenatal clinics, and found that 48% of Pakistani marriages were between first cousins compared with 8% of Indian marriages and 0.5% of non-Asian marriages. Figures for 1985 were 54% of Pakistani marriages, 8.5% of Indian marriages and 0.7% of non-Asian marriages.

Darr and Modell (1988), cast doubt on the reliability of information about consanguinity in hospital case records.

In their study however the results were similar. They found that 55 of 100 Pakistani women on the postnatal wards in Bradford had married their first cousin and thirteen were married within the 'Biraderi' (the wider family group). In only 33 cases had the woman's mother been married to her first cousin. This increasing frequency of close consanguineous marriage has genetic implications.

Hearing loss of genetic aetiology. Fraser (1976) estimates that there are 16-18 types of autosomal recessive deafness, and suggests that 10% or more of the general population are heterozygous carriers of one or more genes.

With fairly recent isolation of a small section of a population with consequent inbreeding, such as may occur when a group emigrates to a country with different social and religious customs, the incidence of recessive genetic disease will increase. However where consanguineous marriages have been a tradition for centuries the incidence of recessive disease is less than expected. Here inbreeding reduces the proportion of heterozygotes, and natural selection eliminates the abnormal homozygotes, resulting in an increase in normal homozygotes. So the frequency of harmful recessive genes in the population falls. Even in this situation, when children are affected by the disorder, their parents are still more often blood relatives than expected from the general rate of consanguinity in the population (Roberts and Pembrey, 1985).

Ben Arab (1990) states that socio-economic isolation of groups may have led to an increased gene frequency owing to genetic drift. Guilford et al. (1994) believe that consanguinity supports the idea of a single gene and founder effect.

Feinmesser et al. (1990), studying deaf children in Jerusalem, found that the prevalence rate of deafness declined at the

same time as the rate of consanguinity of their parents decreased. If this is a genuine trend then it is a major concern that Bradford, which may have an increasing rate of consanguinity (Darr and Modell, 1988), may experience a further increase in the prevalence of deafness.

Looking at Bradford Asian pedigrees in detail has shown that many sibships are from highly complex consanguineous families. Within this type of pedigree the most likely type of inheritance is autosomal recessive.

Amongst the Pakistani Muslims it is likely that even if the parents consider themselves unrelated, they may have common ancestry within the past few generations. Thompson et al (1991) suggest that in 'genetic isolates', even where parents are not, strictly speaking, consanguineous, the chance of mating with another carrier of a particular recessive condition may be as high as that observed in cousin marriages.

Environmental factors When establishing aetiology of hearing loss it may not be valid to make a clear division between genetic and environmental causes.

Lie et al. (1994) state that a high risk of having infants with birth defects can result from maternal or paternal genes, dietary patterns or long-term exposure to environmental teratogens. Possible causes are polygenic inheritance, an interaction between genetic and environmental factors and purely random mechanisms.

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Appendix 1

Aetiology

Autosomal recessive

Autosomal recessive inheritance was presumed if at least one of the following criteria was present.

1. At least two hearing-impaired children in one family without any other apparent cause.
2. Early hearing impairment in two cousins without any other apparent cause.
3. An apparent autosomal recessive form of hearing loss in the subject's relatives.
4. Autosomal recessive syndromes.

Autosomal dominant

Autosomal dominant inheritance was presumed if at least one of the following criteria was present.

1. Hearing impairment in at least three successive generations, according to history.
2. Audiometrically proven hearing loss in at least two generations.
3. Autosomal dominant inherited syndromes.

Perinatal

A perinatal cause was assumed if there were one or more adverse factors during late pregnancy, natal or immediate postnatal period.

Table 1

Numbers of hearing impaired children in Bradford within the different ethnic groups.

Asian Pakistani	119
Asian Indian	7
Asian from Africa	1
Bangladeshi	1
Afro-Caribbean	3
African	1
European	78

Table 2

Aetiology of hearing loss for children born in Bradford

	Genetic	Congenital Infection	Perinatal	Postnatal	Unknown
Asian	54	1	13	3	34
Non-Asian	20	5	15	7	30
Total	74	6	28	10	64

Table 3

Prevalence of hearing loss among children born in Bradford between 1974 and 1987.

	Asian	Non-Asian
Total Bradford births 1974-87	22,377	55,616
Children with hearing loss	105	77
Prevalence rate (per 1000 births)	4.69	1.38
Children with severe/profound loss	60	35
Prevalence of severe/profound loss	2.68	0.63

Table 4

Prevalence of hearing loss amongst all children resident in Bradford born between 1974 and 1987

	Asian	Non-Asian
Deaf children living in Bradford	127	83
Prevalence of hearing loss/1000 population	5.99	1.66

Figure 1

Aetiology of hearing loss
for children born in Bradford

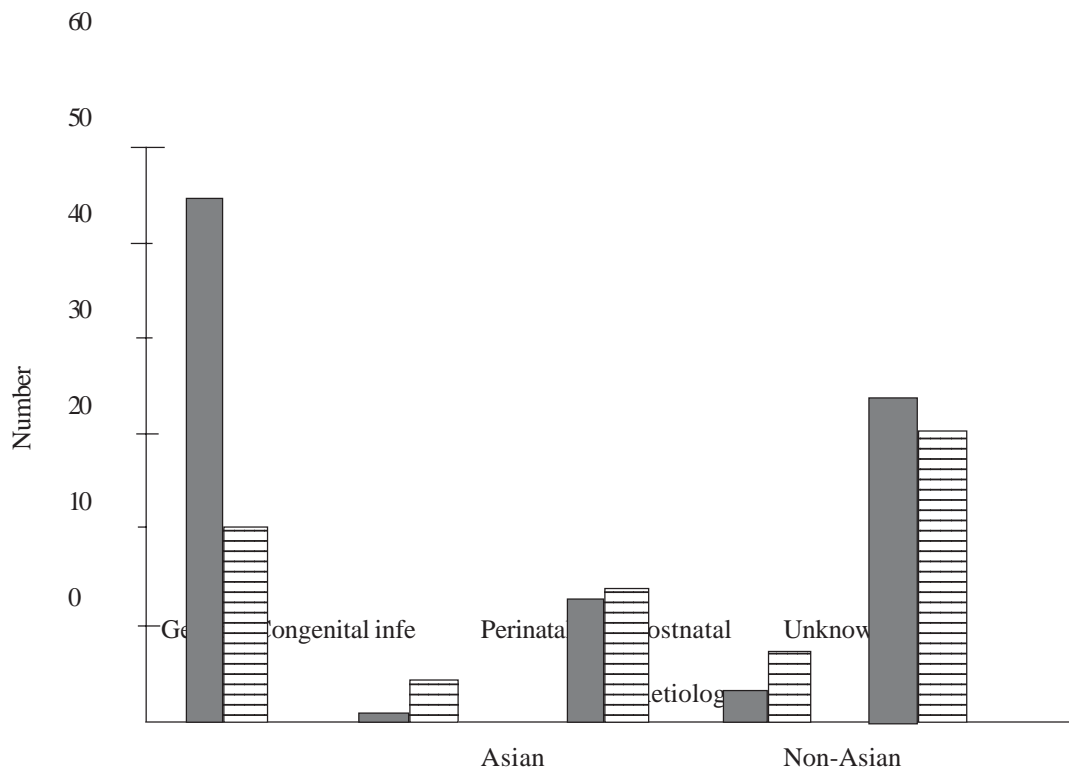


Table 5
Prevalence of hearing loss for different causes (Bradford births 1974-87).

	ASIAN	NON-ASIAN
GENETIC number	54	20
prevalence	2.41 per 1000	0.36 per 1000
CONGENITAL INFECT.	1	5
	0.04 per 1000	0.09 per 1000
PERINATAL	13	15
	0.58 per 1000	0.27 per 1000
POSTNATAL	3	7
	0.13 per 1000	0.13 per 1000
UNKNOWN	34	30
	1.52 per 1000	0.54 per 1000

Table 6
The Bradford children with genetic hearing loss.

	ASIAN	NON-ASIAN
CHROMOSOMAL	1	2
SYNDROMAL	4	5
NON-SYNDROMAL AUTOSOMAL	44	5
RECESSIVE		
AUTOSOMAL	5	7
DOMINANT		
SEX-LINKED	0	1

AUDIOMETRIC DESCRIPTORS AND AUDIOGRAM FORMAT

Danny Lang

BATOD have recently written to BACDA seeking our comments on their proposed standardisation of audiometric descriptors and audiogram format. The recommendations are based mainly on previous publications by the BSA. The BSA recommended procedures are available at a small cost (see references) and prove valuable in standardising a wide range of audiological procedures. It is timely for BATOD to focus on this now as it is clear that different systems are being used. These recommendations were discussed by the BACDA committee. *Comments by the committee appear in italics.*

BATOD recommend that degree of impairment should be defined as
 Mild (20 - 40 dBHL),
 Moderate (41-70 dBHL),
 Severe (71-95dBHL) and
 Profound (>95 dBHL)

and suggest that the degree of impairment should be based on the average of hearing thresholds at 250, 500, 1000, 2000, 4000 Hz. *It should be recognised that in younger children it may not be possible to test all these points.*

Types of impairment should be defined as conductive, sensorineural and mixed. These terms may be qualified as appropriate e.g. bilateral, unilateral, asymmetrical, high / low frequency).

The recommended audiogram format is that set down by the BSA in 1989. The recommended symbols for audiograms are as in the diagram below.

It is recognised that a variety of practice still exists. The recommended symbols are frequently used the other way round. The committee noted that the BSA recommended the term 'not-masked' should be used rather than 'unmasked'. The latter term is more correctly reserved for the very different phenomenon of unmasking associated with suppression (see BSA recommended procedures). It is

clear that the recommendations are based on adult audiology. The symbols do not allow us to show that masking was indicated but could not be applied because of the age or the developmental level of the subject. BSA suggests that additional wording may be added at the foot of the audiogram. Paediatric audiologists could use this to clarify masking issues. The BSA note in their recommended procedures on masking that some centres may find it useful to adopt an "in house" arrangement for half filling air conduction symbols when masking does not change a result (Right O ; Left X). The symbols for loudness discomfort levels are a reminder of the importance of these measurements. The recommended format does not include a schematic speech spectrum (speech banana) which can be useful when explaining results to parents.

Additional recommended standards to define and describe hearing impairment in children where the hearing levels are defined by methods other than pure tone audiometry would be useful.

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British Society of Audiology "Recommended Procedures"
 Available from; Secretariat, BSA, 80 Brighton Rd., Reading RG6 1PS

Dr. Danny Lang

BACDA Committee : 7th September 1996

	Right	Left
Air conduction, masked if necessary	O	X
Air conduction, not masked (shadow point)	●	X
Bone conduction, not masked		△
Bone conduction, masked	[]
Uncomfortable loudness level	L	J

REFLECTIONS BASED ON LITERATURE REVIEW **IS THE DISTRACTION HEARING TEST WORTHWHILE?**

Roshan Ansari SCMO Audiology Tower Hamlets

The distraction test is one of the most widely used tests in the assessment of children's hearing. Historically the approach was first described by Ewing and Ewing in 1944.

1960s

Techniques based on this approach have been used since the early 1960s in paediatric audiology clinics and to this day form an essential part of the test battery.

1970s

In the 1970s questions were being asked about the efficacy of the test for yield. These questions were being directed NOT at the 'diagnostic' clinics but at the Health Visitor 'Screening' clinics in the community. In 1977 the HVA screening survey highlighted a number of areas of concern notably in training offered to Health Visitors.

1980

A pilot study in 1980 from Nottingham (Latham, Haggard) pointed out the difficulties of identifying the cases of sensorineural deafness. The incidence of this disorder was placed at 3 per 1000, which meant that the actual cases seen by a Health Visitor were very few. The screening system had demographic and logistic limitations and high attendance failure rates. The acoustic environment in many clinics and homes was not suitable for testing. Up to 70% of cases of childhood deafness were said to be detected by parents. The results of raising parental awareness were inconclusive, but it was suggested that a combination of professional and parental attention would probably be beneficial.

1983

In July 1983 the HVA Hearing Screening Survey (2) showed that both the provision of routine screening for the under five's and training for Health Visitors had improved but there was still a lot of work required to set standards across the country for the latter.

In December 1983 Kathy Robinson's paper 'The Scandal of Late Diagnosis of Deafness in Children' described 2 cases of severely deaf children (the author's daughters) where routine screening tests in the first year of life had failed to identify hearing loss. Recommendations for improving the effectiveness of screening were made.

About the same time Barry McCormick's paper 'Hearing Screening by Health Visitor: A clinical appraisal of the Distraction Test' was published. Possible reasons for poor performance of the test were said to be lack of training and poor test techniques. Inadequate premises is one problem most of us are aware of and in some areas such as mine (Tower Hamlets), the coverage is poor. He concluded "I believe that despite rapid advances in technology and the

introduction of electronic screening equipment, the Health Visitor distraction test will not be superseded for many years to come because no other method can compare in terms of rapidity, cheapness and validity."

1986

In 1986 another paper from the same author, 'Screening for hearing impairment in the first year of life' considered the methods available for early detection of sensorineural hearing loss, as the concerns about late identification grew in terms of the possible effects this might have on the linguistic, emotional, social and intellectual development of the affected child. It was felt that both hospital and community services needed to develop and had their place.

1989

In 1989 a paper by Brown *et. al.* 'Screening infants for hearing loss' questioned yet again "the value of the Health Visitor distraction test as currently used" and underlined the usefulness of computerisation in the evaluation of screening tests. It discussed the possible changing role of the distraction test as the thinking was that it was also important to detect children with conductive hearing loss. If this was the primary aim of the test then perhaps the test should be carried out later (Haggard and Cannon) as the optimum age for detecting hearing losses is over 8 months. It also stated that there was an 'urgent need for studies to justify the continuation of the 8 month distraction screen, which is a time consuming programme, the resources for which may be put to better use.'

1990

In 1990 Johnson and Ashurst from Oxford in their article 'Screening for Sensorineural deafness by Health Visitors' showed that screening for hearing loss in the first year of life using the distraction test remained the responsibility of Health Visitors in most districts in the UK. They concluded that the Health Visitor distraction test was a sensitive though somewhat non-specific screening test. Screening only high risk babies would miss a high proportion of deaf children, and high risk children who miss being tested are particularly vulnerable. Failure to test all the infant population severely limits the effectiveness of screening.

1990 also saw the paper by Scanlon and Bamford 'Early identification of hearing loss: Screening and Surveillance methods'. "Service monitoring data on the outcomes of Health Visitor's screening for hearing loss at 8 months in West Berkshire indicated low sensitivity and low positive predictive value despite efforts to improve the conduct of the screen". It was decided that the traditional screening method for detection of hearing loss in babies would be

discontinued and efforts concentrated on alternative procedures i.e. neonatal 'at risk' screening and surveillance using parental observation.

We await results of outcome.

1991

In 1991 Tower Hamlets followed suit by dropping their Health Visitor distraction test. The coverage had been as low as 56% by 9 months of age. In one survey, out of 36 children fitted with hearing aids only one child had come having failed a distraction test and this was a high risk infant.

1994

In 1994 Mott and Edmond's paper, 'What is the role of the distraction test for hearing?' reviewed "the use of the distraction test as a screen for hearing loss in infancy in Bristol and District Health Authority". It showed that large numbers of children with minor hearing loss were identified. Referral of these children to secondary services led to delays for those children with more significant loss. The test was said to be "no longer the main method for identifying children with sensorineural hearing loss". It concluded that 'although the distraction test has a place in the assessment of the hearing of children, its role has changed and its function as a universal screening test is questionable'.

1995

Robertson *et al*'s paper, 'Late diagnosis of congenital sensorineural hearing impairment: why are detection methods failing' was published in 1995 and is well worth a read. In their study the median age at diagnosis was 18 months, age

at fitting of hearing aids was 20.8 months, and for commencement of specialised intervention programmes was 22.3 months. Reasons for failure of the method were said to be a "combination of poor screen test efficacy, incomplete population coverage and parental and professional denial".

So therefore *is the distraction test worthwhile?*

1996

It is one of the screening options available in "ascending order of estimated cost" as shown in the Hall report 1996. In the present climate where we have to justify every action in terms of cost efficiency, many questions need to be asked and much more research is needed before the 'ideal' method of screening for hearing impairment in children can be found. The Hall report highlights some of the hottest issues:-

1. What is the most cost effective approach to neonatal screening and to the detection of hearing loss not present at birth? (progressives and mild-moderate conductives)
2. Does early intervention really improve outcome for the affected child?
3. Is this enough to justify universal screening?

The debate continues.....

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SCHOOL HEARING SCREENING IN SOUTH WEST ENGLAND

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Abstract

School provides a good opportunity for population screening. Most districts in the U.K. continue to screen children's hearing when they enter school. Programmes for detection of significant sensorineural hearing impairment in the preschool years should pick up the majority of such cases long before school entry. Mild sensorineural losses may escape preschool screens. Otitis media with effusion will be the commonest cause of loss detected by school hearing screening. Earlier publications have referred to considerable variation in practice of school screening. A number of professional bodies and working parties on Child Health have made recommendations on screening protocols. This is a descriptive survey of the structure and process of school screening programmes in South West England. A variety of practice remains across the Region. Advantages and disadvantages of school age screening are outlined. The need for national consensus on school screening protocols is stated.

Key words: screening, audiometry, school health services; otitis media with effusion, non-organic hearing loss.

Introduction

Most school health services in the UK. continue to screen children's hearing in school. Otitis media with effusion (OME) will be the main cause for failing school age screening. Preschool programmes should catch case of significant permanent hearing impairment before school entry, few cases should remain undetected on admission to school. Such cases may reach school undetected for the following reasons: (a) not screened in the preschool years; (b) false negative result in previous screen; (c) acquired hearing loss since previous screen (e.g. post meningitis); (d) progressive loss; (e) incomplete records / data (e.g. for an in-transfer child). Mild losses are more likely to escape detection in the preschool years.

The ACSHIP report (1981) recommended one universal screen during the first year of school between age 5 and 6, and did not recommend subsequent screening except in special circumstances: e.g. no previous test result, children with special educational needs and children with language delay.

Whitmore & Bax (1986) stated that the whole procedure of school entry assessment (including sweep audiometry) is as much concerned with the confirmation of normality as with detection of abnormality. It is also important to look "beyond the sound booth" as shown in a recent paper on non-organic hearing loss and child abuse by Riedner & Efros

(1995).

Stewart-Brown & Haslum (1987) found considerable variation in practice of school screening in England and Wales. Sweep audiometry was used in 92% of districts. Tympanometry was used in 3.6%. It is not clear from the study as to whether tympanometry was used with, or instead of, sweep audiometry in these districts.

The most common pattern of screening was a single screening at 5 years, then 5 and 7 years, and thirdly 5 and 8 years. Screening at secondary school was reported from 18.2% of districts. The hearing level prompting referral varied among districts. The three most common were: 20 dB (32% of districts), 25 dB (44% of districts) and 30 dB (19% of districts).

The first edition of "Health for all Children" edited by Hall (1989) recommended that the universal school entry screen should be maintained, whilst recognising its limitations and the need for further research.

BAAP (1990) recommended universal school age screening in two stages: a speech discrimination test prior to or at school entry (i.e. Kendall / McCormick test or Manchester junior word lists according to child's ability) and sweep pure tone audiometry at 1000, 2000, & 4000 Hertz. The child must pass all in both ears. It was intended to eliminate the detection of hearing loss limited to low frequencies as these are likely to be due to mild OME. It is rare to find a sensorineural loss only involving 250 & 500 Hertz. There is a special difficulty with screening at 250 Hertz as this frequency is commonly affected by background noise in school.

To consider other countries, ASHA (1990) issued guidelines for screening for hearing impairment and middle ear disorders in the USA. It recommended that screening programmes include procedures for the detection of all peripheral auditory disorders and not just impairments of auditory sensitivity. The screening protocol involved four sources of data: history (from parents / teachers), visual inspection, audiometry and tympanometry. ASHA acknowledged over-referral rates from programmes based on abnormal tympanometric findings alone. These guidelines gave little consideration to the public health elements of screening. The protocol did not define uptake, sensitivity, specificity or yield criteria. It is not clear as to whether the protocol could be applied to both private and state sectors in the USA. The Scandinavian countries still screen three times in the school years.

Haggard and Hughes (1991) stated that for reasons of ease of coverage, screening at school entry would not be supplanted by other ages of screen .The chapter on epidemiology of otitis media referred to the secondary prevalence peak coinciding with the UK age for school entry. They observed that school entry screens had not been subjected to high levels of scrutiny and examination of their role would be more fruitful once arrangements for earlier (preschool) years have stabilised. This publication contained an outline of a study to evaluate combined screening techniques for school entrants. The authors observed that the USA focus on school age screening may be due to the lack of adequate preschool systems.

BACDA (1994), 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. Universal tympanometry was not recommended. BACDA is at present preparing guidelines for training in paediatric audiology which will specify procedures on sweep testing of hearing (in press).

“ The Health Needs of School Age Children” edited by Polnay (1995) 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 tests and a specialist audiology service. The target is to test 98-100 % of all five year olds. Approximately 20-25% of children age five may fail a sweep test and this should be repeated 4- 6 weeks later. Only a small proportion will fail a second test. The criteria for failure are 30 dB at 500Hz, and 25 dB at I, 2, 4 kHz. There should be a well defined policy on hearing screening and referral with methods of monitoring the results . The British Society of Audiology (1995) have recently circulated a document specifying what they consider to be the minimum training requirements for testers involved in hearing screening. The Hall report (1996) has advised that the school entry screen should be maintained and referred to Polnay (1995).

Method

Questionnaires were given to community doctors with responsibility for audiology in 11 districts in the South West of England to obtain descriptive information. The questionnaire occupied one sheet of A4 and was designed to allow quick completion . There were questions on age of screening, who carries out screening, methods of screening, screening intensity level and frequencies screened . Respondents were asked to outline their pass / fail / referral criteria.

Results

Ten questionnaires were completed. The responses to the first four questions outlined in the method are presented in

Table 1 (a-d). Districts most commonly applied a single screen at age five . Two districts continued to screen on transfer to secondary school. .Screening was performed by school nurses and non medical technical officer (MTO) screeners in all but one district where an MTO screened. Pure tone sweep was used in all districts with one district reporting additional use of a speech discrimination test (Table 1 c). Screen intensity level was either 20 dBHL (5 districts) or 25 dBHL (5 districts) . There was considerable variation in frequencies screened. Three screened 1000, 2000 and 4000 Hz; three screened 500, 1000, 2000 and 4000 Hz; three screened 500, 1000, 2000, 4000 and 8000 Hz and one district screened all standard audiogram points. The majority of districts follow up initial failures as recommended in Polnay (1995) i.e.. repeat the test in 4-6 weeks proceeding to threshold testing and additional tests for second fails.

Discussion

This study set out to summarise recent literature on school hearing screening and determine present practice in terms of structure and process. The importance of outcome measures such as coverage, sensitivity, specificity and yield are recognised but it was not possible to address them in this short survey. Screening practice still varies across the South West of England. School nurses were the most common category of screener .With respect to frequencies screened three districts followed the BAAP (1990) pattern as do many others nationally. A further three screen the frequencies as recommended by in Polnay (1995) but do not set a fail level of 30 dBHL at 500Hz. South West districts opt for either 20 dBHL or 25 dBHL. The British Society of Audiology (1988) recommended that mild hearing loss occurred from 20 dBHL upwards, and went on to state that average losses of less than 20dBHL do not necessarily imply normal hearing. It is recognised that children with borderline normal or mild losses may have difficulty hearing in school. A speech in noise test may be more effective in defining these children.

Tympanometry is being used to screen in other areas usually in combination with a pure tone screening. Such screening protocols will have to decide on how to deal with children failing tympanometry and passing audiometry.

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 preschool 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. Confirmation of normality is an important function of school health services as shown by Whitmore and Bax (1986). 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 the school entry screen .The school health services are well placed to consider the physical, psychological and social aspects of such cases. Child protection issues may exist. The school entry

screening test can provide useful audit information to help evaluate preschool screens not only by detecting false negatives, but also by helping to ensure that detected cases are being correctly managed.

Disadvantages of school age screening should 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.

If we accept the recommendations by Haggard and Hughes (1991), BACDA (1994), Polnay (1995) and Hall (1996) we should maintain the school entry sweep test. Each district and health board should define precisely its policy on OME management and to closely monitor the results.

Further evaluation is required and information technology should allow better audit in the future. Methods of detecting hearing loss through observation by parents and teachers linked to a responsive system for assessment may help to detect significant cases. It is important to evaluate efficiency and effectiveness of all medical activity but wider benefits outlined above may not appear in an evaluation based only on sensitivity, specificity and yield.

Different authorities continue to recommend various screening protocols (e.g.. BAAP 1990 and Polnay 1995) which may perpetuate the variation in practice found in this survey. National consensus for school screening protocols should be sought.

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TABLE I (a-d)

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

Age	No. of Districts
5	7
5&8	1
5& 11	2

b. Who carried out screening ? (n = 10)

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

c. Methods of screening (n = 10)

Method	No. of Districts
Pure tone sweep	10
Speech disc. test	1
Tympanometry	0

d. What is the screen intensity level ? (n = 10)

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

Unilateral Hearing Loss in Children
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(God would not “have simply hung a second ear on our heads purely as a mechanical safety factor in a chancy world” (Markides, 1977)

Introduction.

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

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

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

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

possession of two ears.

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

Suggestions to habilitate children with unilateral hearing loss

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

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

At Clinic:

1. Identify a.s.a.p. - especially if attentional or behavioural problems. In the UK these children are frequently picked up by the 5 year school entry audiometry screen. Once identified these children should have a full investigation to establish, where possible, the cause of the unilateral hearing loss.
2. Audiological supervision. There should be an annual review to check there is no change in the hearing level of the poor ear, and to promote the preservation of hearing in the good ear. The opportunity can also be taken to discuss the implications of the unilateral hearing loss with the child and the parents.

3. Counsel re preservation of good ear:
 - early and efficient treatment of ear infections.
 - ear protectors in noise.
 - protective helmets when cycling or skate boarding.
 - advise re scuba diving
4. Counsel child and parents re likelihood of directional difficulties, e.g. in traffic the child may not be aware of direction of unseen oncoming traffic.
5. Alert carers and teachers: advise re classroom placement. i.e. seat with good ear towards sound sources, and bad ear towards unwanted noise.
6. Involve teacher of the deaf, and if appropriate speech therapist.

At home and in School:

1. Gain child's attention before speaking, if necessary by non auditory means, e.g. touching, and ensure attention is maintained.
2. Use familiar vocabulary, and simple sentence structure, if necessary rephrasing, - these children are likely to be language delayed.
3. Use visual supplement to communication.
4. Minimise background noise.

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