ABSTRACTS

The epidemiology of permanent hearing impairment in children: consideration of a cost model for alternative forms of early detection.

Adrian Davis, MRC Institute of Hearing Research, Nottingham

In a number of articles over the last five years I have reviewed the epidemiology of hearing impairment in children (for a review see Davis, A.C. (1993) A Public Health Perspective on childhood hearing impairment. In B. McCormick (Ed.), Paediatric Audiology 0-5 years, 2nd edn., Whurr Publishers, London p1-41). Over the last year with colleagues in the Trent region of the NHS we have undertaken an ascertainment study of permanent childhood hearing impairment in each of the ten health districts in Trent. The provisional results of this study are based on a subset of the 589 children we have on our database born between 1985 and 1993 with a permanent hearing impairment of 40 dB HL or greater, in the better ear, over the mid-frequencies (0.5, 1, 2 and 4kHz or equivalent measure). Prevalences and some of the quality indicators (e.g. age of detection of hearing impairment) can only be estimated from the earlier cohorts we investigated. This is necessary because of the influence of possible late identification on the data. This is one reason why the analysis of children presenting in any one year are going to be misleading. Where possible data should be analysed by birth cohort and NOT year of detection / referral.

We found that the prevalence of 40dB HL+ permanent hearing impairments was about 1.27 (ci 1.2-1.4) per 1000 children born in the region at about 4-5yrs and older. This reduced to 1.16 per 1000 congenital hearing impairments, assuming an equal number of cases transferring into the region as those flowing out. Just under 10% of the impairment are acquired or are progressive in nature. Only 1 in 3300 births result in profound hearing impairments. A further 1 in 4000 have a severe impairment. In this study a smaller number were found in the three major risk categories, than we have found in previous studies. This was due to the difficulty in retrospectively finding out whether the children had been in the NICU for the required amount of time. Thus 20% have a definite history of NICU, but 55% of the children have no information on this important factor. The proportion that reported a family history of permanent childhood hearing impairment was 27% and a further 6% had a craniofacial abnormality noticeable at birth. Therefore, in this study - provisionally - 53% had one of these risk factors. This is lower than others have reported, but is mostly accounted for by the missing data we are currently trying to identify in the NICU records.

It is interesting to look at the NDCS Quality Standards targets and see whether the cohorts born 1985-1990 meet these targets. The target suggests that 80% of the congenital hearing impaired children 50+dB HL, resident in the district at the time of the relevant screens, should be detected by 12 months of age, and 40% by 6 months. In terms of the age referral the 40th and 80th percentile were 8 and 23 months, whilst in terms of age at fitting of hearing aids these percentiles were 15 and 40 months. This is some way out from the targets. However, when analysed by severity, the severe and profound children were 6 and 14 months for these percentiles at age of referral. Moderate impairments were much longer at 9 and 40 months of age.

The data suggest that there is a reasonable meeting of the target for severe and profoundly impaired children - but that there is still some way to go to improve age of detection for moderately impaired children. It is obvious from the data that the districts that have targeted neonatal screening have a higher proportion of the moderate group detected earlier. It would seem that most districts would immediately benefit from introducing targeted neonatal screening, whose marginal 'personnel' cost would appear to be about £50-70 per child screened (1993/94 prices, including allowance for sickness/holiday/study-leave cover). Some of the pros and cons of different approaches to paediatric hearing screening are reviewed in an article currently in press (Current thoughts on hearing screening, Davis, 1995. Ch.4 in Progress in Community Child Health, ed NJ Spencer, Churchill Livingstone).

Finally I would like to emphasise that the epidemiology of hearing impairment and associated research give some clues as to how services ought to be organised (and costed), but the local context is very important in actually determining these. It is quite surprising that almost ten years after the importance of information systems was outlined in the organisation of services for hearing impaired children that so few districts can numerate children that meet a simple criterion (e.g. possess a hearing aid, 50+ dB HL) or can say what the coverage and yield of their Health Visitor Distraction Test programme is. There appears to be a need for further research training (to avoid the pitfalls of ad-hoc 'research') and research taking the current emphasis by the NHS R&D programme on appropriate health services research, development of research protocols into clinical procedures and post implementation evaluation. One area that needs substantial more research is on the further development and use of an integrated strategy to the information needs of professionals in paediatric audiology.

Targeted Hearing Screening in Neonates - Comparison of Follow-up with Neonatal results.

John Stevens and Hilary Webb
Central Sheffield University Hospitals and Sheffield Children's Hospital, Sheffield

About fourteen hundred babies born in the north of the Trent region and considered to be "at risk" of hearing impairment have entered the Sheffield study between 1985 and 1992 to investigate the efficacy of transient evoked oto-acoustic emissions (TEOAE) and the auditory brainstem (ABR) response as methods of detecting hearing impairment in the newborn. Approximately 95% of these infants have been followed up at 8 months and about 40% have returned for a final follow up hearing assessment to date.

Audiometric results are now available up to the age of two. The results of an initial analysis of this data will be presented. Results above the age of two for aided children are available for a proportion of the babies entering the study and these results will also be presented. The main conclusions are as follows.

For the neonatal screen the evoked oto-acoustic emission failure rates were higher than for auditory brainstem response at 50dBnHL. This result and the small numbers involved in a targeted screen indicates the use of ABR for this population.

The evoked oto-acoustic emission screen detected 94% of those failing the auditory brain stem response at three months. This result indicates that the evoked oto-acoustic emission test is sufficiently sensitive to be used as a primary screen. It is concluded that its role is likely to be where large numbers of babies need to be tested.

The results of the follow up from eight months of age to two years of age show a considerably higher proportion failing at 50dBnHL compared to the proportion failing at three months after the neonatal tests. These results confirm the difficulty of differentiating between those infants post eight months who have a permanent hearing loss requiring aiding and those with a transient problem that does not require intervention.

The majority of babies aided by two years (17 out of 21) failed the neonatal ABR screen. However, the follow up results show a further four cases where the ABR test was passed at birth and the infant has been aided. These results indicate the need to re-test all of the target group at eight months.

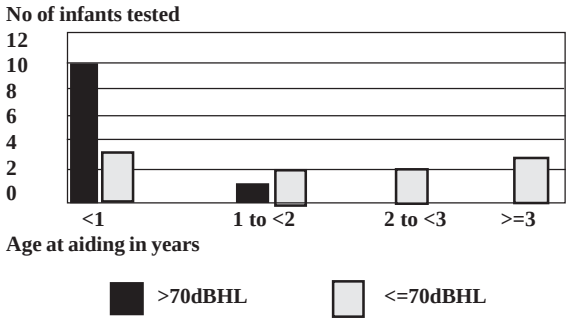
ABR screen against TEOAE screen
BILATERAL
(excluding 4 cases of atresia

		ABR screen result	
TEOAE screen result	**Pass**	**Fail**	**Total**
Pass	**1029**	**14**	**1043 (73.1%)**
Fail	**308(93.8%)**	**75**	**1426**

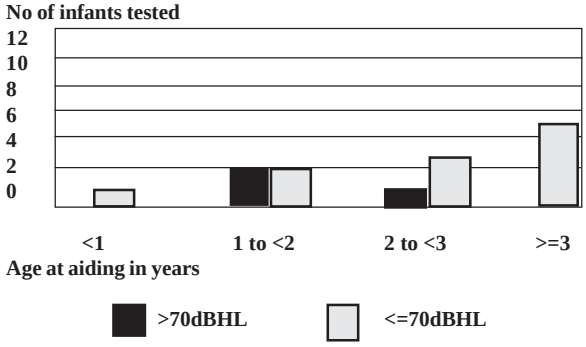
ABR 0utcome at 3 months against TEOAE screen
BILATERAL
(excluding 4 cases of atresia

	ABR outcome	
TEOAE screen result	**Pass**	**Fail**
Pass	**1040**	**3**
Fail	**335**	**48**
Total	**1375(96.4%)**	**51**

Infants failing ABR at 3 months
N = 21 (18 failed TEOAE)



Infants passing ABR at 3 months
N = 14



Outcome of follow up compared to ABR outcome at 3 months
(Numbers of aided children in brackets)

	Outcome of follow up to age 2 years				
ABR outcome	**No result**	**Pass**	**Fail**	**Incon-clusive**	**HV fail**
Pass	**72**	**1103**	**119(4)**	**18**	
Fail	**1**	**25(2)**	**23(4)**		**1(1)**

Outcome of follow up compared to ABR outcome at 3 months
(Numbers of aided children in brackets)

	Outcome of follow up to two years			
ABR outcome	**Pass**	**Fail**	**Fail +10dB**	**Fail +20dB**
Pass		**88(2)**	**24(2)**	**7**
Fail	**19(1)**	**2(1)**	**1**	**3(2)**
Fail +10dB	**2**	**2**		**4(3)**
Fail +20dB	**4(1)**	**1**	**2(1)**	**8(7)**

Bone Conduction ABR Testing in Infants

H.D.Webb and J.C.Stevens

One problem encountered in screening for permanent hearing loss in neonates is the identification of middle ear disorders. It is important that a differential diagnosis should be possible in the first few months of life if infants are to receive appropriate management. Bone conduction (BC) ABR offers a method which could be used in the identification of conductive hearing loss. This study looks at the test procedure for the bone conduction (BC) auditory brainstem evoked response (ABR) test and its clinical evaluation in high risk infants.

The test procedure for the BC ABR test was modified for use in infants. Studies on normal neonates have found that there is attenuation of the BC click across the neonatal skull; the degree of attenuation being approximately 35dB between the mastoid and forehead and 25dB (min. 20dB) between the two mastoids (Ref. 1,2). In view of this phenomenon the response was optimised in infants by using the mastoid as the bone vibrator site. In addition, masking of the non test ear was not routinely carried out unless crossover was indicated. So that the sleeping infant would not be disturbed by fitting a headband firm finger pressure was used to hold the bone vibrator in place.

The results are presented of 1628 infants, admitted to a neonatal intensive care unit at birth or considered to be at high risk of hearing impairment, screened by the air conduction ABR test (AC ABR) between March 1988 and January 1995. Failures (wave V thresholds > 45dB nHL in one ear and 50 dB nHL in the other) were immediately retested by BC ABR (fail criterion > 30 dB nHL). AC ABR failures were retested by ABR at 6 week intervals to 3 months post estimated date of delivery (EDD). All infants entering this study were followed up by distraction testing at the Sheffield Children's Hospital at 7 months post EDD.

Out of the total study population 103 infants (6.3%) failed the AC ABR screen. Of these 50 did not pass BC ABR (3.0%) (43 failed and 7 awoke before testing). By 3 months post EDD the number of failures had reduced to 71 (4.4%) (69 failures and 2 no follow up) for AC ABR and 33 92.0%) for BC ABR (includes 6 unilaterals). Follow up data from the BC ABR failures to date shows that 4 have died, 26 have been hearing aided (includes 3 unilateral failures) and 3 have a unilateral hearing loss. In addition there have been 5 infants who have been aided from the AC ABR fail/ BC ABR pass group.

In conclusion, BC ABR has been found to be a practical test in high risk neonates producing an approximately 50% reduction in failure rate achieved with AC ABR alone. An improvement in BC ABR threshold was shown by some of the initial screen failures tested pre-discharge. However, by 3 months post EDD the BC ABR test identified a high proportion (84%) of the study population hearing aided to date.

Refs:- 1. Yang E Y et al, 1987, Ear Hear, 8, 244-251.
2. Webb HD & Stevens JC, Clin Phys Meas, 1991, 12, 75-86.

Introduction to the Behaviour Orientated Work of the Otitis Media Group at IHR.

Josie Higson, Scientific Officer, MRC, Institute of Hearing Research

Disadvantages with existing behaviour rating schemes.

There are, of course,many existing behaviour rating schemes, but these suffer one or more of nine disadvantages:

- are gross screening instruments.
- involve unduly subjective qualities.
- rely excessively on somatic symptoms confounded in disability or chronic illness.
- are psychometrically oriented to extreme pathological behaviours.
- are unsuitable for preschool children.
- do not exhibit continuity between preschool and school age.
- are unremittingly negative in impact on the rater.
- lack up-to-date UK standardisation.
- have rating scales which make too many demands on the respondent.

The IHR Behaviour Assessment Inventory (BAI) maximally avoids all these disadvantages.

Who would use the BAI?

The BAI and its short form would be an appropriate instrument for use by a variety of professionals, e.g., community paediatricians, health visitors, educational psychologists.Health visitors could use the instrument in the assessment of preschool children suspected of having behaviour problems. Older children may be assessed by any of the appropriate professionals. In practice, this would probably be educationally based, e.g., educational psychologists, community paediatricians or possible school nurses.

What will professionals gain by using BAI?

The BAI is not intended as a screening instrument, though its short form could be used in contexts where the term 'screening' is sometimes used. It is not intended to be applied to unselected (asymptomatic) populations. Standardised age norms will be available. Hence the BAI equips the professional with evidence of behaviour in terms of age equivalents. 'Extreme' scores would need to go on the clinical register. This need not dictate an increase in case load. The accuracy of measurement should reduce occurrence of false positives when extreme is appropriately defined.

Use of the BAI provides an efficient and time-effective behaviour assessment. Although the questionnaire is lengthy, it is a simple instrument to use and is completed by the parent or teacher outside the interview thus reducing the duration of domiciliary/clinic visits.

What is the benefit to patient needs?

Use of the BAI enables the professional to demonstrate to parents an interest in specifying the problem and the ability to provide informed feedback. This can be helpful in at least two ways. Firstly, where necessary, an under-concerned parent is provided with evidence of a behaviour problem and assured of genuine need for intervention. Secondly, the over-concerned parent can be reassured of the lack of need for intervention.

Routes to Identification of Sensori-neural Hearing Loss
Patricia Scanlon

This presentation is an introduction to the work of the Costow Multi-Centre Group which is made up from nine members of BACDA:- Sarita Fonseca, Hope Forsyth, Jackie Grigor, Janet Lowe, Mairi MacKinnon, Susan Rose, Patricia Scanlon and Dolores Umapathy, all S.C.M.O.s working in various parts of the United Kingdom.

Meetings of the group began in May 1992 with a view to sharing experiences about early identification of sensori-neural hearing loss and to find ways of working together. It was decided to consider, as a specific project, routes to diagnosis for all cases of permanent hearing loss identified in the nine districts during 1993 and 1994. Criteria for inclusion in the survey were:- children identified up to the age of 7 with congenital permanent sensori-neural or conductive loss or an acquired sensori-neural loss. Audiological status for inclusion was an average of $\geq$ 50dBHL in the better ear.

Data was (and is being) collected within the nine districts by use of a detailed questionnaire and is being collated using the EPI Info (Version 5.01) database system. The questionnaire aims to show every encounter each child has with the paediatric audiology services in each district so that the route to identification may be followed and delays in the system identified.

Not all the data are available as yet, so the information in this presentation is only preliminary. It is confined to cases of congenital sensori-neural loss which number 58 so far, after the exclusion of 2 which are known to be progressive.

Where there was no delay (i.e. identification by 6 months of age), the median and average age of identification for 12 babies was 3 months. The delays which occurred for the remaining 46 babies and children resulted in average ages of identification for profound losses of 12 months, for severe losses of 15 months, and for moderate losses of 30 months.

The most common causes for delay were found to be service failures, parental reasons and professional inadequacies, the latter including failure to note high risk factors and not listening to parents.

Ninety children are included in the survey so far; it is anticipated that there will be approximately 130 in all. Much work remains to be done and meetings of the group will continue during 1995. It is hoped to publish its findings towards the end of the year.

OF CABBAGES AND KINGS

The time has come, the Walrus said, to talk of other things.....

Enough of all that Audiology stuff ..... now for something completely different.....

It was just over two years ago that my husband came home from a meeting of the local Council, of which he had been a member for just over fourteen years, and announced that he had been chosen as the next Mayor of Calderdale. I had known that he was in the running but hadn't dared to hope too much until it was confirmed.

Nothing happened for a while. Then, about a month before the Mayor-making ceremony, we had to go to the Town Hall for the official photographs. When I saw Tony all dressed up in the scarlet robes, lace cravat and £30,000 worth of gold chain, it really began to hit me that this was "for real" and that there was no escape! The Mayoress didn't have any robes, but a gold chain with a beautiful diamond-studded pendant worth another £30,000 or so.

Tony had chosen another Councillor and his wife as Deputies and our local Vicar as Mayor's Chaplain. The rest of the "team" were the Mayor's Secretary, the Macebearer and the Chauffeur, a six-foot something ex-para. During the Mayor-making ceremony in Halifax Town Hall - lots of speeches from Councillors, the new Mayor, retiring Mayor and Deputy, - we were all invested with our respective chains of office, and after the first of (very) many buffet meals it was time to be driven home in the official Limousine, complete with coat of arms on the side and pennant on the bonnet. The people of the village in which we live got used to seeing the big car, but at first I felt very self-conscious, being stared at and waved at - a far cry from being a Paediatric Audiologist.

Then began a hectic year of varied engagements, both within and outside Calderdale. Each Mayor has a theme, and Tony's was commerce and industry, so he visited over 200 firms and companies and I went with him on as many occasions as possible. I saw Quality Street chocolates and McVities cakes being made, stained-glass windows, tumble driers, fire-extinguishers, and the "cat's eyes" that they put down the middle of roads. We visited spinners, weavers, dyers, carpet manufacturers, makers of luxurious Cashmere scarves (I asked them if they had a factory shop!) and lots of other industries. We went to concerts, both choir and orchestral, plays, amateur operatics, OAP talent competitions, and about seven pantomimes. We got quite good at the "Oh no it isn't" routine. As the Northern Ballet Theatre is a local company, we went to two "Premieres" and saw them on three other occasions, one of which was a Gala performance of their much-acclaimed "Cinderella", which they did to support the Mayor's Charity.

Each Mayor chooses one or two charities to support and to raise funds for during his year of office. Tony chose the Heart Research Unit at Killingbeck Hospital, Leeds and the R.N.L.I., and it was the job of his Charity committee to organize various Fund-raising events. Amongst others we had a Police Band Concert, a Fashion Show, a Cheese and Wine Evening, and the Mayor's Ball. Altogether about £11,000 was raised and Calderdale's name will go on the list of supporters of the new Humber Lifeboat and we hope to be able to go to see it sometime. At Killingbeck we saw the current research into artificial blood vessels which they hope will replace the need to use a patient's veins in coronary bypass surgery.

During the year we met Princess Anne, Prince Edward and the Duchess of Westminster, and saw the Queen, Prince Phillip, and Princesses Margaret and Alexandra. We went to a Buckingham Palace Garden Party and a couple of Bishop's "At Homes" and made two official trips abroad. The first was to Halifax's twin town of Aachen where we were

wined and dined with apparently no expense spared, by their Mayor and other officials, and taken round the sights of the old city and also some of their industry. The second trip was to sign the Twinning Charter with Halifax, Nova Scotia. The two Halifax's have been linked informally since 1917 when an ammunition ship blew up in their harbour, killing over 2,000 people, injuring many more, and devastating a large part of the city. The people of our Halifax sent aid, and thereafter there had been occasional exchange visits of individuals and groups. During our year of office the Councils of Halifax NS and Calderdale W. Yorks, (of which Halifax is the main town) agreed to an official twinning. We had a very busy and interesting week over there, being entertained by their Mayor and Councillors. We saw the opening of the Nova Scotian Parliament, meeting the Prime Minister, and had morning coffee in Government House with the Lieutenant Governor and his Lady. At a Scout Jamboree we met a couple of Mounties and the Grand Chief of the Micmac Indians, native to Nova Scotia. Whilst Tony went on some industrial visits I had a look round the new Paediatric wing of the hospital, and naturally I found my way down into the Audiology dept.

We had a tour of part of the Nova Scotian coastline and countryside - miles of bays, inlets and forests. We were taken round the large Naval Dockyard and inside a Canadian Navy Frigate, and the week ended with the Twinning Ceremony in the Town Hall, transmitted on local TV. We were very impressed both by the city and the friendliness of the people.

During the year we received a visit from the Mayor of Riorges in France, Riorges being twinned with one of the smaller towns in Calderdale and Tony met some Chinese trade delegations. Interspersed with all these big occasions were visits to local hospitals, OPH's elderly couples celebrating their diamond weddings, and even more elderly ladies on their 100th birthday. One of these ladies obviously found the occasion too much and laid about her with the knife with which she was meant to cut the cake! We discovered that there were so many ordinary people voluntarily working hard to help others in many different ways.

The summer was the Garden Fete and Buffet season, followed by the Christmas dinners - we had seven!- and then it was the season of Society Annual dinners. I hardly cooked any meals and we seemed to live on lettuce sandwiches when we were at home. We managed without putting on too much weight but it would have been very easy to put on a stone or two. I, personally, enjoyed the "Churchy" occasions, Civic Services, our own and those of neighbouring Mayors, Inductions of Vicars, and lots of special occasions. On one of these we met the Archbishop of Kirov, in Russia, whose only word of English was "thankyou". On Ascension Day we went to the top of Beacon Hill which overlooks Halifax, to light the beacon, originally put there to warn of the Armada, being lifted to the top of the beacon, carrying a flaming torch, in the sort of hoist that they use to mend street lights.

It was a very busy and interesting year, in which we saw so much and met so many people, but all good things have to end and we were not too sorry when it was all over, as by then we were getting very tired. Both house and garden had been sadly neglected, in fact we said we would have to send out a search party to look for the flower beds as the weeds had had such a wonderful time! Going back to work full-time seemed a bit of a comedown afterwards, but not surprisingly there was a waiting list to tackle, even thought the rest of the staff had done their best to fill the gaps. I was very grateful to them, and to Rochdale Trust for letting me have some extra leave, but if any of you are contemplating a year as one of the Civic Heads of your district, it really is a full-time job! - but though hard going at times, it was a year I wouldn't have missed for the world. The trouble now is that I can't justify buying any more new clothes - at least for a long time

Kathleen Mazey
SCMO Audiology, Rochdale

FOR THE PURPOSE OF AUDIT

What does Audit tell us? How much information needs to be documented to give a clear picture of the situation? In a current Audit, the following information was used to describe my patient at the age of 6 years:

Hearing Loss	***66dB***
Handicapped	
1st time loss of hearing noted	***92 days***
1st failed test	***102 days***
Sensori Neural Hearing Loss	***Detected 842 days***
Impressions Taken	***963 days***
Hearing Aided	***983 days***

The true picture of this little boy takes far more than a single line.

He was born on 11th November 1988. At birth it was noted that he had the following problems:

1 Asphyxia
2 Small jaw
3 Posterior tongue
4 Cleft palate

As a result of this, he was diagnosed as Pierre Robin Syndrome. Because of his disabilities, it was not possible to incubate him and oxygen was administered by mask. In addition to the oral and breathing problems, he had rotation and some deformity of the right hand. He was tube fed.

On several occasions during the succeeding 2 - 3 weeks, he had episodes of cyanosis and aspiration of his feed. Eventually the Oro-Pharyngeal Surgeon fitted him with a feeding plate, and he was discharged home feeding well.

At the age of 2 - 3 months, audiology was requested and he was tested by distraction using voice and high frequency rattle. Although responses were obtained using eye movements, it was noted that he barely turned his head and only with difficulty.

When he was five months old, he had a severe cyanotic attack during which he required artificial respiration and was admitted to a neighbouring hospital where his problems with his neck were noted.

He had further admissions with bronchiolitis and cyanosis and observations were made on him that:

1 He was a Pierre Robin Syndrome with a cleft palate.
2 He had deformities of the neck, spine and right hand.
3 He had slow growth.

It was decided to investigate the Chromosomes.

By the age of 15 months, he had been admitted five times with cyanosis and bronchiolitis.

At the age of 10 months he had a further hearing assessment when his responses were satisfactory on the right side, but less so on the left. It was noted both at this assessment and at the preceding one that he had flat compliance curves at tympanometry.

He continued to develop, and at 14 months he responded to his name, but by the time of a development assessment at age 15 months, he had stopped responding. He did, however respond to Warble Phones at 20dB on both side on that occasion. He was also responding to speech and made no independent sounds, and was a particularly silent child.

At this time the Speech Therapists were unwilling to give therapy to a cleft-palate child pre-repair, so he was not at this stage eligible for therapy. He was referred, however, to the Wolfsen Centre by the local paediatrician for confirmation of the findings. The report obtained on this occasion showed: Nicholas was able to respond to Warbles at 35dB, though the response to the right side was slower than that of the left. Sound localisation was also reported immature. He was thought to have a developmental delay with a DQ of 50.

He was diagnosed as Kippel Feil Syndrome.

As a result of the assessment, he was referred to Michael Baraitser at Great Ormond Street.

About this time, he was observed in the playgroup to mimic sounds. The Playgroup leader produced an apple from which a worm could be enticed by making certain sounds. Nicholas was observed to make each of the sounds in mimicry of the Playgroup Leader.

When he was 2½, his cleft palate was repaired with some difficulty. At about the same time he was found to have a more marked hearing impairment which required hearing aids. At this time, the hearing was tested using Free Field audiometry when levels around 60-70dB were noted in the high frequency range. He was prescribed a B-17 hearing aid.

In July 1988, a developmental assessment was performed, and his profile was found to be:

Locomotion	-	18 months
Social/Personal	-	2 years 6 months
Hearing/Speech	-	Less than 1 year
Vision/Manipulation	-	2 years 6 months
Performance	-	3 years 6 months

His chronological age at this time was 2 years 6 months.

At 3 years he was seen by the Educational Psychologist for assessment. He was found to have scores between 2½ and 5 year level of development.

In January 1989, at the age of 3 years 2 months, an audiogram was obtained showing high frequency hearing loss but incomplete so the low frequency values were not obtained. The bone conduction values, however, showed satisfactory hearing.

In June, he was considered for the Pre-school Language Group but found to be unsuitable because of his degree of language impairment. He was, at this time, using signing and had bilateral hearing aids.

His next contact was a formal assessment medical which was performed in August 1990. The medical merely confirmed the previous findings and gave as his education needs "small class teaching with intensive speech therapy, and the availability of signing".

In November 1990, he was admitted to Great Ormond Street at the request of the Clinical Geneticist. The discharge letter reported:

1 Kippel Feil Syndrome with deafness.
2 Cleft Palate.
3 Right radial ray apeasia for hyper-reflexia.

A magnetic resonance imaging on this occasion was unsuccessful.

Following his admission to the John Horniman School for children with speech and language disabilities, he was seen by the School Doctor who is Dr. Gillian Baird. She arranged to have him checked at the Newcomen Centre where the audiogram obtained showed hearing impairment at 89dB on the left side, and 60-70dB on the right. Bone conduction at this time dropped from 0 at 50dB to 30dB at 2 KHz. All this was at the age of 5 years 2 months.

In January 1992, at the age of 6 years 2 months, an Educational Psychologist report was compiled for his special needs. The report showed that he was of low average performance ability.
His special needs identified as:
1 A specialist language teaching including sign language, probably Paget Gorman.
2 Awareness of hearing impairment and use of appropriate teaching staff.
3 Use of visual awareness.
4 On-going review of hearing.
5 Socialisation.

In January 1992, his first school report was that:
1 He was making greater attempts at signing, and able to act on two signs.
2 Beginning to recognise some spoken words by listening and lip reading.
3 Responding to softer voice.
4 Responding to his name.
5 Responsive in the group was less satisfactory than this.
6 He was beginning to read and write using his left hand.
7 His self-help improving in feeding.

In the succeeding time, he has continued to improve. When the results of his magnetic resonance imaging screening were available these showed that:
1 The ventricles were enlarged with altered signals in the white matter in the periventricular distribution.
2 There was no raised intracranial pressure.
3 There was a small arachnoid cyst at the mid thoracic level with no displacement of the cord.

He continues to have problems with communication. The incontinence which has been a problem began to improve.

At the age of 7 he began to comprehend speech, and communicated using total methods. He has had a pharyngoplasty, despite which he continues to have hypernasality and few speech sounds.

Eileen Waring