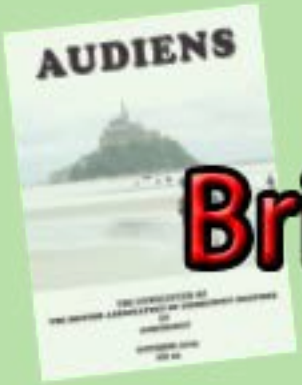

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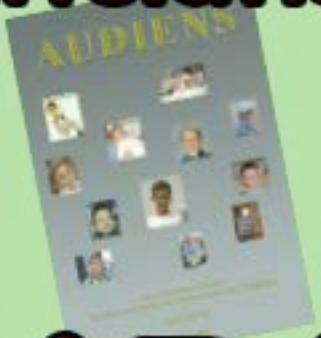

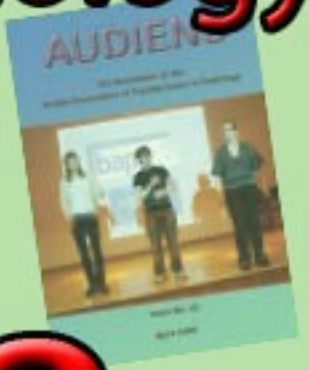

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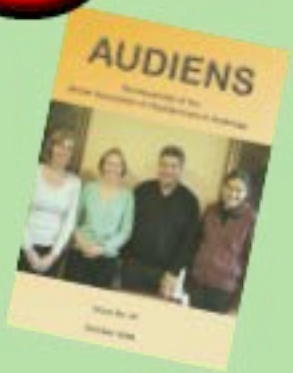

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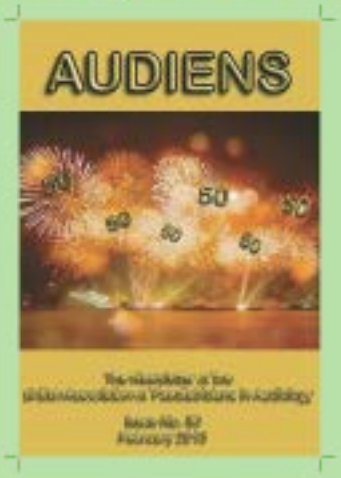

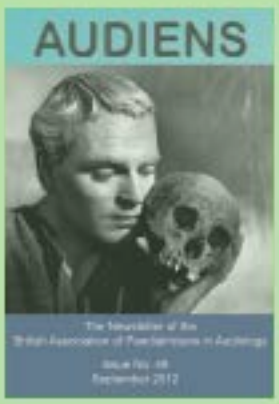

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Editorial:

Dear All

Welcome to the first electronic version of Audiens, and my first as newsletter editor.

A focus of the BAPA calendar is the Annual meeting and AGM, and the reports from this year's AGM are included. The three winning submissions for the BAPA prize are included, with varied topics. I hope reading them encourages you to apply in the future.

NEW for this year is the BAPA Audit prize. Time has been allocated at the January meeting for presentation of the four best submitted (one from each region). With all of us required to be involved in clinical audit, this is a great opportunity to present at a national meeting. So is your opportunity to tell us about your audit(s). For more information, go to the BAPA website, or contact your regional rep, or any of the exec members.

The annual meeting, and regional meetings, are always useful for networking, and finding out what is happening in the world of paediatric audiology. Now Audiens has transformed into electronic format, it will be much easier for you to submit comments, ideas etc. Have you read something interesting? Attended a paediatric or audiology meeting, or seen an interesting patient? Share your ideas (or reflections,) here. How do you update your generic paediatric skills if you only see audiology patients? How is the new NHS affecting your service, have changes led to any new innovative ways of working? We are all facing similar pressures of the effects of austerity on resources and pressures on the workforce. On this theme but from a different perspective, included is some information on Soundseekers working in audiology in parts of the world facing much greater pressures than the UK. Please share this information with anyone you know who may be interested.

On a personal note, when taking over this role, I initially considered whether I had enough time to give. All of us seem to be working at a faster pace and seem to take on more. However, having now attended a couple of exec meetings, I have much more insight into how BAPA functions and the wider picture of paediatric audiology, and I would encourage colleagues to look being invoved as a regional rep or on the exec in the future.

Hope to see as many of you as possible at the January meeting/AGM

Anne Marsden

BAPA Annual General Meeting 2013 **Chair report**

The executive committee has met on four occasions during the year to oversee the business of the association. We meet at RCPCH and have the assistance of the BACCH secretariat with minute taking and maintaining the membership which has been a great help to our secretary.

We have been becoming increasingly concerned about the loss of posts for paediatricians in paediatric audiology; Jane Lyons met Dr Hilary Cass, the president of RCPCH at the BACCH annual scientific meeting in October and was able to discuss this briefly with her. As a follow up to this we have written to Dr Cass detailing our concerns and suggesting ways that the college might be able to help. We also need the help of the members who should let us know if posts are threatened. HAB UK has also written to a number of national bodies expressing concerns about the loss of medical input to paediatric audiology and received a reply from the GMC suggesting a meeting to discuss this.

BAPA and BAAP continue to work together as the Audiovestibular Medical Federation. BAPA have provided two presentations to the BAAP audit meetings this year from Adrian Dighe and Jeanette Nicholls. A number of us were able to contribute to a national audit of aetiological investigations that was instigated by BAAP. I would encourage you to attend the BAAP annual conference which will take place on 14th and 15th March 2012 at Latimer Place in Chesham.

We have provided comments to a number of consultations throughout the year; NHSP targeted follow up guidelines, BSA VRA guidelines, BACCH Family Friendly Framework Commissioning document. We have agreed to collaborate in establishing care pathways and standards for children with microtia. We have also been asked to contribute to a review of the audiology guidance for IQIPS (Improving Quality in Physiological diagnostic Services).

BAPA is organising a session with BACCH, BAAF and the child protection interest group at the RCPCH conference this year in Glasgow on Friday 7th June with the title “Effective Interventions in Vulnerable Children”.

BACCH held its Annual Scientific meeting in October and Dr Ganesh delivered a workshop on hyperacusis. We contributed to the BACCH prospectus with information on paediatric audiology.

It is very disappointing that the vacancies on the executive committee for regional representatives for the regions of South West/Wales and South East have not been filled. In these uncertain times we need regular updates on what is happening in our field. We also need fresh blood on the committee so I would urge any members from these regions to think about whether this is something they could take on. We are a friendly bunch on the committee and the tasks are not onerous. Where there are regional representatives, local meetings are organised which gives members an opportunity to meet.

We are really struggling to get email addresses from all our members. There have been a number of things that we would like to have circulated to the membership this year but have been unable to because of the lack of accurate email addresses. With the rising cost of postage, it is not feasible to post things out to the members and most of the responses these days require electronic communication. We are exploring your views today via a questionnaire which is in your packs on sending out Audiens electronically.

We were very sorry to receive the resignation of Jeanette Nicholls as editor of Audiens. The 50th issue of Audiens will be her final edition. She has made a number of improvements to the newsletter during her tenure, changing the format to A3 and with some very interesting pictures on the front covers.

Lastly I want to thank my fellow committee members for all their hard work and support over the year and particularly Jane Dalzell who, year after year, organises such excellent programmes for the annual conference.

Gill Painter, BAPA Chair

BAPA: Income Tax Relief In Respect Of Annual Membership Subscriptions

We have now obtained the approval form from HMR & C for the above. I think it would be useful to inform members about the following from HM Revenue & Customs:

'Records show that approval was granted to BACDA in October 1989. It has therefore been decided that approval under S201 of ICTA 1988 (now S344 of the Income Tax (Earnings & Pensions) Act 2003) granted to BACDA in October 1989 will continue following the change of name to BAPA. This decision has been made on the basis that there has been no other changes that have a bearing on this legislation. The published list (known as list 3) will be revised later this year and the new title will be added at that time. Please advise your members that until the new title is included they should quote both the old and the new titles when requesting tax relief for their subscription'

Ken Abban
BAPA Honorary Treasurer

Auditory neuropathy spectrum disorder: Examples of poor progress following cochlear implantation

Wanda Neary[1] & Guy Lightfoot[2]

[1]Community Paediatric Audiology Services, Warrington Primary Care Trust, Child Development Centre, Orford, WARRINGTON, Cheshire, and [2]Department of Medical Physics and Clinical Engineering, Royal Liverpool University Hospital, Liverpool, UK

Abstract

The majority of babies with auditory neuropathy spectrum disorder (ANSD) spend 48 h or more in the Special Care Baby Unit (SCBU), and the current UK recommendations for the Newborn Hearing Screening Programme state that babies admitted to the SCBU should undergo hearing screening with transient evoked otoacoustic emissions together with auto mated auditory brainstem response audiometry, in order to identify babies affected with the condition. Current recom mendations propose that individuals affected with ANSD should be considered candidates for cochlear implantation. It has been suggested that in patients with ANSD, the presence or absence of cortical electric response audiometry (CERA) responses can help to predict favourable or unfavourable prognosis in auditory language development and comprehension post implantation. We describe two individuals who had not been in SCBU, but came from the well baby population yet satisfied the diagnostic criteria for ANSD. These two patients underwent cochlear implantation, but made poor progress in auditory language development and speech understanding on subsequent follow-up. We suggest that ANSD should be considered in all cases of infants who undergo cochlear implantation but do not make subsequent good progress in audi tory language development and speech understanding. We further suggest that CERA be performed prior to cochlear implantation in patients satisfying the diagnostic criteria for ANSD, as the results can assist in predicting post implantation progress in auditory language development and speech understanding.

Key words: *well babies, poor language progress, cochlear implantation, cortical electric response audiometry*

Introduction

Auditory neuropathy spectrum disorder (ANSD) is diagnosed when patients are found to have measur able otoacoustic emissions (OAEs) and/or cochlear microphonics (CM), but an absent auditory brain stem response (ABR), or an ABR with severely abnormal morphology at high stimulus levels. On clinical examination patients have poor speech comprehension relative to their pure tone thresholds, which in some cases can be normal. In ANSD it appears that outer hair cell function is intact, but with impaired or absent conduction of synchronous signals by the auditory nerve. Starr et al. (1) observed that clinical testing has indicated that the disruption in the stream of sound information is localized to one or more of three probable sites -the inner hair cells of the cochlea, the synapse between the inner hair cells and the auditory nerve, or a lesion of the ascending auditory nerve itself.

Recommended protocols for the assessment and management of ANSD have been published by the Newborn Hearing Screening and Assessment Team (2). In the UK the hearing of all babies admitted to special care baby units (SCBUs) is screened using OAEs and automated ABR (AABR). Babies found to have OAEs present, but an absent AABR are referred for audiological testing according to the recommended protocols. Babies in SCBU may be premature and of low birth weight, and may have been treated for hyperbilirubinaemia. The prognosis for a child diagnosed with ANSD is uncertain and careful follow-up is necessary. The results of behav ioural testing when the child is able to sit may indi cate varying results ranging from a very significant hearing loss to normal hearing thresholds. The Joint Committee on Infant Hearing (3), together with Berlin et al. (4), at the International

Newborn Hearing Screening Conference, Como, Italy, recommend appropriate amplification, fitting to behavioural hear ing thresholds, and careful review to monitor the speech and language development in children diag nosed with ANSD. Where poor progress is found with respect to auditory language development and speech understanding, regardless of behavioural audiometric thresholds, referral for consideration of a cochlear implant is recommended. The rationale for this approach is that in ANSD it appears that the auditory nerve is not firing synchronously. Fitting a cochlear implant appears to provide a more effective and synchronous firing of electrical impulses in the auditory nerve.

Rance et al. (5) investigated whether CERA responses could be recorded in children with ANSD, and determined the relationship between the pres ence of these responses and speech perception assess ments using PBK words. They found that in approximately 50% of children with ANSD, CERA responses were present at normal latencies, ampli tudes and morphology. In all cases, the presence of response at normal latencies was consistent with reasonable speech perception ability, while response absence was consistent with negligible speech percep tion. They suggested that these obligatory responses could offer a means of predicting percepcual skills in newly diagnosed infants, as their presence was cor related with significant open set speech perception abilities and amplification benefit in their study. The absence of CERA responses, in contrast, indicated profound hearing disability, with extremely poor speech perception and a poor prognosis for normal speech development.

Aims

We report our findings in two individuals affected with ANSD, from two separate and unrelated fami lies. We were interested to note whether their poor progress in speech development could be related to their ANSD and if the severity of their ANSD could be predicted from their electrophysiological test results.

Methods

A family where seven individuals are affected with a sensorineural hearing loss is described. The genetic pedigree for the family is shown in Figure I . The proband was diagnosed with ANSD, because of the finding of normal OAEs in the newborn period, but an absent diagnostic ABR when the family questioned the possibility of hearing loss at the age of two months. This baby was at risk of sensorineural hearing loss because of the family history, but he did not have risk factors for ANSD. The proband had been born at full term by normal delivery, fol lowing a normal pregnancy. The other hearing impaired members of the family were then tested by audiological and electrophysiological methods, to determine whether they were affected with ANSD.

We also report on a profoundly hearing impaired child from an unrelated family, where an ABR was absent bilaterally at the age of 14 months and a unilateral cochlear implant had been inserted soon after the age of two years. Follow ing implantation this child had difficulties with auditory communication, despite having nor mal hearing thresholds when using the cochlear implant. CM testing was carried out at the age of nine years in this patient, to determine whether the findings were consistent with the diagnosis of ANSD.

Electrophysiological methods

Objective audiological testing with transient evoked OAEs (TEOAEs), diagnostic ABR, CMs, and CERA was carried out in the hearing impaired members of the family of patient 1, and the bearing impaired child from a separate family, patient 3.

The test parameters were as follows. Electrode positions for both ABR and CERA were vertex (Cz) posirive and mastoid reference with a forehead com mon. In CERA the two mastoid electrodes were

linked. ABR tests used at least 2000 100-µs alternating polarity clicks presented at 32.1/s; filters were 30 Hz to 1500 Hz and an amplifier rejection level of ±20 µV or less was used. Waveforms were averaged over an 18-ms post-stimulus epoch.

In CERA tests I 000-Hz (in some cases additional frequencies were also used) tone bursts with!0-ms rise/fall time and 60-ms plateau time were presented at 0.7/s and responses averaged over a - 250-ms to +650-ms epoch relative to stimulus onset. EEG was filtered between I Hz and 15 Hz and an artefact rejection level of ± 50 µV was used. Up to 60 sweeps were presented before concluding response absence. In both ABR and CERA tests the objective was to determine whether a repeatable response could be recorded using stimuli up to I OOdB HL. Responses were considered absent only if recording conditions allowed a suitably luw residual noise to be achieved in the recording. Response detection was aided by the availability of statistical response analysis for CERA testing (see www.CorticalERA.com for details).

CM testing employed separate runs of 2000 rar efaction polarity and 2000 condensation polarity 100-µs clicks at I OOdB nHL, 91.1/s recorded using an 8-ms epoch. EEG was filtered between 150 Hz and 5000 Hz and an amplifier rejection level of ±10 µV or less was used. If a CM was thought to be present the tubing of the insert transducer was clamped in order to distinguish a genuine CM from stimulus artefact. Instrumentation was: ABR/CM: Interacoustics Eclipse or Nicolet Spirit evoked potential systems; CERA: Cambridge Electronics Design CERA system.

Results

Patient 1
Patient 1 (the proband) satisfied the diagnostic criteria for ANSD.

Patient 2
The results in patient 2 (grandfather) indicated unilateral (right-sided) ANSD.

Remaining five hearing impaired family members
The remaining five hearing impaired family members showed no evidence of ANSD.

The findings are summarized in Table I. The non-implanted ear of patient 1 had a recordable CM, shown in Figure 2, but no recordable ABR or CERA response using stimuli that were behaviourally supra-threshold. Cortical responses were recorded bilaterally at supra-threshold levels in patient 2. On his right side a clear CM (Figure 3a) but an absent ABR indicates ANSD while on the left an absent CM (Figure 3b) and a recordable ABR suggests a conventional cochlear loss.

The results of the remaining five hearing impaired family members who showed no evidence of ANSD are included in Table I.

Patient 3
The results were consistent with a diagnosis of ANSD.

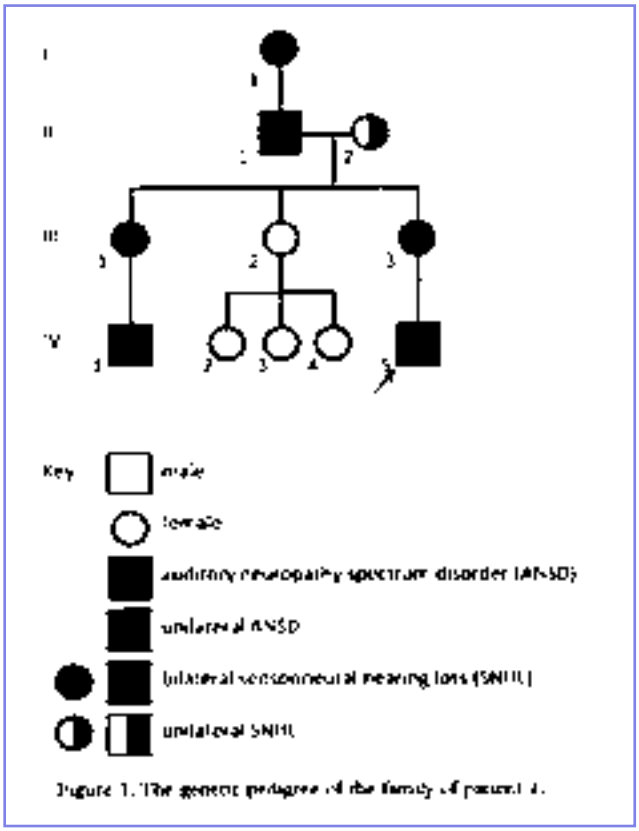
Table II summarizes our results. In the nonimplanted ear a CM was recorded (Figure 4) in the presence of a profound hearing loss. ABR had been absent at age 14 months.

Discussion

The reason for reporting the cases of these particular hearing impaired individuals is to highlight that ANSD may be present when high risk factors for the condition are absent, and to draw attention to the possibility of ANSD in patients who make slow progress in auditory language development and speech understanding following cochlear implantation.

The hearing impaired individuals described are of particular interest, as they do not have risk factors for ANSD. Patient 1 would not have been identified as a newborn affected with ANSD using the protocols for Newborn Hearing Screening. He was a full term baby of a hearing impaired mother and passed his

TEOAE screening. He had been listed for targeted follow-up behavioural testing at eight months of age, in line with NHSP guidance. The reason for his referral for diagnostic electrophysiology was because of his grandmother's concern regarding his hearing responses. We find it interesting that patient 1 had poor language development and no recordable cortical responses at levels that were clearly supra-threshold. Patient 3 was identified as profoundly hearing impaired following behavioural testing at eight months, but the loss was thought to be sensorineural rather than ANSD. Patient 3 was diagnosed with profound sensorineural hearing loss prior to the implementation of the Newborn Hearing Screening Programme.



Betner et al. (6) examined the findinl!" in 37 children diagnosed with ANSD and drew attention to the fact that prematurity and low birth weight was recorded in roughly half of the patients. Hyperbilirubinaemia in the neonatal period was present in 13 children, three had been exposed to intrauterine infection, two had congenital anomalies and two had a positive family history. Seven individuals had no causative factors identified. They suggested that prenatal or perinatal insults, rather than genetic factors, are responsible for the majority of cases of ANSD.

Genetic studies in patients and families with ANSD have demonstrated an autosomal recessive inheritance pattern in some families. Mutations in the OTOF gene, underlying the DFNB9 form of hearing loss have been reported by Yasunaga et al. (7), and mutations in the DFNB59 gene have been identified by Delmaghani et al. (8). The OTOF gene encodes otoferlin, while the DFNB59 gene encodes pejvakin. None of our patients had genetic testing carried out, as testing for mutations in DFNB9 and DFNB59 is not routinely available in the United Kingdom.

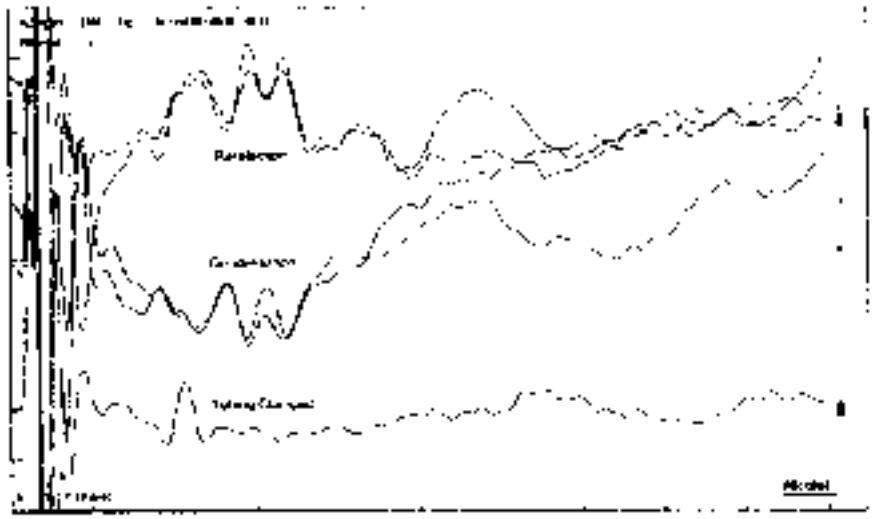
The diagnosis of ANSD was made when patient 1 was two months of age, but in the case of patient 3, this diagnosis was not considered until a much later stage, when he was having educational and behavioural difficulties. Both implanted individuals had made very slow progress with auditory language development and speech understanding, despite consistent use of their cochlear implants, together with appropriate contralateral amplification. For the majority of children who have received cochlear implants, the Manchester Paediatric Cochlear lmplantTeam would be anticipating spoken language development following surgery. The minimum that would be considered as adequate progress would be language development in line with the length of time that the child has had the implant. This time is often referred to as their hearing age. The hope and aim is for spoken language development to be in line with the child's chronological age within three to five years of implantation. The older the children are when they have surgery, the longer it takes them to catch up. A poor outcome would be highlighted if the child's speech and language progress was below that anticipated by considering their hearing age. Standardized language measures are used for assessment. For the young child, the Preschool Language Scales are commonly used. For older children, the Clinical Evaluation Linguistic Fundamental (CELF), or the Preschool CELF for the younger age group may be employed. It should be noted that the standardized tests are not standardized on hearing impaired children (Henderson, personal communication).

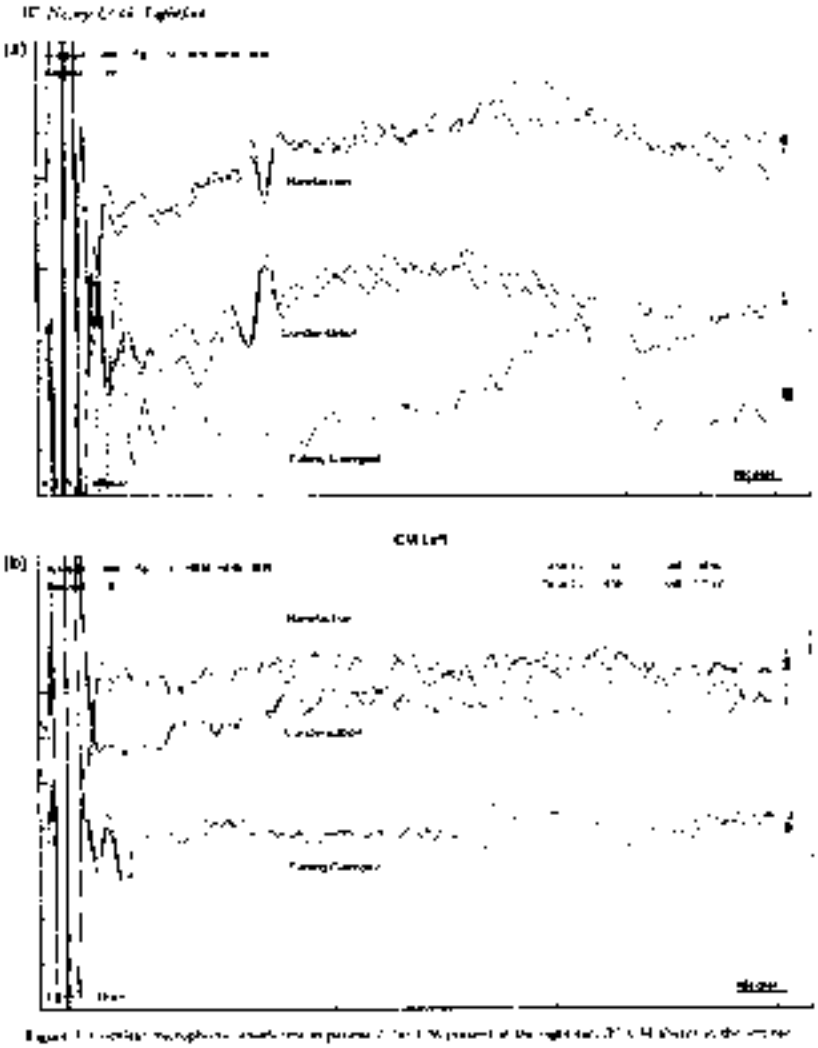
Table I. Test results of the family of patient 1, Member IV, 5 (tympanometry was normal in all family members).

Member	TEOAEs	PTA	CMs	Click ABR	CERA	Conclusion
I,1	Absent R & L	Bilateral moderate sloping sensorineural hearing loss (SNHL)	No obvious CM, but residual noise level was high	Recording conditions too poor to evaluate	Clear response bilaterally using I kHz 60dB HL stimulus	No evidence of ANSD but not excluded
Patient 2 II.1	Absent R & L	Severe bilateral high frequency SNHL	Present on R Absent on R	Absent on R Present on L	Clear response at 1 kHz at 15 to 20dB above behavioural threshold on R & L	Evidence of unilateral (R) ANSD
II,2	Present R Absent L	Mild loss R Dead ear L (MRI no evidence of vestibular schwannoma)	Absent R & L	Present on R Absent on L	Clear response on R Absent response on L	No evidence of ANSD
III,1	Absent R & L	Bilateral profound SNHL	Absent R & L	Absent on R & L	Absent at 1kHz at 100dBHL Clear response on R	No evidence of ANSD
III,3	Absent on non CI side (Left	U-shaped SNHL on left. Right CI present	Present on L (on non CI side)	Response present at 100 dBnHL on L	Clear responses on L	No evidence of ANSD
IV,1	Absent R & L	Bilateral moderate SNHL	Present R & L	Probable response on R	Clear response R & L, in keeping with audiogram	No evidence of ANSD
Proband	Present at birth	Severe R sided SNHL	Present on R	Absent on R	Absent on R, at lOOdB HL at 500 Hz, 1 kHz, 2 kHz & 4 kHz. At lower two frequencies, here was a clear behavioural acknowledgement that the sound was heard	Evidence of ANSD on R Not tested on L (implanted)
Patient 1	Absent at time ofstudy	L CI present				
IV,5						

NB. The degree of severity of the hearing loss was designated as: mild, 20 -40dB HL; moderate, 41 -70dB HL; severe, 71 -95dB HL; profound, > 95dB HL. British Society of Audiology (13)

Gibson and Sanli (9) reported on a cohort of 60 ANSD patients with cochlear implants, and reported that 75% of these individuals had speech perception scores equal to controls with sensorineural hearing loss. Simmons (I0) suggested that one reason for reported success following cochlear implantation for patients with ANSD is related to the site of lesion. For many individuals with the disorder, particularly in young children with ANSD, the pathophysiology involves either the inner





hair cells or the synapse between the inner hair cells and the eighth nerve, rather than the nerve itself. In these cases, the cochlear implant would be expected to bypass the site of the lesion, just as with a patient with a sensorineural hearing loss. Even if the eighth nerve is the site of the lesion, the discrete electrical pulses from the cochlear implant may be less affected by the impaired function, and increase or restore synchronous firing activity in the nerve.

Table II. Test results of patient 3 (tympanometry was normal).

	TEOAEs	PTA	CMs	ABR	CERA	Conclusions
Patient 3	Absent at time of study	Profound B SNHL. L cochlear implant present	Present on R	Not carried out at time of study but ABR absent bilaterally at age 14 months	Not carried out	Evidence of outer hair cell function but profound PTA_4 consistent with the diagnosis of ANSD

NB The degree of severity of the hearing loss was designated as profound, >95dB HL. British Society of Audiology [13].

Our experience is limited but does appear to suggest that CERA may be a helpful predictor of post-implantation performance. In counselling parents of children with ANSD about to receive cochlear implantation, we agree with the recommendation of Rance et al. (5) who suggest that CERA testing should be undertaken to help predict which children will make good progress with speech and language development post implantation, and which children may have difficulty, and benefit from additional, non-auditory communication strategies such as signing or cued speech. Presence of CERA responses would be consistent with reasonable speech perception (i.e. 'mild' ANSD), while response absence could suggest poor speech perception ability (i.e. more severe ANSD). Sharma et al. (11,12) suggest abnormal, or dys-synchronous, patterns of subcortical transmission, which occur in children with more disabling degrees of ANSD, have the potential to disrupt normal cortical development; it is this abnormal cortical development that explains the failure to evoke cortical responses. We would like to proffer an alternative basis for the lack of recordable cortical responses in some patients with ANSD: it requires only a modest degree of temporal dys-synchrony to 'smear' the relatively short latency responses of the ABR but it takes a correspondingly greater degree of dys-synchrony to abolish the longer latency cortical response.

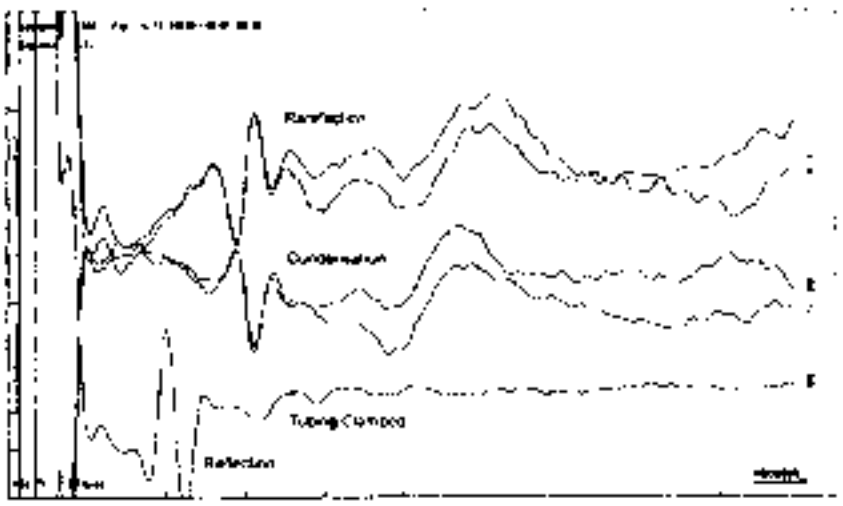


Figure 4. Cochlear microphonic waveforms of the right post-implantation ear of patient 3. The deflection in the response tracing elicited by the tubing control is thought to be an artefact related to reflection of the stimulus from the point of coupling.

Thus, if both the ABR and CERA are absent then we can conclude the dys-synchrony is profound and the prognosis is poor; if the ABR is absent but cortical responses are present we can conclude the degree of dys-synchrony is more modest and therefore the prognosis is better. In this model the cortex may or may not have achieved normal development and function. The clinical implications of the use of CERA responses in children with ANSD have been discussed recently in a publication by Cardon and Sharma (12), who draw attention to the suggestion that CERA responses may provide useful information regarding treatment and behavioural outcomes. We concur with this view.

In summary, we recommend that the diagnosis of ANSD should be considered in individuals who make slow progress with auditory language development, and have difficulties with comprehension of speech in spite of exhibiting satisfactory aided thresholds following cochlear implantation. ANSD should be considered both in risk groups for the condition, and in individuals who do not have risk factors.

Finally, CERA testing appears to offer a predictive factor to speech perception performance, and should be considered in all cases of ANSD.

Acknowledgements

We wish to thank the patients and fami!y members who kindly took part in the study.

We also thank Llse Henderson, Paediatric Coordinator, Specialist Speech and Language Therapist, Manchester Auditory Implant Programme, The University of Manchester, for her advice regarding assessment of progress in language development following cochlear implantation.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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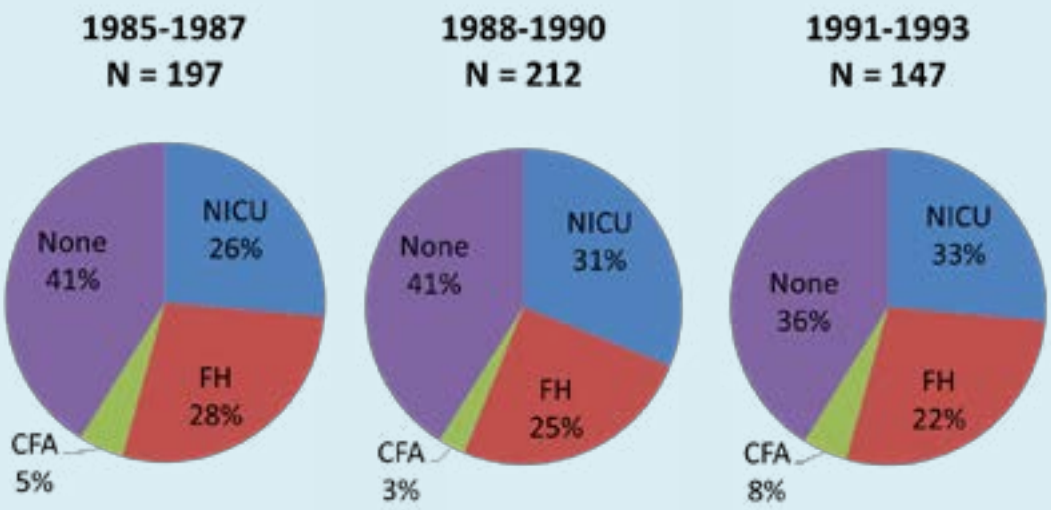
A survey of hearing outcomes in infants at or less than 30 weeks gestation

Dr Glynnis Parker
Sheffield Children's Hospital
Chesterfield Royal Hospital

Evidence of prematurity as a risk factor for permanent hearing loss:

Trent Ascertainment Study, *Fortnum, Davis et al 1996*

Distribution of risk factors among children with PCHI



Das (1996)

- 339 cases of PCHI 1981-1990
  - perinatal factors in 12.8% ……..majority prems
  - particularly RDS/ ventilation, hyperbilrubinaemia

Uus, Bamford (2006)

- 169 infants with PCHI
  - 44% NICU > 48hrs
  - 10% ANSD

Previous studies of hearing outcomes in extreme prems

Robertson et al (2009):

1279 survivors ≤ 28 wks / BW <1250g 1974-2003

- 3.1% (40) PCHI
- 1.9% (24) severe/prof PCHI

Gestation age: mean	Birth weight
No HL 26.6 wks ± 1.3	No HL 929g ± 178
HL 26.2 wks ± 1.4	HL 863g ± 160

Kramer (1989)

667 NICU infants 2.4% had severe bilat PCHI

Lorenz (2001)

3-5% of survivors ≤ 26 wks gest had significant hearing disability

Marlow (2000)

15 infants <33 weeks gestation with PCHI

Risk increased with

- low birth weight (mean 960g),
- prolonged ventilation,
- acidosis, repeated use of dopamine/frusemide

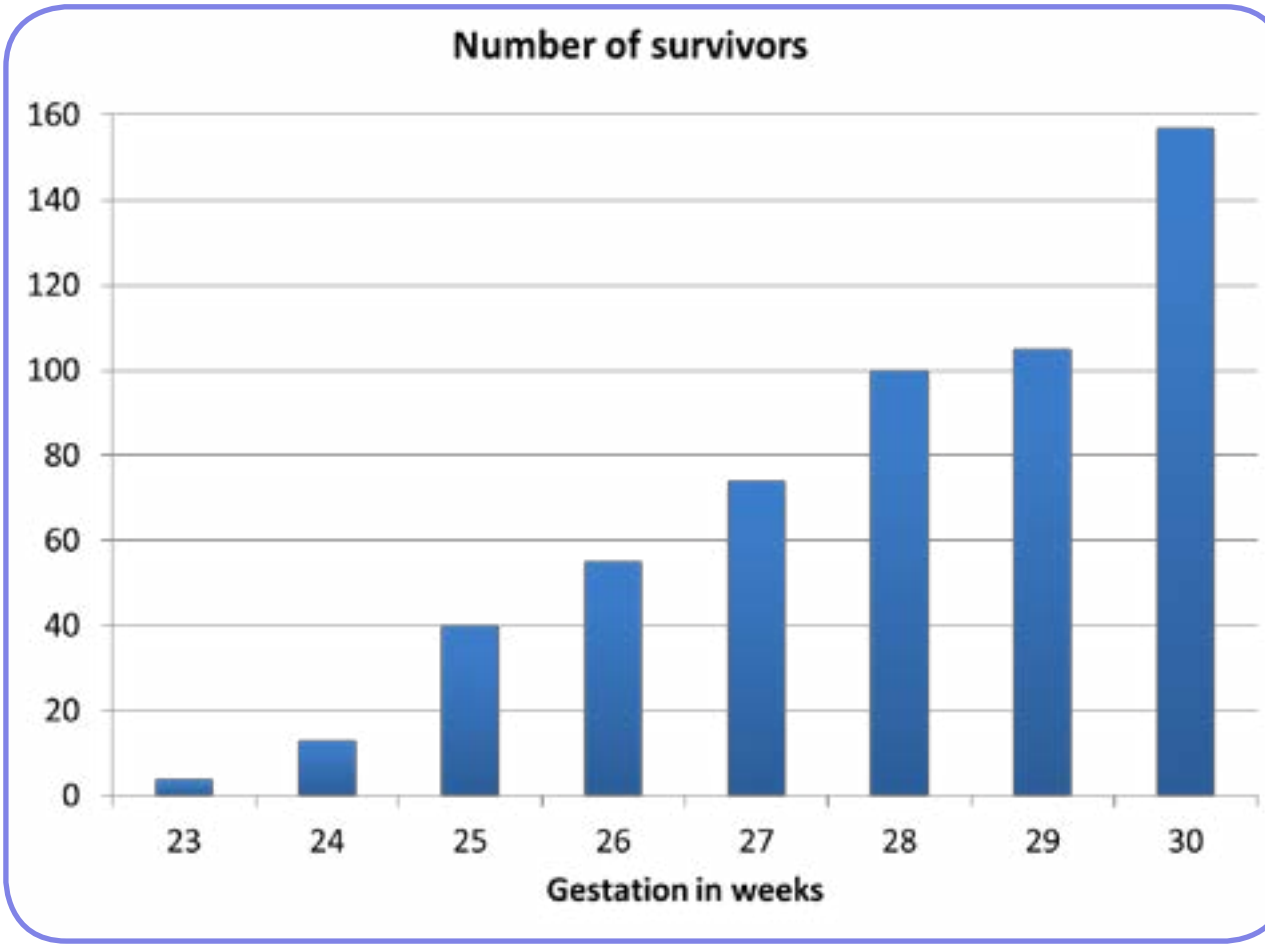
Study method

ESP database used to identify-

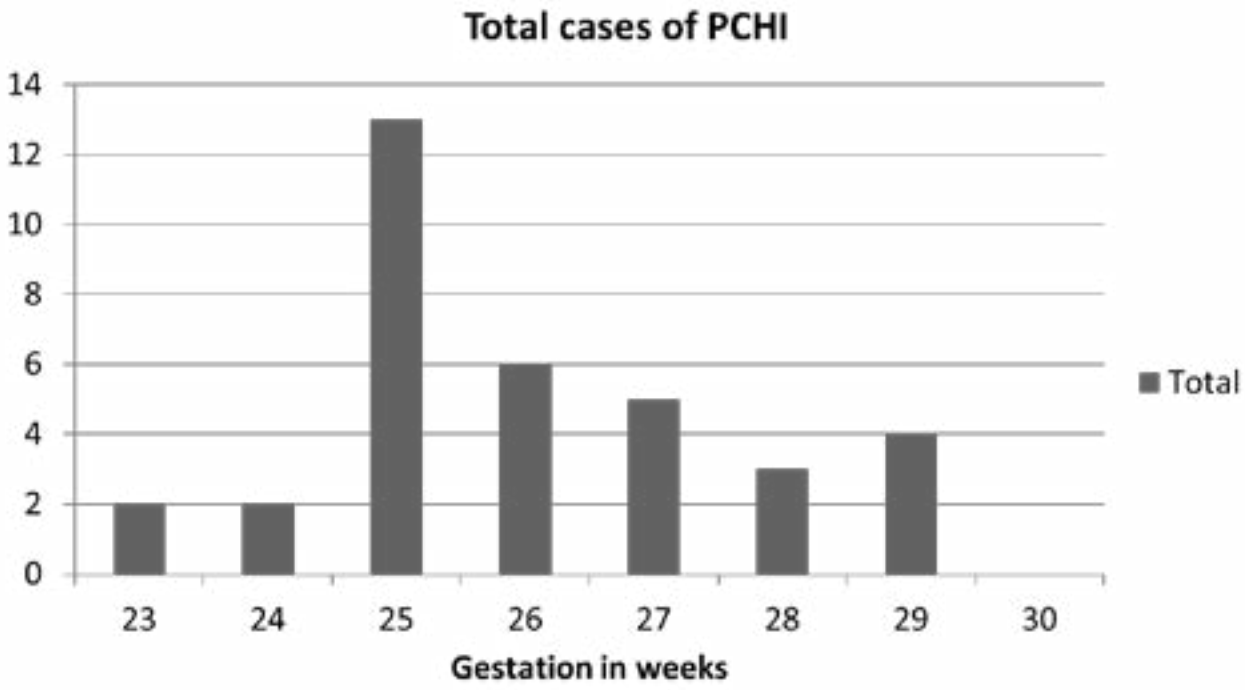
- All infants registered with gestation age = or < 30 weeks under Sheffield and Chesterfield NHSP between 6/10/2003 and 05/10/2010
- Results of subsequent audiological assessments sought for:
  - 'cases' from screen
  - those with no newborn outcome
  - late identified hearing loss
- Exclusions:
  - Deceased
  - Any infant with no newborn or follow up outcome

549 infants fulfilling study criteria

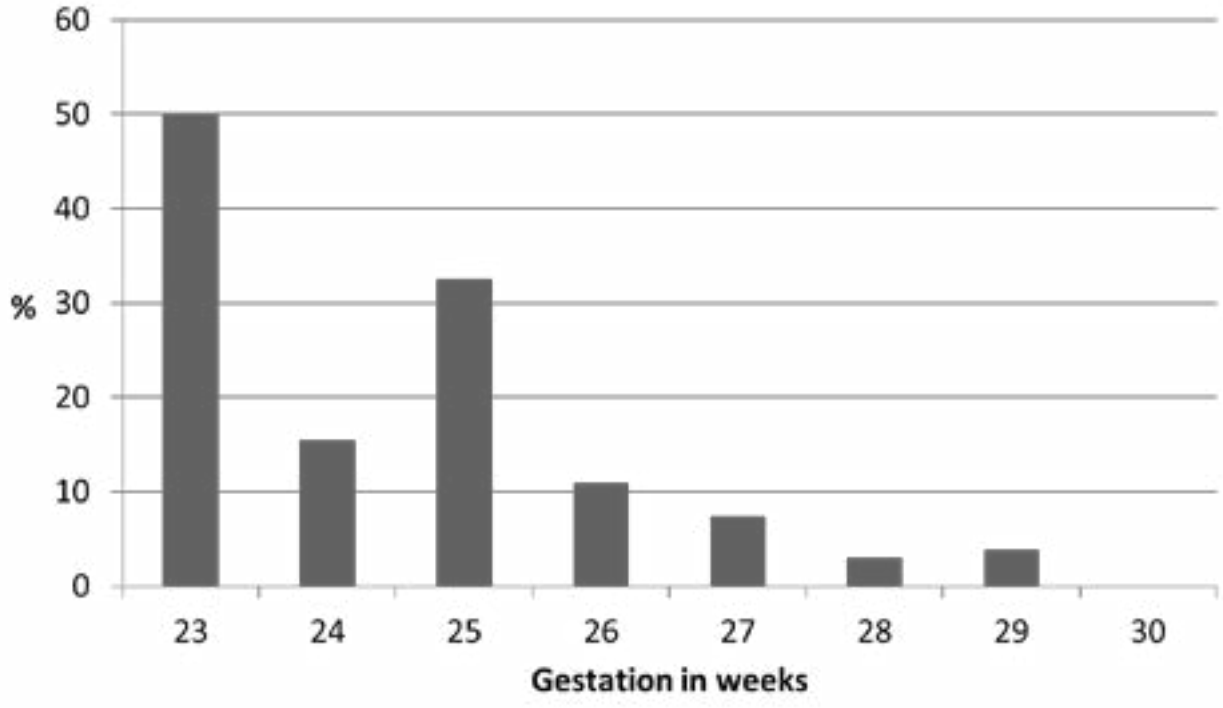
- **34 infants with PCHI** — **Yield = 62/1000**
- **31 bilateral SNHL >40dBHL av** — **Yield = 56/1000**
- **10 severe/profound** — **Yield = 18/1000**
- **3 unilateral SNHL >40dBHL av** — **Yield = 5 /1000**
- **15 bilateral ANSD** — **Yield = 27/1000**

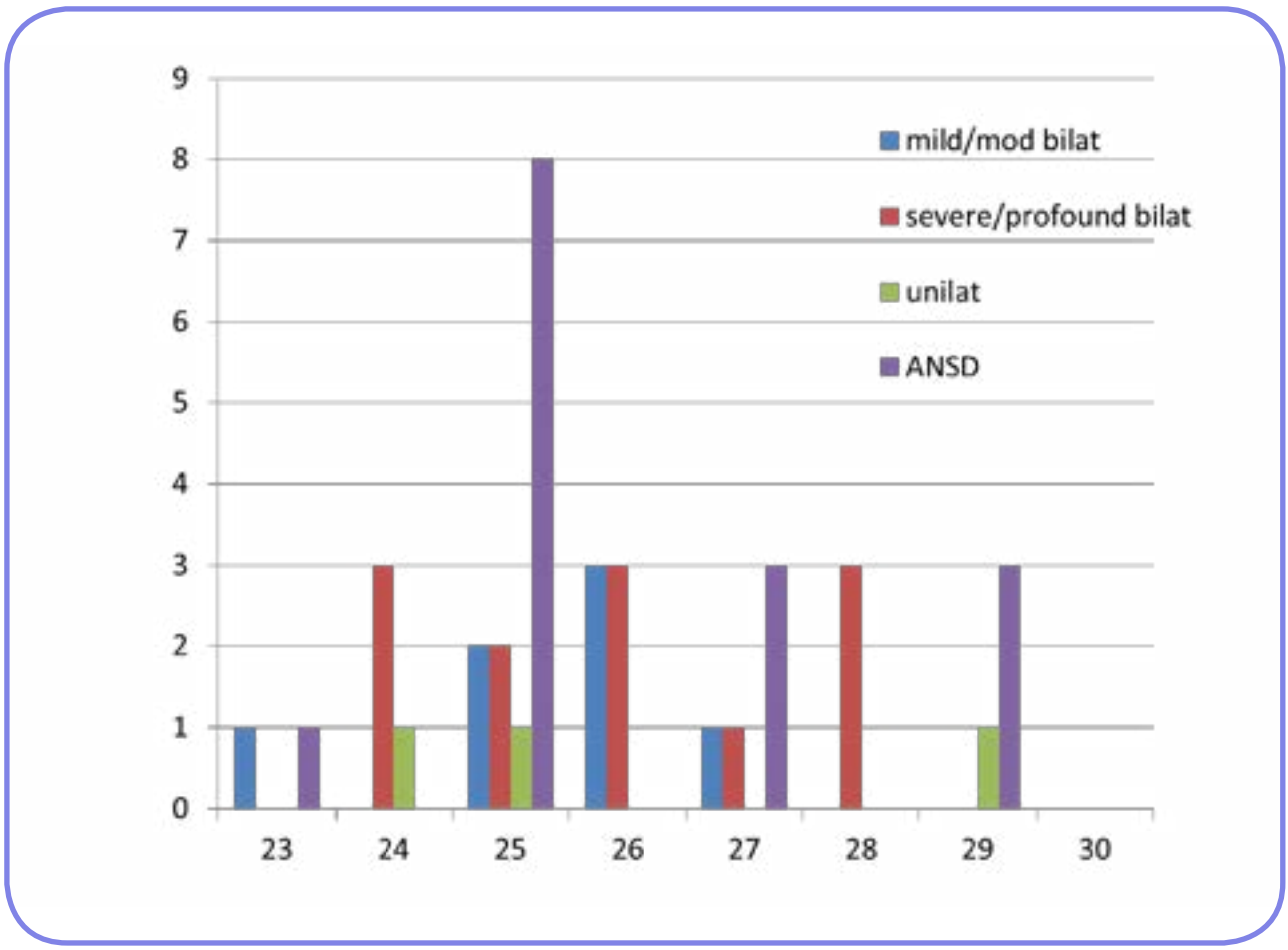


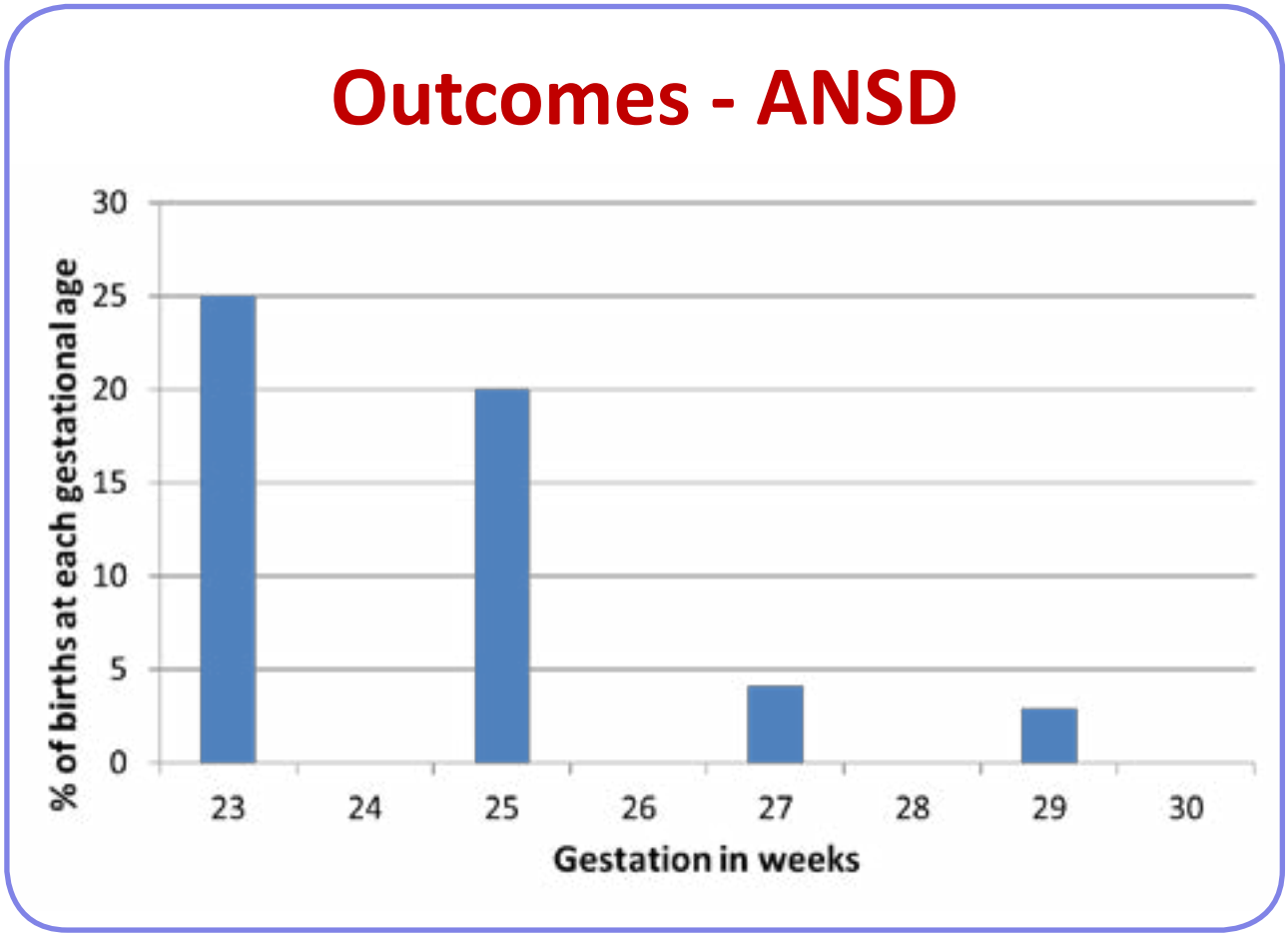
Hearing outcomes



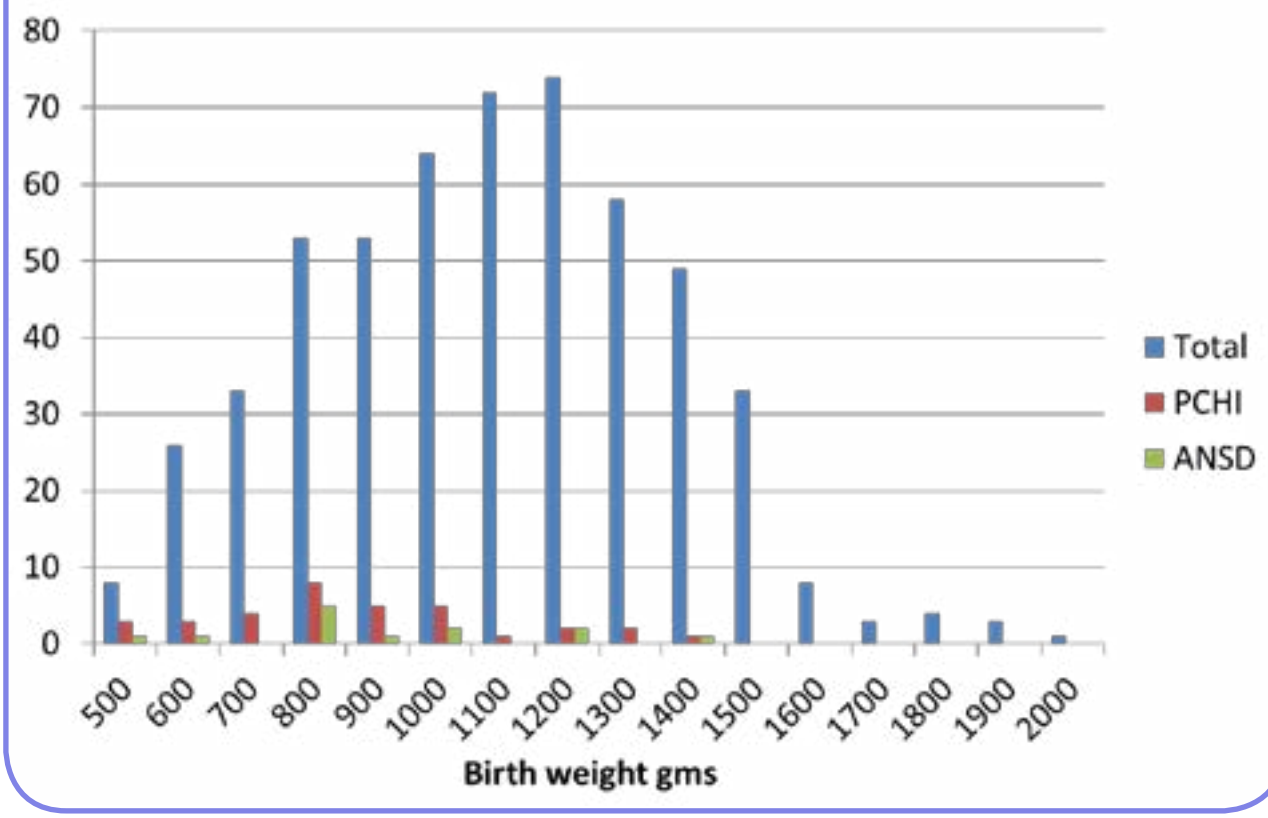
Outcomes – PCHI as %



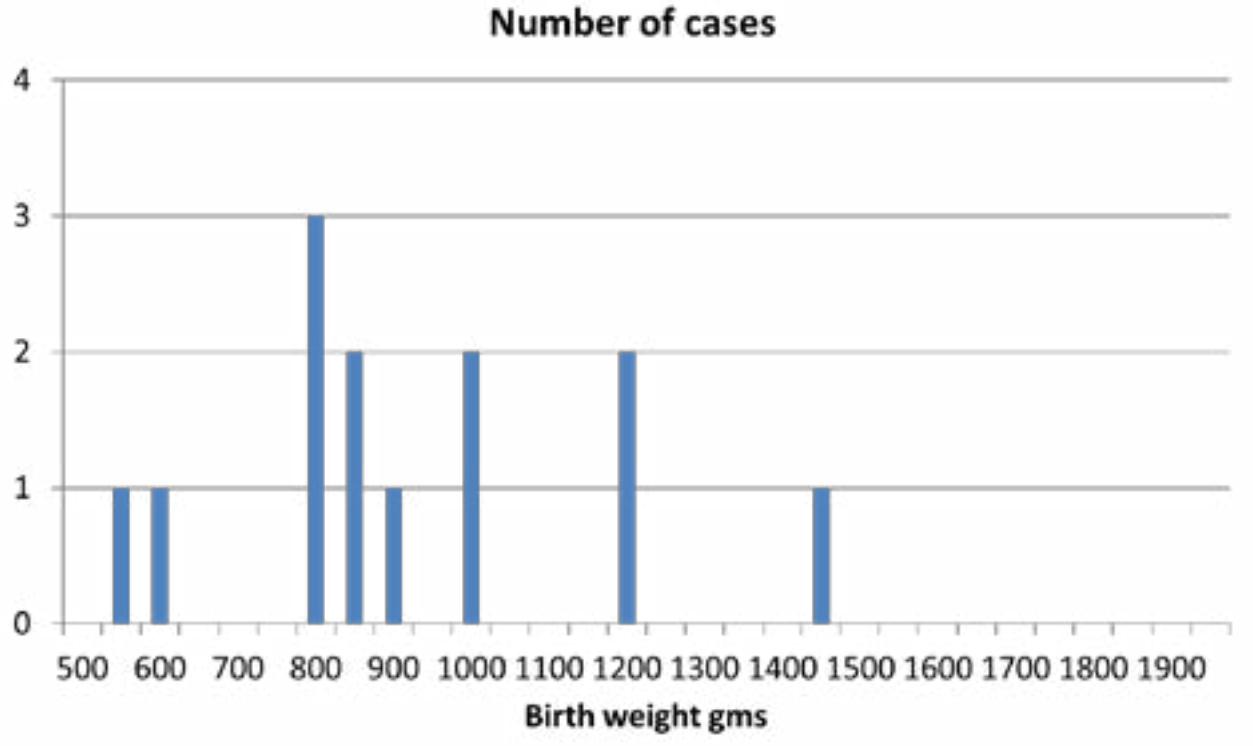

9
8
7
6
5
4
3
2
1
0
23
24
25
26
27
28
29
30
mild/mod bilat
severe/profound bilat
unilat
ANSD


Outcomes - ANSD
% of births at each gestational age
30
25
20
15
10
5
0
23
24
25
26
27
28
29
30
Gestation in weeks

Distribution by birth weight



ANSD as function of birth weight



Prevalence of ANSD identified by newborn screening

In Sheffield *(Parker, Stevens et al, 2000)*

-1 in 7 (14%) 'cases' with bilateral absent ABR had ANSD

From UK NHSP

- ANSD identified in 14% of reported 'true cases' in 'first wave' sites *(Uus et al 2004)*

- AN identified in 37 out of 324 (11.4%) infants identified by screen with moderate or greater bilateral loss – 0.09 per 1,000 infants screened

(NHSP annual report 2006 -2007)

Auditory neuropathy: unexpectedly common in screened population *Dowley et al (2009)*

45,050 infants screened 2002-2007

- 30 cases of severe/profound bilat SNHL

- 12 ANSD

	ANSD	PCHI/non ANSD
NICU	12/12	7/18
Gestation	33wks mean Range 25-40 wks	35 weeks mean Range 23- 40 wks
Mean birth weight	1863 g	1976g
Mechanical ventilation	10/12 Mean duration 9.2 days	7/18 Mean duration 34.3 days
Hyperbilirubinaemia	4/12	0/18
Sepsis	8/12	4/18

Time course of axonal myelination in the human brainstem auditory pathway,
Moore JK et al (1995) Hearing research

" Structures in the human brainstem auditory pathway, from the proximal end of the cochlear nerve to the inferior colliculus, undergo myelination between the 26th and 29th fetal weeks. By the 26th week of gestation, axons in the cochlear nerve and brainstem pathways have acquired linear arrays of oligodendrocytes and faint myelin sheaths can be distinguised. By the 29th week, definitive myelination is present in all auditory pathways…."

Summary

- **PCHI is relatively common in extreme preterm infants, particularly those born at or before 25 weeks, when around 1 in 3 may be affected.**
- **The incidence falls significantly by 30 weeks.**
- **ANSD was diagnosed in over half the cases of PCHI at or under 25 weeks**
- **This may be related to effects on myelination of the auditory nerve**

Further considerations

- **Suitable for a national study using ESP?**
- **Should extreme prems with absent/raised ABR thresholds be initially managed according to the ANSD protocol?**
- **Should extreme prems with satisfactory ABR thresholds as newborns be kept under surveillance for minor manifestations of ANSD eg APD?**

BAPA Audit Prizes - January 2014

Dear BAPA Member,

The 2014 BAPA Conference will have a session for prize winning audit presentations in the afternoon. Members from each region are invited to submit short audits to present (approximately 15 minutes) at the January conference.

There will be 4 prizes. The best from each region will be considered first. In the event that all prizes cannot be allocated to different regions then the remaining prize(s) will be allocated irrespective of the region. If your audit is selected you will be requested to present it* at the BAPA annual conference in order to receive the prize.

To enter your audit for the prize presentation, please provide the following information:-

A Prize Audit application form is also available on the website www.bapa.org.uk

Name of Audit Lead and/or Presenter *	
Title of audit	
Methodology used	
Outcomes of the audit including how will this affects your practice and plans for future audit	
Relevance of your audit to the specialty of paediatric audiology	

*if you are unable to present the audit at the conference another may be selected

Please submit your application to: BAPA Secretary, Veronica Hickson Veronica.Hickson@wales.nhs.uk as soon as possible.

The closing date for submission is 15th November 2013. Entries after this date will not be considered.

All audits welcome!

•Please continue on a separate page if more space required.

Review of Unilateral Referrals from Newborn Hearing Screen to Audiological Assessment.

Author : Adrian Dighé – Team Leader Bath(Wiltshire) NHSP

Aims of Review

Routine data quality checks revealed that for well babies rates of unilateral refer at Automated Auditory Brainstem response screen (AABR) were higher than bilateral referral rates (see chart 1). The aim of this review was to investigate unilateral referrals for all babies from screen completion to Diagnostic Assessment to ensure national protocols are adhered to and to examine diagnostic outcomes.

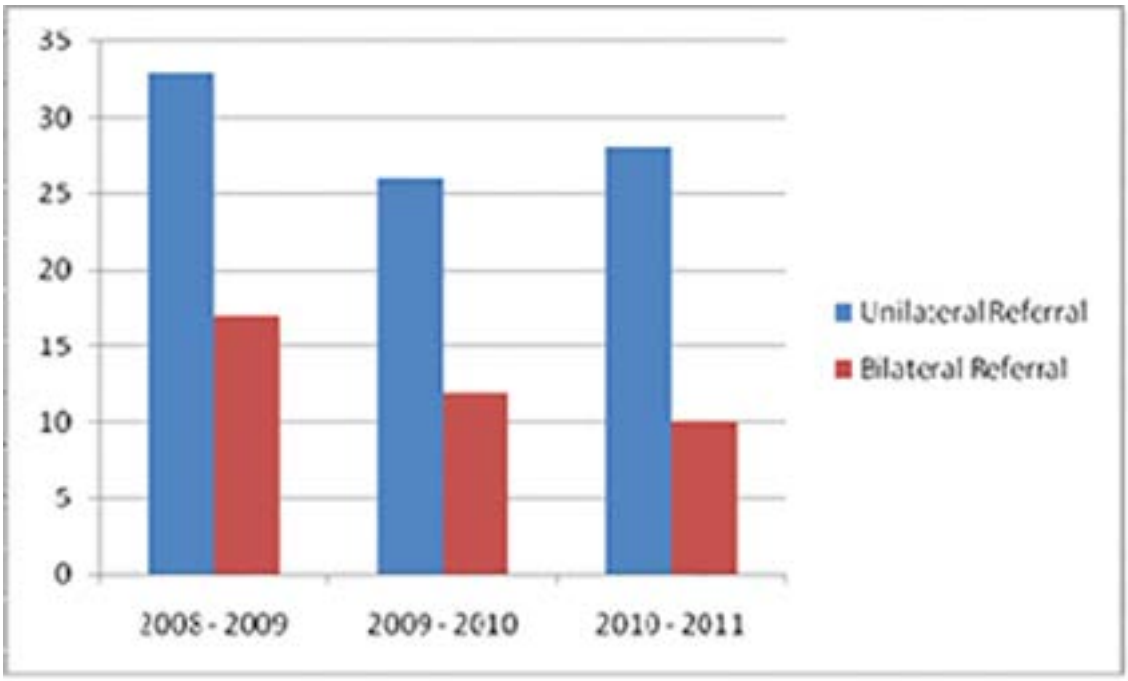


Chart 1: Well Baby Referrals from AABR screen to Diagnostic Assessment

Programme protocol

Bath and North East Somerset is a Community Site with a birth cohort of approximately 5000. Health Visitors (HV) perform the initial screen of well babies using Oto-Acoustic Emissions (OAEs) at approximately 10 – 16 days of age. If no clear response is obtained following a second attempt the Health Visitor refers baby for an Automated Auditory Brainstem response screen (AABR). This is performed by the Local Manager of the Programme who places soft headphones over the baby's ear and a click stimulus containing a range of frequencies is delivered at a predetermined level of 35db. At this point both ears are re-tested.

If 'no clear response' is obtained in either ear (unilateral) or both(bilateral) at AABR, the baby is referred for Diagnostic Audiological Assessment within 4 weeks. Diagnostic Auditory Brainstem Response (ABR) is performed by an Audiological Scientist.

For babies admitted to a neonatal intensive care unit (NICU) for more than 48 hours the screening protocol is slightly different. These babies have two screens, OAE and AABR. These screens can only be performed when baby is over 44 weeks gestational age and deemed well enough for screening. For NICU babies referral for audiological assessment is determined on the results of the AABR, regardless of the results of the OAE. However if the baby has a bilateral no clear response using OAE and a clear response with AABR the baby will be referred for a targeted review when baby is eight months corrected gestational age. The pathways are illustrated in diagrams 1 & 2.

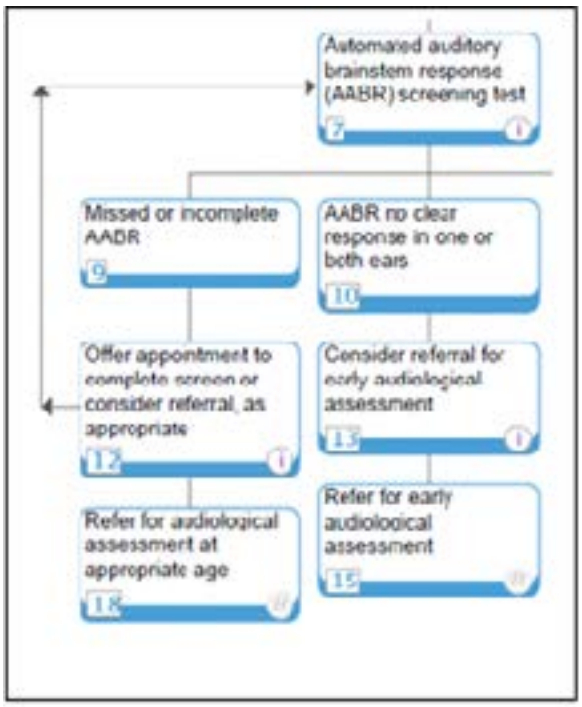


Diagram 1: well baby screen pathway

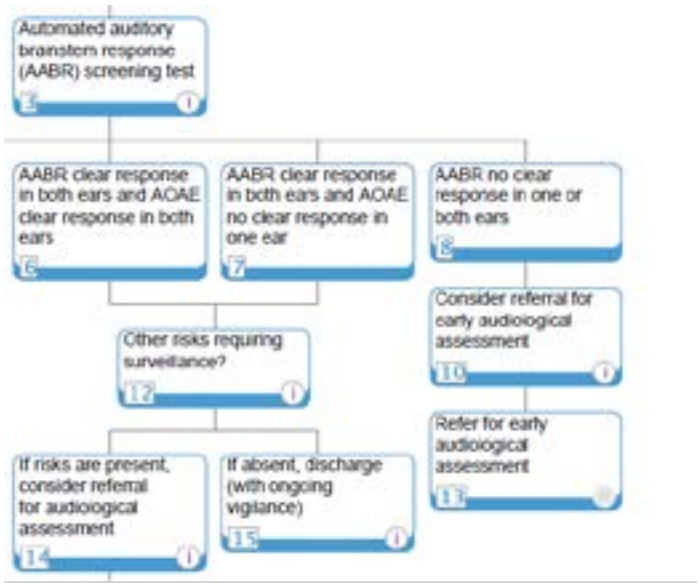


Diagram 2: NICU baby screen pathway

The relevant Standards from the national newborn Hearing Screening Programme that relate to unilateral referrals are Standard 14 & 16b

Methodology

The total numbers of unilateral referrals at each stage of the patient journey over a three year period, from April 2008 to March 2011 were identified from the electronic screening database (eSP). The outcome of audiological assessment for those babies referred was recorded.

Results

During 2008-2011 74 well babies and 36 NICU babies were referred for audiological assessment following a unilateral no clear response at AABR (see chart 2)

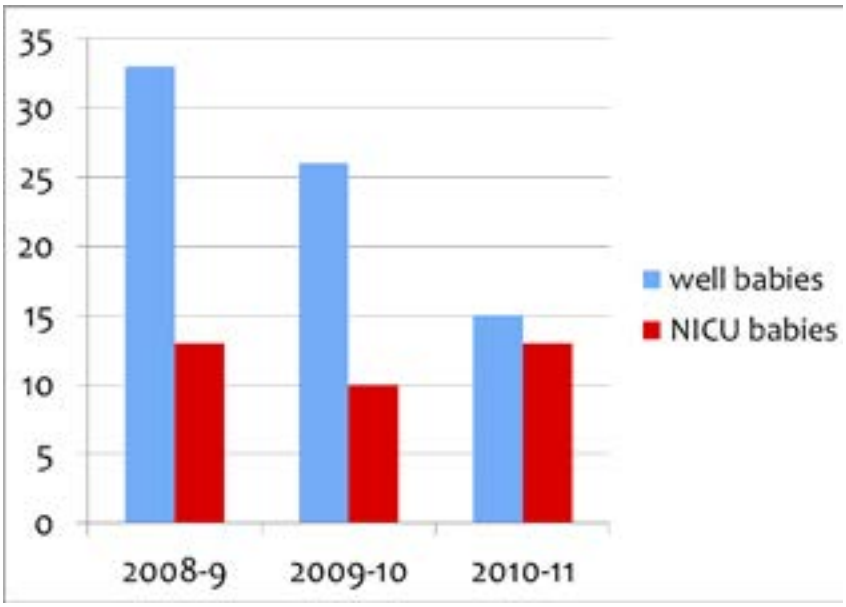


Chart 2: babies referred 2008-2011 at AABR with unilateral no clear response

Outcomes of all unilateral referrals at ABR are shown in table 1.

Outcome	2008	2009	2010
Bilateral Conductive	9	9	10
Unilateral Conductive	8	2	3
Bilateral SN	3	4	2
Unilateral SN	2	5	3
Satisfactory	17	12	6
SN/conductive	1		
Deceased	1		
ANSD			1
Lost to Follow Up			1
Conductive Permanent		1	
Did Not Attend	5	3	2

Table 1: ABR Outcomes of all unilateral referral

The data set is further split into referral outcome for well baby and NICU baby pathways (tables 2 & 3)

Outcome	2008	2009	2010
Bilateral Conductive	7	7	4
Unilateral Conductive	8	2	2
Bilateral SN	2	2	2
Unilateral SN	2	4	2
Satisfactory	11	7	4
Conductive Permanent		1	
Did Not Attend	2	3	
Lost to Follow Up			1

Table 2: ABR outcomes – well baby unilateral referrals

Outcomes	2008	2009	2010
Bilateral Conductive	2	2	6
Unilateral Conductive			1
Bilateral SN	1	2	
Unilateral SN		1	1
Satisfactory	6	5	2
ANSD			1
Deceased	1		
Did Not Attend	3		2

Table 3: ABR outcomes – NICU babies

Details of outcome for unilateral referrals resulting in Bilateral Permanent Childhood Hearing Impairment (PCHI) are given in table 4

Case	Notes	AABR result	Hearing loss	Comments
DJ	Well baby - ?Autism	Bilateral refer	Bilat HF SNHL Moderate	Incorrectly coded
TR	NICU Complex congenital heart disease	R Pass L refer		
	R est HL 60dB L est HL 40dB	Deceased (cardiac surgery)		
MS	Well baby	R refer L pass	Bilat SNHL Insert VRA R 40 30 30 40 L 35 35 35 40	Bilateral aided
CA	NICU Downs Syndrome	R pass L refer	Bilat mixed insert VRA R 60 50 45 55 L 50 35 40 NT	Bilat aided
CB	Well Baby	R pass L refer	SNHL Mild L Mod R	Ongoing review not aided
HL	Well baby	R refer L pass	Bilat HL R NR-100-NR-NR L 40-55-60-60	Bilat aided progressive loss R static L
CK	Leukodystrophy syndrome	R pass L refer	Bilat SNHL Mod	Deceased
JD	NICU baby removal in Known CMV + Cerebral palsy	L pass R refer	L mod R severe SNHL	Aided L only at present
MC	Down's Syndrome	R Pass L Refer	Mod bilat SNHL @ ABR - BC @50dB bilat 4K Tone	Removal OUT

Table 4: Details of cases of Bilateral PCHI after unilateral referral

Table 1 contains data for unilateral referrals who did not attend for diagnostic ABR. Outcome at targeted review for these cases is presented in table 5

Year & Baby	ABR Outcome	Targeted or assessment outcome
2008 all babies (N=5)	DNA	DNA
2009 all babies (N=3)	DNA	DNA
2010 BM	DNA	Satisfactory at review
2010 BC	Attended ABR 1, declined ABR 2	Satisfactory at review, to continue monitoring as Downs
Total N=10		

Table 5: Targeted review outcome of DNA at ABR

Discussion

High levels of satisfactory outcome at ABR testing of unilateral referrals in 2008/9 (17 and 12 respectively) were thought to be due to screener technique. The screener was removed and 2010 levels greatly reduced as expected. Monitoring this trend will continue.

As expected a number of unilateral referrals are confirmed to have a unilateral PCHI (10 of 110 - 9%) on the same side. Not unexpectedly a significant proportion of unilateral referrals have a temporary conductive loss either unilateral or bilateral (41/110, 37%).

The small group of great interest are those cases that show bilateral PCHI at ABR following unilateral referral. The AABR screen delivers 35dB of sound at mixed frequencies. Technically at this level although a clear response is obtained, a mild hearing loss can be present. So it is expected that a unilateral refer could identify an individual with a mild loss in the non- referring ear and a loss in the referring ear. Case MS could conceivably be in this category. Case CB does not fit this category unless there had been an error in recording which ear had been tested (in this case a mild loss on the right is feasible but in fact the loss that side was moderate).

What explanations are there for those remaining 7 cases? Each case would appear to represent a progression during the interval between AABR and ABR. Not a lot is known about progression of hearing loss in the early weeks and months of life. Progression is likely in case JD where a confirmed diagnosis of congenital cytomegalovirus infection was made very early on. All cases had testing for CMV and were negative (investigation records for cases TR and CK were incomplete).

Clearly more research on early progressive hearing loss is required, and each unilateral refer case requires careful assessment to this end. This small study does present evidence to support the practice of screening both ears.

Satisfactory outcome at diagnostic ABR could represent cases where neural pathways continue to mature between the AABR measurement and diagnostic ABR – in which cases satisfactory hearing levels are expected as an outcome. This could be the explanation for the small number of satisfactory

outcomes once screening competence was addressed

Finally the Bath team addressed cases of non-attendance for diagnostic ABR, so that numbers have reduced and assiduous follow up allows targeted review data to be collected on those still not attending diagnostic ABR. Results are encouraging.

Acknowledgements

The author would like to thank Vicky Mainstone Bath NHSP Local Manager, and the audience at the BAAP Audit Meeting September 2011 who offered comments and questions

Adrian Dighé – September 2012

For the 27th AGM (2nd as a company).

January 25th 2013

Reports from around the Regions

Regional Representatives

Liaison between members in the area and reporting back to the Quarterly Executive meeting are the main duties of the Regional Representative. Organisation of local meetings is encouraged.

Report Yorkshire and the North East

We continue to have an active group in this area meeting every six months. Our venue has changed from St Mary's Hospital in Leeds to Doncaster Royal Infirmary, as I have now taken over from Kathleen Coats as regional representative.

This year we have had varied contributions for the meetings from several of our members. Each meeting consists of about 6-8 members. Presentations have ranged from exposing us to the work of professionals in trying to improve services for deaf children and young adults, to case presentations for interest as well as peer review.

In March at St Marys, Hospital, Leeds, members presented ''Audiology Input in Special Schools', 'Social Care Involvement for Children with a Hearing Loss' which both led to discussions and food for thought in these areas. We concluded by talking about auditory processing disorder and some research in this area taking place in Sheffield.

Our latest meeting took place in October at Doncaster Royal Infirmary. We had a 'Case Presentation' afternoon.

There was a presentation of several vestibular cases, a reminder to us that there are several children with vestibular difficulties, as well as an encouragement to try and perfect our own vestibular assessments and go on to further investigations. Another case was a peer review of a possible case of auditory neuropathy spectrum disorder in a well baby. This led to discussions on the timing of the repeat auditory brainstem response, the real meaning of the results of the cochlear microphonics and also further management. The usefulness of speech discrimination was then discussed after a further presentation of a case of a teenager with poor speech discrimination disproportionate to his small level of hearing loss. This patient went on to have an MRI scan and resection of a cerebellar tumour within the next 10 days, illustrating the need to be very astute in our interpretation of symptoms

Winifred Baddoo (North East and Yorkshire Rep.) email:winifred.baddoo@nhs.net

NW BAPA region report

The NW had its second meeting of the year on the 30th Nov 2012 on 'Auditory Processing Disorders-Diagnosis and Management'.

The agenda included an outside speaker- Dr Johanna Barry and Dr Ansar Ahmmed both well-established workers in APD. It was held at the Royal Preston Hospital Education centre 2 and 13 attended. The meeting was interactive and extremely helpful in understanding APD. Dr Johanna Barry, Head of the MRC at NIHR (Nottingham Institute of Hearing Research) who has an extensive APD background, gave a clear and easy to understand outline of her current research project, a questionnaire (ECLIPs), she designed and validated on > 900 children. This is likely to have significant

clinical implications in helping to identify APD on continuum that included Specific language impairment (SLI), ASD and ADHD and compared favourably with current questionnaires – CHAPPS and SCAN C.

It tied in neatly with the talk on management Dr Ansar Ahmmed (Consultant Paediatric Audiovestibular Physician in Preston) also based on his research work in this on APD. We in the NW will be encouraging our kids who fall within this spectrum to have fun - juggling and learning a musical instrument!

I would like to draw attention to the fact that I am retiring from Paediatric AVM after 25 years in Paediatric Audiology, first as a CMO/ SCMO in East Berkshire, then as consultant in Paediatric (and Adult) AVM in Bolton. If anyone is interested in taking over this, now is the time to throw your hat into the ring. As the above meeting ran over time, I was unable to put this to those that attended.

I have enjoyed the one year as NW regional representative and wish it and BAPA on-going success in all its endeavours.

Please contact a member of the Executive Committee with proposals for the next NW Region Representative.

Dolores Umapathy (NW Region)

Report from Midlands

The Regional Representative for the Midland region is Dr M Ganesh. He can be contacted at m.ganesh@telfordpct.nhs.uk

Report from Northern Ireland

No specific BAPA meetings have been arranged in Northern Ireland but the Paediatricians involved in Audiology meet up at the BACCH meetings which recommenced in NI last year. I represent BAPA interests on the Regional BACCH Committee and Dr Anne Dooley and myself will be organising Audiology Workshops at one of the BACCH NI days this year.

Esther Harper. esther.harper@westerntrust.hscni.net

South West and South Wales Region

Please contact any member of the Executive if you are willing to be, or are proposing a colleague with their consen,t to be, the Local Regional Representative. This area has been without a representative for some time.

South East Region

Please contact any member of the Executive if you are willing or are proposing a colleague, with their consent, to be the Local Regional Representative. This area was previously active but has had a reduction in numbers and no representative since the retirement of the previous representative.

Report from Scotland

BAPA Scotland continues to meet regularly, three times a year, at Perth Royal Infirmary.

Our annual regional AGM was held at our meeting on 7th March 2012, with no changes to the current office bearers:

Chair: Ruth Henderson

Vice chair: Christine Niven

Secretary: Martina Stones

Treasurer: Alison Schulga

The business component to our meetings covers a variety of topics and continues to review Newborn Hearing Screening across the country, with particular interest and discussion around changes to the IT support and data collection for the screening programmes. eSP Northgate will no longer be used in all areas and alternative systems are being evaluated. Updated guidelines for surveillance and audiology referral of infants and children following newborn screening, based on the revised NHSP guidance have been rolled out in Scotland.

In the clinical component to our meetings we often have case discussions and peer review/support. This year we have been busy organising a Scottish study day on aetiology of hearing loss, for BAPA Scotland members plus invited medical guests, which went ahead on 28th November. We were particularly pleased to attract interest from paediatric trainees. Our keynote speaker was Dr Breege McArdle from the Royal National Throat Nose and Ear Hospital who gave a great overview of the current guidelines and how to interpret and implement them. Other speakers were local Scottish colleagues from Virology, Radiology, Clinical Genetics and Neonatal Medicine. Initial feedback has been excellent.

The next BAPA Scotland meeting was held on 6th March 2013

Ruth Henderson (ruth.henderson@luht.scot.nhs.uk)

Calling all would-be humanitarian audiologists

Sound Seekers is a small, London-based charity with projects in seven countries in sub-Saharan Africa (Cameroon, Gambia, Ghana, Malawi, Sierra Leone, Tanzania and Zambia). Our work is dedicated to improving quality of life for deaf and hearing-impaired people, in particular by assisting Ministries of Health to establish or develop audiology infrastructure and capacity. In the countries where Sound Seekers works, audiology services are few and far between, and where they do exist they usually lack the necessary equipment and qualified staff. Our work aims to address these gaps through providing training opportunities and audiological equipment.

The number of fully qualified audiologists in poor countries is pitiful. In Zambia, a country of 13 million people, there is only one. In Sierra Leone, a country of six million people, there are none. In several countries that we support, the only fully qualified audiologists are working in the private sector. Although Sound Seekers supports candidates from developing countries to follow courses in audiology, at the moment we are limited to sponsoring participation in a one-year diploma in Clinical Audiology. Those who complete this course usually return to their home countries and are expected to hit the ground running and establish an audiology service from scratch.

This is where you come in!

1) Are you looking for a challenge and can you spare at least two weeks*?

2) Do you enjoy coaching and mentoring?

3) Would you like to use you skills in a resource-poor setting, where little is known about what an audiologist is or does?

4) Are you keen to visit a new country and work with some fabulous people?

… then we would like to hear from you!

Sound Seekers is looking for audiology professionals to go to our project countries for a minimum of two weeks, to support staff on the ground that have received basic training in audiology and need help to establish or develop their service. If you are available immediately or in six months, please do email Emily Bell on projects@sound-seekers.org.uk and include a copy of your CV**.

*but the longer the better!

**If we can work out a suitable placement for you, Sound Seekers will help you raise funds towards your trip.

A boy having his hearing tested in Ndola, Zambia

A student at St. Joseph's School for the Hearing Impaired in Makeni, Sierra Leone

Annual London Conference

Friday 31st January 2014

SOAS, Brunei Gallery,

University of London

Russell Square, London WC1H 0XG

BAPA Conference Programme

09:00	**Registration and Exhibition**	
09:25	**BAPA:**	**AGM**
09:55	**Morning Plenary Session:**	**Chair: TBA**
	Introduction and Housekeeping	
10:00	**Lamb Inquiry: SEN and Parental Confidence**	
	Brian Lamb OBE, South Bank University	Visiting Fellow Centre for Government and Charity Management London
10:40	**Special Educational Needs of Deaf Children**	
	Scottish Speaker Girfec	
11:10	**Coffee and Exhibition**	
11:30	**Cleft Palate: A Surgical Perspective**	
	Mr Adrian Sugar	Consultant Cleft and Maxillofacial Surgeon, South Wales
12:10	**Cleft Palate: A Genetic Perspective**	
	Dr Annie Procter	Consultant in Medical Genetics, South Wales
12:50	**LUNCH**	**Posters and Exhibition**
13:40	**Afternoon Plenary session:**	**Chair: TBA**
13:40	**Current Research on the Neurophysiology of APD**	
	Dr Jennifer Linden,	Reader in Neuroscience, UCL
14:20	**Educational Management of APD**	
	Pauline Grant,	Specialist Education Consultant for Hearing
15:00	**Audit Presentations**	**4 selected presentations**
16:00	**Close of Meeting**	

BAPA Conference Registration Form

BAPA Conference – Friday 31st January 2014

Please reserve me a place at this meeting

Name	
Address, (for confirmation of place)	
Post code	
Work address (for delegate list)	
Post code	
E-mail	

❑ I enclose a cheque/BACS for £120 (BAPA members)

❑ I enclose a cheque/BACS for £130 (Non- members)

❑ Early Bird Offer £95 (BAPA and non-members – payment before 1st Dec)

❑ I enclose a cheque/BACS for £55 – Non-medic BSA, BATOD, BAA RCSLT: membership number _____

Payment options

❑ Cheque with your registration form made payable to BAPA

❑ Bank transfers to be credited to BAPA-RBS Preston Fulwood RBS 2 Lytham Road, Fulwood, Preston, PR2 8JB.

Account details: Sort code 16-20-16; A/C No. 10068508.

Please forward a copy of your payment advice to Mrs. Pam Williams, quoting the Customer name

❑ Please indicate here if you require a loop system

Special dietary requirements _____

Opportunities for poster displays, enquires to:

Dr J Dalzell / Mrs Pam Williams: pamelawilliams@onetel.com

Delegates are invited to submit audit presentations – please see BAPA website www.BAPA.uk.com for details.

Please email Dr V Hickson by 15 November 2013 (closing date)

Veronica.Hickson@wales.nhs.uk

Please return this form by email or post: Mrs Pam Williams

23 Stokesay Road SALE Cheshire M33 6QN, Tel: 0161 962 8915pamelawilliams@onetel.com

Closing date for applications Friday 17th January 2014

Any changes?

If any of your details have changed, please let BAPA know by sending your details to Isabelle Robinson: isabelle.robinson@rcpch.ac.uk
Please be sure to include the following:

Name,

Address,

Post code.

Preferred Email address,

Home Tel. No.,

Work Tel. No.