Auditory processing disorders.
A clinical perspective: controversies, ambiguities & challenges

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Before the introduction...

- “The rustle of grass as a rodent forages for food attracts the attention of an owl. The owl takes flight, using the time and intensity information carried by the displaced foliage to find its next meal. Hearing the owl approach, the rodent has just a few milliseconds to freeze or flee: one strategy leads to survival, the other to near-certain death. How is the prey’s location computed in the owl’s auditory circuits, and how does the rodent’s brain rapidly craft a strategy for survival from sound pressure waves alone? Answering these questions requires extraction of biological meaning from the sound pressure and frequency waves represented in the cochlea. Although medullary and midbrain auditory centers form topographic maps and analyze acoustic parameters underlying this behavior, survival requires that the auditory thalamus and cortex extract and transform information representing biologically and ecologically significant aspects of sound that are essential for perceptual analysis and behavior.”
- Winer et al., TRENDS in Neurosciences 2005

Lecture Outline

- Introduction
- Some insights from auditory neuroscience
- Definitions
- Symptoms, DD with Auditory Neuropathy, Aetiology
- Integrating Information for Diagnosis
- (Current) test battery approach, test categories, (commercially) available behavioural tests and the GOSH/NHNN test battery
- Management
- Final comments

Introduction

- Bocca and colleagues (1954) - brain pathology can lead to hearing difficulties which are not identifiable by conventional audiometry.
- Myklebust (1954) - central auditory function ought to be considered and assessed in children with communication disorders.
- in the last decades the notion that patients may complain of hearing or listening difficulties attributable to disordered auditory processing within the brain has been more widely acknowledged by health professionals.
- Prevalence unknown. “Guessimates” - 5% for school-aged children, higher prevalence that increases with age in the adult population.
Hearing and the brain

- Initial “decomposition” of the auditory signal in the cochlea
- Accurate transmission
- Further analysis (incl summation, subtraction or correlation) by the brain.
- “What” and “where” streams (Rauscheker 1998)
- Auditory cortex - sounds are represented as “auditory objects” (Nelken 2006).
- Neural representation of sound influenced by task-specific demands, expectations, and higher order top down effects at multiple levels of the auditory pathway.
  - Eg, auditory attention may modulate OAEs in a frequency-specific manner,
  - attentive expectation helps to shape cortical responses (Fritz et al., 2007).
- Common representation between auditory function and known cognitive, emotional and visual centres
- Functional organisation of the auditory cortex spans a continuum
  - basic sensory processing in primary areas
  - polymodal integration in non-primary areas
  - behavioural modulation in the limbic areas
  - multisensory processing in multisensory subregions (Winer et al., 2006).

Auditory Processing Disorder

APD results from impaired neural function and is characterized by poor recognition, discrimination, separation, grouping, localization, or ordering of non-speech sounds. It does not solely result from a deficit in general attention, language or other cognitive processes.

http://www.thebsa.org.uk/apd/Home.htm#working%20def
May 2007

Alternative definitions

(C)APD - “a deficit in neural processing of auditory stimuli that is not due to higher order language, cognitive, or related factors”. ASHA, 2005. Deficits in:

- Sound localisation/lateralisation
- Auditory pattern recognition
- Auditory discrimination
- Temporal aspects of hearing (masking, ordering, integration, resolution)
- Processing degraded auditory signals
- Processing the auditory signal when embedded in competing acoustic signals

ASHA, 1996
**Alternative definitions**

- “Disorders of central auditory processing”: “disorders in the processing of sound, after the transduction of the sound into neural activity in the cochlea.”
- Such processing involves the characterisation of auditory patterns in frequency or time that are used to identify and localise sound objects … before the patterns acquire labels or schemata”  
  Griffiths, 2002

**Shortcomings**

- APD (mostly) attributed to pathology of the brain, but contribution of peripheral hearing deficits
- Deficits in specific skills
- BSA definition: APD deficits demonstrable in processing of non-speech sounds. ASHA definition: deficits “should not be due to… language disorders”. TDG definition - APD takes place before semantic processing.
- Neither of the three definitions attempts to define the term “disorder”.

**DSM IV operational definition of a “Mental disorder”:**

- “a clinically significant behavioural or psychological syndrome or pattern that occurs in an individual and that is associated with present distress (e.g., a painful symptom) or disability (i.e., impairment in one or more important areas of functioning or with a significantly increased risk of suffering death, pain, disability, or an important loss of freedom…”
- Whatever its original cause, it must currently be considered a manifestation of a behavioural, psychological or biological dysfunction in the individual”

**Guidelines for inclusion of specific diagnosis in DSM III (1980)**

- at least 50 published articles pertaining to the diagnosis (with at least 50% of these to be empirical)
- specified diagnostic criteria, with available assessment to treatment linkages
- at least two empirical studies conducted by independent groups showing Kappa coefficients ≥ .70
- the proposed diagnostic category represents a syndrome of frequently co-occurring symptoms and
- there are at least two independent studies demonstrating that this diagnostic category is separate and distinct from other diagnoses.
What are the symptoms?

1. Adult studies

Validation of CAT studies + hearing questionnaire
- Neijenhuis et al 2003: 24 adults with suspected APD
- 68% abnormal CAT results
- subjects with suspected APD higher scores in Amsterdam Inventory
- speech in noise and sound localization most prevalent

- Blattner et al (1989) - patients with unilateral cerebrovascular lesions of CANS
- auditory perceptual problems reported in questionnaire:
  - 49% overall, 79% with abnormal CAT
  - situations with simultaneous speakers most prevalent difficulty
  - Of interest, these patients “often stated that they did not have any hearing complaints” prior to the questionnaire being administered.

What are the symptoms?

2. Children studies

  - 64 children with APD.
  - Listening difficulties in quiet, in noise, ideal listening condition, with multiple inputs present. problems with auditory memory & attention
  - children with APD showed a wide inter- and intra-subject variability in listening skills

- Meister et al (2004): parent-answered questionnaire to evaluate differences between children with a “suspected” APD (identified as clinical suspicion of APD and failure in non-validated tests, unspecified by the authors) vs. a control group.
  - APD group gave significantly poorer scores
  - Factor analysis - 7 main components: speech understanding in demanding situations (speech in competing speech, speech in noise, degraded speech), speech/language production abilities of the child, general behavioural issues (aggression and frustration), difficulties of the children with reactions to (orally given) questions and demands, reproduction of musical cues, discrimination of speech sounds, and loudness perception.

(C )APD-symptoms and behaviours

- Poor performance in confusing environments
- Difficulties following oral instructions
- Difficulties with rapid/degraded speech
- Difficulties in background noise

- Language, reading and spelling disorders
- Inattention, Distractibility
- Academic difficulties
- Higher likelihood of behavioral, emotional, and social difficulties.
- Not 1 to 1 correspondence between deficits and sequelae

Frequent Complaints

- “Can’t understand what people are saying in background noise”
- “Hear, but don’t understand”
- “Highly distractible child”
- “Child acts like she/he can’t hear, but has passed hearing screening”
- “Can’t remember series of instructions”
- “Difficulties learning a foreign language”
- “Difficulties understanding if someone talks quickly”
Presentation in children

• Symptoms may become apparent in the early school years or
• may be perceived as a problem at a later academic stage of the child's life, due to changes in the acoustic environment or to increased academic demands
• In rare cases, these symptoms may be the first manifestation of a neurological disorder

Behavioural indications of APD

• Infancy to early childhood:
  – baby not as alert/responsive to sound
  – less talkative
• Pre-school/Kindergarten
  – difficulty learning rhymes
  – difficulties sitting for story time
  – need tactile/visual cues to attend when spoken to
  – difficulties with oral instructions
  – “tune out”, “day-dreamers”
  – difficulties with phonemic awareness tasks

Behavioural indications of APD

• Primary school
  – difficulties with telephone conversations,
    announcements over loudspeakers, understanding tape-recorded speech
  – slower with phonics
  – problems grasping abstract concepts
  – failing behind peers
  – poor self-esteem
  – APD may be hidden until middle school years

Aetiology of (C)APD

• 1. Neurological conditions
  – stroke, hypoxia (e.g. prematurity), head trauma
  – brain tumours
  – meningitis
  – epilepsy
  – adrenoleukodystrophies, multiple sclerosis
  – GENETIC
    – heavy metal exposure
• 2. Auditory deprivation/Delayed central nervous system maturation
  – Glue ear, other types of hearing loss, idiopathic
• 3. Other Developmental Disorders
  – ADD, Dyslexia, Specific Language Impairment, Autism
• 4. Age related changes
• 5. “Positive” disorders of auditory processing
  – tinnitus, musical hallucinations
  Bamiou et al., 2001; Griffiths, 2002
Differential Diagnosis?
Auditory Neuropathy

- In 1996, Starr et al. described 10 subjects with hearing loss, with normal outer hair cell function and abnormal ABR and other reflexes that indicated an auditory nerve lesion. The term "auditory neuropathy" was coined.

- Nerve biopsies in AN accompanied by peripheral neuropathy show demyelination, axonal loss and reduced numbers of auditory fibers of the auditory nerve.
- AN could also be due to
  - a disorder of the synapse between inner hair cell and the auditory nerve,
  - or to selective inner hair cell loss.

AN Diagnosis. all three of the following:

- a. **evidence of poor hearing in the presence of normal or abnormal audiometric thresholds.**
- b. **evidence of poor auditory neural function**: elevated or absent auditory brainstem reflexes such as middle ear muscle reflexes and OAE suppression by noise and abnormal ABR.
- c. **evidence of normal hair cell function** such as normal OAE or cochlear microphonics.

50% of children with AN will have event related potentials to tone and speech stimuli, regardless of the degree of the hearing loss. This correlates with good speech perception ability.

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<tr>
<th>Table 2. Causes of auditory neuropathy</th>
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<td><strong>Autoimmune</strong></td>
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From Bamiou & Luxon, In: Dyck & Thomas, Peripheral Neuropathies, chapter 51.
### Diagnosis of APD requires a multidisciplinary assessment

- Detailed audiometry (including tympanometry, acoustic reflexes, OAEs and suppression and ABR) to check peripheral hearing and auditory neuropathy/dyssynchrony
- APD tests should include ≥ 2 non-speech & speech tests
- Tests of language, cognition (e.g. verbal and non-verbal reasoning), and short term auditory memory
- Other: observation of the child in the classroom record of academic attainment etc


### Behavioural Test Categories (ASHA, 2005)

- Binaural Interaction tests
- Dichotic tests
- Monaural Low Redundancy Speech Tests
- Temporal tests
- Auditory discrimination tests

### Integrating Information for Diagnosis

- Case History.
- Examination.
- Observation of auditory behaviours.
- **Audiologic test procedures- behavioural and electrophysiological**
  - Speech and Language tests
  - Psychology Assessment
  - ADHD checklist (by 2 individuals)
  - Other

### Which test?

**Test considerations**

- **Sensitivity** and **specificity**. Standardisation? Lesions of CANS/normal population/suspected APD
- Test-retest **reproducibility**
- **Age appropriateness**
- Verbal/nonverbal material. Control for **linguistic variables**.
- Memory load
- Response mode
- Test duration
- **Normative data**
Minimal APD Test Battery

- Behavioural
  - PTA
  - Performance – intensity functions for word recognition
  - Dichotic tests
  - Duration pattern sequence test
  - Temporal gap detection

- Electrophysiological
  - Immitance audiometry
  - OAEs
  - ABR and MLR

- Consider testing other modalities, eg vision

Consensus Conference on APD in Children, 2000

Range of responses in ERA plotted on a Logarithmic time scale

Otoacoustic emission suppression by contralateral noise
**Masking Level Difference**

- Presentation of pulsed tone (500 Hz) or spondee word to both ears simultaneously with binaurally presented masking noise.
- Find threshold of signal for homophasic (S and N in-phase) and antiphasic (S and N out of phase) condition.
- Difference of the two = MLD
- Brainstem lesions give abnormal results, cortical lesions normal. Good correlation between MLD and ABR
- can only be administered if hearing thresholds symmetrical between the two ears.
- Development of more sophisticated, “challenging” paradigms to tax the system and tease out the deficits, e.g. LISN by Cameron et al, use of different types of noise

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**Speech in noise/babble**

- White noise/speech noise/multitalker babble
- Contralateral deficits in temporal lobe lesions, but these patients may score within normal limits of variability
- BS lesions – abnormal results but no laterality pattern
- Test does not identify site of lesion

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**Dichotic Digits** (Musiek, 1983).

Sensitivity>70%, specificity>90%. Low linguistic load, resistant to cochlear SNHL

R ear advantage

**Dichotic CVs test** (Noffsinger et al., 1994)

6 CV syllables. Stop plosive (b, p, t, d, g, k) and the vowel /a/. A different CV is presented to each ear, with the syllables precisely aligned in time, at 50 dBSL. The listener is instructed to report what he/she hears.

**Dichotic Rhyme test** (Wexler and Halwes, 1983)

15 dichotic pairs of monosyllabic CVC words that begin with a stop consonant (b, p, t, d, g, k). Words in each pair matched for frequency content - difference in the initial consonant. The listener reports only one word.
Dichotic tests in patients with commissurotomy – an explanation

- Language perception takes place in the left hemisphere
- In the monaural situation both the ipsi- and contralateral pathway are functional for sound transmission
- In the dichotic situation, the contralateral pathway becomes dominant in auditory speech signal transmission.

Attentional modulation of dichotic speech test results

- R ear advantage may increase by focused attention to the right ear. Focused attention to the left ear will result in an increased left ear score (Hugdahl and Andersson, 1986).
- A PET study: decreased activation of primary and secondary auditory cortices in the forced vs. the non-forced listening condition. Facilitation of callosal transfer during focused attention (Hugdahl et al, 2000).
- Patients with lesions of the corpus callosum show lack of benefit from focused attention to the left ear on a dichotic CV task, (Pollman et al., 2002).
- Left ear performance in dichotic tests under the left ear forced attention condition may reflect callosal transfer efficiency (Hugdahl 2003) – both sensory and other input

Frequency and Duration Patterns

- Three tone burst sequences of Long (500 ms) and short (250 ms) tone
- Subject requested to label the sequence
- Pattern recognition on R, labelling on L hemisphere
- Not affected by SNHL. >80% sensitivity, ~90% specificity
- Requires complex response – effect of top down processes
- Taps into >1 auditory processes

Frequency/Duration Patterns

- Subject requested to label the sequence (high-high-low, or long-short-long)
- Not affected by SNHL
- >80% sensitivity, >90% specificity
Gaps in Noise test

- Monaural presentation of a 6 s white noise burst, in which 0 - 3 gaps of varying duration (2ms to 20ms) are embedded.
- Patient has to identify number of gaps in each noise burst.

Gaps in noise

- Leading and trailing markers stimulate same set of auditory nerve fibres. Representation of gap stimulus in time course of spike discharges of single cochlear fibres. Dip or discontinuity in ongoing discharge rate of cochlear nerve fibres corresponds to behavioural gap thresholds Zhang et al., 1990
- Some elderly listeners may exhibit loss in temporal resolution which is unrelated to hearing loss (Schneider et al., 1993)
  - Sensory processing, peripheral or central
  - Cognitive factors
- Longer gap detection thresholds correlate with poorer speech discrimination in noise, even when the SNHL has been controlled for (Tyler et al., 1982)
- Longer gap detection thresholds correlate with poorer reverberant speech discrimination (Gordon-Salant and Fitzgibbons, 1993)

Screening procedures: SCAN and SCAN-A Keith 1995

Filtered words: reflects ability to understand distorted speech in a poor acoustic environment (auditory closure)

Auditory Figure ground: reflects ability to understand speech in background competing noise

Competing Words/Sentences: indicates auditory maturation

SCAN and SCAN-A

- Easy to administer, quick
- From 3 years onwards

But:
- Test-retest reliability
- Validity (not validated on CANS lesions)
- Small normative data sample
- Screening, not diagnostic procedure
Test protocol at GOSH/NHNN

- The rationale behind the choice of tests is:
  - Baseline testing should provide an accurate assessment of middle ear & cochlear function and hearing sensitivity. Normal middle ear & cochlear function and hearing sensitivity is desirable to proceed to testing for (C)APD, however, testing for (C)APD can be modified to accommodate for findings of abnormal middle ear & cochlear function and of reduced hearing sensitivity.
  - A minimum of tests are administered to “screen” for (C)APD.
  - Additional tests may be given
    – in order to characterize a specific auditory deficit in more detail (with the view to inform the management plan)
    – if the minimum test battery is negative while the patient’s history is highly suggestive of (C)APD,
    – if the reported symptoms are lateralized to one ear.

Test protocol at GOSH/NHNN

- **Baseline tests**
  - PTA
  - Word recognition in quiet
  - OAEs, OAEs with suppression
  - Tymps & ARTs
  - ABR
  - (Note: OAE Suppression, ARTs and ABR are thought to assess overlapping but not identical brainstem pathways, and may underline different patient reported hearing deficits. Also, because auditory neuropathy may need to be DD from (C)APD).

Test protocol at GOSH/NHNN

- **Behavioural central auditory tests**
  - Dichotic digits
  - Frequency patterns (Monaurally conducted)
  - Duration patterns (Monaurally conducted)
  - (Note: Both FPT and DPT should be conducted, as a dissociation has been observed in these two test results and in left vs. right ear results in neurological patients).
  - Gaps in Noise and/or Random Gap Detection (Monaurally conducted)
  - Speech in Babble * (Monaurally conducted) or SCAN-A

Test protocol at GOSH/NHNN

- **Electrophysiological central auditory tests**
  - N1 P2 to fast temporal transitions of sound or MMN

Test protocol at GOSH/NHNN

- **Additional tests**
  - Dichotic rhyme tests and CVs
  - Why? If pt reports difficulties with integration of sounds/switching attention to 1 of 2 speakers/difficulties in background competing noise. Because of ceiling effects of DDT. Because DDT affected by attention, DR CVs not.
  - Note: norms with both non-focused and with directed attention.
  - Masking Level Difference
  - Why? If pt reports difficulties with localization, past history glue ear, suspected brainstem problem.
  - Note: LISN (listening in spatialised noise) test by Cameron et al., 2005
  - Environmental sound recognition
  - Why? Some pts with neurological impairment may have environmental sound agnosia (may be partial), eg Landau-Kleffner, CANS stroke etc.
Test protocol at GOSH/NHNN

- CHAPS /Kramer or Gatehouse validated questionnaires
- SLT assessment
- Ed Psych assessment/Cognitive Psychology
- Before clinic appt (children) after clinic appt (adults)

**Tymp**

**OAEs**

**ABR**

**OAEsupp**

**ART**

**PTA**

**Central Auditory Tests**

CAPE(IMAP) testing battery (IHR, UK)

- 14 measures of auditory processing

Normative study incl:
- peripheral hearing tests
- speech in noise and in quiet
- performance IQ, language
- memory, attention

Simple and composite measures of auditory processing abilities

Dissociations with other related skills

Moore, Aud Medicine 2006

The Newcastle Auditory Battery (NAB) Griffiths et al., 2001

Aim of battery:
- to define dissociated deficits in certain auditory functions.
- to allow inference about the existence of discrete psychophysical mechanisms for the perception of certain types of complex sound.
- to allows inference about the likely neural substrate for the perceptual process by comparison of the deficits with the known anatomical lesion
The Newcastle Auditory Battery (NAB)

- two-alternative-forced-choice (2AFC) psychophysics, full psychometric function
- carried out binaurally over headphones, at a sensation level of 60 dB,

- Norms on 30 naive normal subjects.
- personal computer software platform software implemented within Matlab

Montreal Battery of Evaluation of Amusia

“Multiple disorders of musical abilities can occur after brain damage. Conversely, early brain anomalies or vast brain injuries may sometimes spare ordinary musical skills in individuals who experience severe cognitive losses. To document these incidences, comprehensive behavioral testing is required. We propose to use the Montreal Battery of Evaluation of Amusia (MBEA) because it is arguably the best tool currently available. Over the last decade, this battery was developed and validated in populations with brain damage of various etiologies. Furthermore, the MBEA is theoretically motivated and satisfies important psychometric properties. It is sensitive, normally distributed, reliable on test-retest, and correlates with Gordon’s Musical Aptitude Profile, another more widely used battery of tests. To promote its wide usage, the MBEA is now available upon request. In addition, individual MBEA data of 160 normal participants of variable age and education have been made available to all via the internet”


How can APD be managed?

- Management strategies
  - Signal enhancement strategies
  - Auditory Training
    - Formal
    - Informal
  - Linguistic, cognitive, metacognitive and educational strategies

Signal enhancement strategies

- Clear speech
- Minimise background noise
- Minimise reverberation levels
- Assistive listening devices:
  - personal FM systems
  - Classroom FM systems

Bamiou, Campbell, Sirimanna Audiological Medicine 2006;4:46-56.
Auditory Training: Formal

- AT as indicated from auditory test battery in clinic
- Earobics (Cognitive Concepts, Inc 1997)  
  http://www.earobics.com/
- FastForWord (Scientific Learning Corporation, 1997)  
  http://www.scilearn.com/
- Phonomena (Mindweavers, 2005)  
  http://www.mindweavers.co.uk/main.asp

Auditory training - informal

- Vowel training
- Auditory directives
- Sequences in piano, Simon game
- Localisation of sounds in park
- Parent reads a story – child asked to raise hand everytime the target word/sound occurs – same in noise.
- Listen to lyrics of song – English –1 singer, 2 singers, 3 singers, American –1 singer etc.
- Listen to a story with headphone on one ear, music in background, gradually increase volume of music
Metacognitive strategies

- 'executive ' function which encompasses planning for learning, thinking about the learning process as it is taking place, monitoring and evaluating learning (Brown, 1997).
- Metacognitive approaches for APD include cognitive problem solving, self-planning/monitoring, assertiveness training.
- Cognitive problem solving: a. identify the key words and think about their meaning, b. think about the context of the sentence, and what similar experience he/she has to understand this c. think about the main message of the sentence and predictions or inferences that are needed to understand this (Chermak 1998).
- Assertiveness training refers to teaching the client to become effective by communicating what he/she feels, thinks and wants.

Cognitive strategies

- May be task-specific, and often refer to direct manipulation of the learning material itself (Brown, 1987).
- Examples of cognitive strategies are note-taking, repetition, guessing meaning from context, or using mnemonic devices.
- Since these strategies are not task-specific, it is quite likely that once taught, they may be employed to resolve difficulties in varied communication situations (Chermak, 1998).

Final Comments

- There is a pressing need for reliable diagnostic tools and for uniform diagnostic criteria for APD in order to facilitate research, which would translate into evidence-based clinical practice (Bamiou and Luxon, 2008). This has acknowledged by several multiprofessional consensus conferences, both in the UK and abroad (e.g., BSA 2007; ASHA 2005). In the UK, the British Society of Audiology established a multidisciplinary APD Interest Group in October 2003 with an elected steering committee (of which the first author is current chair and the second author is past chair), which aims to define guidelines and identify research needs via a multidisciplinary forum (please see http://www.thebsa.org.uk/ on how to become involved if interested). The clinician who deals with the assessment of suspected APD cases is faced with challenges and scientific uncertainties. However, this relatively new field is rapidly expanding, as basic scientific findings are being translated into clinical practice, and clinical questions and ambiguities are identified and addressed by scientific studies.
- Bamiou & Sirimanna, ENTNews 2009

THANK YOU FOR YOUR ATTENTION!

Any questions (I can answer)?