Audiology for Adults with Learning Disabilities

Siobhán Brennan
Definition

British Institute for Learning Disabilities [2011]:

- Intellectual Impairment
- Social or adaptive dysfunction
- Early Onset
Prevalence

Learning Disabilities in England - 985,000 - 2% (Mencap)

- Increasing

Hearing Loss in Adults with Learning Disabilities - 30 - 40%

- Highly variable
- Can depend on pathology
- May be higher with increasing intellectual impairment
Challenges to Audiology

- Highly variable carer support
- Lack of validated outcome measures
- Lack of Research
- Wax
• 28% of excessive / impacted wax in ALD (with or without DS)
  – (2-6% in general population)
  – (Brister et. al. 1986)

A third of Appointments incomplete due to wax
(Sheffield Audit 2010)
How responsive are we?
How responsive should we be?

- Low levels of self-report
- High levels of diagnostic overshadowing by health professionals and carers
- Impact of undetected loss is greater
Why not use paediatric techniques?

- Different developmental trajectories
  - (Paterson 2001)

- Ethical considerations

- Impact of long term undiagnosed HL

- Consider but with additional factors taken into account
Syndrome
Specific
Factors

Alertness

Social
Interaction

Confidence

Co-morbidity

Aquiescence

Capacity
Capacity – Assessment

- Reduced Executive function
- Difficulty integrating information from senses
- Reduced understanding the purpose of a request
- Short term Auditory Memory difficulties
- Problems understanding and generalising between objects & situations
Teaching at home prior to assessment could help including:

- The vocabulary and objects you will/may use
- How to wait and listen for the sound before the action

“Once testing, if [they] don't understand the 'rules of the game' and its one they have been rehearsing at home do not change the game, it will only confuse them further. If they have been practising men on the boat don't switch to pegs on the board”

(Amanda Glennon: Member of HaLD)
Capacity – VRA Impact

- Head turn to image responses: 28%
- Head turns but not to images: 10%
- Clients with purely stilling or other subtle responses: 28%
- Clients with combination of responses: 17%
Capacity – OAE

- OAE Success highly dependant on patient group

  - Driscol (2008) – 80% feasible - children wLD
  - Starska (2006) – 75% pass rate - Olympians
Reduced social skills

Reduced ability to generalise between situations

Reduced ability to tolerate change

Reduced understanding
Capacity – Reviewing

Limited self-report
Continuity of Care

“All patients with learning disabilities and hearing impairment should be reviewed at regular (yearly) intervals”

GUIDELINES FOR MEETING AUDIOLOGICAL NEEDS OF ADULTS WITH LEARNING DISABILITIES [2009]
Confidence – Behavioural Test

Quickly find their level of difficulty and alternate between a challenging sound and a heard sound level for them: Children with Down Syndrome have poor concentration, they often will get bored waiting to hear a sound and pretend they have heard one or get disengaged testing Alice with a sound she might not hear quickly followed by a sound she can hear keeps her engaged, her game moving on and she doesn't lose confidence in her ability.

Children with Down Syndrome hate to get things wrong, if they do get it wrong ignore and remodel correct way to do it

(Amanda Glennon: Member of HaLD).
Confidence – Behavioural Test

- Begin without headphones
- Extended conditioning + encouragement
- Pace
- Repetition
Alertness
Alertness

- [PwLD] operate on a low arousal level as a result of dysfunction & neurological mechanism
  - Clausen (1965) (supported by Myklebust 1954, Wolfe & Macpherson 1959)

- The monotonous pure tone needs to increase in intensity for subjects to response because of low basal arousal levels and accompanying inability to direct attention to the stimuli for any length of time.
  - Clausen (1965)
Behavioural Sound type

- PwLD more sensitive to pulsed tones than continuous stimuli
  - (Fulton 1967)

- Warble tone more effective than pure tones in PwLD
  - (Reichstein & Rosenstein 1964)

- Performance improved with increasing stimulus level
  - (Werner et al 1996)
Alertness – Information Delivery

Children with LD VRA, thresholds matching previous tests:

- Air Puff: 91%
- Visual Reward: 65%

(Lancioni et al 1989)

Tactile Instruction

People with DS are often good visual learners (Mills 2014)
Aquiescence
Acquiescence

- [People with learning disabilities] have an increased likelihood of acquiescent responses
- Exacerbated if ill at ease
- Unfamiliar environment
- Unfamiliar interviewer

(Cummins 2003)
Acquiescence – PTA

- May be Suprathreshold
- Patients’ willingness to please
- Lateralisiation to confirm
Co-morbidity
Comorbidity

- Impact of additional health needs
  - on assessment
  - on optimum management
  - high rates in PwLD

- Diagnostic Overshadowing
Social Interaction
Social Interaction

- Tactile Defensiveness
- High Rates of Autism in PwLD

“Many people with an ASD are thought to be visual learners” (National Autistic Society)

Visual Supports / Objects of Reference / Makaton
Communication
Communication and people with the most complex needs: What works and why this is essential [Mencap 2010]:

- Intensive Interaction
- Music and other creative arts-based approaches
- Picture Exchange Communication System (PECS), Visual timetables
- Signing, including Makaton and Signalong

Sheffield Teaching Hospitals
NHS Trust
Communication

• “Language difficulties in people with Down’s Syndrome are disproportionate in relation to non-verbal activity”

Expressive language skills are poorer than receptive language skills

ABR & Down’s Syndrome

• Latencies Shorter
  • (Widen et.al. 1987) (Ferri et. al. 1995)

• Peaks Smaller
  • (Widen et.al. 1987) (Ferri et. al. 1995)

• Factors are sufficient to alter threshold estimation
  • (Widen et. al. 1987)
CERA & Down’s Syndrome

- Latencies Increased

- Peaks Larger
  (Yellin et al. 1980) (Straumanis et al. 1973)

- No habituation
  (Schafer & Peeke 1982) (Vieregge et al. 1992)

Waveforms often Atypical
(Sheffield Audit 2009)
Syndrome Specifics – Speech Discrimination Assessment

• Pw DS can have smaller receptive vocabulary than intellectual level suggests (Paterson 2001) (Price et al 2007)

• PwDS can have greater difficulty storing, understanding and retrieving phonological information
  – (Stoel & Gammon 1997)

• PwDS can have poorer verbal than visual short term memory which may affect ability to perceive acoustic information
  – (Chapman & Hesketh 2001)
Syndrome Specifics

AB Word List
– Impact of reduced vocabulary

Manchester Picture Test
- Abstract vocabulary an issue

HaLD Speech Test
Reduced strength and reduced height of auricles in Down Syndrome. (Miller 1997)

Shallow concha in Down Syndrome. (Miller 1997)

PwDS – More difficulties with HA pushing down on auricle and earmould pushed out

– Risks losing aid / not wearing / feedback
Reduced pinna size in Down Syndrome
Reduces localisation  (Porter & Tharpe 2010)
Concentration of sound  (Schwartz 1978)

High instance of canal stenosis  (Maurizi 1985)
Hourglass shape  (Pappas 1994) (Kanamori 2000)
RECD more vital but more challenging
Audiology for ALD

• “Use a multiple-test protocol which increases the reliability & validity relative to a single test”

• (Gans & Gans 1993)
www.hald.org.uk

• “Become a member” on the website - add a document

www.facebook.com/groups/HaLDgroup/

• “Join” group - leave comment

www.facebook.com/HaLDgroup

• “Like” page - leave comment

www.twitter.com/HaLDgroup

• “Follow” - tweet us

www.linkedin.com

Interests – Groups – Hearing and Learning Disabilities SIG

• “Join” group – leave comment
From Finding to Fitting  
10th October 2014  
Sheffield  

Sheffield Hearing and Learning Impairment Service