Practice Guidance:

Audiological Assessment for Adults with Intellectual Disabilities

Date: April 2021

Due for review: April 2026

OD104-90

General foreword

This document presents practice guidance by the British Society of Audiology (BSA). Practice guidance provides a reference standard for the conduct of an audiological intervention that represents, to the best knowledge of the BSA, the evidence-base and consensus on good practice given the stated methodology and scope of the document and at the time of publication. Although care has been taken in preparing this information, the BSA does not and cannot guarantee the interpretation and application of it. The BSA cannot be held responsible for any errors or omissions, and the BSA accepts no liability whatsoever for any loss or damage howsoever arising. This document supersedes any previous practice guidance by the BSA and stands until superseded or withdrawn by the BSA.

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Published by the British Society of Audiology

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Declaration of Interests: None Declared

Citation

Please cite this document in the following format:

BRITISH SOCIETY OF AUDIOLOGY (*2019*), Audiological Assessment for Adults with Intellectual Disabilities [Online]. Available at: https://www.thebsa.org.uk/resources/ [Accessed *date*]

Contents

1. Introduction

This document gives guidance on the audiological assessment of adults with intellectual disabilities. In this document the term “adults” is used to denote the period after 16 years of age while it is acknowledged that many individuals with intellectual disabilities do not transfer to adult services until later.

It is proposed that the reader considers this document alongside companion BSA guidance for working with individuals with intellectual disabilities such as Position Statement: Audiological Care for Adults with Intellectual Disabilities, Access to Audiology Services for Adults with Intellectual Disabilities and Audiological Rehabilitation for Adults with Intellectual Disabilities. It should also be read in conjunction with the latest NICE Guidance and other guidance and protocols relating to healthcare for adults with intellectual disabilities some of which are referenced in this document.

This document is not intended to provide guidance on specific circumstances or on interpretation of results. It is important that the competent person carrying out, or responsible for, the audiological care of the client (the ‘clinician’) uses professional judgement when deciding on the particular approach to be used with each person being provided for (the ‘client’), given the specific circumstances and the purposes of the care, and the carer’s level of competency.

The term ‘shall’ is used in this document to refer to essential practice, and ‘should’ to refer to desirable practice. Unless stated otherwise, this document represents the consensus of expert opinion and evidence as interpreted by the Professional Guidance Group of the BSA in consultation with its stakeholders. The document was developed in accordance with the BSA Procedures for Processing Documents (BSA).

2. Definitions

Intellectual Disabilities

The World Health Organisation defines Intellectual disability as *"a significantly reduced ability to understand new or complex information and to learn and apply new skills (impaired intelligence). This results in a reduced ability to cope independently (impaired social functioning), and begins before adulthood, with a lasting effect on development.*

Whilst the term "learning disability" is the preferred term in the UK, (superseding historic terms such as mental handicap or mental retardation), "intellectual disability", "developmental disability" and "learning difficulty" are also found in the literature. The term "intellectual disability" will be used throughout this document for consistency and to reflect global preference.

There are aspects of this document that may be applicable to adults with cognitive needs that have been acquired after childhood, including traumatic head injury or dementia, but the reader is advised to consult the relevant BSA guidance specific to these groups.

Carer

Throughout this document the term "carer" is used in a general context to refer to any individual providing support to a person with intellectual disabilities, either paid or unpaid. Typical, unpaid carers are family members or friends of people with intellectual disabilities. Paid carers (often known as support workers) are employed to provide the levels of support required, which may vary from occasional input, to full support including personal care or feeding. Many people with intellectual disabilities rely on carers for advocacy, detection and management of health issues, including hearing (McShea et al, 2015).

Audiometry

The majority of people with intellectual disabilities can have their hearing assessed successfully using audiometry. For many this may be pure tone audiometry, with no adjustments. For others, reasonable adjustments can result in a successful and comprehensive assessment. Audiology service providers have a duty of care to make any reasonable adjustments required. This may involve adjustments to appointment length or location, or alternate assessment techniques e.g. visual reinforcement audiometry. The misconceptions around hearing assessment for people with intellectual disabilities has been documented in the literature such as the belief that hearing assessment is unproductive in people with intellectual disabilities (e.g. Bent et al, 2015; McShea, 2015b).

3. Background

There are just over 1 million people in England with intellectual disabilities, which equates to approximately 2% of the population (Emerson et al 2008). This prevalence is increasing. Hearing impairment is common among adults with intellectual disabilities, with estimates of the prevalence of hearing loss in adults with intellectual disabilities at 40 - 45% (Neumann 2006, Evenhuis 2001) and the prevalence of hearing loss in people with profound and multiple intellectual disabilities is likely to be even higher (Evenhuis 2001). Despite the high prevalence of hearing loss in adults with intellectual disabilities the rate of hearing assessment is low (Strydom et al., 2005; Van Buggenhout et al., 1999). Reasons attributed to this include logistics (Evenhuis et al., 2004) and the misconception that hearing assessment is not effective for adults with intellectual disabilities (Andersson et al., 2013). Appendix 1 shows a summary of recommendations for the frequency of hearing assessment for adults with intellectual disabilitiesby The European Federation of Audiology Societies Working Group for Intellectual Disabilities.

4. Mental Capacity Act 2005

The Mental Capacity Act (MCA) came into force in 2007 in England and Wales and provides the legal framework for acting and making decisions on behalf of those individuals who may lack the capacity to make particular decisions for themselves. Everyone working with or caring for an adult (aged over 16) in England and Wales who may lack the capacity to make decisions for themselves must comply with the act. In Scotland the Adults with Incapacity Act came into force in 2000. In Northern Ireland the applicable legislation is the Mental Capacity Act 2016.

4.1 Capacity Underlying Principles

There are 5 statutory underlying principles:

1. The assumption of capacity – every adult must be assumed to be able to make decisions for themselves unless it can be evidenced that they lack the required capacity. Capacity is time and decision specific, therefore it must not be assumed that because an individual lacks the capacity for a particular decision that this will apply to all decisions. Capacity can fluctuate over time making the time a decision needs to be made crucial in that assessment of capacity.

2. A person must not be treated as unable to make a decision unless all practicable steps to help them have been taken without success. The assessor must ensure that all support appropriate communication tools and possible assistance must be offered to the individual. Adults with intellectual disabilities may be able to demonstrate their consent to some areas of assessment/intervention by cooperation, behaviour e.g. pushing away. It is important to gather as much information as possible from those who know the client well, to determine if their behaviour demonstrates that they are acquiescent or giving informed consent to the procedure. A compliant client does not necessarily mean that they are consenting to a procedure.

3. A person is not to be treated as unable to make a decision merely because he makes an unwise decision, the issue of an unwise decision can be subjective, what one person thinks is unwise another may not in the context of their life and experience. It is important to take an objective and concrete view of the facts.

4. An act done, or decision made on behalf of another under the act must be evidenced as being made in the person's best interests taking account of the individual factors and impacts specific to the person.

5. Before an act is done or decision made consideration must be given to how the desired outcome can be achieved in the least restrictive manner, and that impacts least on that person's rights and freedoms.

An individual must never be deemed incapable of making a decision based simply on:

- Age
- Appearance
- Assumptions about their condition/diagnosis
- Any aspects of their behaviour

The Act prescribes a test of capacity that must be evidenced by all people undertaking an assessment of capacity, anyone undertaking this assessment with an individual must provide documentary evidence of their findings and conclusions.

4.2 The statutory test of capacity

Stage1

Does the person have an impairment of, or a disturbance in the functioning of their mind or brain?

Stage 2

Does that impairment or disturbance mean that the person is unable to make the specific decision at the time they need to make it?

The act determines that a person is unable to make a decision if they cannot:

- Understand the information relative to the decision – stick to the salient points covering the nature of the decision, why the decision needs to be made and the likely effect of making a decision one way or another or not making a decision at all.
- Retain the information for long enough to make the decision – the person only needs to hold the information in their mind for long enough to make the decision.
- Use or weigh that information as part of the decision making process
- Communicate their decision by any means available to them.

Evidence of the findings of the test must be documented, a yes or no answer is not satisfactory. It is important to consult with others who may have information/evidence to assist.

4.3 Best Interests

No adult is able to consent on behalf of another adult. If an adult is assessed to not have capacity for a procedure, the best interest process must be followed. Where an assessment has concluded that the person does lack the capacity to make the decision, the best interests process as prescribed by the act lays down the steps that must be taken to determine best interests.

When trying to work out what is in a person's best interests it is important to:

- Encourage the person to participate or improve their ability to participate in making the decision wherever possible or appropriate.
- Identify all the circumstances that are relevant by identifying the things the person would take into account themselves and the things that are important to them.
- Find out the views of the person including their past and present wishes and feelings, these could have been expressed verbally or written down.
- Consider any religious or cultural beliefs that would be likely to influence the decision.
- Include any other factors the person themselves would be likely to consider.

It is important to consider whether or not the person may regain capacity and if so can the decision wait until that time without the clients suffering any serious or irreversible effects.
If the decision involves life-sustaining treatment, assumptions must not be made about the person's quality of life.

It is important to consult with others wherever practical to do so, people such as relatives, carers and friends who may be significant in their lives. Also consider any healthcare and social care staff. They may have valuable information and views that are important to the decision making process.
Where there is a Lasting Power of Attorney for healthcare and welfare of a Court of Protection Deputy they will be the decision maker in respect of the person, however the best interest's process still applies.
https://www.gov.uk/government/publications/mental-capacity-act-code-of-practice
http://www.legislation.gov.uk/ukpga/2005/9/contents

Consent and any assumptions about consent must be clearly documented in the clinical notes. For example:

- Consent was assumed as client allowed me to look into their ears.

- Examination modelling was used e.g. Client initially pushed me away. When I allowed him to feel the auriscope and put it against the carer's ear, he allowed me to examine his ear.
- Client repeatedly said 'no' but allowed me to examine his ear.

5. Profound and Multiple Intellectual disabilities

There is a group of people who have a range of complex needs commonly associated with pronounced developmental delay, significant physical and sensory impairments and epilepsy. Most people will also have difficulties in eating and drinking, and problems with their breathing. People with Profound Intellectual and Multiple Disabilities (PIMD)[1] have a range of complex disabilities which may include:

- profound intellectual disabilities
- physical disabilities that limit them in undertaking everyday tasks and often restrict mobility
- sensory impairment
- complex health needs, i.e. epilepsy or respiratory problems, eating & drinking problems
- restricted communication, i.e. pre-verbal, though a small number have some spoken or key-word signed language or use other forms of Alternative and Augmentative Communication
- 'coping behaviours' (for example, communication or other difficulties, avoidance or self-injurious actions) which may present as challenging

[1] The terms *profound intellectual and multiple disabilities (PIMD)* and *profound and multiple learning difficulties (PMLD)* are used interchangeably for the same group of people, however, we will use the former as international research is moving towards this terminology.

- Mental ill health

http://www.gov.scot/resource/0042/00424389.pdf

In the United Kingdom people with IQ below 50 refer to as having PIMD. The causes of PIMD are many and varied. They include genetic disorders, acquired brain injury or brain damage as a result of infection or anoxia. Causation may be ante-, peri- or post- natal. For many there is no known causation. It is estimated that the prevalence of PIMD in the general population is 0.05 per 1,000. This figure is derived from a survey undertaken in Scotland and would lead to a figure of 2,600 people with PIMD in the country. In England, Emerson (2009) estimated the number of adults with PIMD to 16,000 and anticipated this to increase by 1.8% each year, on average. This is possibly an underestimate and a useful working figure would be 3,000. These numbers will increase with better survival rates, not only in the neonatal period but into childhood and adulthood, due to advances in medical care. The Department for Education figures demonstrate the significance of this rise, in England, based on their annual data collection. For example, 2009 statistics identified those with PIMD to be 9,400 aged between 5-16yrs. The 2017 report from the Department for Education, notes 10,981 pupils aged 5-16yrs are identified with PIMD as their primary need.

All people who have PIMD will have great difficulty communicating. Many people will have additional sensory or physical disabilities, complex health needs or mental health difficulties. The combination of these needs and/or the lack of the right support may also affect behaviour.

Communication and interaction barriers prevent people with PIMD from having full opportunity to inclusion and engagement in health care. This group of people, however, can benefit greatly from good health care and are able in various ways to communicate their views. It is essential to remember that they share the same general intentions as other adults, that is, they need to be involved to the best of their ability.

People with PIMD can and do lead meaningful lives but they require a high level of support with respect to all activities of daily living. Not only do people with PIMD require fully trained staff with

specialised knowledge of their healthcare and communication needs but communities need to be made fully inclusive through the provision of both intellectual and physical access. Despite improvements in service delivery in the last decade, people with PIMD still confront barriers to good quality health care, education, leisure activities and support services.

It is crucial to highlight the potential of undiagnosed sensory, especially hearing, loss for this group of people, especially those who are unable to gain functional use of their senses because of neurological/processing issues. Estimates continue to suggest 50-70% of the general population of learning disabled experience sensory loss, and many will fall into the PIMD sector. Fluctuating hearing loss is also a real probability for this group of people therefore, access to audiology services is vital for people with PIMD. There are two areas to consider when people with PIMD are referred to Audiology services:

5.1 Communication and Consensus

Effective communication and support will be in place throughout the referral, assessment, and review periods to ensure the person is ready and any changes in their need is met. It is important the information is provided to family and carers in advance. The service needs to ensure effective communication is achieved with all relevant professionals, and with the family and carers of the individual to share knowledge and information about the person's medical background and their wellbeing.

The individual should have a Healthcare Passport which is a document about the individual and their specific health needs. In addition to describing health needs, this document also provides information for healthcare providers including likes, dislikes and preferred method of communication. This is shared in advance with the service. The relative or carer of the individual is involved to ensure adjustments are made to suit the needs of the person and quality of the service provision. The assessor and other audiology staff are aware of what is

‘usual’ for the individual in terms of their health and wellbeing, and are able to identify and respond swiftly to indicators of physical, emotional or mental challenges to their health during all stages of contact. Staff should seek advice on and are aware of the person’s communication and consensus needs, including indicators of discomfort or distress and are responsive to these communication methods.

It is recommended the use of techniques such as interactive, profiling and consensus approaches are used (Thurman et al 2005). Interactive approaches involve intensive interaction techniques with the person, for example, spending time with the person helps to relate and communicate more effectively and to get to know the person better enables you to interpret their needs and wishes better. These approaches help the person and their communication partner to relate to each other and to establish a communication link by building on the person’s communication abilities. Due to time restrictions, such methods might not be possible for the audiologist to do during the visit, so, instead, they can obtain information and support from the key-worker or carer. Profiling approaches include communication profiles, passports, and multimedia profiling such as videos, audio files, online media etc. Profiling methodology provides key information about the person’s day to day ‘need to know’ habits, routines and patterns. Finally, consensus approaches involve careful observation and analysis of information about the person’s communication abilities and practice to create a synthesis of information. For all the above methods, the role of key people in an individual’s life is essential. The person is deemed able to exercise choice and control over decisions about their health and wellbeing and MCA and best interest decision processes are in place, where appropriate, to evidence assessment of capacity.

Access is also related to information distributed, e.g. easy read and explained in a manner that the person will understand. People with intellectual disabilities are often put-off visiting Audiology (and other health) services simply because they don’t understand the procedure or they don’t have enough information about the assessment (what is required, what is it for, how they will benefit etc.). Information and support for families and carers typically does not include

specific information on profound and multiple disabilities. So, it is important that carers get information on intellectual disabilities and physical disabilities and sensory impairment.

5.2 Physical Environment

The second area of consideration is the physical environment. This needs to be appropriate and personalised to respond to individual needs and preferences e.g. sufficiently spacious to ensure people can move freely, adapted to meet sensory needs, use of assistive technology.

- The environment is equipped appropriately with hoisting, changing and other mobility equipment people require. It also meets the person's individual needs such as sensory needs and preferences, being functional and personalised at the same time.
- People visiting the service have easy access to all facilities and there is evidence that these facilities are well maintained at all times.
- People have access to appropriate transport, which is available when the person needs it.

Working with carers is vital for all health appointment, especially though for Audiology. By carers we mean both paid support staff and relatives/friends of the person. These people are vital to convey and gather information about the person, particularly when the person cannot communicate such information. An open approach is necessary to ensure the right amount of personalised support is provided to the individual and to ensure a successful assessment and overall process is achieved. A holistic management plan should follow the assessments and it should contribute in sufficient detail to establishment of aetiology, prognosis and further management.

6. Hearing Assessment

"Audiological certainty can be defined as when ear specific and frequency specific information across the speech frequencies has been reliably established." (McCracken et al 2008). This is ideally achieved using a battery of assessments. The tester should make the client and their carer aware that audiological assessments may take multiple sessions. Prior to beginning hearing assessment, the priorities should be established with the client and their care team. These could include;

- What are the quietest sounds this person can hear?
- Can this person discriminate sound sufficiently to understand speech and enjoy sound?
- Does this person have sufficient hearing to lead the life they want to lead?
- What auditory inputs are most important to the client?

There is extensive existing guidance available from the British Society of Audiology on the assessment of hearing which should be consulted in the first instance. Then consideration should be given to any additional adjustments that may be useful to complete the hearing assessment as accurately as possible with adults with intellectual disabilities. This section provides guidance on adjustments that could be utilised.

6.1 Who Should Lead this Process?

There are a variety of models of service, many of which are led by audiologists. Audiologists assessing adults with intellectual disabilities must have the competencies required to assess children and adults, and be able to perform complex hearing assessments. It is good practice to also have an ENT champion as part of the team to enable coordination of the pathway.

6.2 Wax impaction

The prevalence of wax impaction in adults with intellectual disabilities is high. The cause for this is unclear although it is known that in some syndromes it is associated with the anatomy of the ear. Fransman (2006) identified a link with the lack of back teeth and wax impaction. In addition to the implications on health, the presence of wax can alter the accuracy of assessment of a client's underlying hearing.

Many adults with intellectual disabilities find it difficult to tolerate ear syringing, therefore requests to visit the GP for wax clearance before attending an appointment is often unsuccessful. It is good practice for staff to be available and time to be allocated during an assessment to complete wax removal at the same visit, as this is usually more effective and reduces the number of visits clients have to make to the hospital. Dry removal of ear wax with for example micro forceps by suitably trained staff should also be considered.

6.3 Sensory Profile

There is a higher prevalence of sensory modulation disorder in adults with intellectual disabilities, particularly those with a comorbidity of Autism, than in the general population (Joosten & Bundy 2010). This can have significant impact on both assessment and rehabilitation.

Some clients may be distressed, confused or indeed comforted by certain sounds, sights, smells, movements or textures. Appendix 2 presents a sample checklist for tactile issues that may justify a referral to Occupational Therapy.

6.4 History Taking

Communication factors need to be considered to maximise the information gathered when taking a history for adults with intellectual disabilities. It is noted that self-report may not be

accurate (Emerson et al 2013) and that carers tend to overestimate hearing ability (Kerr et al 2003). Possible indicators for the tester that this may be the case include:

- Evidence that carers are unaware that they are providing necessary visual cues.
- Vague descriptions of possible responses to sound.
- Reports from the carer that the client "can hear but sometimes ignores people".

Expressive communication factors can also influence the accuracy of history taking unless they are taken into account. The expressive communication skills are reported to be less developed than receptive language in individuals with Down Syndrome (Abbeduto 2003). Additionally, echolalia (the repetition of another person's spoken words without meaning) and acquiescence may be misinterpreted by the clinician. Tactics such as cross-questioning, informant checks and asking for examples can all assist with these issues. Open questions for adults with intellectual disabilities can result in fewer details, but increased accuracy (Finlay & Lyons 2002). Reported wishes in communication with healthcare professionals include:

- Enough time for questions and repetitions
- Demonstration of physical examinations before carrying them out
- Addressing the person with ID as the principal communication partner in triadic communication (Wullink et al 2009)

Further resources on optimising communication with adults with intellectual disabilities from organisations such as Mencap are recommended.

6.5 Ear Examination

Ear examination should be carried out according to the British Society of Audiology recommended procedure however it is acknowledged that there may be modifications required for the client's comfort. There is a higher instance of sensory processing disorder in individuals with intellectual disability who also have comorbidity of autism. This sensory processing disorder may include hypersensitivity to a range of inputs including tactile stimuli. This may cause the client to be resistant to various Audiological tests including otoscopic examinations

(Baranek & Berkson 1994). The risks associated with incomplete otoscopy should not be underestimated and a desensitisation programme considered. Facilitating the client seeing and, if possible, feeling the otoscope and speculum can help, as can showing the light on the back of the client's hand before placing in the ear canal. Additionally, modifications could include:

- Carrying out otoscopy towards the end of an appointment as opposed to the start.
- Breaking the examination down into parts instead of the full examination of the ears in one go.

6.6 Behavioural Assessment

The basic principles of behavioural assessment are the same in adults with intellectual disabilities as for the general population; i.e. can it be noted that the client's behaviour changes when sound is presented? The client may or may not be consciously aware of this change in behaviour or not. Assessments involving a conscious change in behaviour involve asking the client to wait for the sound and then to react. If the client does not have the capacity to wait (Evenhuis et al 2001), or to behave in a consistent way when the sound is presented involuntary reactions to sound are observed instead. This latter type in adults with intellectual disabilities tends to be less accurate because:

- The accuracy depends on the tester subjectively interpreting an involuntary response.
- Involuntary responses to sound tend to vary considerably in their presentation.
- The quietest sound to which there is a response may not be true threshold and is generally referred to as a "minimum response level".

An attempt to carry out "Stimulus Response Audiometry" where an instruction is given to the client prior to sound presentation should be attempted before moving onto Behavioural Response Audiometry (BOA) methods.

While modifications to standard protocol are frequently required when carrying out behavioural assessment with this client group it is imperative that the tester considers whether the modifications made are such that conclusions should not be drawn from the results. The following section presents elements of the testing that could be adjusted during behavioural assessment and what impact this may have on the validity of the result;

- Starting with presentation of the sound through a speaker as opposed to headphones allows the client to associate the sound with a source which can improve relating the sound to an action.

- Neither the headphone nor bone conductor have to be attached to the headband for testing if the client is more comfortable without them. This can lead to a source of error if the bone conductor is not held sufficiently firmly to the skull. This should not be done by a carer if possible. Insert phones should also be considered if headphones are not tolerated.

- Due to the higher risk of false positives additional no-sound trials should be considered to minimise tester bias and error.

- Responses that require a larger client movement than button pressing can increase accuracy. Using items that the client is familiar with such as hair clips can increase the likelihood of engagement. Age appropriate response activities could also include using 'thumbs up' or clothes pegs into a basket. Trying to find a special interest of the client and incorporating that in testing can also improve engagement in the test.

- Stimulus presentations may need to be longer than typical BSA Audiometry to allow for auditory processing delay (Glasgow 1997).

- A client's possible lack of confidence can impact the accuracy of a behavioural assessment as the client may not feel sure enough of the presence of sound to respond and suprathreshold responses may result in overestimation of a hearing loss. Frequent encouragement is recommended during testing. The duration of the test may also be affected so breaking testing into 2 or 3 episodes may be required. These episodes should not be more than 2 weeks apart to reduce the possibility that hearing thresholds have changed between tests.

- Lateralisation, or asking a client which ear a sound is coming from, can direct the client's attention away from "can you hear the sound" to "where is the sound". This method as a verification of minimum response levels can also improve accuracy in clients for whom lack of confidence is an issue. Appendix 3 outlines considerations for this methodology.

- Sounds that are more interesting (e.g. LING sounds) or familiar may be more likely to elicit a response. Care should be taken to consider what frequencies are being tested and calibration factors should not be disregarded when using non-standard stimuli.

- Due to frequently reduced attention span in adults with intellectual disabilities using larger step sizes could speed the test e.g. presenting the sounds down in 20dB steps and up in 10 dB steps. This is likely to introduce an error of at least 5dB around threshold above the usual test-retest reliability; however this could be improved on when the client has become familiar with the test method.

- The use of longer headphone and bone conductor leads to allow clients to sit on floor/bean bags if this is where they are more comfortable. If sound field testing is being used the sound field should be calibrated for different sitting positions.

- Behavioural assessment relies heavily on the subjective opinion of the tester as to whether a movement in the presence of sound, or shortly afterwards is in response to sound. As with other areas of audiological assessment that rely on tester interpretation, such as ABR, the use of systematic peer review is advised (BSA Peer Review statement). In the case of BOA this can be facilitated by either dual testers or use of video recording facilities. The client's carer (if they know the client well) should also be asked for their opinion. Care should be taken however as previously highlighted that carers can be overly optimistic regarding the presence of a response to sound.

- Adults with intellectual disabilities with severely limited sight or who are registered blind will need alternative rewards for turning to stimuli. If they have limited sight they may be able to use a flashing orange light as a reward, although the lighting in the test environment will need to be dimmed for this to be effective. Avoid using a red light as this may be misunderstood as a signal of danger. If adults with intellectual disabilities are registered blind, a tactile reward is appropriate. This could involve reaching out to touch an interesting texture or vibrating pad or air-puff when a sound is heard. The reward will therefore need to be removed and only present for the person to touch when the stimulus is presented.

In addition to responses to quiet sounds, the presence or absence of hypersensitivity to loud sounds is of use. The reader is directed to the relevant BSA guidance on Uncomfortable Loudness Levels however modifications should be considered;

- Discussion with the patient and their care team how discomfort is typically conveyed. This should have been established at the start of the appointment.
- A scale of discomfort could be used by individuals who may not be able to verbally articulate their perception of sound, such as the Wong-Baker FACES™ Pain Rating Scale.

- The sounds should be presented more slowly than typically used to give the individual ample opportunity to respond.

6.7 Observation Questionnaires

In addition to a formal assessment of hearing in an Audiology clinic there is merit in observing a client's behaviour to sound in their familiar surroundings such as home or day services. There is a lack of validated auditory behaviour questionnaires for adults with intellectual disabilities, however there is an example in Appendix 4 which could be used with the necessary caution. It is recommended that the examiner uses a sound level meter in the location where the observation is taking place to accurately document the level of sound to which a response or lack of a response has been observed. It would be even more beneficial if this sound level meter provided information on the frequency spectrum of the sounds, however it is appreciated that equipment of that type is not widely available.

6.8 Objective Assessment

Objective measures should be carried out and interpreted according to the relevant BSA Guidance, please see sections below. High levels of client movement and background noise can affect the accuracy of this form of assessment. In the case of individuals who have involuntary movement or make involuntary sounds advice should be taken from the client, their carers and/or their family about techniques that should be employed to reduce these during testing. The tester should be familiar with Mental Capacity Act Code of Practice guidance on restraint (Department of Constitutional Affairs 2007). Additionally, the tester should consult BSA guidance for strategies that can be employed to reduce the impact of these factors such as pausing the test as required, adjusting test parameters and facilitating testing at optimum times of day.

6.8.1 Otoacoustic Emissions

While the likelihood of successfully completing a test of Otoacoustic Emissions (OAEs) is less in the population with IDs than in the wider population (Andersson 2000) due to the higher risk of inaccurate behavioural assessment it should be offered to every client for whom it is clinically appropriate. The tester should be aware of the risk of false positive results from this test, and if no OAEs are recorded other forms of assessment should be attempted. Heightened anxiety may lead to the individual finding it more difficult to sit still, so techniques to reduce this would be worth incorporating. These could include the use of visual distractions during testing (Gravel, Dunn, Lee and Ellis, 2006) and providing individuals, their families or carers with information about the technique prior to the appointment so that they can prepare as they see fit.

6.8.2 Tympanometry

Caution should be applied to the interpretation of tympanometry results in individuals with Down Syndrome and it has been suggested that a group specific normative range for compliance is needed. One study has suggested a normative range for compliance of 0.2-0.9ml (Kirkland 2017). The impact of cranio-facial abnormalities associated with other syndromes may also influence a tympanometry result; however there is currently limited literature on this.

6.8.3 Auditory Electrophysiology

Electrophysiological assessment should be considered in the event of unreliable responses to behavioural assessment. Behavioural assessment should be attempted in the first instance irrespective of the extent of a client's intellectual impairment.

If electrophysiological assessment is required it is necessary to refer to a specialist familiar with auditory electrophysiology. This specialist should be familiar with relevant guidance regarding working with adults with intellectual disabilities. The outcomes of auditory electrophysiological

tests should only be considered in the context of other tests and reports from the client and their carers. The most commonly used auditory electrophysiological methods in the UK are auditory brainstem response (ABR), cortical auditory evoked potentials (CAEP) and auditory steady states responses (ASSR). The ABR test could be considered for assessment under general anaesthetic (GA). The 40Hz ASSR may be affected by syndromes associated with intellectual impairment so may not be successful whereas 90Hz "sleeping adult" ASSR may be considered, as an alternative to ABR, under GA. If testing under GA is required attempts should be made to coincide this with other tests required under GA to limit the number of periods under GA if possible.

There are syndrome specific factors which have been reported that should be considered. These include:

- Differences have been observed in the ABR and CAEP morphologies in individuals with Down Syndrome (Kittler et al 2009) (Arisi et al 2012). These differences can include response amplitude, latency and morphology. Additional care should be taken in interpretation of waveforms due to these factors as they can be sufficient to affect threshold estimation (Widen et al 1987).
- A person's level of alertness can influence the accuracy of the CAEP (BSA 2016). This can be monitored by someone who knows the client well and the test should be paused when there are doubts about whether the client is alert. Additionally, test duration should be limited for this reason.
- Free-field CAEP should be available for hearing assessment for clients who do not wish to wear headphones and the limitations of audiological information from free field assessment be considered.

Appendix 1: European Federation of Audiology Societies Working Group of Intellectual Disabilities Screening Recommendations

AUDIOLOGICAL CARE	PERSON WITH ID IN GENERAL	PERSON WITH DOWN SYNDROME	PERSON WITH ID ELIGIBLE FOR HEARING DEVICE
EARWAX REMOVAL	Annual	2x/ year	2x/ year
HEARING SCREENING	Neonatal Hearing Screening	Neonatal Hearing screening	2-4x/year < age 6 2x/year ages 6 < 10 Annual hearing evaluation > age 10
	Annual screening < age 6	2x/ year < age 6	
	Every 3 years from age 6 to 18	Every 2 years from age 6 to 18	
	Every 5 years from age 18 to 50	Every 3 years from age 18 and 35	
	Every 3 years > age 50	Annual > age 35	
	Annually if 8h/ day noise exposure (>75dBA)		

Appendix 2: Tactile Defensiveness Checklist

The following are indicators that a referral to Occupational Therapy may be beneficial to address possible tactile issues that may affect audiological assessment. Consultation with local Occupational Therapy teams is advised to identify local referral criteria. In isolation each of these may not suggest a significant difficulty, but may warrant further discussion.

- The client shows dislike of physical contact sufficient to limit behaviour or life choices
- The client avoids activities that makes hands dirty
- The client avoids certain clothes because of the fabrics they are made from
- The client avoids wearing hats or scarves irrespective of how cold it is outside
- The client declines hair cuts
- The client selects food based on texture as opposed to taste
- The client tends to sit away from people including those they are familiar with

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Appendix 3: Verification using Lateralisation

Audiometry should be carried out according to British Society of Audiology Recommended Procedure “Pure-tone air-conduction and bone-conduction threshold audiometry with and without masking” as far as possible and taking factors outlined in section 7.6 into account. This methodology is recommended only for verification after attempts have been made to identify threshold as discussed in Section 7 of this guidance. This methodology is only appropriate for air-conducted stimuli.

A3.1 Preparation

Any items that have been used to identify a response to sound, such as a button or pegs, should be removed. The headphones or insert earphones should be removed to provide instructions, then repositioned.

A3.2 Instructions

Instructions shall give clear information about the task. This could be as follows:
“Tell me where the sound is coming from. Every time you hear the sound point to the side where it is. Point to the side if the sound is quiet or loud. If there’s nothing there you do nothing”. Alternative wording is acceptable providing the same points of instruction are included. These instructions should be accompanied by appropriate communication methods as required. They should also be told that they can stop at any time.

A3.3 Client’s response

The subject’s response to the test tone should clearly show which ear they can hear the sound by pointing to one of their ears or saying left or right. The response method should be recorded.

A3.4 Test order

Start with the better-hearing ear (according to the audiometry up to this point) at 1000 Hz 5dB above the previously given threshold. Randomise the ear, frequency and level of presentation. Include presentations 10dB below previously identified threshold. If the correct ear is identified at the same level on two out of two, three or four (i.e. 50 % or more) responses, presentations should also be presented at 10dB below this level. The lowest level at which this may be taken to be within 10dB of the hearing threshold level. It is often better to test fewer frequencies accurately than to attempt all frequencies typically seen on an audiogram.

Appendix 4: Observation Questionnaire

Home Observation Form

Individual's Name:	**Date of Assessment:**
Carried out by:	**With:**
Location of Assessment:	

Indicate whether the individual was aided or unaided during the observation session	Unaided Aided Right Aided Left Aided Bilateral Type of aid: Volume or other setting:
Describe the environment: whilst the observation was taking place. How quiet or noisy was it? How many other people were present in the room? What was the individual's general mood like during the observation?	_____ _____ _____ _____ _____
Condition	**Comment**
Awareness to sound (close): During the observation session, did the individual indicate the presence of a sound presented from a distance of less than 1 meter, *without visual cues and without being prompted*? Describe the sound and the individual's reaction (e.g. stilling, eye widening, head turn). Note whether or not the behaviour could be repeated.	_____ _____ _____ _____ _____ _____ _____
Awareness to sound (distance): During the observation session, did the individual indicate the presence of a sound presented from a distance, *without visual cues and without being prompted*? Describe the sound and the individual's reaction	_____ _____ _____ _____ _____ _____
Attention to sound: During the observation session, did the individual pay attention to a sound *without visual cues*, for a few seconds or longer?	_____ _____ _____ _____ _____ _____ _____ _____

Searching for a sound: Did the individual attempt to look for a sound presented *without visual cues*?	
Localisation of a sound: Did the individual correctly identify the source of a sound?	
Responding to name: Did the individual respond to their name being called, *without visual cues*? Did the individual respond the first time they were called?	
Responding to familiar sounds: Did the individual respond to a familiar sound presented without visual cues? E.g. a familiar song or music	
Meaning from environmental sounds: Did the individual recognise and know the meaning of an environmental sound? E.g. pedestrian crossing, fire alarm, animal noise.	
Meaning from speech: Did the individual recognise and know the meaning of a word presented without any visual cues? E.g. lunch, stop	
Additional comments	

Individual Name: **Date:** **Completed By:**	**Aided** Right Left Binaural Indicate how many hours per day the amplification is worn:
When the hearing aids/implant are first switched on, do you notice any change in the individual's vocalisations? Do vocalisations change in frequency/quality? Do vocalisations change if the battery isn't working? Does the individual test the device by using their voice when the device is first switched on?	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____ _____
With the hearing aids switched on, does the individual indicate the presence of a sound presented from a distance of less than 1 meter, *without visual cues* and without being prompted?	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____
With the hearing aids switched on, does the individual indicate the presence of a sound presented from a distance, *without visual cues* and without being prompted?	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____
With the hearing aids switched on, does the individual turn their head towards a sound, or try to search for a sound?	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____ _____
With the hearing aids switched on, does the individual respond to their name being called *without visual cues and without being prompted?*	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____
With the hearing aids switched on, does	___0 Never

the individual respond to a familiar sound presented without visual cues and without being prompted? E.g. a familiar song or music	___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____
With the hearing aids switched on, does the individual recognise and know the meaning of an environmental sound without being prompted? E.g. pedestrian crossing, fire alarm, animal sound, microwave, telephone ringing	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____
With the hearing aids switched on, does the individual recognise and know the meaning of a word presented without any visual cues? E.g. lunch, stop	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____
Does the individual recognise if the device isn't working? e.g. the battery has run down.	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____
With the hearing aids switched on, does the individual show any dislike of sounds? e.g. loud sounds made by other individuals, fire alarm etc.	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____
Does the individual ever ask for their hearing aids?	___0 Never ___1 Rarely ___2 Sometimes ___3 Frequently ___4 Always Notes:_____ _____

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