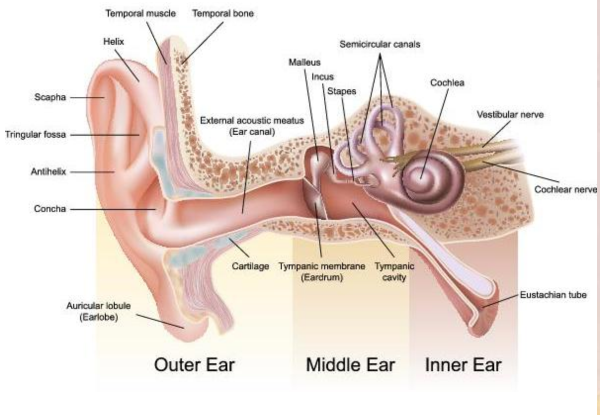
DESCRIPTION OF AUDITORY DISABILITIES

OVERVIEW

For humans the primary, though not the only, use of the sense of hearing is in social communications.For this reason we focus primarily on the impact of hearing difficulties on language acquisition and development and social communications. Although a delay in language acquisition does not necessarily lead to grave consequences, age-appropriate verbal communications with adults and peers also promotes the development of general cognitive and social skills, so any language delays can impair other skills too, even those not directly linked to hearing. In addition, problems with language acquisition can be indicative of more serious developmental issues such specific language impairment, dyslexia or autism spectrum disorder. Therefore, early diagnosis of hearing impairment is important and has given rise in many countries to neonate hearing screening programmes.

In considering auditory disabilities we distinguish between *afferent problems* that relate primarily to mechanistic audibility; i.e. whether and to what extent the acoustic signal is received and processed within the cochlea and correctly transmitted to the rest of the auditory system via the auditory nerve, and *integrative problems* that apply to the perception of complex sounds in general. In the next section problems relating specifically to language impairment will be discussed.

The auditory system is rather different to the visual system in that a great deal more processing occurs before the signal reaches the cortex. In addition, there are no good animal models of auditory cortical processing due to the huge expansion of communication sounds in humans. Consequently rather less is understood about the cortical processing and representation of sounds than visual images. Figure 1 below shows the key processing stages of the subcortical auditory system.



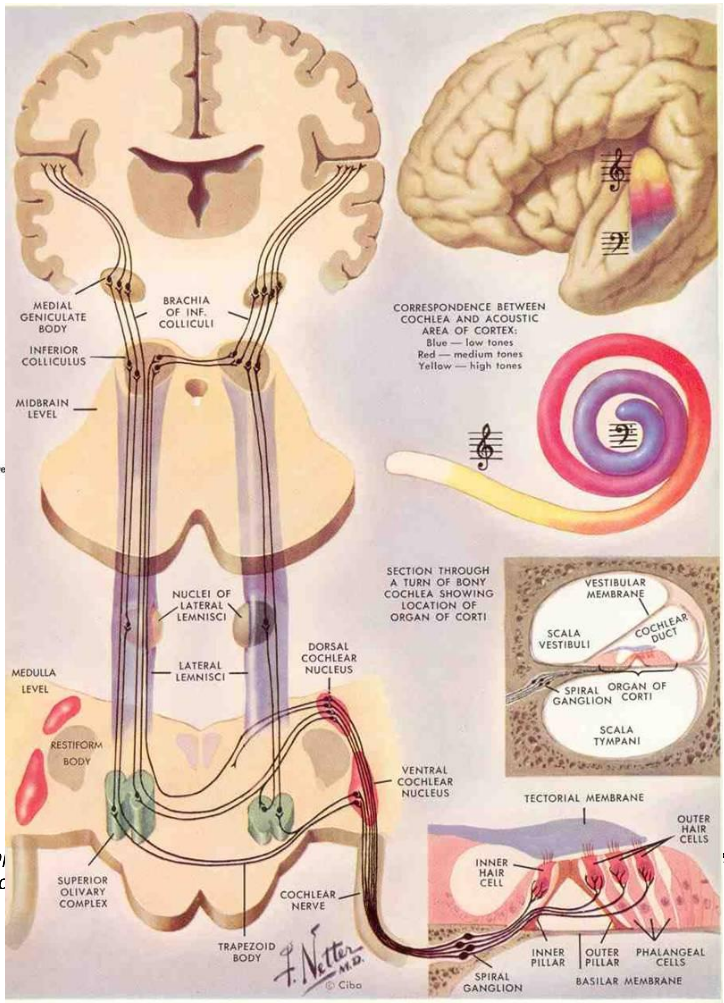


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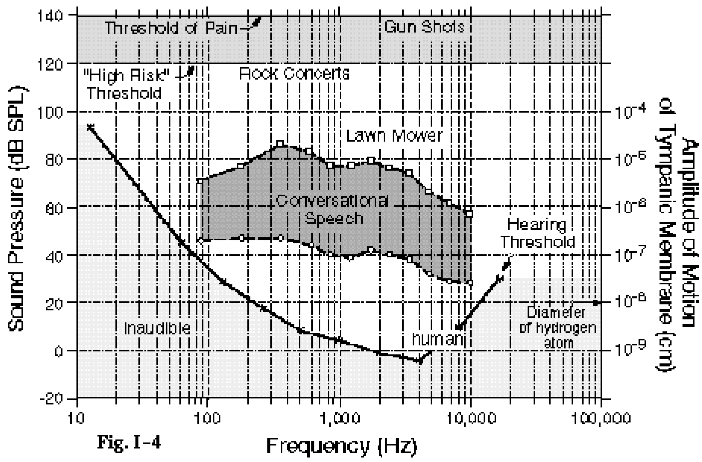
Figure 1. Key processing stages in the auditory system. Left: the anatomy of the hearing organ, the ear. Right: Processing stages in the subcortical auditory system, from the organ of Corti (lower right), located within the cochlea, to the primary auditory cortex (top right).

GENERAL HEARING LOSS AND CONSEQUENCES

Due to the huge dynamic range of sound intensities which we can hear, perceptual sensitivity is usually measured in terms of decibels (dB), the unit of relative sound pressure level (where the reference pressure in air is set to the minimum typical threshold of perception, 20 mPascals). Figure 2 shows the frequency



sensitivity of normal hearing and the sound levels of typical everyday sounds.

Figure 2.Frequency dependence of hearing sensitivity in normal listeners and the typical sound levels of some exemplar sounds.

Hearing loss is usually expressed in terms of the number of decibels (dB) needed to amplify a sound above normal hearing levels at a number of pure tone probe frequencies (typically 125, 250, 500, 1000, 2000, 4000 and 8000 Hz) before it is audible; the larger the amplification the worse the loss. Levels of impairment are classified as mild (26-40 dB), moderate (41-70 dB), severe (71-90 dB) and profound (> 90 dB)(Alzahrani, Tabet et al. 2015). People with severe or profound hearing loss are commonly referred to as deaf and those with mild or moderate hearing loss as hard of hearing.

Hearing loss has a number of functional consequences for children, depending on the severity of loss and the frequencies affected; e,g, as consonants tend to be high frequency noisy sounds thaey are affected more by high frequency loss than vowels. For mild loss, nearly all speech can be heard in quiet, but some sounds, especially the relatively quieter consonants may be misheard in noisy environments unless the child is looking at the speaker. For moderate loss, there is some difficulty in hearing others even when they are close by. For this reason, children need visual cues (lip-reading) to guide hearing, and when speaking may miss some word endings and articles (e.g. a, the). Hearing aids and visual cues are essential for those with severe loss, and their speech is markedly affected too. For the purposes of this project, we assume that any hearing loss in the children using the proposed adaptive system will not exceed mild to moderate loss.

Communication difficulties caused by childhood hearing impairment can have long term impact on social skills and academic achievement. Observable effects on social behaviour includedifficulties in comprehending communicative intent, frequent requests for repetition of messages, abnormally intense focus on the face of lips of a speaker, failure to focus attention on a speaker, a tendency to speak more loudly than appropriate,

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and ultimately social withdrawal. In studies of preschool hearing impaired children it has been found that they had fewer and shorter social interactions than normal hearing children (Brown, Rickards et al. 2001, Brown, Bortoli et al. 2008) and they often had fewer friends and felt rejected or neglected more often than their hearing peers, leading them to feel isolated and lonely ((Wauters and Knoors 2008). In addition, children with hearing loss often have associated vestibular and motor problems (e.g. balance deficits and clumsiness) (Rajendran and Roy 2011, Rajendran, Roy et al. 2012).

MEASURING HEARING LOSS

Hearing loss can be measured in a number of ways, and include both subjective and objective measures (Smith, Bale et al. 2005). Subjective tests included pure tone audiometry described above and behavioural tests, although need to be adjusted according to the maturational state of the child. Physiological (objective) measures include auditory evoked responses, otoacoustic emissions, and auditory steady-state response, and tympanometry; all of which assume normal middle ear function. Tympanometry is used to assess functioning of the middle ear.

Otoacoustic emissions are sounds originating within the cochlea that are thought to activity of outer hair cells. Transient evoked otoacoustic emissions (elicited by short sound bursts) are generally absent whenhearing loss is greater than 40–50 dB, except in the case of auditory neuropathy, which is characterised by the presence of otoacoustic emissions and the absence of a normal auditory brainstem response. The auditory brainstem response (see Figure 3) is a stimulus evoked electrophysiological measure of activity in the auditory nerve (VIIIth cranial nerve), and provide a clear indication of various processing stages in the subcortical auditory system. The auditory steady-state response is another electrophysiological measure which assesses the extent to which brain responses phase lock to the stimulus.

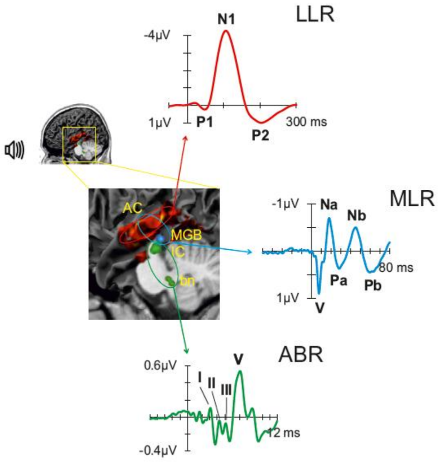

Figure 3. Auditory event related potentials, and the auditory nuclei thought to correspond each peak in the evoked response.

SENSORINEURAL HEARING LOSS (SNHL)

Description

Sensorineural hearing loss generally arises from damage to, or abnormalities of, the hair cells of the organ of Corti, although occasionally the problem originates in the auditory nerve or more central areas. SNHL can be congenital, having a genetic basis, or acquired, caused by factors such as infectious diseases (e.g. rubella, bacterial meningitis) and noise trauma(Smith, Bale et al. 2005).

Effects

Hearing loss depending on severity can affect social skills, academic achievement, employment prospects, and even life expectancy (Carvill 2001). The consequences for children with mild hearing loss that remains stable and only affects a few frequencies is not well understood, as it may remain undetected for some time (Smith, Bale et al. 2005), but there may be societal and scholastic effects that are not properly appreciated. Recent studies have shown around 7% of 7 year olds may suffer from mild high frequency hearing loss which can affect perception of some speech sounds.

Diagnosis

Usually by a trained audiologist using one or more of the methods described in the section, 'Measuring hearing loss'.

CONDUCTIVE HEARING LOSS

Description

Conductive hearing loss refers to any condition which impairs the transfer of sound energy from the world to the cochlea, including fluid in the middle ear, infections in the ear canal or middle ear, poor eustachian tube function, perforated eardrum, impacted earwax (cerumen)absence of malformation of occurs when there is a problem conducting sound waves anywhere along the route through the outer ear, tympanic membrane (eardrum), or middle ear (ossicles).

Effects

Conductive hearing loss increases hearing thresholds, i.e. the sound level necessary to detect a faint sound.

Diagnosis

Usually by a trained audiologist using one or more of the methods described in the section, 'Measuring hearing loss'. To distinguish sensorineural from conductive loss a differential test such as one or more of the ones in the table below is used.

Criteria	Sensorineural hearing loss	Conductive hearing loss
Anatomical site	Inner ear, cranial nerve VIII, or more central processing nuclei	Middle ear, tympanic membrane, or external ear
Weber test[1]	Sound localizes to normal ear	Sound localizes to affected ear
Rinne test2	Positive Rinne; air conduction is better thanor bone conduction	Negative Rinne; bone conduction is better than air conduction

Sensorineural and conductive hearing loss comparisons

OTITIS MEDIA

Otitis media refers to inflammatory disease of the middle ear which may be acute or chronic (otitis media with effusion (OME), chronic suppurative otitis media). The chronic forms of the disease seem to be related to viral

[1]The *Weber* test uses a tuning fork touched to the midline of the forehead.
[2]The *Rinne* testtests air conduction vs. bone conduction.

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respiratory infection or allergies, and worldwide affects about 80% of children at some point in time under the age of ten (Monasta, Ronfani et al. 2012, Minovi and Dazert 2014).

Effects

The chronic forms of the disease may result in hearing loss that affects the child's social communications (Minovi and Dazert 2014).

Diagnosis

Since chronic otitis media does not respnd to antibiotics, it is importantto distinguish it from acute otitis media. Diagnosis is typically performed by general practitioners, otolaryngologists or paediatricians.

Auditory Neuropathy (Auditory Agnosia)

Auditory neuropathy is a general term denoting a failure in the transmission of sound-related signals from the inner ear to the brain. The condition may stem from a variety of causes relating to signal transmission in the afferent pathway, or,for the auditory agnosias, within the cortex. However, the outer hair cells, which are part of the efferent auditory system, appear to function as normal.Auditory neuropathy in children has a strong genetic basis (Manchaiah, Zhao et al. 2011) but has also been linked to perinatal problems (e.g. jaundice, premature birth, low birth weight, inadequate oxygen supply). There may also be genetic links with the disorders, Charcot-Marie-Tooth syndrome (a group of inherited diseases that damage the peripheral nerves) and Friedreich's ataxia.

Effects

People with auditory neuropathy may even have normal hearing according to pure tone audiometry, but are unable to recognise or interpret sounds.Medical treatment of auditory neuropathy is not currently available. Some children benefit from teaching that focuses only on learning to listen and speak. However, sometimes a child with auditory neuropathy has great difficulty understanding what is heard and benefit more from visual communication approaches.

Diagnosis

The key diagnostic is a negligible or abnormal auditory brainstem response in combination with normal otoacoustic emissions.Diagnosis is typically performed by general practitioners, otolaryngologists or paediatricians.

Hyperacusis

Hyperacusis refers to the over-sensitivity to sounds(Coelho, Sanchez et al. 2007), which may even be painful at normal levels. Certain groups of children, e.g. those with Autistic Spectrum Disorder or Down's syndrome, are particularly prone to hyperacusis, which may also be accompanied by tinnitus. In some cases there hyperacusis may exhibit as an intense dislike or fear, termed phonophobia.

Effects

The main effect of hyperacusis is an emotional one, common signs include crying in noisy environments, clasping hands over the ears, fear of noise or noisy objects, or even self-harm when exposed to loud noise, and reluctance to participate in noisy or loud activities.

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Diagnosis

As it is essentially subjective, hyperacusis is difficult to diagnose, but generally a trained audiologist or hearing specialist would be involved.

TINNITUS

Tinnitus is the perception of sounds, including buzzing, ringing or noise,in the absence of external sounds. It is not a disease per se but typically results from damage to the auditory system involving hearing loss, e.g. noise-induced hearing loss, neurological damage (multiple sclerosis), ear infections, oxidative stress, side effectsfrom certain medications(Levine and Oron 2015). Children with hearing loss have a high incidence of tinnitus, even though they seldom report it (Shetye and Kennedy 2010).

Effects

The effects of tinnitus range from slight to catastrophic according to the impact it has on day-to-day life, e.g. only perceptible in quiet through to sleep disturbance and interference with social communications(Baguley, McFerran et al. 2013). Among those children who do complain of tinnitus, there is an increased likelihood of associated problems such as migraine, juvenile Meniere's disease or chronic otitis media(Shetye and Kennedy 2010).

Diagnosis

Tinnitus is generally also a subjective phenomenon which cannot be measured objectively. Generally a trained audiologist or hearing specialist would be involved in the diagnosis. It affects roughly 12% to 36% of normal hearing children and up to 66% of children with hearing loss(Shetye and Kennedy 2010)but there are no effective medications(Langguth, Kreuzer et al. 2013).

Delays in Language acquisition

Language delay or deviation: Overview

Children's language ability develops continually into adolescence and beyond. The vocabulary size and the syntactic abilities also develop across the life span into later adulthood. Therefore, the continuous language maturations have lasting effects on reading and writing skills. Continuous language development is based on a complex integration of three major aspects: sensory-motor (e.g. vocal and motor production), neuro-cognitive (e.g. audio-motor attention) and social-emotional (e.g. use of language in interacting and manipulating other people and environments).

The typical average language development of a child serves as a base to consider how a language outcome is considered in an atypical development. This helps to identify the differences in term of language delay or language deviation from the normal pathway. There are two main categories of difference: in the first case all sub-areas (e.g. phonology, syntax, semantics and pragmatics) of language are delayed compared to a typical normal or 'average' child but the developmental patterns are similar to the language of typically developing children, in the second case, a language disorder or deviation, is noted when the pattern of impairments are different across sub-areas.

Congenital language disorders (Children with Down syndrome, Williams Syndrome, X-fragile, and Autistic Spectrum Disorders)

In children with congenital developmental disorders the difficulties in language development are paralleled by a pervasive neuro-developmental disorder affecting both linguistic and non-linguistic processing. So, for these children their cognitive abilities are a limiting factor on any development of their language abilities. Children with learning difficulties typically show a later onset of language learning, and the overall level of language ability may be limited(Dick, Leech et al. 2008). Whilst language difficulties are easily identified at early stages, the developmental pathway diverges according to a disorder-specific profile, with its weaknesses and strengths, and is not constant throughout the development of the child. Language development during childhood is affected by a number of factors that include the mental ages (assessed through standard testing), severity of language impairment during the early stages of development, frequency of communicative acts, ability to understand the thought and the intentions of others, and the influence of additional non-linguistic problems.

Effects

Pervasive cognitive and/or social disorders impact on different areas of language to different degrees depending on the syndrome. For example, with the moderate cognitive impairment, children with Down's syndrome show better visuo-spatial abilities than verbal encoding compared with typically developing children that are matched with the same mental age(Varnhagen, Das et al. 1987, Jarrold and Baddeley 1997, Vicari, Bellucci et al. 2000).However, these strengths and weakmesses are not constant in the child's life.

Diagnostic & Implications

For these children constant and regular assessment help to identify at specific moments in time the strengths and weaknesses of the linguistic, cognitive and social abilities of that child. The general rule is to simplify the text adjusting the difficulty to the child's ability.

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Specific Language Impairments

Specific language impairment (SLI) is defined as a delay in the understanding and production of language in the absence of known neurological impairment, intellectual disability, hearing loss, autistic spectrum disorders or severe social or emotional problems. The language delay is expressed in itself without any other apparent overt impairment in other areas. However, the delay is manifested in all sub-areas of language including reading and writing. Particularly strong weakness is evident in grammatical morphology (e.g. use of verbs inflections for example the use of the verb tense, and detecting grammatical violations).

The problem for children with SLI is that they do not move through the optional infinitive stage as quickly as typically developing children. Evidence for this account comes from studies that have shown that English-speaking children with SLI use morphemes that are unrelated to tense (e.g., regular plural –s, -ing) with much higher accuracy than morphemes that are related to tense (e.g., third person singular –s) (Rice and Wexler 1996).

The strict meaning of specific disorder has to be taken with caution as associated non-linguistic deficits such as working memory, motor skills and reaction speed may also be observed. In particular a non-linguistic factor identified is limited phonological working memory, assessed usually by the non-word repetition and sentence repetition task (Paul 2007).

Effects

Working memory capacity is reduced. Children with SLI have general difficulty in mastering language abilities in all sub-areas, and have great difficulty in understanding grammatical rules and figurative language.

Diagnostic & Implications

Best practice principles in the UK and USA are delineated by the findings of meta-analyses of early intervention and screening studies, but much work remains to be done(Law, Garrett et al. 2004, Nelson, Nygren et al. 2006). There is a need to regularly assess the language abilities and phonological working memory of the child using the non-word and sentence repetition tests to indicate in which level the child should be placed. Support should be given to assist in improving working memory abilities with particular attention to phonological working memory. For reading and writing, help given is to reduce working memory load using simple words and sentences. In addition appropriate simplification of texts to the level at which the child currently performs is helpful.

Children with multiple physical and sensorial (MDs) disabilities

For children with multiple physical and sensory disabilities (MDs) such as quadriplegic cerebral palsy, it is often not possible to communicate verbally; speech may be limited, hard to interpret, or extremely effortful. Children with MDs often develop their own basic form of communication using facial expressions, movement, gestures and sounds which may enable them to convey simple emotions, wants or needs to their family or close friends.

Recently, various Augmented and Alternative Communication (AAC) devices have been developed to assist these children in communication; to build upon their existing vocabulary (whether verbal or non-verbal), to form new sentences and to facilitate interactions and social engagement (Sigafoos, W. et al. 2013).

Effects

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Among the various AAC aids, Voice Output Communication Aids (VOCAs) are high technology communication systems that provide a 'voice' output for the individual that is easily understood by others. VOCAs allow young people with MDs to use varying methods (e.g., direct selection, pointers, scanning, etc.), depending on their abilities to select what they would like to communicate on their VOCA from an array of options programmed onto the machine. The VOCA then turns their input into a recognisable verbal output or sentence that almost everyone can understand and interpret.

Diagnostic & Implications

The linguistic, cognitive and social abilities of children with multiple disabilitiesare severely impaired so that social communication is almost absent. Non-verbal form of assessment tailored for these children should be used (Cattani and Carroll in preparation).

BILINGUAL CHILDREN AND 'NORMAL' LANGUAGE DELAY

The bilingual population is increasing worldwide. According to the Office for National Statistics (ONS, 2011) the number of births to non-UK born mothers in England and Wales has seen a marked rise over the last decade. These births accounted for 25.1% of all live births in 2010 compared with 15.5% in 2000. This proportion has increased every year since 1990, when it was just under 12%.

It is well-established that simultaneous bilingual children may have smaller vocabularies in each of their two languages when compared to monolinguals learning one of those languages in isolation (Bialystok 2009, De Houwer 2009). They also take a little longer to reach the same level as monolinguals on various grammatical tasks, cf. (Nicholls, Eadie et al. 2011). Nonetheless, amongst the general population the impression remains that one should expect bilingual children to be delayed – even quite dramatically delayed – in the early acquisition of language (Stow and Dodd 2003) and later development of grammar (Nicholls, Eadie et al. 2011). This is considered 'normal' delay.

Limited expressive vocabulary skills in young children are considered to be the first warning signs of a potential Specific Language Impairment (SLI). However,since there is limited knowledge as to what bilingual vocabulary size should be considered as a risk factor for SLI, the effects of bilingualism and language-learning difficulties on early lexical production are often confounded.

It has been found that the extent of English language mastery is strongly predicted by the amount of exposure to the host country (in this case English). Although there is a general consensus that it is usually invalid to compare bilingual children to monolingual norms, Cattani, Abbot-Smith et al. (2014)showed that at and above 60% of exposure to English, measured using a language exposure questionnaire (see also (Gatt, O'Toole et al. 2015)), bilingual children are comparable to their monolingual peers and can be assessed using the monolingual norms so that a child with language disorder can be identified. For children with less than 60% of exposure to English, the findings are less clear but the suggestion is to test the child in the additional language. However, the general assessment does not give any indication of areas where the child has full knowledge competence in the vocabulary for that language (for example, at home the topics of conversation might not overlap in what happens at school, so the words within those topics might not be learnt in the host language). Increasing the amount of reading (in either language) on a vast array of topics will help the child to expand their vocabulary.

Effects

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In bilingual language learning environments, the expressive vocabulary size in each of the child's developing languages is usually limited compared to the number of words produced by their monolingual peers. The large variability depends primarily on the exposure to the language of the host country.

Diagnostic & Implications

It is difficult to disentangle the effect of bilingualism from the effect of specific language impairment. Vocabulary tests normed specifically for bilingual children do not currently exist. However, there is currently work on designing Cross-linguistic Lexical Tasksfor bilingual preschool children(Haman, Łuniewska et al. 2015).

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