# MASKING LEVEL DIFFERENCES AND BINAURAL INTELLIGIBILITY LEVEL DIFFERENCES IN CHILDREN WITH DOWN SYNDROME

By

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# CHAPTER I

# INTRODUCTION

Binaural hearing results in a number of listening advantages relative to monaural hearing, including enhanced hearing sensitivity, more accurate localization of auditory signals, and better speech understanding in adverse listening conditions. These advantages are facilitated by the ability to detect and utilize interaural cues. However, some listening advantages can be captured monaurally, such as attending to the ear with the better signal-to-noise ratio (SNR). To assess binaural abilities using interaural cues, one must first account for any monaural advantage due to improved SNR within each ear. Some studies have used clever techniques to allow independent manipulation of the various cues underlying speech recognition (i.e., by recording through a manikin's ears and presenting the signals to subjects via headphones). Such techniques enable the investigation of the relative influence of various listening cues for reducing the interference of background noise on a signal of interest in the free-field (e.g., Bronkhorst & Plomp, 1988). Listening cues that include monaural components have a greater relative influence on speech understanding in noisy situations than binaural cues for adult listeners; however, this has not been investigated for children (Bronkhorst & Plomp, 1988). Auditory perceptual abilities develop throughout childhood; therefore, it is possible that the relative influence of various listening cues differs for children and adults (Cameron et al., 2009; Hall, Buss, Grose, & Dev, 2004; Hall & Grose, 1990a; Van Deun et al., 2009).

The auditory perceptual abilities of children with developmental disabilities can also be expected to develop throughout childhood, though it is reasonable to suspect that their auditory perceptual development might vary from that of typically-developing children. For example, it is well understood that infants and children with Down syndrome have unique auditory structures resulting from congenital and acquired influences (e.g., Balkany, Mischke, Downs, & Jafek, 1979; Becker, Armstrong, & Chan, 1986; Bilgin, Kasemsuwan, Schachern, Paparella, & Le, 1996; Blaser et al., 2006). Binaural processing of auditory stimuli begins subcortically with the most peripheral site of human binaural interaction being the superior olivary complex. Aberrant subcortical anatomy, such as that seen in children with Down syndrome, might result in atypical binaural processing. It is possible that these structural aberrations contribute to the differences in the auditory perceptual capabilities of children with Down syndrome and their typically-developing peers that have been discussed in speech perception studies (e.g., Cardoso-Martins & Frith, 2001; Jarrold, Baddeley, & Phillips, 2002; Keller-Bell & Fox, 2007). The following review of the literature will describe current knowledge regarding the binaural capabilities of typically-developing children as well as provide a rationale for why the binaural capabilities of children with Down syndrome might differ.

# CHAPTER II

# REVIEW OF THE LITERATURE

## Binaural Release from Masking

Listening in noise is more difficult than listening in quiet, even for robust signals of interest like speech. Even individuals with normal hearing experience difficulty attending to a single speaker in a room when many sound sources are present. However, it has been historically well documented that the effects of interfering background noise decrease when listening with two ears rather than one (e.g., Carhart, 1965; Moncur & Dirks, 1967). The central auditory system is able to utilize information from signals arriving at each ear to reduce the interference of background noise on a signal of interest. This phenomenon is known as binaural release from masking.

Various models have been proposed to describe binaural release from masking including an Equalization-Cancellation model (EC; Durlach, 1972), a "vector" or lateralization model (Hafter, 1971; Jeffress, Blodgett, Sandel, & Wood, 1956; Webster, 1951), and a cross-correlation model (Colburn, 1973, 1977; Jeffress, 1948). Though these models vary slightly from one another, they can each be described under a framework that includes a comparison of signals within the central auditory system after processing by the peripheral auditory system (Stern & Trahiotis, 1995). Whereas cross-correlation and lateralization models suggest that the central auditory system compares timing information from responses of the fibers from the two ears as a function of characteristic frequency, the EC model does not suggest specific neural mechanisms underlying binaural processing. However, the mathematical equations proposed by Durlach (1972) successfully describe many binaural phenomena.

## Mechanisms of Binaural Hearing

### Interaural Cue Detection

Sounds originating from a single location in space will arrive at each ear at a particular time and intensity. The relative time and intensity that a sound arrives at each ear varies based on the position of the sound source relative to head orientation. The central auditory system can use these relative differences between ears for localization, spatial awareness, orientation to sound, and minimization of the effects of background noise on a signal of interest. Differences in sound between ears are described in terms of phase (for periodic signals), or interaural phase differences (IPDs), time, or interaural time differences (ITDs), and level, or interaural level differences (ILDs).

The ITD of a sound can vary from 0 ms for sound originating at 0° azimuth to approximately 0.6 ms for sounds generated at 90° or 270° azimuth. Interaural time or phase cues are most reliably detected for low frequency sounds as their relatively long wavelength reaches each ear at a different phase. To contrast, the short wavelength of high frequency sounds is susceptible to head shadow and as a result, interaural level cues are most reliably detected for high frequency sounds. The maximum ILD for a 500 Hz tone is 4 dB compared to the maximum ILD for a 6000 Hz tone, which is nearly 20 dB (for review see Grantham, 1995). The central auditory system uses multiple cues (both interaural cues as described above and monaural spectral cues) for localizing sound and minimizing the effects of noise on a signal of interest.

Anatomy and Physiology of Binaural Hearing

Multiple nuclei and pathways are involved in binaural hearing occurring at three primary levels of the brainstem: the superior olivary complex, the nuclei of the lateral lemniscus, and the inferior colliculus.  Figure 1 illustrates major auditory brainstem nuclei and their projections.

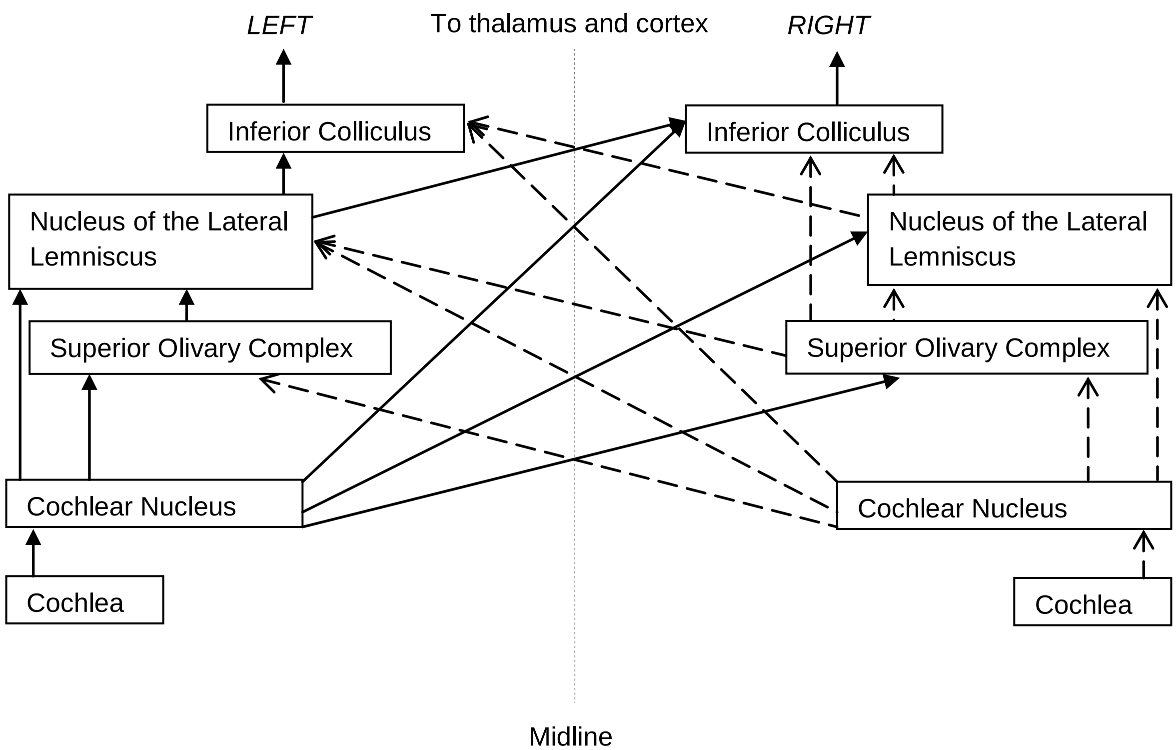


*Figure 1.*  Major auditory brainstem nuclei utilized for binaural hearing.

The anterior ventral cochlear nucleus projects to the ipsilateral and contralateral superior olivary complex and the ipsilateral nucleus of the lateral lemniscus.  In addition, some cochlear nucleus axons also project to the contralateral nucleus of the lateral lemniscus and the inferior colliculus. The superior olivary complex projects bilaterally to the nucleus of the lateral lemniscus and ipsilaterally to the inferior colliculus.  The nucleus of the lateral lemniscus projects bilaterally to the inferior colliculus. The inferior colliculus is unique in that it is innervated by every nuclear group within the auditory brainstem. Axons from the inferior colliculus project to the thalamus and cortex.

There is evidence to support the involvement of brainstem structures including the superior olivary complex and inferior colliculus in speech-in-noise discrimination and the analysis of ITDs and ILDs (Litovsky, Fligor, & Tramo, 2002; Moore, 1991; Song, Skoe, Banai, & Kraus, 2010). Brainstem encoding of the fundamental frequency (F0) is associated with speech discrimination in background noise (Song, Skoe, Banai, & Kraus, 2011). In addition, the superior olivary complex is predominately involved in analyzing ITDs and ILDs (For review see, Moore, 1991). However, the inferior colliculus is also sensitive to ITDs and ILDs in addition to mediating other aspects of binaural processing such as localization of sound in a hemifield and echo suppression (Litovsky et al., 2002).

## Measurement of Binaural Release from Masking

### Pure Tone Stimuli

A typical measure of binaural release from masking is based on the detectability of pure tone signals in the presence of noise maskers presented over headphones. For example, detection threshold can first be measured in a reference condition in which the noise and the signal are presented identically to the two ears (a condition referred to as N0S0). If the interaural relationship of the noise or the masker is changed, there can be an improvement in threshold, or a release from masking. The difference in threshold between the dichotic configuration and the reference condition is commonly referred to as the masking level difference (MLD). The maximum MLD – about 15 dB for a low frequency tone – is typically observed when the noise is presented identically to the two ears while the signal tone is presented 180° out of phase in one ear relative to the other (a condition referred to as $N0S\pi$).

The MLD can be affected by many variables, such as signal frequency, signal interaural phase, noise intensity, and noise bandwidth (for review see Green, 1976). For example, low frequency signals yield greater improvements than high frequency signals. For the $N0S\pi$ configuration, an MLD of 15 dB can be expected for adult listeners with normal hearing for a low frequency pure tone signal, but the MLD decreases with increasing frequency to about 3 dB at approximately 1500 Hz and higher (Hirsh, 1948). The largest $N0S\pi$ MLDs occur when the noise is presented at a moderate to loud sensation level that is similar in intensity between ears (Hirsh, 1948). Though some binaural benefits can be expected for noise intensity differences of 10 dB between ears, benefits diminish for larger differences (Blodgett, Jeffress, & Whitworth, 1962). Finally, the MLD for low frequency signals increases as noise bandwidth decreases, at least for adults (e.g., Grose, Hall, & Dev, 1997). This result could be due to the inherent amplitude fluctuations present in narrowband noise. Mature binaural auditory systems might be able to take advantage of the relatively good signal-to-noise ratio in the minima (troughs) of ongoing narrowband noise.

Speech Stimuli

Binaural release from masking for speech signals is often measured by comparing the change in detection or intelligibility of speech signals in noise presented in varying binaural configurations. This can be done in the free-field by spatially separating signal and noise sources or under headphones by varying parameters such as ITD and ILD. In free-field situations it is not possible to independently manipulate the ILD and ITD cues, because they vary together as a sound source is moved. Also, it must be noted that moving a sound source away from midline, in addition to producing ITD and ILD cues, results in changing SNRs at each ear. Thus, an improvement in threshold may reflect the fact that the spatial configuration allows a listener to attend to the ear with the better SNR, which requires only monaural abilities.

Several studies have demonstrated the positive effects of binaural release from masking on listening to speech in adverse listening situations (Bronkhorst & Plomp, 1988; Carhart, 1965; Harris, 1965; MacKeith & Coles, 1971; Moncur & Dirks, 1967). Advantages of 3 to 9 dB can be expected for conditions similar to real-life binaural hearing situations in which ITD and ILD are able to be utilized by listeners (e.g., Carhart, 1965; Levitt & Rabiner, 1967). By recording sounds through a manikin's ears, processing these recorded left ear and right ear signals, and presenting them over headphones, Bronkhorst & Plomp (1988) were able to investigate the separate contributions of ILDs and ITDs to spatial release from masking. They found that adults with normal hearing have greater average binaural release from masking for speech with only ILD cues (ITDs set to 0) than with only ITD cues (ILDs set to 0). These results suggest that head shadow is a significant determinant of binaural advantage for speech intelligibility in free-field conditions, at least for adults.

## Development of Binaural Release from Masking

### Pure Tone Stimuli

The ability to use interaural cues to detect signals in noise develops throughout childhood. Infants and children have been shown to have smaller MLDs than adults for many signal and noise configurations (Grose et al., 1997; Hall & Grose, 1990a; Nozza, 1987; Van Deun et al., 2009). For example, typically-developing infants with normal hearing have smaller MLDs than adults for 500 Hz pure tones presented in broadband noise (i.e., 600 Hz; Nozza, 1987). However, by about 5 to 10 years of age, typically-developing children have adult-like MLDs for similar signal and noise configurations (e.g., 500 Hz pure tones presented in broadband noise; Grose et al., 1997; Hall & Grose, 1990a; Moore, Cowan, Riley, Edmondson-Jones, & Ferguson, 2011). Similar results have been observed for filtered click trains presented in broadband noise (Van Deun et al., 2009).

Though the discussion above suggests that children have adult-like MLDs by school-age, the bandwidth of the masking noise can affect MLDs in children. That is, children have smaller MLDs than adults for signals presented in narrower bandwidths of noise, such as 20 Hz and 40 Hz (Grose et al., 1997; Hall & Grose, 1990b). Children seem to have difficulty optimizing binaural temporal window placement, but the ability to take advantage of temporal minima in masker noise to detect a signal improves with age (Hall, Buss, & Grose, 2007; Hall et al., 2004).

Speech Stimuli

Some developmental changes in binaural advantage for the perception of speech sounds and sentences presented in noise have been demonstrated (Cameron & Dillon, 2007; Nozza, Wagner, & Crandell, 1988). Infants have smaller MLDs than adults for the speech sound /ba/ presented in broadband masking noise; however, adult-like MLDs can be expected in this condition for children between the ages of 3.5 and 4.5 years (Nozza et al., 1988). Increased binaural advantage with age for understanding sentences presented in multi-talker noise can be expected for children between the ages of 5 to 11 years (Cameron et al., 2009; Cameron & Dillon, 2007). However, children as young as 2.5 years of age have demonstrated binaural benefit similar to adults for understanding speech in noise in some circumstances (Garadat & Litovsky, 2007). For example, preschool and school-aged children exhibit adult-like binaural benefits for understanding closed-set spondees presented in competing speech stimuli (Garadat & Litovsky, 2007; Litovsky, 2005).

## Binaural Hearing in Children with Down Syndrome

With the exception of a few studies (e.g., Glenn, Cunningham, & Joyce, 1981; Werner, Mancl, & Folsom, 1996), little work has been done to understand the auditory abilities of infants and children with Down syndrome. However, some broad conclusions about the perceptual auditory capabilities of infants and children with Down syndrome can be made from speech perception studies. Results from speech perception studies suggest that infants and children with Down syndrome have poorer auditory perceptual capabilities than their typically-developing peers even when they are matched by equal reading ability, developmental age, and chronological age (e.g., Cardoso-Martins & Frith, 2001; Glenn et al., 1981; Jarrold et al., 2002; Keller-Bell & Fox, 2007; Roch & Jarrold, 2008; Yoder, Camarata, Camarata, & Williams, 2006).

Although one might suspect that the relatively poor speech perception abilities characteristic of children with Down syndrome are the result of cognitive deficits, their ability to listen in noise could be influenced by less-than-optimal binaural processing abilities. Though the binaural processing capabilities of children with Down syndrome have not been explored specifically, other auditory capabilities have been investigated, such as the development of auditory sensitivity, phonological discrimination, and stimulus complexity discrimination. A comprehensive summary of these studies is provided in Table 1.

Table 1

*Research Relative to Auditory Processing Capabilities for Children with Down Syndrome*

| Task | Performance | References |
|---|---|---|
| Sensitivity | DS<TD detection performance and threshold; similar developmental trajectory | (Werner et al., 1996) |
| Localization | DS begin to perform VRA at older ages than TD | (D. Greenberg, Wilson, Moore, & Thompson, 1978; Wilson, Folsom, & Widen, 1983) |
| Stimulus discrimination | DS preference for complex stimuli | (Glenn et al., 1981) |
| Speech perception – word stress | DS<TD | (Pettinato & Verhoeven, 2009) |
| Phoneme detection | DS<TD | (Cardoso-Martins & Frith, 2001; Cardoso-Martins, Michalick, & Pollo, 2002; Roch & Jarrold, 2008; Snowling, Hulme, & Mercer, 2002) |
| Initial phone detection for simple words embedded in noise | older DS = TD children | (Jarrold, Thorn, & Stephens, 2009) |
| Phoneme deletion (unspecified) | DS<TD | (Roch & Jarrold, 2008) |
| First syllable recognition | DS<TD | (Verucci, Menghini, & Vicari, 2006) |
| Rhyme detection | DS<TD | (Boudreau, 2002; Cardoso-Martins et al., 2002; Gombert, 2002; Roch & Jarrold, 2008; Snowling et al., 2002; Verucci et al., 2006) |
| Alliteration judgment | DS<TD | (Boudreau, 2002; Snowling et al., 2002) |
| Speech discrimination : /i/ - /u/ and /dab-i-ba/ - /dab-u-ba/ | DS<TD | (Keller-Bell & Fox, 2007) |
| Speech sound discrimination (passive task) | Positive correlation for DS for grammatical comprehension and ERP for speech stimuli | (Yoder et al., 2006) |

*Note*. < = poorer performance; = = similar performance; DS = individuals with Down syndrome; TD = typically-developing individuals, VRA = visual reinforcement audiometry; ERP = event-related potential.

Auditory Sensitivity

The development of auditory sensitivity in infants with Down syndrome is similar to that of typically-developing infants in that younger infants respond less consistently and louder sounds are required to elicit responses compared to older infants (Werner et al., 1996). The auditory sensitivity of infants with Down syndrome has been shown to be up to 10 to 25 dB poorer than their typically-developing peers (Werner et al., 1996). However, it should be noted that children with Down syndrome were less attentive than typically-developing infants enrolled in that study. Levels of attentiveness can be modeled by examining an infant's maximum percentage of correct responses, or $p(C)_{max}$. Typically-developing 6 to 9 month old infants are expected to achieve maximum performance levels of 0.89, but younger (i.e., 2 to 3 months of age) and older (i.e., 4 to 12 months of age) infants with Down syndrome in this study only achieved $p(C)_{max}$ levels between 0.70 and 0.80, respectively (Werner et al., 1996). Thus, infants with Down syndrome are less attentive than typically-developing infants and this non-sensory factor could have contributed to these auditory sensitivity estimations.

Phonological Discrimination

The ability to perceive weak syllables (e.g., the second syllable in the word "hearing") and discriminate between speech sounds is often compromised for children and adults with Down syndrome compared to typically-developing peers (e.g., Keller-Bell & Fox, 2007; Pettinato & Verhoeven, 2009). Though phonological discrimination can be influenced by reading ability, mental age, cognitive characteristics, and chronological age, individuals with Down syndrome continue to demonstrate poorer abilities than typically-developing individuals even when groups are matched by these factors (For review see, Lemons & Fuchs, 2009). Furthermore, physiologic measures of passive phoneme discrimination are related to speech discrimination in children with Down syndrome providing more evidence for the influence of perceptual auditory abilities on phoneme discrimination (Yoder et al., 2006).

## Stimulus Complexity Discrimination

Stimulus complexity discrimination has been investigated in 12 month old infants with Down syndrome using behavioral test paradigms that utilize toys producing sound in response to physical manipulation (i.e., touching a switch; Glenn et al., 1981). Developmental age matched infants with Down syndrome and typically-developing infants had some response similarities. For example, both groups of infants responded with equal frequency to both simple stimuli (e.g., a single repeated piano tone) and complex stimuli (e.g., a song) suggesting similar motor skill ability. In addition, both groups of infants demonstrated a preference for complex stimuli over simple stimuli by triggering complex stimuli more often and for longer durations than simple stimuli. Interestingly, research utilizing visual paradigms has shown that infants with Down syndrome remained engaged longer and were less likely to habituate than typically-developing infants (For review see, Wagner, Ganiban, & Cicchetti, 1990). However, for this complex versus simple stimuli task, infants with Down syndrome remained engaged longer than their typically-developing peers only when listening to complex stimuli. The exact relationship of stimulus complexity and response tendency is unknown; however, it is possible that the enhanced preference for complex over simple stimuli seen for infants with Down syndrome could be the result of unique perceptual auditory capabilities.

## Anatomy of Binaural Hearing for Children with Down Syndrome

It is well known that many infants with Down syndrome have stenotic ear canals and congenital malformations of the ossicles, cochlea, and internal auditory canals (e.g., Balkany et al., 1979; Bilgin et al., 1996; Blaser et al., 2006; Satwant, Subramaniam, Prepageran, Raman, & Jalaludin, 2002; Strome, 1981). Alterations in peripheral auditory structure might influence auditory input, such as spectral characteristics of signals, and confound auditory functions such as localization and recognition of speech in noise. In addition, alterations in peripheral auditory structures and, consequently, in the auditory input signal could influence the development of the central auditory system (Wilmington, Gray, & Jahrsdorfer, 1994).

A large body of histologic, radiologic, and electrophysiologic evidence describes altered anatomy and synaptic communication within the central auditory system for children with Down syndrome (e.g., Banik, Davison, Palo, & Savolainen, 1975; Becker et al., 1986; Colon, 1972; Crome, Cowie, & Slater, 1966; Kittler et al., 2009; Pinter, Brown et al., 2001; Pinter, Eliez, Schmitt, Capone, & Reiss, 2001). Specifically, histologic and radiologic evidence shows that individuals with Down syndrome have reduced cortical and subcortical structures and reduced cortical white and gray matter in selected brain regions compared to individuals without Down syndrome (Colon, 1972; Crome et al., 1966; Davidoff, 1928; Frangou et al., 1997; Haier, Cheueh, Touchette, & Lott, 1995; Jernigan, Bellugi, Sowell, Doherty, & Hesselink, 1993; Pinter, Brown et al., 2001; Pinter, Eliez et al., 2001; Takashima, Becker, Armstrong, & Chan, 1981; Weis, Weber, Neuhold, & Rett, 1991; Wisniewski, 1990; Wisniewski, Laure-Kamionowska, & Wisniewski, 1984). For example, one of the earliest studies of subcortical structures of infants, children, and adults with Down syndrome noted reduced brain weight, small brainstem and cerebellum size, embryologic simplicity, and cell poverty (Davidoff, 1928). More recent research has also described smaller brain structures (e.g., ventral pons, hippocampal formations) and overall smaller brain volumes for children, young adults, and adults with Down syndrome compared to typically-developing peers (e.g., Pinter, Eliez et al., 2001; Raz et al., 1995; Wisniewski, 1990). This altered anatomy within the central auditory system for children with

Down syndrome suggests that they could process binaural auditory information differently than their typically-developing peers.

Differences in neural development between individuals with Down syndrome and their typically-developing peers could appear early in life. Dendritic trends toward less expansion often occurs earlier for infants with Down syndrome than their typically-developing peers (Becker et al., 1986). Molecular research using mouse models for Down syndrome has shown that the Minibrain (Mnb) gene on chromosome 21 is involved in neurogenesis and could be involved in developmental alterations, such as relative decreases in the number of neurons seen for individuals with Down syndrome (Hämmerle et al., 2003). Furthermore, adults with Down syndrome have reduced myelin compared to their typically-developing peers (Banik et al., 1975). Taken together, these results could explain findings of altered neural cell density and cytoarchitecture for individuals with Down syndrome (Colon, 1972; Takashima et al., 1981; Wisniewski, 1990; Wisniewski et al., 1984). It is clear that there are differences in the size and cytostructure of cortical and subcortical structures between individuals with Down syndrome and their typically-developing peers. It has been suggested that these differences might be influenced by impairments in neurogenesis and early programmed cell death that typically-developing individuals require for optimal neuronal development.

Electrophysiologic studies also converge on the notion that individuals with Down syndrome have differing auditory capabilities from those of their typically-developing peers. In general, auditory brainstem response (ABR) latencies are shorter for infants, older children, and adults with Down syndrome compared to their age-matched, typically-developing peers (Arao & Niwa, 1991; Folsom, Widen, & Wilson, 1983; Forti et al., 2008; Jiang, Wu, & Liu, 1990; Kittler et al., 2009). Figure 2 illustrates a sample ABR evoked using moderately-high level stimuli for infants with Down syndrome and typically-developing infants.

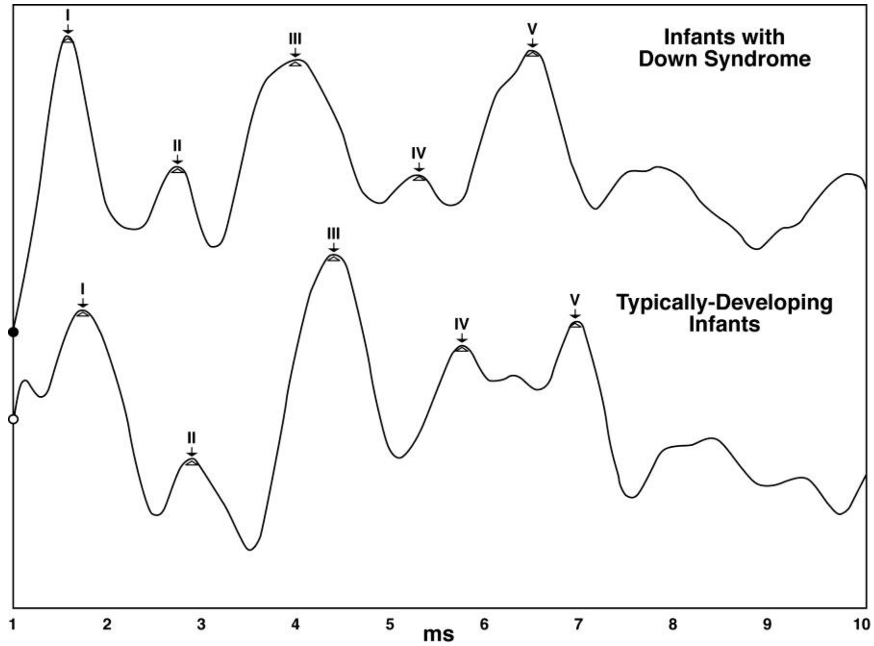


*Figure 2.* A sample auditory brainstem response (ABR) evoked using moderately high intensity level stimuli for infants with Down syndrome and typically-developing infants. Image from "Hearing Loss among Persons with Down Syndrome," by H. Porter and A.M. Tharpe, 2010, *International Review of Research in Mental Retardation, 39,* p. 214. Copyright 2010 by Elsevier, Inc.

Though anatomic contributions to decreased brainstem latencies for individuals with Down syndrome have been suggested including decreased size of auditory structures (e.g., cochlea, central auditory pathway) and shorter neural conduction times, calibration techniques have not been widely reported in the literature (Diaz & Zurron, 1995; Ferri, Gracco, Elia, Musumeci, & Stefanini, 1995; Forti et al., 2008; Kakigi & Kuroda). Individuals with Down syndrome often have stenotic ear canals, which can result in higher stimulus sound levels at the tympanic membrane relative to normative data and decreased ABR latencies measures in the abovementioned studies. However, assuming appropriate calibration, evidence of decreased

ABR latencies for individuals with Down syndrome suggests altered responses to auditory stimuli measured at the brainstem, the first site of binaural interaction.

Though children with Down syndrome have shorter latencies than their typically-developing peers for measures of early-latency evoked potentials, evoked potentials measured at later latencies reflecting more central processing are often delayed for children and young adults with Down syndrome.  Seidl et al. (1997) reported longer N1, P2, N2, and P3 latencies for children and young adults with Down syndrome compared to typically-developing peers.  Delays in these components could reflect a stimulus processing deficit as they are sensitive to changes in stimuli such as frequency, amplitude, auditory motion, and speech (Stapells, 2002).  Other studies that have used electrophysiologic methods to examine passive neural responses to auditory stimuli suggest impairments in preattentive auditory processing and lateralization of speech and non-speech stimuli for individuals with Down syndrome (Groen, Alku, & Bishop, 2008; Pekkonen, Osipova, Sauna-Aho, & Arvio, 2007).

Structural and neurochemical deficits have been suggested as contributors to differences in auditory processing between individuals with Down syndrome and age- and gender-matched peers measured using electrophysiologic test methods (Groen et al., 2008; Pekkonen et al., 2007; Seidl et al., 1997).  Deficits of the cholinergic system, which is utilized for synaptic transmission, and the histaminergic system, which regulates brain activity, have also been suggested as contributors to differences in auditory processing abilities between individuals with Down syndrome and their typically-developing peers (Pekkonen et al., 2007; Seidl et al., 1997).  There is clear evidence to suggest that differences in neural processing of auditory stimuli exist for individuals with Down syndrome and their typically-developing peers though the specific mechanism responsible for these differences remains undefined.

## Purpose

Structural anomalies within the central auditory system and alterations in synaptic communication could adversely affect various aspects of auditory processing for children with Down syndrome, including binaural hearing. However, binaural hearing capabilities have not been examined in these children. Therefore, this study sought to determine if the binaural abilities of children with Down syndrome are compromised relative to those of typically-developing children on two different tasks:

1.) a masking level difference (MLD) task using pure tone stimuli in 300 Hz wide masking noise; and

2.) a binaural intelligibility level difference (BILD) task using speech stimuli in the presence of broadband noise for natural (i.e., simulated free-field) and isolated cue (i.e., ILD and ITD) conditions.

Hypothesis: Children with Down syndrome will have poorer MLD and BILD scores than typically-developing children for all experimental conditions.

This hypothesis was based on evidence of altered subcortical anatomy and deficits in synaptic communication for children with Down syndrome (e.g., Colon, 1972; Kittler et al., 2009; Pinter, Eliez et al., 2001). In addition, children with Down syndrome have poor speech perception abilities (e.g., Keller-Bell & Fox, 2007; Pettinato & Verhoeven, 2009), which could be the result of poor auditory perceptual abilities, including binaural hearing.

# CHAPTER III

# METHODOLOGY

## Participants

Fifty-two typically-developing children and 14 children and adolescents with Down syndrome were consented to participate in this study. However, six typically-developing children and three children with Down syndrome were excluded based on results from pre-experimental testing. As such, 46 typically-developing children ( $\overline{X}$ =8 years, 6 months; range=3 years, 4 months to 12 years, 11 months), nine children with Down syndrome ( $\overline{X}$ =10 years, 2 months; range=6 years, 6 months to 11 years, 11 months), and two adolescents with Down syndrome ( $\overline{X}$ =16 years, 9 months; range= 16 years, 8 month to 16 years, 11 months) were included in the data analysis. These age ranges were chosen to facilitate the identification of developmental trends in binaural release from masking for both groups. Typically-developing children from a younger chronological age range than children with Down syndrome were included to facilitate comparisons based on estimates of developmental age (i.e., receptive language age equivalent).

The majority of children and adolescents with Down syndrome included in this study had Trisomy 21 identified by karyotype according to the medical record or parental report (81%). However, one child was noted to have translocation of chromosome 21 to chromosome 14 and one child's karyotype information was unavailable in the medical record. The average receptive language age equivalent for children and adolescents with Down syndrome was 6 years, 7 months (*SD*=5 years, 9 months). A greater percentage of children and adolescents with Down syndrome were reported by their parents to have a significant history of otitis media (64%) than typically-developing children (22%).

Seven typically-developing adults ( $\overline{X}$ =27 years, 6 months; range=23 years, 1 month to 35 years, 9 months) and three adults with Down syndrome ( $\overline{X}$ =28 years, 1 month; range=22 years, 3 months to 38 years, 4 months) were consented to participate in the study to obtain an estimate of adult performance for the experimental tasks. One typically-developing adult was excluded based on use of a primary language other than English.

All study participants had bilateral pure tone threshold averages (PTAs) of ≤30 dB HL measured at 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz and PTAs. Threshold at 500 Hz was verified to be ≤30 dB HL bilaterally to ensure audibility of the stimuli used for the MLD task as it is possible to have a PTA of ≤30 dB HL and hearing loss ≥30 dB HL at any one frequency.

As no gender effects in binaural abilities have been documented in children, gender was not considered in the selection of participants. Children with additional significant disabilities affecting gross motor control or a known receptive language age of less than 3 years were excluded from recruitment for this study. All participants were recruited according to principles and procedures approved by the Institutional Review Board at Vanderbilt University Medical Center. All participants were recruited from the Down Syndrome Association of Middle Tennessee, the Monroe Carell Jr. Children's Hospital at Vanderbilt Down Syndrome Clinic, the Vanderbilt Bill Wilkerson Center, and the Vanderbilt Kennedy Center.

## Materials and Instrumentation

All auditory stimuli were presented through Sennheiser HD 265 linear headphones (Sennheiser Electronic Corporation; Old Lyme, CT) using MATLAB 7.1 (The MathWorks, Inc.;

Natick, MA) to generate and control auditory stimuli and visual reinforcement when applicable. Visual stimuli were routed from a main computer to a secondary computer monitor used for the singular purpose of response reinforcement for paradigms in which visual reinforcement was used (i.e., audiometric testing and MLD tasks).

Masking Level Difference (MLD)

Stimuli consisted of a 500 Hz pure tone presented either interaurally in phase (S0) or 180° out of phase (S$\pi$) based on data in the literature suggesting that MLD is greatest for these signal configurations and low-frequency stimluli (e.g., Hirsh, 1948). The masker was a 300 Hz wide Gaussian noise, centered at 500 Hz, presented interaurally in phase (N0), similar to other research (e.g., Hall & Grose, 1993). The signal was approximately 400 ms in duration with a 50 ms rise/fall time, temporally centered within the 800 ms masker noise. Stimulus presentation rate was adapted based on individual responses; that is, presentation of stimuli on each trial began approximately 500 ms following participant response to the stimuli on the previous trial.

Binaural Intelligibility Level Difference (BILD)

Speech material consisted of recorded words from the Northwestern University-Children's Perception of Speech test (NU-CHIPS; Elliott & Katz, 1980). This test was designed to be used with children whose language age 3 years or above. Speech stimuli were approximately 500 ms to 800 ms in duration and noise remained constant for each condition. Noise stimuli consisted of broadband noise that contained the same long-term spectral average of the speech stimuli (Figure 3).

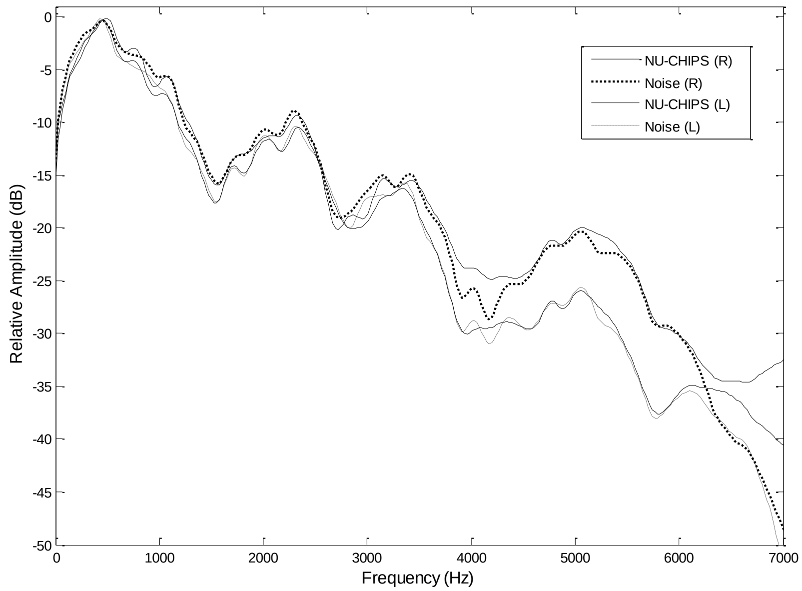


*Figure 3.* Long-term spectral averages of speech stimuli obtained from the Northwestern University-Children's Perception of Speech test (NU-CHIPS; Elliott & Katz, 1980) and broadband noise stimuli created for use in the binaural intelligibility level difference (BILD) task shown here as recorded from the left (L) and right (R) ears of a Knowles Electronic Manikin for Acoustic Research (KEMAR).

Speech and noise stimuli were presented from a Tannoy Precision 6P loudspeaker (Tannoy, Ltd.; North Lanarkshire, SCT) and recorded from two ER-11 microphones (Etymotic Research, Inc.; Elk Grove Village, IL) placed in the ears of a Knowles Electronic Manikin for Acoustic Research (KEMAR) that was positioned in an anechoic chamber at the Vanderbilt Bill Wilkerson Center. Speech material was presented from 0° azimuth (i.e., directly in front of KEMAR) at a distance of 1.5 m. Noise was presented from 0°, 45°, or 90° azimuth by rotating the head and torso of KEMAR while keeping the position of the loudspeaker fixed. An equalization filter was applied to recordings as described by Killion (1979). This was necessary to account for the addition of the ear canal resonance of the listener when playing this type of recording through headphones. Some research has described similar head-related transfer functions (HRTFs) for children and adults by the age of 3 years (Kruger, 1987); therefore, no additional filters were applied to stimuli for presentation to children.

Using signal processing techniques described by Bronkhorst and Plomp (1988), three conditions were derived from the free-field recordings to be presented through headphones: 1) simulated free-field (i.e., unprocessed recordings, preserving the natural ILD and ITD cues), 2) isolated ILD (i.e., recordings processed such that the ITD was equal to 0 μs for all presentations), and 3) isolated ITD (i.e., recordings processed such that the ILD was equal to 0 dB for all presentations). A sample of recorded signals and derived signals is shown in Figure 4.

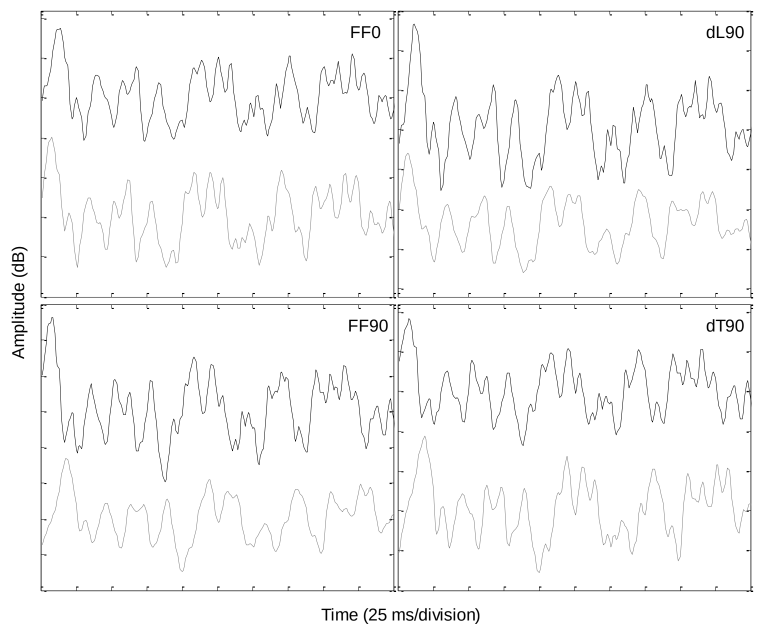


*Figure 4.* Sample of recorded noise signals (left panels) and derived signals (right panels) illustrating the effect of signal processing. The upper and lower traces in each panel correspond to the recordings made ipsilateral and contralateral to the loudspeaker, respectively.

These conditions were chosen to examine the individual contributions of ILD and ITD to free-field performance since performance in the isolated ILD condition relies mainly on monaural or "best ear" performance as a result of head shadow, whereas performance in the isolated ITD condition utilizes purely binaural interaction.

## Pre-Experimental Procedures

### Audiometric Testing

Hearing thresholds were obtained for octave frequencies 500 Hz to 4000 Hz bilaterally using the modified Hughson-Westlake method in 5 dB steps (Carhart & Jerger, 1959). Visual reinforcement consisted of a series of pictures and animated scenarios that were revealed on a computer screen as children correctly identified that they heard an auditory stimulus. Audiometric testing lasted approximately 15 minutes.

### Vineland Adaptive Behavior Scales

Parents of children with Down syndrome were asked to complete the Vineland Adaptive Behavior Scales Parent/Caregiver Report Form (Second Edition; Sparrow, Cicchetti, & Balla, 2005), which is a measure of personal and social skills required for daily living (i.e., adaptive behavior) appropriate for individuals aged 3 years, 0 months to 21 years, 11 months. Age equivalent scores from the receptive communication subdomain were used as an estimate of developmental ability as it pertains to the experimental tasks used for this study.

### Otologic Case History

Recurrent otitis media has been shown to reduce MLD in children (Hall & Grose, 1993; Hall, Grose, & Pillsbury, 1995). Chronic middle ear disease afflicts approximately 70% of children with Down syndrome compared to about 40% of typically-developing children (e.g., Auinger, Lanphear, Kalkwarf, & Mansour, 2003; Mitchell, Call, & Kelly, 2003; Strome, 1981). As such, it was anticipated that many children with Down syndrome recruited for this study would have a history of otitis media. A brief survey was constructed to obtain demographic information regarding each participant's otologic health history (Appendix A).

This survey was completed by the parent on the day of testing. A significant history of middle ear disease was defined as the occurrence of ≥ five episodes of middle ear disease before the age of 5 years or the surgical placement of pressure equalization tubes. Although not used as exclusionary criteria, otologic health history information was considered during data interpretation.

### Familiarization and Feasibility Testing

Procedures for feasibility testing and experimental testing were identical with the exception that signal level was greater than noise level for both tasks to facilitate ease of signal identification. Five trials were used for feasibility testing for each experimental procedure. Feasibility testing for the MLD task was completed in the N0S$\pi$ condition and for the BILD task in the FF0 condition. If behavior (e.g., wiggling, inattention, coughing) was observed by the examiner to have influenced responses, another five stimuli were presented and included in the calculation of accuracy. Participants must have demonstrated at least 80% accuracy to have been included in the corresponding experimental task.

## Experimental Procedures

### Masking Level Difference (MLD)

Participants were told a story by the investigator about an owl (500 Hz signal) that lived in a windy forest (masking noise). The story script is included in Appendix B. A computer monitor was used to generate two identical forest images located on the left and right of the screen. The images were highlighted individually to correspond to the presentation of noise to demark temporal intervals. A series of owl and forest images were randomly presented to increase visual interest for this task. Participants were asked to identify by pointing in which forest the owl was hiding (i.e., left or right). The location of the owl (i.e., correct response) was revealed following the participant's response.

Data were collected using a two-alternative forced-choice (2AFC) paradigm, two-down one-up adaptive procedure to estimate the 70.7% point in the psychometric function (Levitt, 1971). Noise level was fixed at 85 dB SPL (i.e., 60 dB/Hz SPL). The first stimuli were presented at 0 dB SNR. Signal level was adjusted in steps of 4 dB for the first two reversals, then in steps of 2 dB for the remaining six reversals. The mean of the last six reversals was used to estimate an MLD threshold for each trial. Two runs were administered for each condition for most children and adults included in this study. In this case, threshold was determined for N0S0 and N0S$\pi$ conditions by averaging results from each run by condition. Threshold for N0S0 and N0S$\pi$ conditions was determined as the result of one run for each condition for five children with Down syndrome due to test fatigue. The test time for the MLD procedure was approximately 20 minutes, not including breaks or instruction.

## Binaural Intelligibility Level Difference (BILD)

Children heard monosyllabic words presented in noise and they were asked to identify the word they heard by pointing to black and white picture plates that are included with the standard NU-CHIPS test materials. Stimuli consisted of the speech and noise signals recorded through KEMAR as described above. Speech stimuli recorded at 0° azimuth was presented in all conditions. Broadband masking noise recorded from 0°, 45°, and 90° azimuth was presented for the free-field condition and from 45° and 90° azimuth, for the isolated ITD and ILD conditions.

Note that isolated ITD and ILD noise conditions were not necessary at 0° as both cues are expected to be zero at this position. Mean speech reception threshold (SRT) was determined for a total of seven listening conditions (3+2+2=7). These conditions are listed in Table 2.

Table 2

*Listening Conditions Derived from Free-Field Recordings*

| Noise Stimulus | Presentation Azimuth (°) | | Listening Condition |
|---|---|---|---|
| | Signal | Noise | |
| Simulated free-field | 0 | 0 | FF0 |
| Simulated free-field | 0 | 45 | FF45 |
| Simulated free-field | 0 | 90 | FF90 |
| Isolated ILD | 0 | 45 | dL45 |
| Isolated ILD | 0 | 90 | dL90 |
| Isolated ITD | 0 | 45 | dT45 |
| Isolated ITD | 0 | 90 | dT90 |

Data were collected using a single-interval 4AFC, two-down one-up adaptive procedure to estimate the 70.7% point in the psychometric function (Levitt, 1971). Noise was fixed at 85 dB SPL. The spoken token on the first trial was presented at 0 dB SNR. Signal level was adjusted in steps of 4 dB for the first two reversals, then in steps of 2 dB for the remaining six reversals. The mean of the last six reversals was used to estimate SRT. One run was completed per condition and total test time for the BILD lasted approximately 30 minutes, not including breaks or instruction.

## Statistical Analysis

Standard procedures were used to calculate MLD and spatial release from masking for simulated free-field conditions at 45° and 90° azimuth (i.e., BILD FF45 and BILD FF90). That is, MLD was calculated by subtracting N0S$\pi$ threshold from N0S0 threshold. Similarly, BILD FF45 was calculated by subtracting FF45 threshold from FF0 threshold and BILD FF90 was calculated by subtracting FF90 threshold from FF0 threshold. Analyses of variance (ANOVAs) were used to examine overall group differences between conditions. Group differences were examined for children with Down syndrome and typically-developing children as well as adults with Down syndrome and typically-developing adults. Group analyses did not include adolescents with Down syndrome. Levene's tests revealed the assumption of homogeneity of variance was not violated for group comparisons (Levene, 1960).

The impact of chronological age on binaural abilities was examined using regression curves to describe MLD, BILD for simulated free-field conditions (i.e., BILD FF45 and BILD FF90), and SNR for simulated free-field conditions for typically-developing individuals and individuals with Down syndrome. These analyses included the groups noted above as well as adolescents with Down syndrome. The impact of developmental age for children and adolescents with Down syndrome was examined using regression curves that considered age-equivalent scores for the receptive language subdomain of the Vineland Adaptive Behavior Scales. The proportion of variance accounted for, $R^2$, was calculated as well as a $p$-value indicating goodness of fit. In cases in which $p \geq 0.05$, we attempted to apply a linear function. In cases in which $p \leq 0.05$ for more than one curve, $R^2$ indices were compared between curves to determine differences in their ability to describe the association between variables.

Psychometric functions were generated for children and adults with Down syndrome and typically-developing children and adults. The computed slope of these functions provided information about the relative influence of non-sensory factors (e.g., attention). Psychometric functions were normalized by log transformation of SPL values relative to SPL values obtained at threshold (i.e., 70.7%) using the following equation: *log[SPL/ΔSPL(70.7%)]*. The slope of the psychometric function was defined in terms of the *k*-parameter in the following equation: $f(x) = 1 + \frac{a}{1+e^{-(x-\mu)/k}}$, where *a* is the maximum *y*-value, $\mu$ is the midpoint of the function, and *k* is the slope parameter.

Results from one-way ANOVAs suggested that children with a history of otitis media did not have scores that differed significantly from children without a history of otitis media within their respective groups (i.e., children with Down syndrome and typically-developing children) for any measure. Therefore, otitis media was not considered as a separate predictor of performance on the experimental tasks. However, a post hoc power analysis revealed risk of a Type II error (i.e., $[1 - \beta] < .80$) for all calculations indicating that additional children with and without histories of otitis media would need to be included in this study to avoid the potential for Type II error (Cohen, 1992; Table 3).

Table 3

*Statistical Power for Analyses of Variance (ANOVAs) Comparing Mean Scores for Children with and without Histories of Otitis Media*

| Condition | Children with DS | TD Children |
|---|---|---|
| | Power ($1 - \beta$) | |
| N0S0 Threshold | .09 | .12 |
| N0S$\pi$ Threshold | .07 | .12 |
| MLD | .16 | .06 |
| FF0 Threshold | .18 | .05 |
| FF45 Threshold | .10 | .10 |
| FF90 Threshold | .25 | .13 |
| BILD FF45 | .35 | .41 |
| BILD FF90 | .10 | .07 |

*Note*. TD = typically-developing; DS = Down syndrome.

Statistical analyses were performed using SigmaPlot® (San Jose, CA) and IBM® SPSS® (Armonk, NY). All $p$ values were two-sided and significance was at the 0.05 level. Cohen's $d$ was used as a measure of effect size and large effect sizes were defined as ≥0.40 (Cohen, 1992). Fidelity of data entry was verified prior to data analysis by a trained undergraduate research assistant.

# CHAPTER IV

# RESULTS

## Masking Level Difference (MLD)

Individual thresholds for the N0S0 condition and the N0S$\pi$ condition are shown in Figure 5, plotted as a function of chronological age.

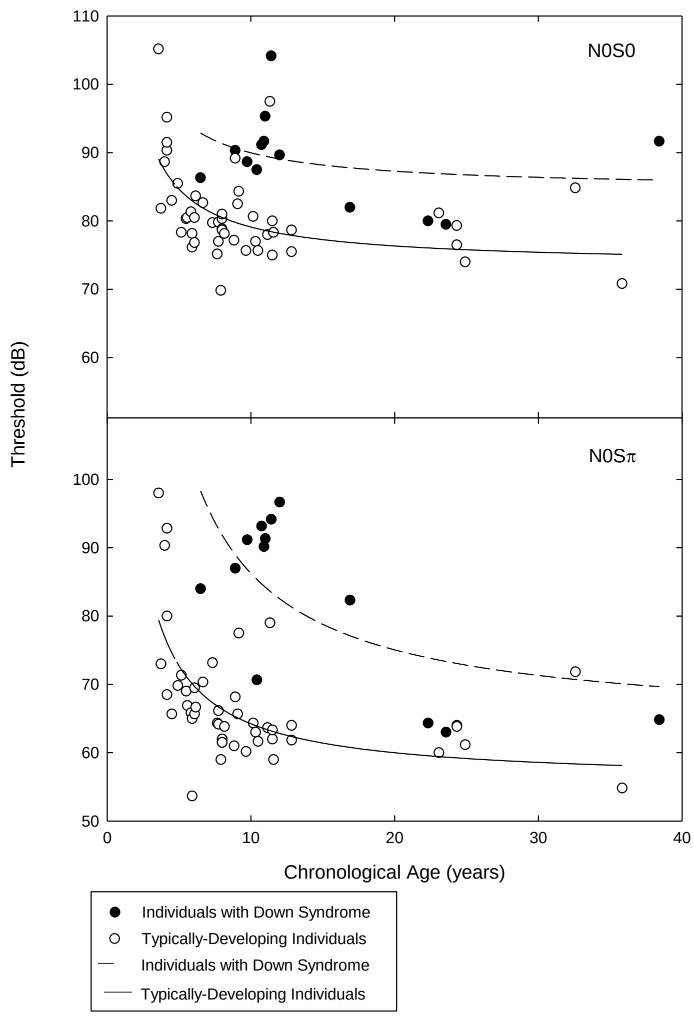


*Figure 5.* N0S0 and N0Sπ threshold for typically-developing individuals and individuals with Down syndrome.

When averaged across chronological age, children with Down syndrome had greater average thresholds for the N0S0 condition and the N0S$\pi$ condition than typically-developing children ($F[1,54]$=4.62, $p \leq 0.05$, $d$=1.70 and $F[1,54]$=15.32, $p \leq 0.05$, $d$=2.51, respectively). Inverse 1st order curves were fit to the variables chronological age and absolute threshold for both stimulus conditions for typically-developing individuals and the N0S$\pi$ condition for individuals with Down syndrome (N0S0, $R^2$=0.29, $p \leq 0.05$; N0S$\pi$, $R^2$=0.38, $p \leq 0.05$; and N0S$\pi$, $R^2$=0.37, $p \leq 0.05$, respectively).

Children with Down syndrome had significantly poorer MLD scores averaged across chronological age than typically-developing children ($F[1,54]$=18.90, $p \leq 0.05$, $d$=1.79). A subset of data from typically-developing children aged 3 to 5 years (i.e., the youngest children recruited to participate) were selected to represent the performance of typically-developing children within a narrow age range. Though no statistically significant differences were seen between means for children with Down syndrome ( $\overline{X}$ = 2.94, SD = 7.07) and typically-developing children aged 3 to 5 years ( $\overline{X}$ =10.38, SD=7.85), a large effect size was noted ($F[1,19]$=1.77, $p$=0.20, $d$=1.00) and a post hoc power analysis revealed risk of Type II error for this comparison ($[1 - \beta]$ = .55). Average MLD scores for typically-developing adults and adults with Down syndrome did not differ significantly, though a large effect size was also noted for this comparison ($F[1,8]$=2.17, $p$=0.18, $d$=0.95). A post hoc power analysis was not completed for this comparison as MLDs for adult participants in this study (i.e., with and without Down syndrome) reached expected levels for typically-developing adults. Table 4 includes means and standard deviations for absolute thresholds measured at 500 Hz, threshold for each masking condition (i.e., N0S0 and N0S$\pi$), and MLD.

Table 4

*Absolute Threshold for 500 Hz, Masked Thresholds in N0S0 and N0Sπ Conditions, and MLDs*

| | $\overline{X}$ (SD) | | | |
|---|---|---|---|---|
| | 500 Hz | N0S0 | N0S$\pi$ | MLD (SD) |
| TD Children | -0.27 (4.87) | 81.48 (6.63) | 67.77 (8.96) | 13.71 (4.94) |
| Children with DS | 12.38 (10.07) | 91.65 (5.37) | 88.70 (7.74) | 2.94 (7.07) |
| Adolescents with DS | 7.50 (0.00) | 82.00[a] | 82.33[a] | -0.33[a] |
| TD Adults | -2.08 (3.38) | 77.78 (6.56) | 62.61 (5.62) | 15.17 (3.26) |
| Adults with DS | 2.50 (5.26) | 83.72 (6.88) | 64.06 (0.95) | 19.67 (6.22) |

*Note*. TD = typically-developing; DS = Down syndrome, MLD = masking level difference.
[a]No standard deviation is shown as only one adolescent with Down syndrome successfully completed this task.

Individual MLD scores (i.e., the difference between the N0S0 and N0S$\pi$ thresholds) are shown as a function of chronological age with the 95$^{th}$ percentile range for typically-developing adults (CI=15.17 ± 2.38) in Figure 6.

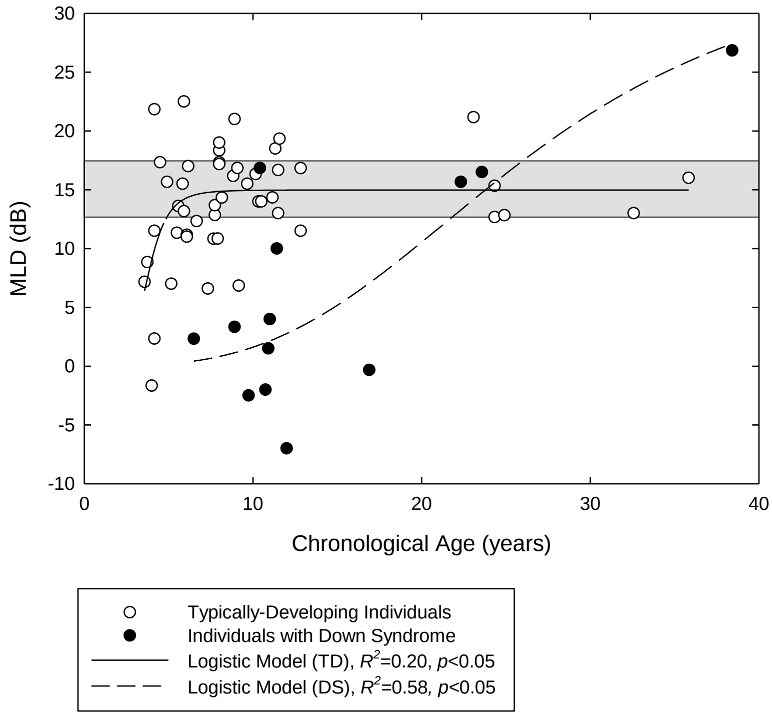


*Figure 6.* Masking level difference (MLD) as a function of chronological age for typically-developing individuals and individuals with Down syndrome. The shaded area represents the 95th percentile for typically-developing adults.

A logistic curve was fit to the variables chronological age and MLD for typically-developing individuals and individuals with Down syndrome ($R^2$=0.20, $p \leq 0.05$ and $R^2$=0.58, $p \leq 0.05$, respectively). The MLD scores of 10% of children and adolescents with Down syndrome and 65% of typically-developing children were within or above the 95th percentile range for typically-developing adults. All adults with Down syndrome were within or above the 95th percentile range for typically-developing adults.

Figure 7 illustrates MLD scores as a function of receptive language age equivalent for children and adolescents with Down syndrome with the 95th percentile range for typically-developing adults (described above). In this case, no logistic equation provided a reasonable fit, so the best-fitting linear function is shown ($R^2$=0.17, $p$=NS).

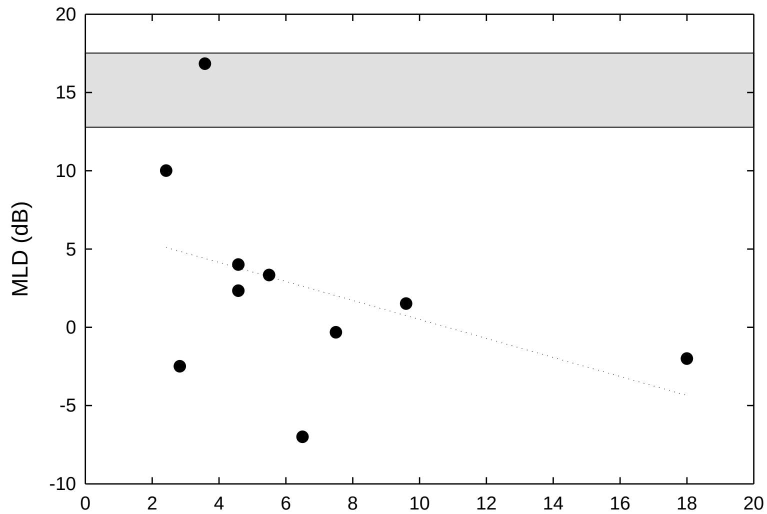


*Figure 7.* Masking level difference (MLD) as a function of receptive language age equivalent for children and adolescents with Down syndrome. The shaded area represents the $95^{th}$ percentile for typically-developing adults.

## Binaural Intelligibility Level Difference (BILD)

### Spatial Release from Masking

Individual thresholds for simulated free-field conditions used for the BILD task (i.e., FF0, FF45, and FF90) are shown in Figure 8, plotted as a function of chronological age.

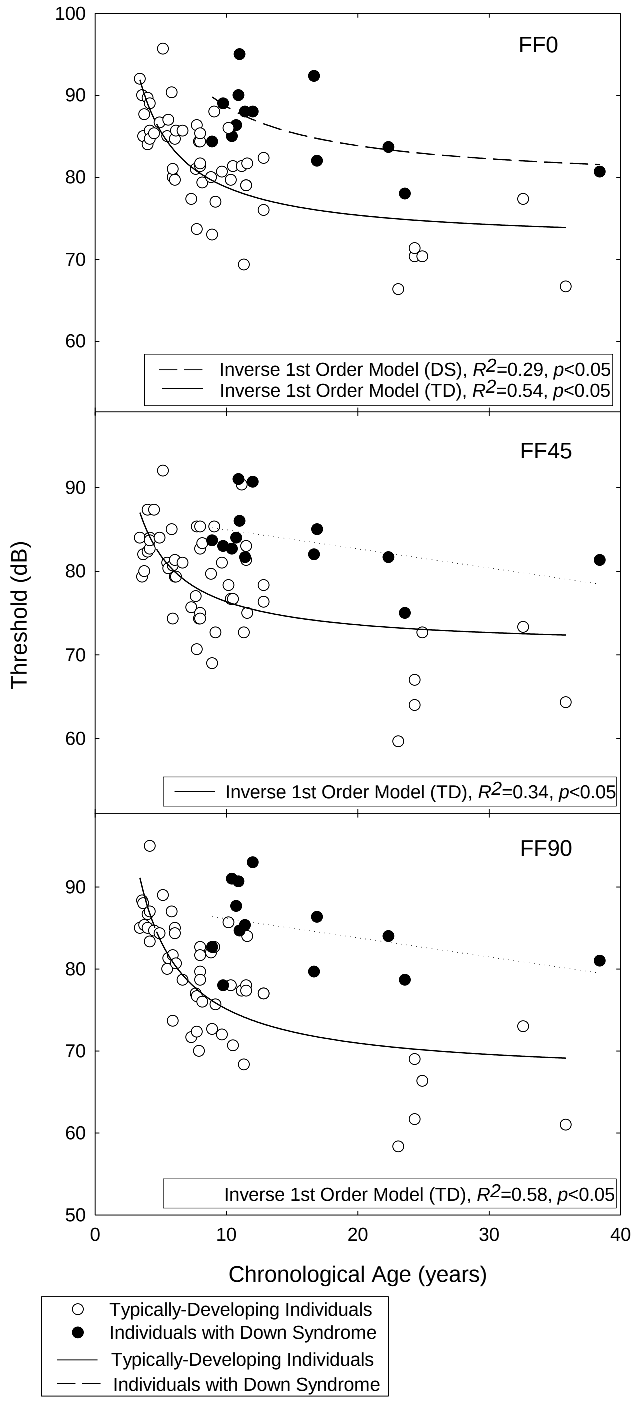


*Figure 8.* FF0, FF45, and FF90 threshold for typically-developing individuals and individuals with Down syndrome.

When averaged across chronological age, individuals with Down syndrome had higher average thresholds than typically-developing individuals for all conditions (FF0: $F[1,64]$=6.04, $p \leq 0.05$, $d$=0.84, FF45: $F[1,64]$=6.73, $p \leq 0.05$, $d$=0.93, and FF90: $F[1,64]$=7.78, $p \leq 0.05$, $d$=1.00). Inverse 1st order curves were fit to the variables chronological age and threshold for all BILD conditions for typically-developing individuals and the FF0 condition for individuals with Down syndrome (FF0, $R^2$=0.54, $p \leq 0.05$; FF45, $R^2$=0.34, $p \leq 0.05$; FF90, $R^2$=0.58, $p \leq 0.05$; and FF0, $R^2$=0.29, $p \leq 0.05$, respectively). No linear or non-linear equations including chronological age and FF45 or FF90 resulted in significant associations for individuals with Down syndrome. For these cases, the best-fitting linear functions are shown ($R^2$=0.21 , $p$=NS, $R^2$=0.16 , $p$=NS, respectively).

BILD FF45 and BILD FF90 scores for children with Down syndrome and typically-developing children averaged across chronological age did not differ by a statistically significant amount. Furthermore, average BILD FF45 scores for adults with Down syndrome and typically-developing adults did not differ by a statistically significant amount, though a large effect size was observed for this comparison ($F[1,8]$=1.01, $p$=0.35, $d$=0.81). In addition, average BILD FF90 scores for adults with Down syndrome were poorer than typically-developing adults ($F[1,8]$=11.07, $p \leq 0.05$, $d$=3.73). Table 5 includes bilateral PTA, thresholds for free-field BILD conditions (i.e., FF0, FF45, and FF90), and BILD for free-field conditions (i.e., BILD FF45 and BILD FF90).

Table 5

*Bilateral PTA, and Masked Thresholds for the Three Free-Field Conditions (FF0, FF45, and FF90), and BILDs for the FF45 and FF90 Conditions*

| | $\overline{X}$ (SD) | | | | | |
|---|---|---|---|---|---|---|
| | PTA | FF0 | FF45 | FF90 | BILD FF45 | BILD FF90 |
| TD Children | 1.10 (3.45) | 83.12 (5.11) | 80.24 (4.97) | 80.41 (5.83) | 2.88 (4.17) | 2.71 (4.28) |
| Children with DS | 11.05 (6.08) | 88.67 (3.43) | 84.96 (3.56) | 85.85 (5.19) | 3.70 (4.47) | 2.81 (6.98) |
| Adolescents with DS | 7.25 (1.77) | 87.17 (7.30) | 83.50 (2.12) | 83.00 (2.12) | 3.67 (9.43) | 4.17 (12.01) |
| TD Adults | 7.83 (4.25) | 70.39 (3.98) | 66.83 (5.33) | 64.89 (5.54) | 3.55 (3.29) | 5.50 (2.98) |
| Adults with DS | 1.33 (2.04) | 80.78 (2.84) | 79.33 (3.76) | 81.22 (2.67) | 1.45 (1.89) | -0.44 (0.20) |

*Note.* PTA = pure tone average; BILD = binaural intelligibility level difference, TD = typically-developing; DS = Down syndrome.

Individual BILD FF45 and BILD FF90 scores (i.e., the differences between thresholds in these conditions and in the reference FF0 condition) are shown as a function of chronological age in Figure 9 with the 95$^{th}$ percentile range for typically-developing adults (CI=3.55 ± 4.26 and CI=5.50 ± 2.59, respectively). No logistic equation provided a reasonable fit for the variables chronological age and BILD FF45 or chronological age and BILD FF90 for typically-developing individuals or individuals with Down syndrome, so the best-fitting linear function is shown ($R^2$=0.00, $p$=NS, $R^2$=0.22 , $p$=NS, $R^2$=0.04 , $p$=NS, $R^2$=0.02 , $p$=NS, respectively).

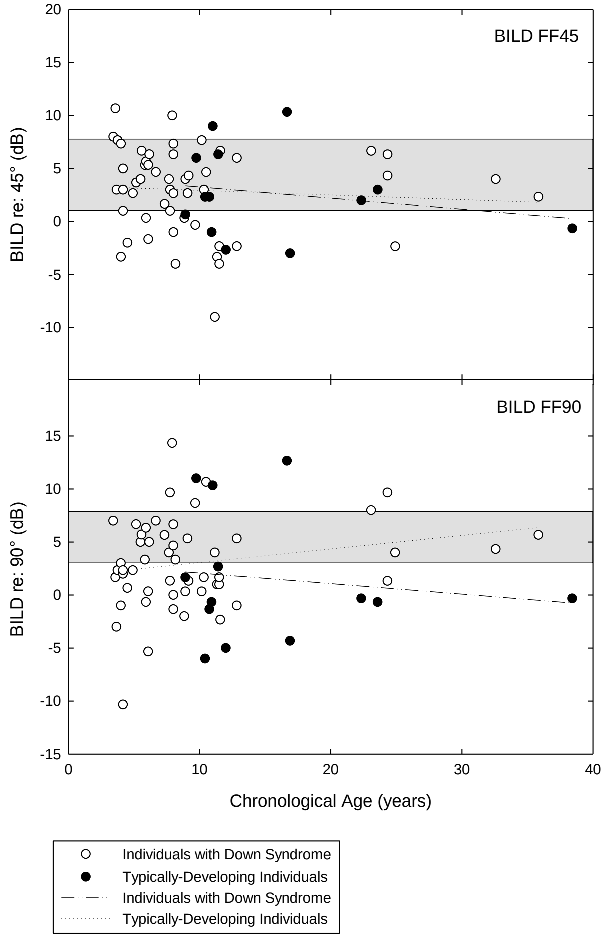


*Figure 9.* Binaural intelligibility level difference (BILD) obtained for simulated free-field conditions at 90° (BILD FF90) and 45° (BILD FF45) as a function of chronological age for typically-developing individuals and individuals with Down syndrome.

The BILD FF45 scores of 67% of children and adolescents with Down syndrome and 72% of typically-developing children were within or above the 95th percentile range for typically-developing adults. Likewise, the BILD FF90 scores of 11% of children and adolescents with Down syndrome and 46% of typically-developing children were within or above the 95th percentile range for typically-developing adults. Scores of 67% adults with Down syndrome were within or above the 95th percentile range for typically-developing adults for the

BILD FF45 condition. However, no adults with Down syndrome had scores that were within or above this range for the BILD FF90 condition.

Figure 10 illustrates BILD FF45 and BILD FF90 scores as a function of receptive language age equivalent for children and adolescents with Down syndrome with the 95th percentile range for typically-developing adults (described above). Non-linear equations including the variables receptive language age equivalent and BILD FF45 or BILD FF90 did not result in significant associations for children and adolescents with Down syndrome, so the best-fitting linear function is shown ($R^2$=0.05, $p$=NS, $R^2$=0.04 , $p$=NS, respectively).

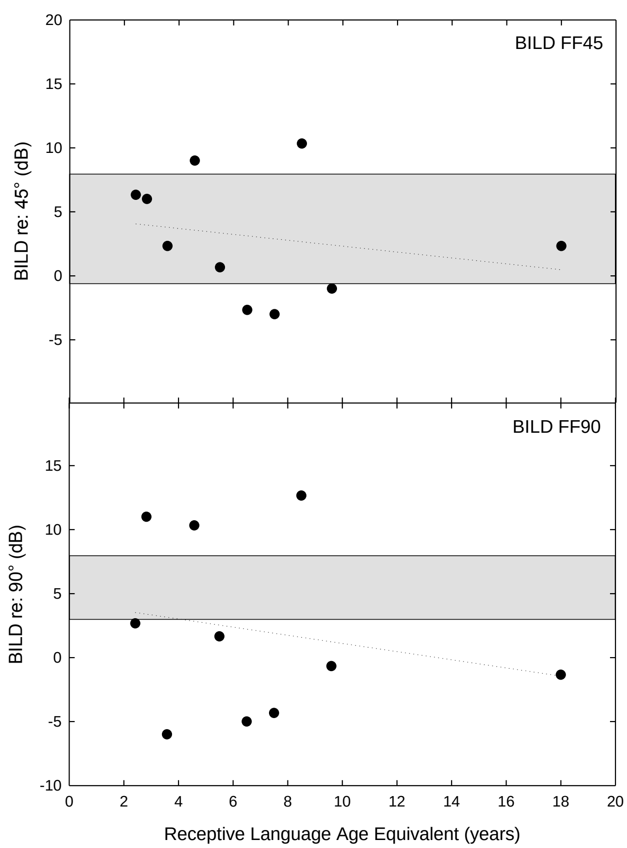


*Figure 10.* Binaural intelligibility level difference (BILD) obtained for simulated free-field conditions at 90° and 45° (i.e., BILD FF90 and BILD FF45, respectively) as a function of receptive language age equivalent for children and adolescents with Down syndrome.

Association between MLD and BILD

The association between MLD and BILD for simulated free-field conditions (i.e., BILD FF45 and BILD FF90) was examined using Pearson correlations.  Figure 11 shows a scatterplot with MLD for individual participants plotted versus BILD FF90.

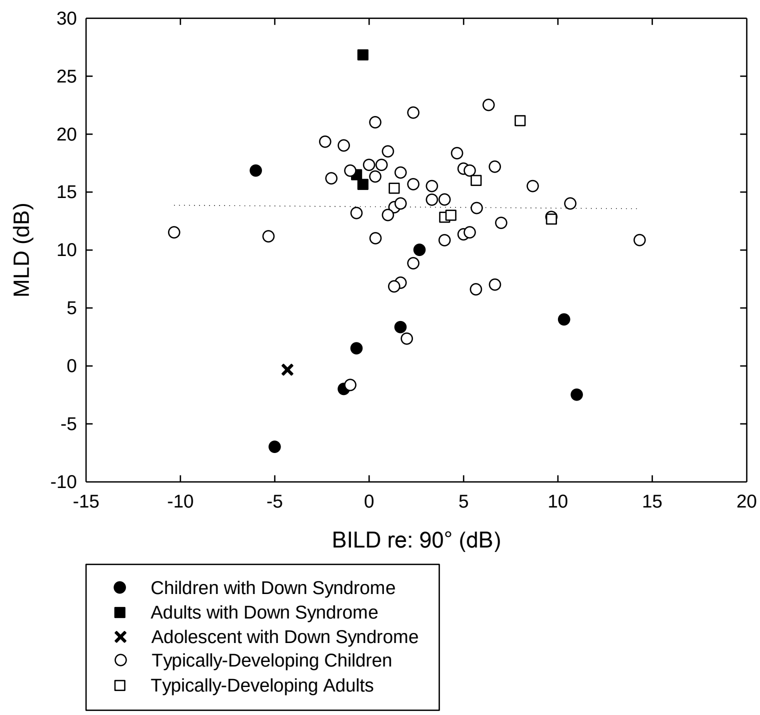


*Figure 11.* Scatterplot showing masking level difference (MLD) scores for individual participants versus binaural intelligibility level difference obtained for the 90° simulated free-field condition (BILD FF90).

Associations were not observed between MLD and BILD FF45 or between MLD and BILD FF90 when data from all participants were included in a single correlation analysis ($R^2$=0.00, $p$=NS, $R^2$=0.00, $p$=NS, respectively), nor when within-group correlations were performed for children with Down syndrome ($R^2$=0.33, $p$=NS, $R^2$=-0.17, $p$=NS, respectively), typically-developing children ($R^2$=-0.16, $p$=NS, $R^2$=0.01, $p$=NS, respectively), adults with Down syndrome ($R^2$=-0.95, $p$=NS, $R^2$=0.44, $p$=NS, respectively), or typically-developing adults ($R^2$=0.54, $p$=NS, $R^2$=0.21, $p$=NS, respectively).

Signal-to-Noise Ratio (SNR)

Figure 12 illustrates SNR thresholds as a function of chronological age for all simulated free-field conditions (i.e., FF0, FF45, and FF90).

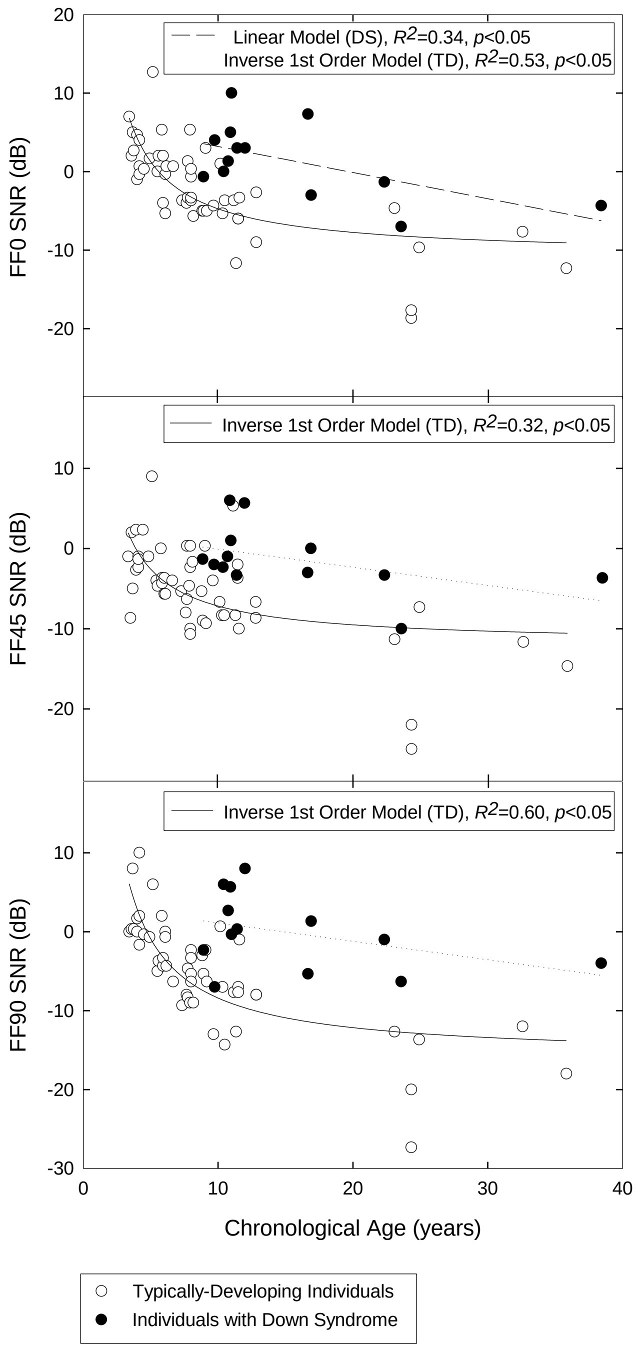


*Figure 12.* Signal-to-noise-ratio (SNR) threshold for simulated free-field conditions FF0, FF45, and FF90 as a function of chronological age for typically-developing individuals and individuals with Down syndrome. TD = typically-developing; DS = Down syndrome.

An inverse 1st order curve was fit to the variables chronological age and SNR threshold for all simulated free-field conditions for typically-developing individuals (i.e., FF0, FF45, and

FF90; $R^2$=0.53, $p \leq 0.05$, $R^2$=0.32, $p \leq 0.05$, $R^2$=0.60, $p \leq 0.05$, respectively). A linear model was fit to the variables chronological age and SNR threshold at FF0 for individuals with Down syndrome ($R^2$=0.34, $p \leq 0.05$). No linear or non-linear equations including chronological age and SNR threshold for FF45 or FF90 resulted in significant associations for individuals with Down syndrome, however the best-fitting linear functions are shown ($R^2$=0.21 , $p$=NS, $R^2$=0.16 , $p$=NS, respectively).

Figure 13 illustrates SNR threshold as a function of receptive language age equivalent for all simulated free-field conditions for children with Down syndrome.

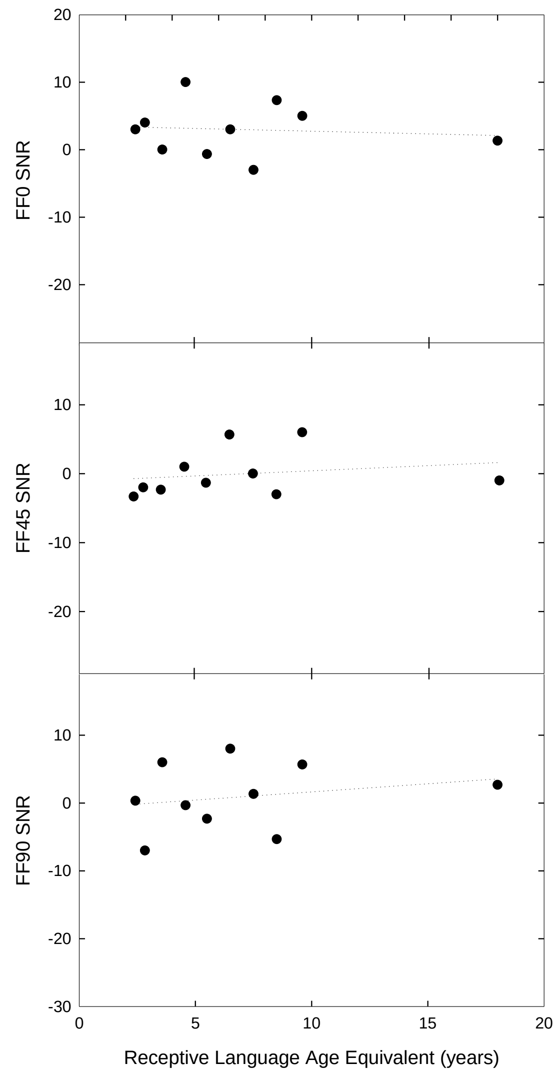


*Figure 13.* Signal-to-noise-ratio (SNR) threshold for simulated free-field conditions FF0, FF45, and FF90 as a function of receptive language age equivalent for children and adolescents with Down syndrome.

Non-linear equations including receptive language age equivalent and SNR threshold at FF0, FF45 or FF90 did not reveal significant associations for children with Down syndrome. As such, the best-fitting linear functions are shown ($R^2$=0.01, $p$=NS, $R^2$=0.04, $p$=NS, $R^2$=0.05, $p$=NS, respectively).

Figure 14 illustrates SNR thresholds as a function of azimuth for simulated free-field conditions for each group.

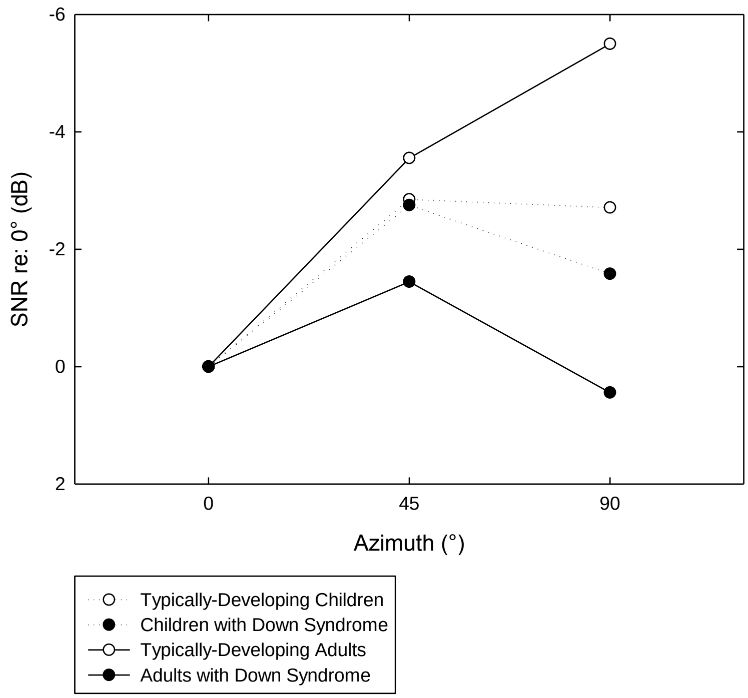


*Figure 14.* Signal-to-noise-ratio (SNR) threshold referenced to SNR at 0° as a function of azimuth for simulated free-field conditions for typically-developing children, children with Down syndrome, typically-developing adults, and adults with Down syndrome.

Within-group differences in SNR thresholds across conditions were analyzed to determine the effect of spatial separation of signal and noise on speech recognition. Differences for SNR thresholds obtained at FF0 and FF45 were observed for typically-developing children and typically-developing adults ($F[1,45]$=21.93, $p \leq 0.05$, $d$=0.66 and $F[1,5]$=6.97, $p \leq 0.05$, $d$=0.57, respectively). In addition, differences were observed for SNR thresholds obtained at FF0 and FF90 for typically-developing children and typically-developing adults ($F[1,45]$=18.42, $p \leq 0.05$, $d$=0.56, and $F[1,5]$=20.34, $p \leq 0.05$, $d$=0.96, respectively). No differences between SNR thresholds obtained at FF0 and FF45 were seen for children with Down syndrome or adults with

Down syndrome, though large effect sizes were noted ($F[1,7]$=4.18, $p$=0.08, $d$=0.82 and $F[1,2]$=1.75, $p$=0.32, $d$=0.44). In addition, a post hoc power analysis revealed risk of Type II error for comparisons of SNR FF0 and FF45 for children with Down syndrome and adults with Down syndrome ($[1 - \beta] = .26$ and .04, respectively). Statistically significant differences in SNR thresholds obtained at FF0 and FF90 were not observed for children with Down syndrome or adults with Down syndrome.

Location-specific SNR thresholds were calculated to simplify characterization of performance at a particular simulated source azimuth. That is, SNR threshold for FF45, dL45, and dT45 were averaged to generate SNR 45, an estimate of ability at this azimuth. Likewise, SNR thresholds for FF90, dL90, and dT90 were averaged to generate SNR 90. The performance of individuals with Down syndrome relative to typically-developing individuals was of interest. In this case, subsets of data from typically-developing children aged 3 to 5 years and 10 to 13 years (i.e., the youngest and oldest children recruited to participate) were selected to represent performance of typically-developing children within a narrow age range and used to provide an estimate of the relative performance of children and adults with Down syndrome.

Figure 15 shows SNR threshold for FF0, SNR 45, and SNR 90 for typically-developing children aged 3 to 5 years, typically-developing children aged 10 to13 years, children with Down syndrome, typically-developing adults, and adults with Down syndrome.

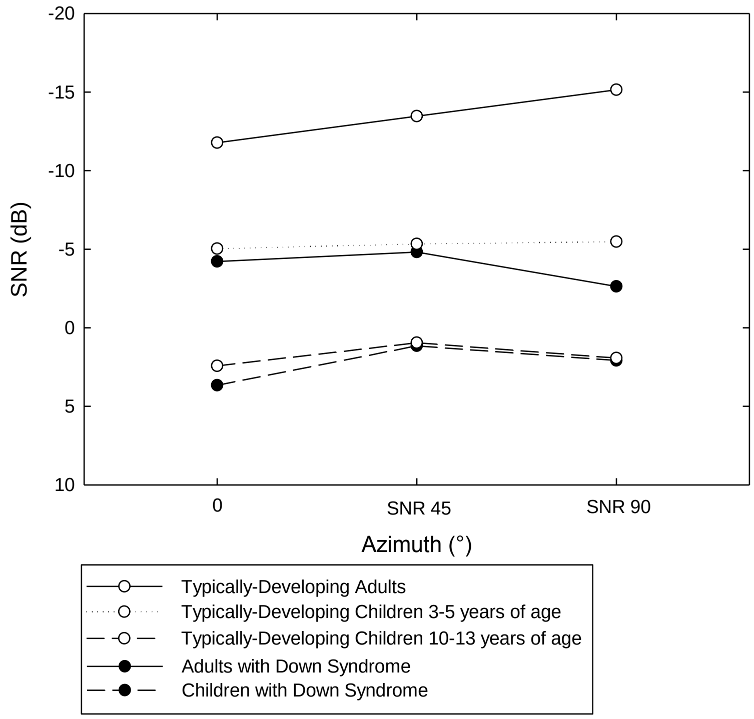


*Figure 15.* Signal-to-noise-ratio (SNR) threshold for FF0, SNR 45, and SNR 90 for typically-developing children aged 3-5 years, typically-developing children aged 10-13 years, children with Down syndrome, typically-developing adults, and adults with Down syndrome.

Adults with Down syndrome had poorer SNR thresholds than typically-developing adults for FF0, SNR 45, and SNR 90 ($F[1,8]$=4.96, $p \leq 0.05$, $d$=1.80, $F[1,8]$=5.76, $p \leq 0.05$, $d$=2.05, and $F[1,8]$=16.46, $p \leq 0.05$, $d$=3.46, respectively). However, no statistically significant differences were observed between the performance of adults with Down syndrome and typically-developing children aged 10 to13 years for FF0, SNR 45, and SNR 90, though a large effect size was observed for SNR 90 ($F[1,12]$=2.82 , $p$=0.12, $d$=1.16).

Children with Down syndrome had poorer SNR thresholds than typically-developing children aged 10 to 13 years for SNR FF0, SNR 45, and SNR 90 ($F[1,17]$=25.21, $p \leq 0.05$, $d$=1.88, $F[1,17]$=24.61, $p \leq 0.05$, $d$=2.92, and $F[1,17]$=19.73, $p \leq 0.05$, $d$=2.94, respectively). However, a statistically significant difference was not observed in the SNRs of children with

Down syndrome and typically-developing children aged 3 to 5 years for the same conditions (i.e., FF0, SNR 45, and SNR 90).

Effect of Cue Isolation

Recall that in addition to creating signals to simulate free-field listening conditions, noise signals recorded at 45°and 90° were processed to isolate ILD cues (i.e., ITD was equal to 0 μs for all presentations) and ITD cues (i..e., ILD was equal to 0 dB for all presentations). As such, four isolated cue conditions were created including isolated ILD cues for each azimuth (i.e., dL45 and dL90) and isolated ITD cues for each azimuth (i.e., dT45 and dT90). Figure 16 illustrates average SNR threshold for each group as a function of azimuth for simulated free-field and isolated cue conditions.

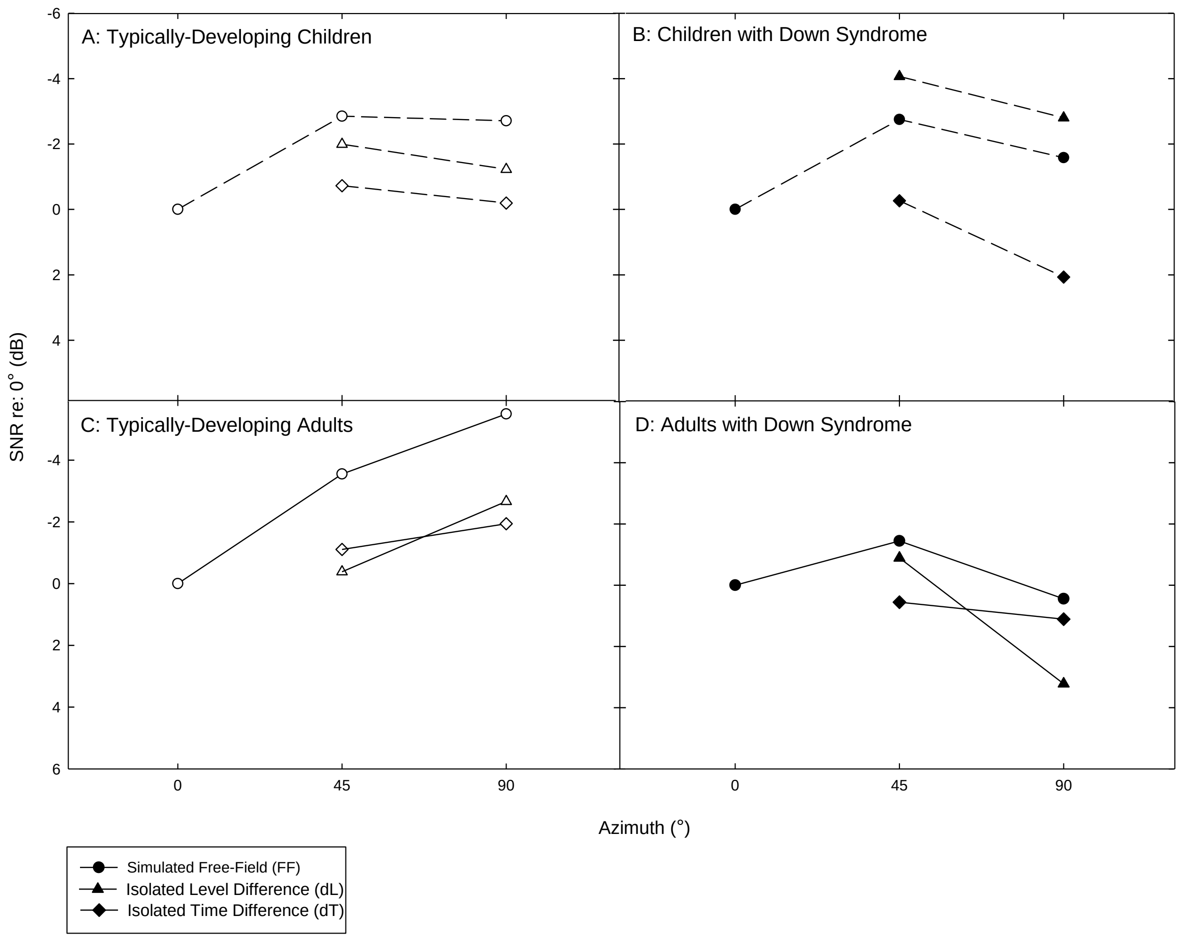


*Figure 16.* Signal-to-noise-ratio (SNR) threshold referenced to SNR at 0° for each group as a function of azimuth for simulated free-field and isolated cue conditions for (A) typically-developing children, (B) children with Down syndrome, (C) typically-developing adults, and (D) adults with Down syndrome.

Differences in SNR threshold were analyzed by condition for each group to determine the relative influence of ITDs and ILDs.

*Isolated Interaural Time Difference (ITD).* As shown in Figure 16A, performance for typically-developing children was poorer for the isolated ITD cue condition at 45° (i.e., dT45) than performance when all cues were present in the signal at this azimuth (i.e., FF45;

$F[1,45]=7.67$, $p \leq 0.05$, $d=0.44$). Statistically significant differences were not observed between SNR thresholds obtained at FF45 and dT45 for typically-developing adults (Figure 16C), children with Down syndrome (Figure 16B), or adults with Down syndrome (Figure 16D), though large effect sizes were noted for each group ($F[1,5]=4.56$, $p=0.06$, $d=0.40$, $F[1,4]=3.68$, $p=.13$, $d=0.52$, and $F[1,2]=0.60$ , $p=0.52$ , $d=0.77$ , respectively). Performance for typically-developing children and typically-developing adults (Figure 16C) was poorer for the isolated ITD cue at 90° (i.e., dT90) than performance when all cues were present at this azimuth (i.e., FF90; $F[1,45]=11.31$, $p \leq 0.05$, $d=0.49$, and $F[1,5]=7.24$, $p \leq 0.05$, $d=0.61$, respectively). Differences in SNR thresholds obtained at FF90 and dT90 were not observed for children with Down syndrome or adults with Down syndrome, but a large effect size was noted for children with Down syndrome ($F[1,4]=2.04$ , $p=0.23$ , $d=0.72$).

*Isolated Interaural Level Difference (ILD).* As seen in Figures 16A, 16B, and 16D, statistically significant differences in SNR threshold were not observed for typically-developing children, children with Down syndrome, or adults with Down syndrome between the isolated ILD cue condition at 45° (i.e., dL45) and performance when all cues were present at this azimuth (i.e., FF45), though a large effect size was noted for children with Down syndrome ($F[1,4]=0.00$, $p=0.98$ , $d=0.47$). Typically-developing adults had poorer performance for dL45 compared to FF45; ($F[1,5]=6.27$, $p \leq 0.05$, $d=0.40$; Figure 16C). Statistically significant differences were not observed for the isolated ILD cue at 90° (i.e., dL90) compared to performance when all cues were present at this azimuth (i.e., FF90) for any of the participant groups, though large effect sizes were noted for all but the typically-developing children: typically-developing adults, ($F[1,5]=1.12$ , $p=0.34$, $d=0.46$), children with Down syndrome ($F[1,4]=0.49$, $p=0.53$, $d=0.47$), and adults with Down syndrome ($F[1,2]=8.23$, $p=0.10$, $d=0.79$).

The relative impact of ILD cues (i.e. head shadow) on speech recognition performance was examined by calculating the difference between SNR for simulated free-field and isolated ITD conditions. This provided an estimate of the relative influence of ILD cues on speech recognition performance. Variables were created to represent the contribution of ILDs to free-field performance at each azimuth. Specifically, the contribution of ILDs to free-field performance was represented at 45° by subtracting dT45 from FF45. In addition, the contribution of ILDs to free-field performance was represented at 90° by subtracting dT90 from FF90. Differences in the relative influence of ILD cues were not observed between children with Down syndrome or typically-developing children at 45° or 90°.

## Psychometric Functions

The group psychometric functions for MLD conditions (i.e., N0S0 and N0S$\pi$) and selected BILD simulated free-field conditions (i.e., FF0 and FF90) are shown in Figures 17 and 18, respectively.

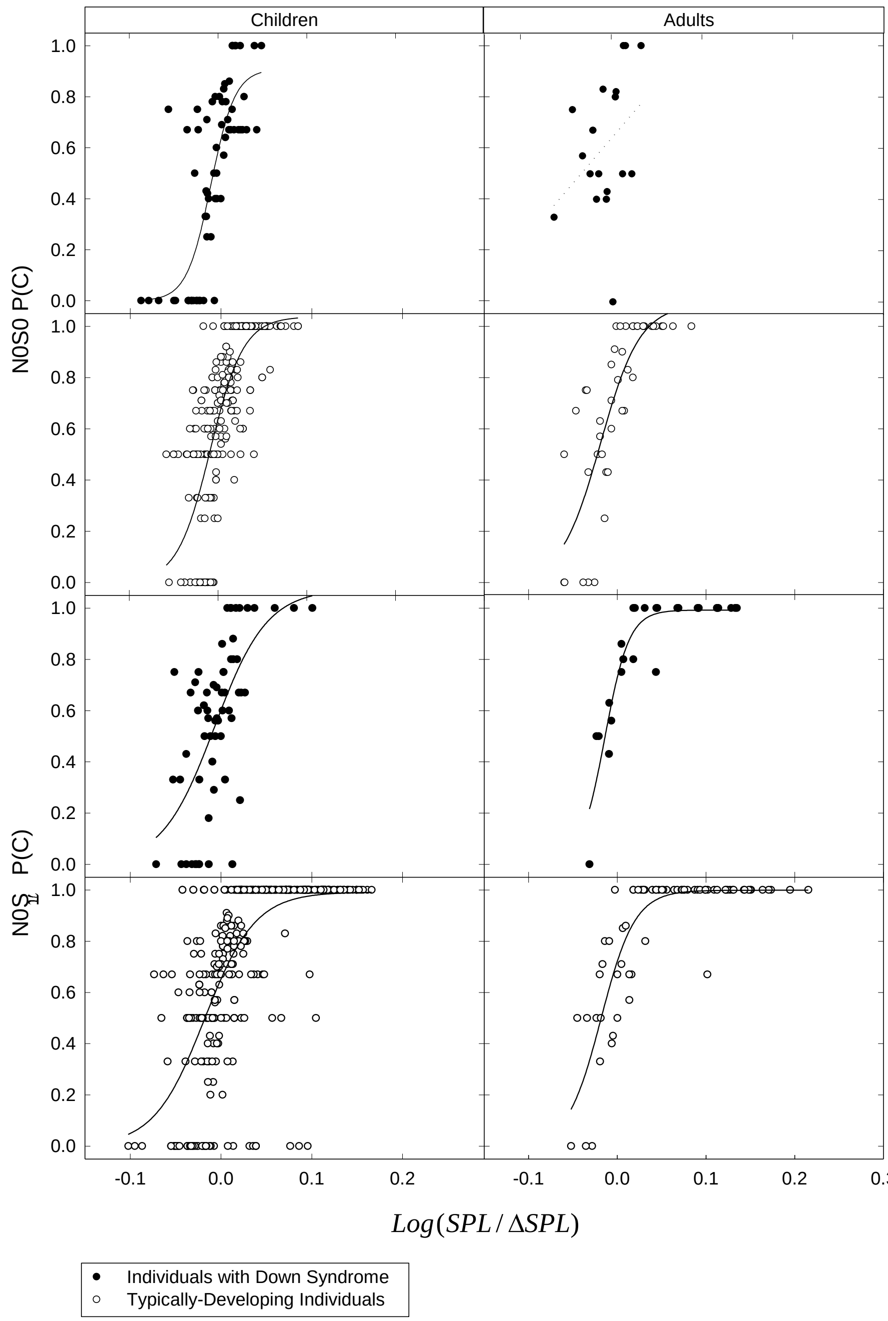


*Figure 17.* Percent correct as a function of log[SPL/ΔSPL(70.7%)] for N0S0 and N0Sπ conditions for children and adults with Down syndrome and typically-developing children and adults.

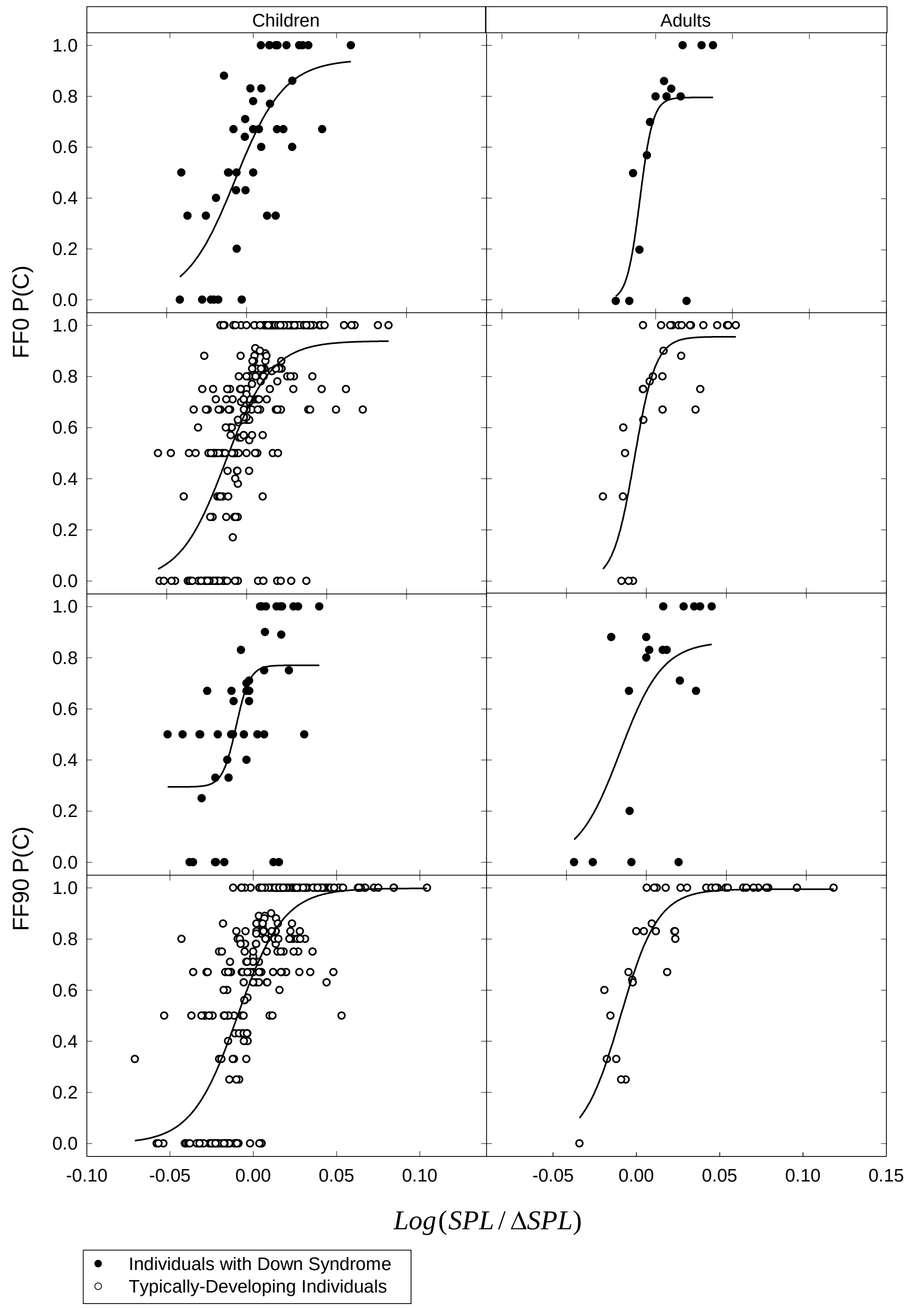


*Figure 18.* Percent correct as a function of log[SPL/ΔSPL(70.7%)] for FF0 and FF90 conditions for children and adults with Down syndrome and typically-developing children and adults.

The individual slopes of psychometric functions of children with Down syndrome and typically-developing children were similar for N0S0, N0S$\pi$, FF0, and FF90 conditions suggesting that that attentiveness and motivation contributed equally to the performance of both groups of children for these tasks (*F[1,35]*=0.00, *p*=NS, *F[1,32]*=0.26, *p*=NS, *F[1,33]*=0.10,

$p$=NS, and $F[1,37]$=0.06, $p$=NS, respectively). Similarly, the individual slopes of the psychometric functions of adults with Down syndrome and typically-developing adults did not differ for N0S0, N0S$\pi$, FF0, and FF90 conditions ($F[1,4]$=1.99, $p$=NS, $F[1,5]$=0.18, $p$=NS, $F[1,4]$=0.60, $p$=NS, and $F[1,7]$=0.32, $p$=NS, respectively). It should be noted that no logistic function was found to fit group data for the N0S0 condition for adults with Down syndrome. As such, the best fitting linear function is shown for this condition.

# CHAPTER V

# DISCUSSION

Our results align well with past studies that have examined MLD for typically-developing adults and children and BILD for typically-developing adults (e.g., Bronkhorst & Plomp, 1988; Hall & Grose, 1990a; Hirsh, 1948; Moore et al., 2011). Specifically, average MLD for typically-developing adults in our study was 15.17 dB, which is consistent with past results of MLDs of 15 dB for adult listeners with normal hearing for low frequency pure tone signals (Hirsh, 1948). Also similar to past research, larger MLDs were seen with increasing age with an asymptote in performance noted at about 5 to 6 years of age for typically-developing children in our study (Grose et al., 1997; Hall & Grose, 1990a; Moore et al., 2011). Furthermore, average SNR threshold for the BILD simulated free-field condition at 90° (i.e., FF90) was -17.28 dB for typically-developing adults in our study, similar to the average SNR threshold of -16.6 dB obtained by Bronkhorst and Plomp (1988) for this condition. Though Bronkhorst and Plomp did not include a simulated free-field condition at 45°, the average of reported SNR thresholds for 30° and 60° was -14.3 dB, similar to our average SNR threshold result of -15.33 dB for this condition. Similarities between results from our study and past results validate our research paradigm. However, some differences were noted between results from our study and Bronkhorst and Plomp (1988). For example, average SNR threshold for the simulated free-field condition at 0° was -11.78 dB for the typically-developing adults in our study, which represents better speech recognition in noise than the -6.4 dB reported for this condition by Bronkhorst and Plomp (1988).

Results of the present study show a marked difference in the binaural abilities of individuals with Down syndrome compared to their typically-developing peers. First, children with Down syndrome included in this study had higher thresholds than typically-developing children for all experimental conditions in both the detection task and the speech recognition task (i.e., N0S0, N0S$\pi$, FF0, FF45, and FF90; see Figures 5 and 8 and Tables 3 and 4). Similarly, individuals with Down syndrome had higher thresholds than typically-developing individuals for the speech recognition conditions FF0, FF45, and FF90. Despite normal hearing sensitivity, individuals with Down syndrome in this study had higher thresholds for detection of signals in noise than typically-developing individuals. Second, individuals with Down syndrome experienced less binaural benefit than typically-developing individuals for both experimental paradigms; both MLDs and BILDs were smaller for those with Down syndrome relative to typically-developing individuals. This suggests that cues resulting in release from masking for typically-developing individuals, such as ITD cues and spatial separation of signals and noise, benefit individuals with Down syndrome to a lesser extent. These differences likely occur due to sensory influences, such as auditory filter width and resultant frequency resolution capabilities, and non-sensory influences, such as attention and motivation. The following discussion elaborates on each of these issues.

## Sensory Influences

### Signal Detection in Noise

*Frequency Resolution.* It is possible that the higher absolute thresholds for the MLD task conditions (i.e., N0S0 and N0S$\pi$) observed for children with Down syndrome in this study were the result of poor frequency resolution capabilities. An individual's physiologic capability to distinguish one frequency from another has direct implications for their ability to distinguish signals from noise. Frequency resolution abilities measured from the cochlea and the brainstem using physiologic techniques (e.g., otoacoustic emissions and ABR testing) and those measured behaviorally have been shown to be mature in typically-developing infants by about 6 months of age (Abdala & Folsom, 1995; Abdala & Sininger, 1996; Folsom & Wynne, 1987; Spetner & Olsho, 1990). Immature frequency resolution in typically-developing infants has been attributed to middle-ear, cochlear, and neural factors such as synaptic efficiency, myelination, and dendritic architecture (Abdala & Folsom, 1995; Abdala, Keefe, & Oba, 2007). As previously discussed, individuals with Down syndrome are affected by anatomic differences of the middle ear, cochlea, and neuronal structure and function. As such, it is reasonable to suspect that their frequency resolution capabilities may be compromised as a result. Our data suggest there is an improvement in the ability to detect signals in noise for children with Down syndrome with increasing age (Figure 5). This suggests immaturities in the frequency resolution of children with Down syndrome relative to typically-developing children, not an absolute deficit that persists into adulthood.

*Temporal Resolution*. Temporal resolution refers to the ability to hear changes in a sound over time and is another likely contributor to the relatively elevated thresholds exhibited by individuals with Down syndrome for signals presented in noise. It includes the ability to detect gaps between stimuli as well as the ability to identify changes in signal amplitude and has long been associated with speech understanding (S. Greenberg, 1996; Pisoni, 1977). Furthermore, temporal resolution is thought to contribute to improved thresholds when listening in noise (Hall et al., 2007; Peters, Moore, & Baer, 1998). Similar to frequency resolution, temporal resolution develops in infancy and early childhood for typically-developing individuals (Hall & Grose, 1994; Whightman, Prudence, Dolan, Kistler, & Jamieson, 1989). Changes in temporal resolution ability can be attributed to the development of physiologic aspects within the auditory system (He, Hotson, & Trainor, 2007). As such, delayed physiologic development might contribute to the differences observed between individuals with Down syndrome and their typically-developing peers. Indeed, when thresholds in noise and SNR thresholds are plotted as a function of chronological age for experimental conditions within the BILD task, we see an improvement in the ability to detect signals in noise with age for individuals with Down syndrome for the FF0 condition and typically-developing individuals for all simulated free-field experimental conditions (i.e., FF0, FF45, and FF90; Figure 12).

## Binaural Release from Masking

*Synaptic Transmission.* Recall that binaural hearing is thought to be facilitated within the central auditory system by comparison of timing information from the two ears as a function of characteristic frequency after processing by the peripheral auditory system. Accurate coding of temporal information within the peripheral auditory system and sensitivity to temporal differences between ears within the central auditory system is essential for binaural hearing to occur. This system relies on synaptic transmission. Electrophysiologic test paradigms, like ABR testing, can be considered an indirect measure of collective neural synchrony and synaptic transmission (Ponton, Moore, & Eggermont, 1996). As previously discussed, compared to typically-developing individuals, individuals with Down syndrome have delayed late-latency auditory evoked potentials (i.e., greater than approximately 50 ms) and accelerated early-latency auditory evoked potentials (i.e., ABR waveforms). Known deficits in the structure and function of myelin for individuals with Down syndrome likely impacts synaptic transmission and electrophysiologic testing as a result.

Electrophysiologic test results (i.e., ABR) have been associated with MLDs for typically-developing children and adults with neurologic impairments (Hall & Grose, 1993; Hannley, Jerger, & Rivera, 1983; Noffsinger, Martinez, & Schaefer, 1982). For example, smaller MLDs have been associated with poor integrity of ABR waves I, II, and III (Hannley et al., 1983; Noffsinger et al., 1982). Furthermore, large interaural asymmetries of interpeak latencies I-III and I-V have been associated with smaller MLDs (Hall & Grose, 1993). These results suggest that smaller MLDs are related to anomalous brainstem processing. Considering differences noted in previous research between ABR test results for individuals with Down syndrome and typically-developing individuals, it is reasonable to suspect that the smaller MLDs observed for children in this study could be the result of aberrant synaptic transmission and brainstem processing of auditory signals.

Spatial Release from Masking

*Isolated ILD Cue Use.* Recall that we examined the relative impact of ILD cues (i.e., head shadow) by calculating the difference between SNR for simulated free-field and isolated ITD conditions. We anticipated that children with Down syndrome would show greater reliance on ILD cues than typically-developing children suggesting poor utilization of binaural cues (i.e., ITD cues). However, children with Down syndrome and typically-developing children had similar performance when the relative impact of ILD cues was assessed. Though the similar relative utilization of ILD cues observed for these groups could support that children with Down syndrome have similar binaural abilities to typically-developing children, MLD results do not corroborate this conclusion. Rather, this finding is likely influenced by our experimental paradigm, which is discussed in a later section of this paper.

*Signal-to-Noise Ratio (SNR).* Additional support for developmental improvements in the ability to detect signals in noise was apparent in our finding that SNR threshold for the FF0 condition showed a gradual decrease as a function of age for typically-developing individuals and for individuals with Down syndrome (Figure 12). In fact, our results suggest developmental improvements for speech recognition in noise for typically-developing individuals in a variety of listening conditions and for individuals with Down syndrome when the signal and noise originate from similar spatial locations. Spatial separation of signals and noise resulted in improved SNR threshold for typically-developing adults and typically-developing children. Effect sizes for children with Down syndrome and adults with Down syndrome were large and a post hoc power analysis revealed risk of Type II error for comparisons of SNR at FF0 and FF45. As such, it is likely that including more participants with Down syndrome in the analysis would indicate a pattern of results consistent with changes in SNR with separation of signals and noise.

*Relative Performance.* Children with Down syndrome included in this study were aged 6 to 11 years, but their collective performance mirrored that of typically-developing children aged 3 to 5 years. That is, as a group, the ability of children with Down syndrome to take advantage of spatial release from masking was similar to that of typically-developing children aged 3 to 5 years. Similar to typically-developing individuals, this ability likely improves over time for individuals with Down syndrome. However, results from adults with Down syndrome included in this study are similar to that of typically-developing children aged 10 to 13 years. Though the performance of children with Down syndrome can be expected to improve with development, the spatial release from masking experienced by adults with Down syndrome remained poorer than that experienced by typically-developing adults.

Interestingly, this pattern of results was not observed for the MLD task. Differences in MLD between children with Down syndrome and typically-developing children were observed, but results from typically-developing adults and adults with Down syndrome were similar. This could suggest that the MLD and BILD tasks rely on different binaural processes. The idea that different aspects of binaural hearing are unrelated has also been proposed by others (e.g., Wilmington, 1994). Whereas MLD takes advantage of a binaural cue (i.e., ITD), the data from our study and the work of others suggests that spatial release from masking may rely in part on the monaural contribution of improved SNR in one ear. Furthermore, MLD results are likely to be more closely related to an isolated auditory function (e.g., frequency resolution or temporal resolution) than spatial release from masking results, which employed a speech recognition task and was therefore susceptible to a broad range of abilities including auditory function, but also non-sensory influences such as phonological awareness and vocabulary.

*Associations between Tasks.* Our data show no significant correlation between MLD and BILD (Figure 11). However, the range of BILD scores we documented was relatively limited and could increase given a listening task that better described a wide range of speech recognition in noise abilities. Alternatively, this could provide additional evidence that MLD and BILD rely on different binaural processes.

## Non-sensory Influences

### Experimental Parameters

We employed experimental paradigms that have been shown to obtain robust results in past research examining binaural abilities in typically-developing adults and children (e.g., stimulus types and presentation levels) and employed psychophysical methods to reduce the variability between individuals. Auditory stimuli in this study were expected and predictable as they were accompanied by a visual stimulus or signaled by a page turn thereby reducing the likelihood of adverse effects due to temporal unexpectedness. Potential differences in threshold criteria were accounted for by using a 2AFC paradigm, as listeners were required to determine in which interval a sound was presented, not indicate the presence of a sound. Furthermore, inclusion criteria included feasibility testing to rule out the possibility that scores could be influenced by an inability to perform experimental tasks. Efforts to maintain attention and motivation were employed throughout experimental testing by offering frequent verbal and tangible rewards (e.g., stickers). Visual reminders of the task were offered (e.g., pointing to each picture as sound played) and sometimes duplicated by the children. Disproportionate effects of attention and motivation were not evident in the slopes of the psychometric functions for typically-developing individuals and individuals with Down syndrome included in this study.

Internalization of Signals

The use of non-individualized head-related transfer functions (HRTFs) could have resulted in most subjects hearing signals internalized, rather than externalized and this could have influenced our spatial release from masking results. Individual HRTFs provide listeners with spectral information similar to that which they receive in natural conditions whereas non-individualized HRTFs rely on an approximation of spectral information based on measurements made from a mannequin (e.g., KEMAR). It is not clear that individualized HRTFs would have influenced our results to a measureable effect as past research has shown adequate performance using estimated HTRFs and simulated free-field conditions, at least for typically-developing adults and children (Bronkhorst & Plomp, 1988; Cameron & Dillon, 2007; Wenzel, Arruda, Kistler, & Wightman, 1993). Nonetheless, the potential effect of non-individualized HRTFs for individuals with Down syndrome should not be dismissed. However, the use of individual HRTFs represents an ideal that was not feasible. Obtaining HRTFs requires that listeners remain still for a period of time, which would have been difficult to sustain in young children and children with Down syndrome. Moreover, generating individualized simulated free-field and isolated cue conditions for each participant in this study would have added prohibitive time demands.

Surprisingly, observations of Figure 15 indicated that adults and children with Down syndrome had similar or poorer performance when the signal and noise were maximally separated compared to conditions in which they originated from more similar locations. It is possible that individuals with Down syndrome were affected to a greater degree by a maximally spatially separated noise than noise that was perceptually closer to a signal. That is, a displaced masker could be perceptually distracting for individuals with Down syndrome. It is not clear if this pattern of results would persist if the signals were perceptually externalized affording a closer approximation to natural environments. However, this might additionally account for some of the difference observed for spatial release from masking between individuals with Down syndrome and typically-developing individuals.

## Developmental Ability

An attempt to quantify the developmental abilities of the children with Down syndrome included in this study was made by using age-equivalent scores from the receptive language subdomain of the Vineland Adaptive Behavior Scales (Sparrow et al., 2005). The receptive language subdomain was thought to be the most closely related to our experimental tasks of all subdomains on this measure, but it proved to be insensitive to a range of abilities. This was partly due to the limited number of questions included in this subdomain. This subdomain is comprised of 20 questions and results in a range of 0 to 40 total possible points. Furthermore, it does not afford the opportunity to describe specific receptive language abilities, such as vocabulary acquisition, rather it queries more general abilities, such as the ability to follow instructions and listen to stories for specified amounts of time. Lastly, the translation of raw scores to age equivalents was found to be insensitive to describing incremental changes in age-equivalent scores as a small change in raw score corresponds to a large change in age equivalent score. For example, whereas a maximum raw score (i.e., 40) corresponds to an age equivalent score of 18:0, a raw score just one point lower (i.e., 39) corresponds to an age equivalent of 11:0. The use of this tool limited our ability to adequately estimate the developmental age and cognitive function of the individuals with Down syndrome included in this study.

Accurate estimates of developmental age notwithstanding, efforts to include typically-developing children within the same chronological age range as projected estimates of the developmental age range of children with Down syndrome included in this study were successful and resulted in the inclusion of typically-developing children as young as 3 years of age. As such, we were able to describe binaural abilities with development for typically-developing children adding further support to our knowledge base suggesting adult-like MLD by about 5 to 6 years of age for some signal and noise configurations (Grose et al., 1997; Hall & Grose, 1990a; Moore et al., 2011). Including children with Down syndrome aged 6 to 12 years did not offer a broad enough range of abilities to define a developmental pattern considering chronological age or receptive language age equivalent contrary to estimations at the outset of this study. However, the inclusion of a subset of adolescents and adults with Down syndrome afforded us the opportunity to begin to see how binaural abilities might change with development.

## Vocabulary and Auditory Comprehension

A developmental change in spatial release from masking was not evidenced in our data for any group, which was surprising. This result might have been influenced by the use of vocabulary words that were easily recognized in the most difficult listening condition (i.e., FF0). Recall that results from typically-developing adults included in our study correspond relatively well to those found by Bronkhorst and Plomp (1988) for conditions in which the signal and noise are separated. Relatively good speech recognition in the most difficult listening condition could result in less relative improvement in speech recognition as listening conditions improve. Past research supports this hypothesis demonstrating adult-like spatial release from masking for young children for a paradigm that also utilized monosyllabic words in a closed-set task (Litovsky, 2005).

Differences were not seen between children with Down syndrome and typically-developing children on this task, yet they were observed for adults in these respective groups. Conceivably, vocabulary could have influenced performance for adults with Down syndrome for our speech recognition task and been the catalyst for poorer performance relative to typically-developing adults. However, this is not thought to be the case as the words utilized for this study were obtained from a test developed to be used with typically-developing children as young as 3 years of age. It is more likely that adults with Down syndrome have a lower ceiling for this task than typically-developing adults based on SNR threshold, yet the words included in the task were simple enough to obtain near ceiling results in the most difficult listening condition. The performance for adults with Down syndrome was not affected by removal of level or time cues, which might have also been influenced by ceiling effects. A more sensitive task, one that is able to reveal a range of abilities, would likely produce a pattern of results for adults similar to those reported by Bronkhorst and Plomp (1988).

Known deficits in the phonological awareness abilities of individuals with Down syndrome could be partly responsible for the poorer spatial release from masking relative to typically-developing individuals included in this study. Corruption of part of a word might not influence the performance of one with high phonological awareness skills, but could render a word indistinguishable for someone with less capability in this area. The possible influence of phonological awareness ability on the performance of individuals with Down syndrome is likely closely related to temporal resolution abilities, a concept discussed in a previous section of this paper.

### Selective Listening

The ability to detect signals in noise could be influenced by selective listening abilities. Typically-developing adults selectively attend to relevant auditory filters when listening for expected sounds whereas typically-developing infants employ a broad listening strategy monitoring all auditory filters (Bargones & Werner, 1994). Though this affords typically-developing infants the opportunity to attend to expected and unexpected signals with equal success, it may reduce the improvement in threshold for expected signals experienced by typically-developing children and adults. It is possible that individuals with Down syndrome do not selectively attend to auditory filters associated with expected frequencies and therefore do not benefit from such strategies.

Behavioral studies have not been done to determine selective attention and habituation for auditory stimuli. However, electrophysiologic measures of these processes suggest delayed cortical processing for stimulus detection and no habituation in response to repeated stimulation (Pekkonen et al., 2007; Seidl et al., 1997). Furthermore, studies to examine visual selective attention and habituation have suggested that infants with Down syndrome have longer sustained levels of attention, slower habituation, and fewer shifts in gaze from preferred visual patterns (for review, see Zelazo & Stack, 1997). In addition, individuals with Down syndrome experience distraction from visual targets by distal stimuli to a greater degree and experience less benefit when distracters are categorically similar than typically-developing individuals (Merrill & O'dekirk, 1994). As such, it is likely that behavioral measures of auditory selective attention and habituation would reveal similar patterns for individuals with Down syndrome as well.

# CHAPTER VI

## CONCLUSION AND DIRECTIONS FOR FUTURE RESEARCH

Children with Down syndrome are at greater risk for hearing loss and other auditory perceptual difficulties as a result of anatomical differences within the peripheral and central auditory systems (e.g., Bilgin et al., 1996; Blaser et al., 2006). However, with the exception of a few studies (e.g., Glenn et al., 1981; Werner et al., 1996), little work has been done to understand the auditory abilities of infants and children with Down syndrome beyond hearing sensitivity. This study was designed to determine if the binaural abilities of children with Down syndrome are compromised relative to those of typically-developing children.

Children with Down syndrome experienced less binaural benefit than typically-developing children evidenced by results from the MLD task. This could be the result of inefficient use of ITD cues for children with Down syndrome. Importantly, adults with Down syndrome and typically-developing adults had similar MLDs. This suggests a delay in release from masking abilities for children with Down syndrome rather than an absolute deficit in this area.

Despite normal hearing sensitivity, individuals with Down syndrome had poorer thresholds in noise than typically-developing individuals. However, improvements in SNR threshold were observed for typically-developing individuals and individuals with Down syndrome with increasing age. Currently, the frequency resolution and temporal resolution abilities of individuals with Down syndrome are unknown. Studies to explore these auditory capabilities should be undertaken to determine their influence on hearing thresholds in noise for individuals with Down syndrome.

Separation of signal and noise stimuli improved speech recognition in some capacity for all individuals included in this study, but the performance of individuals with Down syndrome remained poorer than typically-developing individuals. This could be related to the temporal resolution capabilities of individuals with Down syndrome and future studies should be undertaken to address this issue. In addition, individuals with Down syndrome have known difficulties with phonemic awareness (e.g., Cardoso-Martins et al., 2002; Snowling et al., 2002) and this could have influenced estimations of spatial release from masking. Interestingly, individuals with Down syndrome had the tendency to perform poorer for conditions in which the signal and noise were maximally separated than those in which they were spatially adjacent. The reasons for this are currently unclear. It would be useful to implement a study of spatial separation of speech and noise in the free-field for individuals with Down syndrome to determine the influence of internalized signals and the possibility of perceptual distraction.

Our observations suggest that children with Down syndrome have similar speech recognition in noise as typically-developing children aged 3 to 5 years. It has long been established that speech recognition in noise improves with age for typically-developing children and that young children require more favorable SNRs than adults for optimal performance (e.g., Elliott et al., 1979; Mills, 1975). Results from this study suggest this is also true for individuals with Down syndrome through adolescence.

Motivation and attention did not appear to influence results for individuals with Down syndrome to a greater degree than typically-developing individuals as evidenced by group psychometric functions. Future research should be done to obtain more specific information regarding attention and selective listening for individuals with Down syndrome. Furthermore, techniques that do not require behavioral responses from participants, such as ERP testing and measures of the binaural interaction component (BIC), could determine if physiologic correlates support our behavioral findings.

In conclusion, results from this study showed that children with Down syndrome had higher masked thresholds for our experimental stimuli and exhibited less release from masking than typically-developing children. The binaural advantages afforded to typically-developing children, such as enhanced hearing sensitivity and better speech understanding in adverse listening conditions, were not present for children with Down syndrome in this study. The reduced binaural benefit experienced by children with Down syndrome suggests that they will require more favorable SNRs than typically-developing children to achieve optimal performance in adverse listening conditions. This has important implications for the planning of educational and therapeutic interventions for individuals with Down syndrome.

APPENDIX A

OTOLOGIC CASE HISTORY

Parent Survey: Ear Infections

Participant Code: _________________

Date of Birth: _________________

Today's Date: _________________

About how many ear infections has your child had since he/she was born? ______

About how many ear infections did your child have before his/her fifth birthday? ______

When was the last time your child had an ear infection? ____________

When was the last time your child took prescription medicine to treat an ear infection? ____________

Has your child ever had tubes surgically placed to help treat chronic ear infections? ______

If so, how many times did your child have this surgery? ______

When did the last surgery occur? ______

Do you think the tubes are still in place? ______

Thank you very much for your time!

# APPENDIX B

## MASKING LEVEL DIFFERENCE STORY SCRIPT

| Experimenter Script | Experimenter Actions |
|---|---|
| There are some owls hiding in a windy forest. I want you to find them. | |
| You are going to see two pictures and hear the wind in each picture. | Point to blank computer screen. |
| There is an owl in one of the pictures. It is your job to tell me which one. | |
| Is he in the first or second picture? | Point to approximate future position of visual stimulus on blank computer screen. |
| Just point. Is the owl here or here? | Point to approximate future position of visual stimulus on blank computer screen. |
| I'll show you the answer after you guess. Let's try a few! | |

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