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ABSTRACT

This study explored the hearing capabilities of Down Syndrome (DS) adolescents and young adults relative to a matched sample of non-DS trainable mentally handicapped (MH) individuals, and examined the relationship between hearing ability and performance on several cognitive tasks. Samples of 26 DS and 26 MH individuals were matched on intelligence quotient (IQ) and chronological age (CA). Audiometric data revealed greater DS than MH hearing losses at five of the six tested frequencies, more DS conductive and mixed hearing losses, and particularly high DS losses in the high frequency range. Measurement of the speech reception threshold revealed poorer reception of speech by the DS than the MH group. Classification of tympanograms indicated fewer normal ears and twice as many DS ears with middle ear problems reflecting no mobility or retraction of the tympanic membrane. Presence of DS middle ear difficulties was also confirmed by poorer elicitation of the acoustic (stapedius) reflex in DS subjects. Correlation of hearing variables with seven cognitive tasks revealed only one significant relationship after statistical removal of the effects of CA and IQ: DS subjects with poorer hearing identified fewer words in a task in which a masking noise quickly followed a spoken word. (45 reference) (JDD)

Hearing Abilities of Down Syndrome and Other Mentally Handicapped Adolescents

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Abstract

Down Syndrome (DS) individuals are prone to auditory processing difficulties in a variety of audiological, short-term memory, and expressive language tasks. The present study explored: a) the hearing capabilities of DS adolescents and young adults relative to a matched sample of non-DS mentally handicapped (MH) individuals, and b) the relationship between hearing ability and performance on several cognitive tasks. Samples of 26 DS and 26 MH trainable mentally handicapped individuals were matched on IQ and CA. Subjects were tested individually in a comprehensive audiological assessment, computer-based memory and word identification experiments, and standardized language subtests. Audiometric data revealed greater DS than MH hearing losses at five of the six tested frequencies, more DS conductive and mixed hearing losses, and particularly high DS losses in the high frequency range. Measurement of the speech reception threshold revealed poorer reception of speech by the DS than the MH group. Classification of tympanograms indicated fewer normal ears and twice as many DS ears with middle ear problems reflecting no mobility or retraction of the tympanic membrane. Presence of DS middle ear difficulties was also confirmed by poorer elicitation of the acoustic (stapedius) reflex in DS subjects. These findings replicated those of Dahle and McCollister (1986) [the only other such study with an IQ-matched control group] and extended knowledge about DS hearing ability to an older age group (19 years), more extreme pure tone frequencies (250 and 8000 hz), and the speech reception threshold. Correlation of hearing variables with seven cognitive tasks administered on the same day revealed only one significant relationship after the statistical removal of the effects of CA and IQ: DS subjects with poorer hearing identified fewer words in a task in which a masking noise quickly followed a spoken word. These results suggest that DS auditory verbal processing difficulties may stem, in part, from subjects with hearing difficulties needing more time to identify spoken words.

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Hearing Abilities of Down Syndrome and Other Mentally Handicapped Adolescents

Considerable evidence has accumulated in three separate literatures--cognitive psychology, language development, and audiology--to suggest that Down Syndrome (DS) individuals have difficulty processing auditory stimuli. Their difficulties have been documented in tasks that focus on auditory short-term memory (e.g., Marcell & Weeks, 1988; McDade & Adler, 1980; Varnhagen, Das, & Varnhagen, 1987), successive processing ability (e.g., Ashman, 1982; Hartley, 1982, 1985; Snart, O'Grady, & Das, 1982), language comprehension (e.g., Bridges & Smith, 1984; Burr & Rohr, 1978; Marcell, Croen, & Sewell, 1990a), and language expression (e.g., Andrews & Andrews, 1977; Cornwell, 1974; Marcell, Harvey, & Cothran, 1987; Miller, 1987). Furthermore, research in the realm of audiology indicates the presence of mild to moderate hearing impairments and conductive (middle ear) problems in a majority of the DS population (Balkany, 1980; Brooks, Wooley, & Kanjilal, 1972; Cunningham & McArthur, 1981; Davies & Penniceard, 1980; Downs, 1983; Fulton & Lloyd, 1968; Glovsky, 1966; van Gorp & Baker, 1984; Keiser, Montague, Wold, Maune, & Pattison, 1981; Schwartz & Schwartz, 1978; Wilson, Folsom, & Widen, 1983). Few psychologically-oriented investigators of DS cognition (including the present authors) have seriously considered hearing loss in relation to their research. This problem may stem from a general lack of familiarity with the terminology and concepts of audiology, an inability to meet the time constraints and economic costs associated with comprehensive audiological evaluations, and a tendency to rely on readily-available school records of simple pure-tone screenings--a procedure which may underestimate DS hearing loss (cf., Cunningham & McArthur, 1981).

The present study reports findings from the first year of a three-year longitudinal study undertaken to explore relationships among auditory memory, language, and hearing abilities in a population of DS adolescents and young adults. Detailed accounts of receptive (Marcell et al, 1990a) and expressive (Marcell, Sewell, & Croen, 1990) language results have been given elsewhere, and a summary of findings from four computer-based auditory memory and speed of word identification tasks is in preparation (Marcell, Croen, & Sewell, 1990b). The current report concentrates on two different questions. First, are the hearing abilities of the DS group different from those of a non-DS mentally handicapped (MH) control group matched on age and intelligence? As Dahle and McCollister (1986) noted, the failure of previous studies (e.g., Brooks et al, 1972) to control for DS and MH intelligence differences presented an ambiguity of interpretation: Were group differences due to the type of mental retardation or differential understanding of test procedures? Dahle and McCollister--the only other investigators, to our knowledge, who included an IQ-matched MH group--found more hearing impairment and otologic disorders in the DS children. The present project attempted both to replicate their findings in an older age group and to expand our understanding of DS hearing difficulties by using additional measures of hearing acuity.

The current report also concentrates on a second question: Does hearing ability in DS subjects correlate with performance on measures of language ability, auditory short-term memory, and speed of auditory word identification? Middle ear infections, for example, are frequently associated with delayed language acquisition and lowered academic achievement in nonretarded children (e.g., Holm & Kunze, 1969; Needleman, 1977; Silva, Kirkland, Simpson, Stewart, & Williams, 1986; Zinkus, Gottlieb, & Schapiro, 1978). Although it is not yet clear how much of a hearing loss will cause a cognitive-linguistic impairment (Ruben, 1984), or which linguistic abilities will be affected (Teele, Klein, & Rosner, 1984), the relationship between hearing and linguistic abilities has been clearly established (Paul & Quigley, 1987). Three studies suggest that such a relationship also holds for the DS population: DS adolescents with abnormal tympanograms scored lower in intelligence than those with normal tympanograms (Libb, Dahle, Smith, McCollister, & McLain, 1985), DS adolescents who received early surgical intervention for otitis media had higher language scores than those with otitis who had not been surgically treated (Whiteman, Simpson, & Compton, 1986), and DS adults with higher pure tone hearing losses had lower receptive vocabularies (Nolan, McCartney, McArthur, & Rowson, 1980). Similarly, the present study attempted to explore the relationship between hearing and several measures of auditory language processing. We also hoped to determine whether such hearing-language relationships, if present, are syndrome-specific; i.e., would similar patterns be found in an age- and intelligence-matched MH sample?

Method

Subjects

Fifty-nine trainable mentally handicapped subjects were recruited in a three-county area surrounding Charleston, South Carolina. An attempt was made, through initial letter and telephone communication with parents, to recruit participants with understandable speech and knowledge of the numbers 1-9. However, 4 individuals (3 DS and 1 MH) with difficult-to-understand speech (subjectively determined by the experimenters) and 2 individuals (both MH) unable to recognize, by sight, all nine digits were included in the final samples. These individuals were kept in the study because of their ability to perform the tasks and to communicate well enough to be understood by the experimenters. Seven other participants were excluded from the study because of no speech (2 MH), an inability to understand (or failure to cooperate during) 40% or more of the tasks (3 DS and 1 MH), or later discovery of educable (rather than trainable) school placement (1 MH).

The final samples consisted of 26 DS (14 males and 12 females) and 26 MH (18 males and 8 females) adolescents and young adults. The samples were matched on Stanford-Binet IQ, $\underline{t}$ (50)=.616 [DS mean=39.7 (SD=7.3); MH mean=40.9 (SD=6.2)] and chronological age, $\underline{t}$ (50)=.096 [DS mean=226.1 months (SD=40.3); MH mean=225.0 (SD=40.5)]. Subjects were recruited from the following locations: eleven public schools (N=44), one residential institution (N=2), and two community programs for mentally handicapped adult citizens (N=6 public school graduates). School records, discussion with parents, and medical histories (when available) were used in an attempt to determine etiologies of mental retardation in the MH group. The "causes" (or relevant associated medical conditions) were as follows: unknown (12), seizure disorders (5), early concussion (1 with and 1 without seizures), microcephaly (1 with early malnutrition and 1 with early high fever), cerebral paslsy (1 with and 1 without seizures), early oxygen deprivation (1), early meningitis, seizures, and blood clot (1), and Williams Syndrome (1).

Apparatus, Tasks, and Dependent Variables

Audiological Tests. Audiological testing was accomplished at the Charleston Speech and Hearing Center by three clinical audiologists experienced in working with the mentally handicapped. Stimuli were presented in a double-walled Industrial Acoustics Controlled Acoustical Environment. Audiometric and impedance measurements were made possible by a Maico Model MA32 Audiometer and a Madsen Model Z073 Electroacoustic Impedance Bridge. The following audiological assessments were made:

1. Pure Tone Audiometry. Pure tone air conduction measurements of auditory sensitivity were made in the frequency range of 250-8000 Hz; pure tone bone conduction measurements were made in the frequency range of 250-4000 Hz. Forty-five of the subjects responded by hand raising and seven (3 DS and 4 MH) were conditioned to respond with standard play audiometry techniques. Air conduction measurements for the left and right ears were combined to produce mean decibel hearing loss (dbHL) scores for each of the frequencies; these six scores were also averaged to produce one overall measure of pure tone hearing loss for correlational analyses (Coren, 1989). Bone conduction measurements were used in conjunction with air conduction measurements to produce, for each ear, an auditory classification of type of hearing loss as conductive, sensorineural, mixed, or none. Hearing loss was defined conservatively by a sensitivity of 20 dbHL or poorer.

2. Speech Audiometry. The mean speech reception threshold (SRT) for left and right ears provided an index of the lowest intensity level (in db) at which common, two-syllable spondaic words were "just intelligible". Live voice was used to present the words at 5 db increments; the subject responded either by repeating the word (N=47) or by pointing to its depiction (N=2 MH and 3 DS). After SRT was established, the audiometer was reset at 40 db above sensation level and a list of words was spoken in a quiet background for a measure of speech discrimination. The mean speech discrimination score for left and right ears reflected the percentage of words correctly recognized out of 25.

3. Impedance Measurement. Impedance techniques provided a relatively objective assessment of the integrity of middle ear functioning. Compliance tympanograms were plotted for each ear across a +200 to -200 air pressure range. Each tympanogram was categorized as representing a middle ear that was either functioning normally or showing reduced mobility, no mobility, retraction, or hyper-flaccidity. Acoustic (stapedial) reflex thresholds were elicited in each ear by contralateral acoustic stimulation at 500, 1000, 2000, and 4000 Hz. A reflex was considered absent if it could not be elicited within the 85-115 db range. A summary acoustic reflex score represented the number of times out of eight that the reflex was elicited in the two ears across the four frequencies.

Cognitive Tests. All cognitive tasks were presented in a suite of sound-attenuated observation rooms at the College of Charleston. Assessments of short-term memory and speed of word identification occurred in a room containing an IBM PS/2 Model 30 computer, an Animated Voice Corporation Professional System Loudspeaker, and a Realistic 33-992B microphone.

Digitized stimuli for the computer-based experiments were created and editted with software produced by the Animated Voice Corporation. Language testing occurred in a second room containing a Panasonic tape recorder and a Realistic 33-2001 microphone. The following cognitive assessments were made and the results correlated with audiological measures:

1. Auditory Digit Span. In this traditional measure of auditory short-term memory, increasingly-long random digit sequences (two examples of each length) were presented freefield until the subject incorrectly repeated two consecutive sequences. Subjects were awarded 1 point for each item correctly reported in its proper location and 1 point for each pair of items recalled in their correct relationship.

2. Receptive Vocabulary. One measure of language comprehension was the Picture Vocabulary Subtest of the Test of Language Development-2 Primary (TOLD-2P; Newcomer & Hammill, 1988). The subject pointed to one of four drawings that best represented the word spoken by the examiner. Subjects were awarded 1 point for each item out of 35 correctly identified.

3. Grammatical Understanding. A second measure of language comprehension, the Miller-Yoder Language Comprehension Test (Miller & Yoder, 1984), required the subject to point to one of four drawings that best represented the sentence spoken by the examiner (e.g., "Mother is kissed by father."). The subject was awarded 1 point for each pair of sentences correctly identified out of 42 sentence pairs.

4. Expressive Vocabulary. One measure of language expression required the subject to define the first eight items (common words such as "cow" or "rest") on the TOLD-2P Oral Vocabulary Subtest. The subject's tape-recorded responses earned 1 point per item for either a precise definition or two descriptive characteristics (Newcomer & Hammill, 1988).

5. Sentence Imitation. A second measure of language expression, the first eight items of the TOLD-2P Sentence Imitation Subtest, required the subject to repeat a sentence spoken by the examiner (e.g., "Yesterday my aunt forgot her lunch."). The subject's tape-recorded responses earned 1 point per item for a correct imitation of the entire sentence (misarticulations were ignored). The subject's responses were also scored for oral response time, the time lapse between the beginning of the examiner's last spoken word and the beginning of the subject's response.

6. Backmasking. In this measure of auditory word identification speed, a concrete noun spoken by the computer was followed at varying brief intervals by a burst of white noise. The purpose of the masking noise was to interfere with the subject's attempt to identify and repeat the word. The interval between word and mask was systematically varied (40, 80, 160, and 320 msec) in order to determine the amount of time needed to identify words. Dependent variables were the number of words identified (out of seven) in each masking interval condition and a control condition with no mask.

7. Gating. A second measure of auditory word identification involved presentation of progressively larger amounts of a spoken word for identification (Grosjean, 1980). Testing always began with the briefest gate

(the first 32% of the word) and proceeded to increasingly longer gates (each representing an additional 17% of the word). The subject attempted to guess what word the computer was saying; the dependent variable was the mean earliest gate (of five gated words) at which the subject first offered a meaningful word phonetically compatible to the sound spoken by the computer.

Although subjects were given a third word identification task--speed of judging whether two spoken items were the same or different--this task is not reported here because of the large number of participants (13) who were unable to make reliable same-different judgments during a training procedure. Because the 13 excluded subjects had a significantly lower mean IQ than the remaining subjects, any correlations using same-different performance data would likely be attenuated by restriction of range.

General Testing Procedure

Each subject spent one day at the College in an individualized program that included the above-mentioned comprehensive audiological assessment, standardized language subtests, and computer-based memory and word-identification experiments. [Only one subject received the audiological and cognitive assessments on different days.] The day generally began at 9 am, ended at 2:30 pm, and included a variety of nonresearch activities (e.g., outside play, computer games, picnic lunch, crafts project, visit to a marine biology specimen display) scheduled at the discretion of the experimenters to provide a break from the testing routine. In addition to the two testing rooms at the College, a third room--brightly decorated and containing a sofa, lounge chair, radio, and games--was used as a rest, play, and crafts area. Each room had an adjacent observation area equipped with one-way mirror and microphones; parents were invited to observe at any time during the day-long session.

The audiological assessment was typically the last test administered; the other tests were administered in a random order. Forty-eight of the subjects visited the College in pairs; the other four subjects, due to scheduling difficulties, visited individually. Three well-practiced experimenters shared the testing duties and each test was individually administered . All visits were made during 1989 in the months of June, July, or August, thus reducing the likelihood of audiological scores being influenced by upper respiratory and middle ear infections (Keiser et al, 1981).

Results and Discussion

Analyses of Audiological Measures

Pure Tone Audiometry. Pure tone air conduction measurements (see Figure 1) were submitted to a 2 (group) x 6 (frequency) ANOVA. The analysis revealed a significant main effect of group, F (1,50)=7.51, p=.008, in which the DS group showed a greater overall hearing loss (20.8 dbHL) than the MH group (12.7 dbHL). The analysis also revealed significant effects of frequency, F (5,250)=22.92, p=.0001, and group x frequency interaction, F (5,250)=3.60, p=.004. Newman-Keuls post hoc comparisons (at alpha=.05) on the interaction effect indicated that the DS group had greater hearing losses than the MH group at each frequency except 2000 Hz. Furthermore, in the DS group, dbHL for the 8000 Hz frequency was worse than for every other frequency, and dbHL at the 4000 Hz frequency was worse than for every frequency below it. This pattern supports both Davies' (1985) contention that high frequency hearing loss is prevalent in the majority of DS adolescents and adults, and Widen et al's (1987) report that DS sensitivity is lowest in the 8000 Hz range. [In contrast, the hearing sensitivities of MH subjects in the current study did not differ at 2000, 4000, and 8000 Hz]. When Widen et al (1987; Wilson et al, 1983) classified DS subjects by hearing losses of 10 db or greater, they found that 13 of 15 subjects (87%) had 8000 Hz hearing losses. A similar classification of DS subjects in the current study revealed 23 of 26 (89%) with 8000 Hz hearing losses. [In contrast, 17 of 26 MH subjects (65%) showed such a loss.] Auditory classification of a subject's hearing loss revealed that relative to the MH group, the DS group had fewer normal audiograms (52% vs 79%) and more hearing losses of the conductive (19% vs 2%) or mixed (17% vs 4%) type, $\chi^2(3)=14.99$, $p=.0023$.

Speech Audiometry. The DS group had a higher mean SRT (i.e., poorer reception of speech) than the MH group (15.0 db vs 8.4 db), $t(50)=2.68$, $p=.01$. Analysis of speech discrimination scores indicated that the two groups did not differ in accuracy (DS mean=92.0% and MH mean=94.1%), $t(50)=1.109$. This finding is not unexpected given that words were presented at sufficient intensities (40 db above each subject's threshold).

Impedance Measurement. Relative to the MH group, the DS group had fewer normal tympanograms (50% vs 100%) and more middle ear problems indicative of no mobility (21% vs 0%) or retraction (21% vs 0%) of the tympanic membrane, $\chi^2(4)=34.67$, $p=.00001$. DS middle ear problems tended to be bilateral: 12 subjects had abnormal tympanograms in both ears and 2 subjects had an abnormal tympanogram in one ear. The overall incidence rates were similar to those reported by Davies and Penniceard (1980), who found that 55% of their DS children and 100% of their severely retarded children (not matched on IQ) had normal tympanograms. Acoustic reflexes were less likely to be elicited in the DS (mean=3.8) than the MH (mean=6.9) group, $t(50)=3.59$, $p=.001$.

To summarize, our audiological findings replicate and extend those of the only other study, to our knowledge, that includes a mentally handicapped control group equated on intelligence (Dahle & McCollister, 1986). We found that DS adolescents and young adults had worse hearing on virtually all measures than their mentally handicapped peers. They showed a strong tendency towards conductive/middle ear difficulties and lowered sensitivity across most frequencies. New audiological findings from our study include the following: a) audiological description of older (19 vs 10 years) DS and MH samples, b) documentation of DS hearing losses at more extreme frequencies (250 and 8000 Hz), c) confirmation of a strong tendency towards a high frequency loss in DS individuals, and d) descriptions of DS difficulties with both the reception threshold for familiar speech and elicitation of the acoustic reflex.

Correlational Analyses of Audiological and Cognitive Measures

An attempt was made to replicate findings of two of the three previously-described studies of DS individuals that reported relationships between tympanogram type and IQ (Libb et al, 1985) and pure tone hearing loss and receptive vocabulary (Nolan et al, 1980). [The third study (Whiteman et al,

1986) could not be evaluated because we did not have access to detailed records of our subjects' early medical histories.] A partial correlation between number of abnormal ears (as revealed by tympanograms) and Stanford-Binet IQ (with the effects of age partialled out, as performed by Libb et al) was in the predicted direction for DS subjects, r (23)=-.373, although not significant (critical value at alpha=.05 is .396). The relationship could not be explored in MH subjects because of the absence of variability in the tympanometry data. To evaluate the Nolan et al study, a correlation was performed between overall puretone hearing loss and receptive vocabulary score. Although this correlation did not approach significance, other correlates of receptive vocabulary did achieve or approach significance in the predicted direction for DS (but not MH) subjects: number of abnormal ears (tympanogram), r(50)=.454, and acoustic reflex score, r(50)=.373. However, these correlations (like others) disappeared when the influences of IQ and CA were removed. The likelihood that Nolan et al's finding is an artifact of uncontrolled concurrent variation due to age and intelligence should be raised.

The major analysis of the current study involved correlating the three numerical audiological measures possessing the greatest variability--overall pure tone hearing loss, SRT, and acoustic reflex score--with the previously described language, memory, and speed of word identification measures. Due to the known relationships between age, intelligence, and cognitive performance, partial correlations were used to statistically remove the influences of IQ and CA. The analysis yielded significant correlates from only one task--the backmasking experiment. As can be seen in Table 1, DS (but not MH) subjects with greater hearing losses, higher (worse) SRTs, and poorer acoustic reflexes identified fewer words when the items were followed quickly (40 or 80 msec) by masking noises. Neither DS nor MH subjects showed this pattern when there was no mask or a longer pause (160 or 320 msec) between the word and the mask. These results suggest that DS subjects with hearing difficulties need more time to identify spoken words--a finding that may help explain the typically low DS performance in auditory verbal information-processing tasks. Because the relationship did not hold for MH subjects, it may be tentatively concluded that hearing difficulty interacts with etiology; i.e., hearing loss appears to be more detrimental to speed of word processing in mentally retarded individuals with Down syndrome.

Table 1

Relationships between Audiological Measures and Number of Words Identified in Speed of Auditory Word Processing Task

	Masking Interval			
	DS Group		MH Group	
Audiological Measures	40 Msec	80 Msec	40 Msec	80 Msec
Pure Tone Hearing Loss	-.485*	-.469*	-.199	-.094
Speech Reception Threshold	-.447*	-.524**	-.317	-.087
Acoustic Reflex Score	.455*	.377	.129	.179

Note. Partial correlations with effects of IQ and CA removed were used.

* $p < .05$. ** $p < .01$.

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Figure 1. COMPOSITE AUDIOGRAM OF DS AND MH PURE TONE HEARING LOSSES