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AUDITORY SYSTEM FUNCTION IN INDIVIDUALS WITH WILLIAMS SYNDROME

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of the requirements for the

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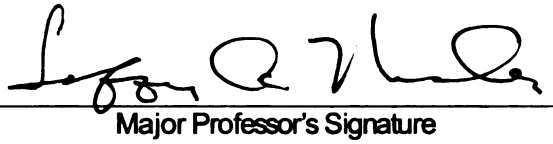

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AUDITORY SYSTEM FUNCTION IN INDIVIDUALS WITH WILLIAMS SYNDROME

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ABSTRACT

AUDITORY SYSTEM FUNCTION IN INDIVIDUALS WITH WILLIAMS SYNDROME

By

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Hypersensitivity to sounds is a well-known characteristic of individuals with Williams syndrome (WS); however, auditory system function in individuals with WS has not yet been thoroughly evaluated. The purpose of this research project was to test objectively whether individuals with WS are more sensitive to auditory stimulation (have better-than-normal thresholds). Seven individuals with WS, between the ages of 7 and 43 years, were recruited for the current study. Audiological and physiological status was examined using air- and bone-conduction thresholds, tympanometry, loudness discomfort level measures, distortion-product otoacoustic emissions (DPOAEs) and a hyperacusis questionnaire that was adapted from Klein, Armstrong, Greer, and Brown (1990). The results of this study revealed that five of the seven individuals with WS showed pure-tone thresholds poorer than expected when compared to age peers from the general population. Only one of them had a documented diagnosis of permanent hearing loss before this study. The empirical evidence presented here not only suggests that reports of hypersensitivity to sounds do not justify a conclusion that overall hearing is intact or better than normal in individuals with WS, but also addresses the importance of early diagnosis of hearing loss in this population.

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CHAPTER 1

Introduction

Williams syndrome (WS), also known as Williams-Beuren syndrome, was reported by Fanconi (1952), and Williams, Barratt-Boyes, & Lowe (1961). Typical features of Williams syndrome include specific facial dysmorphology ("elfin" features), renal and cardiovascular anomalies, failure to thrive in infancy, transient-neonatal hypercalcemia, elastin arteriopathy, mild-to-moderate mental retardation or learning difficulties, delayed language and motor development, a unique cognitive profile, a distinctive personality, and hypersensitivity to speech and abnormal (aversive) sensitivities to other classes of sounds (Morris, Demsey, Leonard, Dilts, & Blackburn, 1988; Lenhoff, Wang, Greenberg, & Bellugi, 1997; Bellugi, Lichtenberger, Jones, & Lai, 2000; Morris & Mervis, 2000).

Historically, the incidence of WS has been reported to be at least 1/20,000 individuals (Morris et al., 1988). Recently, a Norwegian population prevalence study has estimated a prevalence closer to 1/7,500 live births (Morris, 2004). The current understanding of genetic causes of WS involves a micro-deletion of the long arm of chromosome 7 (Ewart et al. 1993; Korenberg et al., 2000). This micro-deletion includes gene-encoding elastin (ELN) and 17-20 other genes (Bellugi et al., 2000; DeSilva et al., 2002; Ewart et al., 1993; Merla, Ucla, Guipponi, & Reymond, 2002; Osborne, 1999) and can be detected by fluorescent *in situ* hybridization (FISH) in 99% of individuals with WS (Lowery et al., 1995).

Relative to their overall level of intellectual ability, individuals with WS typically show strengths in the areas of phonology (Grant et al., 1997; Volterra, Capirci & Caselli,

2001), auditory (verbal) short-term memory (Grant et al., 1997, Mervis & Klein-Tasman, 2000), and expressive vocabulary (Mervis & Robinson, 2000; Singer-Harris, Bellugi & Bates, 1997). The relative strengths in phonology and vocabulary have been attributed to auditory processing strengths (Karmiloff-Smith et al., 1997; Mervis & Klein-Tasman, 2000).

Both auditory verbal strengths (Karmiloff-Smith et al., 1997) and auditory language comprehension abilities (Neville, Mills, & Bellugi, 1994) of individuals with WS have been attributed to hypersensitivity to sound. When data collection for this project was begun, these inferences were supported solely by anecdotal reports of auditory processing strengths and surprising musical abilities (Lenhoff et al., 1997; Mervis & Klein-Tasman, 2000) and brief papers and questionnaire-studies reporting high rates of extreme oversensitivity to sound (Van Borsel, Curfs, & Fryns, 1997; Klein, Armstrong, Greer and Brown, 1990; O'Reilly, Lacey and Lancioni, 2000; Johnson, Comeau & Clarke, 2001). Two descriptive studies recounting audiometric performances of a small number of individuals with WS (Ruangdaraganon, Tocharoentanaphol, Kotchabhakdi, & Khowsathit, 1999; Johnson et al., 2001) indicated that 33% had sensorineural hearing loss. These two recent reports of significant rates of hearing loss in WS are difficult to reconcile with the popular understanding of superior auditory sensitivity. The literature available provides only a limited foundation for exploring this inconsistency.

Literature Review

Behavioral assessment of hearing

Audiometric performance in individuals with WS, or those suspected of having WS (i.e., Nigam & Samuel, 1994), has been reported in three descriptive studies (Nigam & Samuel, 1994; Ruangdaraganon et al., 1999; Johnson et al., 2001). Nigam and Samuel (1994) reported a case study of a 21-month-old girl with WS in Sunderland, Britain. She was seen by a geneticist, and a diagnosis of WS was based on her facial characteristics rather than on a FISH confirmation. She was referred to an ENT clinic because of intermittent otalgia, drooling and snoring accompanied by a clear nasal discharge. The authors reported observation of dull tympanic membranes and collapsed nasal alae, and that, "on a performance test a 35-40 dB loss was found on the left side and impedance testing gave a flat trace on the left but normal responses on the right" (p. 494). They also reported that, six months after adenoidectomy and bilateral myringotomies, her response to everyday sounds, including the ringing of a doorbell or telephone, was to clasp her hands over her ears. A follow-up audiologic assessment revealed hearing within normal limits for both ears.

Ruangdaraganon et al. (1999) reported a case study of two individuals with WS who were identified by the FISH technique. Participants were a 15-month-old girl and a 16-month-old girl seen in Bangkok, Thailand. Ruangdaraganon et al. reported that one girl had auditory brainstem responses (ABR) indicative of a bilateral sensorineural hearing loss. However, they did not report the severity of the hearing loss or the status of the middle ear function. They did not include otoacoustic emissions measures (OAE) in their test battery, although a combination of ABR and OAE is commonly used for early

identification of hearing loss. Such data would have provided confirmation of the ABR results and important information about the site of lesion. For the other participant, they reported no information concerning hearing assessment or her hearing status.

Johnson et al. (2001) contacted 23 families from the Atlantic Provinces of Canada. Nine individuals with WS, ages 9 to 25 years, from these families underwent a full battery of audiologic tests. Their test battery included pure-tone audiometry, speech recognition thresholds, acoustic reflex thresholds, tympanometry, and transient evoked otoacoustic emission (TEOAE) measures. Researchers found that sensorineural hearing loss was present in three participants ages 18, 24, and 25 years within this group. All three showed moderate sensorineural hearing loss; one had bilateral hearing loss and two showed unilateral hearing loss. Seven of the nine participants, including these three participants with sensorineural hearing loss and four participants with normal pure-tone thresholds, were reported to have absent TEOAEs.

Johnson et al. (2001) failed to define what constituted "normal" hearing thresholds for a group of adults with mild-to-moderate mental retardation, did not report speech recognition thresholds, neglected to quantify the magnitude terminology (e.g., "moderate"), and did not describe the configuration of the hearing thresholds. Furthermore, they did not include the pass/fail criterion for the OAE measures. Absent TEOAEs suggested abnormal outer hair cell function. The absence of TEOAEs in the individuals with normal hearing (4 of 6 subjects) suggests a lack of sensitivity in the measurement of hearing loss.

Marler, Elfenbein, Netzloff & Liu (2004) investigated hearing acuity in 21 individuals with WS at the 2004 National Williams Syndrome Conference in Grand

Rapids, Michigan. Fourteen children, ages 7 to 9 years, and seven adults, ages 21 to 48 years, participated in their preliminary study. The test battery included pure-tone, air-conduction screening at the frequencies of 500, 1,000, 2,000, 4,000, and 8,000 Hz, screening tympanometry, and DPOAE measures. The criterion for failure of the pure tone hearing screening was failure to respond to a 20 dB HL signal at any target frequency in either ear for the adults, and failure to respond to a 15 dB HL signal at any target frequency in either ear for the children. The criterion for a DPOAE indicative of hearing loss was an emission below the 5th percentile point for normally hearing ears reported by Gorga et al. (1997). One of the adult participants was not included in the analyses because a second genetic condition (Waardenburg syndrome) was later confirmed. Sixteen of 20 individuals (80%) with WS in that study showed normal bilateral tympanograms (Type A). Two more individuals had normal unilateral tympanograms, but tympanograms could not be obtained on their other ears. No tympanograms were obtained for two other individuals due to excessive cerumen or complaint of ear pain. Marler et al. reported that 84% of their participants failed the hearing screening and 74% showed DPOAEs indicative of impaired cochlear function.

Hyperacusis and otitis media

Jastreboff & Jastreboff (2000) defined hyperacusis as "abnormally strong reactions occurring within the auditory pathways resulting from exposure to even a moderate sound level" (p. 156). They defined "phonophobia" as "abnormally strong reactions of autonomic and limbic systems without abnormally high activation of auditory system resulting from enhanced connections between the auditory and limbic

system" (p. 156). They noted that individuals with phonophobia have developed fear of sound as the result of their hyperacusis.

Dix, Hallpike, and Hood (1948) reported that patients with sensorineural hearing loss who complain of discomfort from loud sounds are diagnosed as having "loudness recruitment." Loudness recruitment has been defined as "an abnormally rapid growth of loudness" (p.219) in individuals with cochlear impairment (Mendel, Danhauer, & Singh, 1999) and is a phenomenon similar to the hypersensitivity to loud sounds seen in hyperacusis. The commonality between hyperacusis and auditory recruitment is intolerance of loud sounds.

The phenomenon of hyperacusis has been reported in greater than 95% of the individuals with WS (Klein et al., 1990; Nigam & Samuel, 1994; Van Borsel et al., 1997). Klein et al. investigated the prevalence of hyperacusis and otitis media in individuals with WS by conducting a survey. They noted that earlier attempts to obtain audiometric data from individuals with WS had been unsuccessful because of factors related to age, hyperactivity, and/or developmental age. Thus, they used a survey approach rather than a complete audiological assessment (i.e., threshold audiogram, high-level speech discrimination, and acoustic reflex measures).

Participants for the survey were recruited from a Williams Syndrome Association membership list of four southeastern states. A questionnaire was sent to 100 parents of individuals with WS. Sixty-five surveys (65%), covering 36 male and 29 female individuals ages 1 to 28 years, were completed and returned. Children attending a pediatric, general dentistry clinic served as a normal control group. The control group included 32 males and 33 females between the ages of 2 to 17 years. According to

parental report, prevalence for hyperacusis was 95% in the individuals with WS and was 12% in the control group. Specific information about offending sounds and the reactions to these sounds were also reported in this study. Firecracker, power saw, electronic drill, fire engine siren, motorcycle, loud auto muffler, and blender were each selected by more than 60% of the parents in this survey. Thunder, the most common write-in answer, was reported by 19% of parents. Sounds that are not usually associated with threatening events such as automatic icemaker, television test tone, newspaper crackling, and church bells were added to the list by some parents. Loud (73%) and sudden (74%) were the descriptive characteristics of offending sounds most frequently selected by parents.

In the Klein et al. study, the prevalence of otitis media in the WS population (61%) was significantly greater than the prevalence in the normal group (30%). The Spearman rank-order correlation coefficient was used to investigate the relationship between frequency of otitis media and the overall hyperacusis rating. A significant relationship could not be established ($r = .156$, $p < .10$) in that study. The authors reported that, among individuals with WS, boys tend to have more episodes of otitis media than girls, and girls tend to have the placement of tympanostomy tubes slightly more often.

In the case study presented by Nigam and Samuel (1994), a left middle ear pathology, left conductive hearing loss and the phenomenon of hyperacusis were reported in a 21-month-old child. They described the subject's responses to sounds as a result of lack of exposure to environmental sounds due to temporary conductive hearing loss secondary to middle ear pathology. Nigam and Samuel reviewed the previous WS research, but failed to find support for their lack-of-exposure hypothesis.

Van Borsel et al. (1997) obtained survey data on hyperacusis and otitis media in individuals with WS. Their questionnaire was similar to the questionnaire used by Klein et al. (1990). The survey was mailed to all parents of children in the Dutch and the Flemish WS associations. The number distributed was not reported. Eighty-two usable questionnaires were returned from the parents. The ages of the individuals with WS ranged from 1 year 10 months to 45 years 5 months. Only 4 (5%) out of 82 subjects had "never been frightened or bothered by sounds" (p.128). The majority of the participants (83%) reported "oversensitivity" to sounds and the remaining participants (12%) reported that they had experienced this problem in the past. Parental report showed that individuals in that 12% had outgrown their fear of sounds at ages ranging from several months to 22 years. The authors did not differentiate hyperacusis and phonophobia in their subjects.

Fifty-four percent of the WS subjects in the Van Borsel et al. study noted above were reported to have had episodes of otitis media, and 76% were reported to show behavior indicative of attention deficit hyperactivity disorder. Van Borsel et al. reported that the tendency for boys to have more episodes of otitis media and girls to have more placements of tympanostomy tubes, reported by Klein et al. (1994), could not be observed in their study. The Spearman rank-order correlation coefficient was used to investigate the relation between frequency of otitis media and the overall hyperacusis rating, and no significant relation could be established ($r = -.1285$, $p = .253$).

Auditory verbal strengths

Individuals with WS show a relative strength in verbal short-term memory (Mervis et al., 2000; Udwin & Yule, 1991). This ability has been hypothesized to provide

an important foundation for later vocabulary acquisition (Mervis, Robinson, Rowe, Becerra, & Klein-Tasman, 2003). Even when demonstrating mild-to-moderate mental retardation, children with WS will frequently score within 1 standard deviation of the general population on standardized measures of auditory rote recall and working memory (Mervis et al., 2003).

Neville et al. (1994) investigated the impact of auditory sensitivity on the development of individuals with WS using the event-related potential (ERP) technique. Four children (two males and two females) ages 10 to 14 years and four adults (one male and three females) served as subjects for their study. One of the adults was tested both as a child at the age of 14 and as an adult at the age of 18. They also recruited normal control children and adults. Approximately half of the control subjects were male. The paradigm of Auditory Recovery Cycle was designed to study the early stage of auditory sensory processing and the neural basis for the hypersensitivity to sounds that individuals with WS showed. One thousand and 1,500 Hz tonal stimuli were presented through earphones. Ninety percent of the stimuli were 1,500 Hz tones, and the others were 1,000 Hz tones. The subjects were asked to respond the target tones (1,000 Hz) by pressing a button. The second part of the research was to investigate the early and late neural systems that may be important to auditory language processing in individuals with WS and also to compare the organization of neural systems important to reading in those who learned to read earlier versus later. In separate conditions, subjects listened to or read sentences and then judged whether the sentences made sense. Sample sentences provided by authors included, "After the snow they clapped their hands" and "I don't go to school

on Saturday or floor" (p. 71). ERP measures were recorded during both the tone and the sentence tasks.

The results showed that tone-elicited ERPs from the individuals with WS showed a normal morphology; however, they were considerably less refractory than tone-elicited ERPs of the normal controls. That is, a short interstimulus interval reduced the amplitude of response in the normal controls but not in the individuals with WS. During the sentence task, the ERPs elicited with visually presented words were the same for the control subjects and the individuals with WS. However, the morphology of ERPs elicited during listening tasks in individuals with WS were described as highly abnormal and characterized by a greater positive/enhanced response to related lexical items. The pattern of results in this study suggested that the organization and operation of cognitive systems could be influenced by alterations in early sensory processing. Neville et al. concluded that the ERP evidence of hypersensitivity in the auditory systems of individuals with WS may provide important clues to the processes underlying the precocious nature of language skill development in these individuals. They also noted that the ERP data provide insight into differences in auditory and visual processing by individuals with WS.

Audiological evaluation of individuals with cognitive impairment

Individuals with WS and individuals with Down syndrome (DS) demonstrate an equivalent cognitive impairment (Bellugi, Wang, & Jernigan, 1994) and a high prevalence of otitis media (Balkany, Downs, & Jafek, & Krajicek, 1979; Klein et al., 1990; Van Bosel et al., 1997). Research in developing hearing assessment strategies for use with individuals with WS has not yet been initiated. As was discussed earlier, researchers have reported difficulties in conducting audiological assessment in

individuals with WS (Klein, 1994). Therefore, data concerning individuals with DS will be used as a foundation for determining procedures to be used with individuals with WS.

Kile (1996) provides a summary of the types of modifications that can be made to standard assessment procedures so that the procedures are appropriate given the behavioral characteristics of children with DS (e.g., social responsiveness, tendency to produce echolalic imitations). He reported that children with DS usually show compliant, interactive behavior during audiologic assessment, and that a wise use of social and tactile interaction and reinforcement often keeps them focused and increases time available to obtain reliable results. Children with DS tend to make more echolalic imitations than do normally developing children (Rondal, Lambert, & Sohier, 1981); therefore, Kile suggested that teaching children with DS to mimic auditory stimuli can increase consistency of response when conventional behavioral testing procedures are not successful. Kile also reported success using this modification in a 12-year-old child with DS who was seen in his clinic. This child could not be conditioned to do play audiometry and did not show consistent responses in VRA; however, he responded to pure tones by saying "Beep Beep." Kile noted that pure-tone threshold measures were obtained using this modification.

Kile conducted the ear canal volume measures for 10 children with DS and 10 children with normal development, and reported that children with DS showed a smaller ear canal volume than normally developing children. He recommended that the establishment of the ear canal volume measures of children with DS can be helpful when comparing pre- and post-tympanometry tube placement measurements. In order to investigate the prevalence of otitis media in children with DS, Kile evaluated 47 children

(mean age = 3.6 years) with DS in his clinic. He reported that 74% of the children with DS in his project demonstrated abnormal tympanograms that were consistent with the presence of otitis media, and that 58% of them had this condition bilaterally.

Kile also suggested that otoacoustic emission (OAE) measures may be useful in assessing cochlear function in children with DS. However, OAEs can only provide accurate information when individuals do not have middle ear disease. Kile recommended that, due to the high incidence of middle ear disease and the risk of hearing impairment in children with DS, early identification of hearing loss in children with DS is critical and audiologic assessment should be performed every four to six months once the hearing loss has been diagnosed.

Dille (2003) summarized strategies for effective testing, as well as common findings, for individuals with DS across the lifespan. Her report was based on a review of previous research concerning hearing assessments of individuals with DS. Dille reported that behavioral testing procedures such as VRA are successful when the appropriate developmental criterion is used. She noted that VRA procedures provide reliable audiometric information when children with DS have obtained a developmental age of 10 to 12 months, due to delayed sound localization skills in children with DS (Greenburg, Wilson, Moore, & Thompson, 1978). Dille reported "pure-tone testing to be reliable for children with DS only after 8 years of age, whereas objective procedures are more reliable for the younger population" (p. 204).

Dille recommended the use of objective techniques (e.g., ABR, DPOAE) to diagnose hearing loss in adults and children with DS, and she also noted that even when

individuals with DS have normal tympanometry and audiometry, their DPOAE measures demonstrated lower amplitude than normal age-peers.

Keiser, Montague, Wold, Maune and Pattison (1981) collected complete sets of audiometric information, including air- and bone-conduction thresholds, tympanometry and acoustic reflexes from 51 adults with DS (aged 15.1 to 51.6 years). They obtained pure-tone thresholds using standard pure-tone audiometry (hand-raising response) or conditioned play audiometry from 46 of the 51 participants. They used 15 dB HL as the criterion for identifying hearing loss, and found that 75% of the participants exhibited hearing loss in at least one ear. This study showed a sloping sensorineural hearing loss above 4,000 Hz, which is indicative of early presbycusis, and this configuration became significant by adolescence and progressed throughout their lifespans. They also reported that tympanograms from 39 % of the tested ears suggested abnormal middle ear function. Their study also indicated that reliable thresholds could be obtained using conventional pure-tone audiometry both for older children (15 years old and older) and for adults with DS.

Review of the available literature showed that procedures for the assessment hearing loss in individuals with DS have been well established. Because characteristic similarities between WS and DS have been reported in the literature (Bellugi et al., 1994; Balkany et al., 1979; Klein et al., 1990; Van Bosel et al., 1997), hearing assessments for individuals with DS may be modified for use with individuals with WS. Modified behavioral assessments described by Kile (1996) and Dille (2003) provide important information on how to approach behavioral assessment of individuals with WS based on the characteristics of individuals with DS. Once children with WS have developed the

skill of sound localization, VRA can be introduced to estimate behavioral thresholds. Modified VRA using earphones can be introduced to obtain behavioral thresholds in young children who are not able to do conditioned play audiometry. Conditioned play audiometry can be used to obtain thresholds in children by age 8 years who are able to demonstrate reliable responses to pure-tone stimuli. For adults, conditioned play audiometry and conventional pure-tone audiometry with verbal or hand-raising response can be used for hearing assessment.

Summary

In summary, previous research reports from independent labs have indicated a prevalence of sensorineural hearing loss in individuals with WS ranging from 33% (Ruangdaraganon et al., 1999; Johnson et al., 2001) to 74% (Marler et al., 2004). These studies have included small numbers of participants, provided incomplete data about test protocols, failed to define the degree of hearing loss, or have reported auditory information gathered in acoustically uncontrolled environments. As a result, interpretation of these data is confounded and application of the data to a clinical setting is limited.

Rationale for the current study

The hearing acuity of individuals with WS has not yet been well documented. It is necessary to establish audiometric performance in individuals with WS before the strengths or weaknesses of auditory processing and language abilities in WS can be accurately evaluated. The purpose of this research project was to test objectively whether individuals with WS are more sensitive to auditory stimulation (i.e., have better-than-normal thresholds) than their normally developing age-peers.

The goal of this project was to answer the following questions:

1) Do individuals with WS have normal hearing?

   Although the data available regarding prevalence of hearing loss in individuals with WS are incomplete, there is growing evidence that a significant portion of this population demonstrates hearing loss. Therefore, it was hypothesized that the prevalence of hearing loss is greater in individuals with WS than in normally hearing age peers.

2) Is there normal outer hair cell function in the individuals with WS?

   Although hyperacusis has been seen as a phenomenon similar to loudness recruitment, few studies have used OAE data to investigate the outer cell function of individuals with WS who demonstrated hyperacusis. Johnson et al. (2001) had subjects with normal hearing and absent OAEs; Marler et al. (2004) did not. Neither set of authors presented hyperacusis data for their subjects. It was hypothesized that the outer hair cell function measured by DPOAE in the current study would be consistent with audiometric data.

3) Do individuals with WS report discomfort from sound levels that are below the loudness discomfort level typically reported by individuals with loudness recruitment associated with sensorineural hearing loss?

   Although hypersensitivity to sounds has been reported in previous studies, information on loudness discomfort levels in individuals with WS is not available. It was hypothesized that individuals with WS will demonstrate lower loudness discomfort levels than individuals with sensorineural hearing loss.

CHAPTER 2

Method

Participants

Seven individuals with WS, between the ages of 7 to 43 years, were recruited through contacts with the Great Lakes Region branch of the National Williams Syndrome Association, special-needs educators, geneticists, and private physicians, and through advertisements placed in local and regional association newsletters. All subjects participated in a broad study of communication skills conducted under the direction of thesis co-advisor Jeffrey Marler. Only the audiological data gathered are reported here.

FISH was previously completed on all the participants in the current study. The results were either reviewed by Michael Netzloff, M.D., Director of Clinical Genetics at Michigan State University (MSU) for confirmation of the diagnosis of WS, or were confirmed by previous participation in the longitudinal projects of Carolyn B. Mervis, Ph.D., Director of the Cognition and Language Laboratory at the University of Louisville. In all cases, FISH revealed the absence of one copy of the elastin gene on chromosome 7 and all had clinical phenotype characteristics of WS.

Following explanation of the study, a parent or a legal guardian of each participant was asked to read and sign an informed consent form. In addition, all participants were asked to sign an assent form. Consent procedures were approved by the MSU University Committee on Research Involving Human Subjects (UCRIHS IRB #02-534). Each participant was paid $15 for participating in this study.

Due to the low incidence of WS, participants were recruited from a wide geographic area; some of the participants had to travel hundreds of miles and stayed in a

hotel the night before attending this study. Thus, it was not possible to recruit each participant for a second visit to assess test-retest variability in performance. Table 1 provides gender and age data for the seven participants in this project.

Table 1. Gender and age data for each participant (M: Male; F: Female).

Participant #	Gender	Chronological Age (years; months)
WS001	M	43;1
WS002	F	39;2
WS003	M	17;5
WS004	M	26;2
WS016	M	7;1
WS018	F	7;8
WS028	F	11;8

Procedures

Table 2 provides a summary of the test battery administrated to each participant. The test battery was modified to meet the individual needs of the participants (i.e., short attention span, difficulties in understanding the task, fatigue, fear of the probes placed in their ears). All the audiometric data were collected by the author under the supervision of an ASHA certified speech-language pathologist and/or an ASHA certified audiologist.

Table 2. Summary of quantitative measures administrated to participants in the current study.

Measures	Independent Variable	Dependent Variable
Air-conduction thresholds	dB HL (.5 to 8 kHz)	Auditory sensitivity
Bone-conduction thresholds	dB HL (.5 to 4 kHz)	Auditory sensitivity
Tympanometry	daPa at tympanic peak	Middle ear status
Speech recognition threshold (SRT)	dB HL	Speech perception ability
Word recognition score (WRS & NU-6 list)	% Correct	Speech perception ability
DPOAE	dB SPL	Outer hair cell function (Cochlea integrity)

Pure-tone thresholds

The goal was to obtain air-conduction thresholds for the frequency range of 500 to 8,000 Hz, and bone-conduction thresholds for the frequency range of 500 to 4,000 Hz

at octave intervals. For the first four participants, these measures were made after the language measures that were part of the broader study. After that portion of data collection was completed, it became clear that to reduce the impact of factors such as fatigue and short attention span on the participants' responses, the audiometric data needed to be collected at the beginning of each participant's session before the language measures were administered. For this reason, the audiometric data were gathered first for the last three participants.

The signals for the standard pure-tone audiometry were pulsed tones generated by a GSI-16 audiometer and delivered by TDH-49 earphones or mastoid placement of a bone conduction vibrator. Participants were seated in an Industrial Acoustics Corporation test booth during this portion of the study. Adult participants were asked to respond to stimuli by raising a hand, and children performed a play audiometry task in which they put a block in a puzzle board. Thresholds $\geq$ 20 dB HL at any frequency in adults and 15 dB HL at any frequency in children were considered to indicate hearing loss. The degrees of hearing impairment were defined using the categories recommended by Jerger and Jerger (1980) for adult participants and by Northern and Downs (2002) for child participants. The criterion for identifying hearing loss as sensorineural was that there be no more than a single air-bone gap greater than 10 dB HL in the pure-tone thresholds. For the individuals for whom the masked bone-conduction thresholds could not be obtained, the results of tympanometry were used to rule out middle ear pathology.

One individual in the current study was able to participate in extended high-frequency air-conduction audiometry in addition to the basic audiometric test battery. Extended high-frequency air-conduction audiometry, gathered for 10,000, 12,500, and

16,000 Hz stimuli, was conducted using a Madsen Orbiter 922 Version 2 audiometer and Sennheiser HDA200 earphones. Both GSI-16 and Orbiter 922 Version 2 systems were calibrated to the ANSI-1989 audiometric standard (ANSI, 1989).

Otoscopy and tympanometry

Otoscopy was performed to evaluate the status of the participants' external ear canals and tympanic membranes. Middle ear function in six of seven participants was evaluated using a GSI-33 middle ear analyzer using a 226 Hz probe tone and a pump speed of -200 daPa/s. Data for the seventh participant (WS018) were obtained using a GSI 28 handheld tympanometer due to technical problems with the GSI-33 at the time of data collection. To avoid the possibility that participants' concerns about this procedure would interfere with other aspects of data collection, these data were collected at the end of the assessment session.

The "pass" for tympanometry for adults was a static admittance (Peak Y) occurring between +100 and -150 daPa and peak compliance $\geq$ 0.3 mmho, an equivalent ear-canal volume (Vec) between 0.6 and 1.5 ml (ASHA, 1990), and a gradient > 0.2 (Gelfand, 2001). The "pass" criteria for children was a static admittance occurring between +100 and -150 daPa and peak compliance $\geq$ 0.2 mmho, an equivalent volume (Vec) between 0.4 and 1.0 ml (ASHA, 1990), and a gradient > 0.2 (Nozza, Bluestone, Kardatzke, & Bachman, 1992).

Speech audiometry

The goal was to measure speech threshold and understanding of speech at suprathreshold levels in individuals with WS. Speech audiometry was performed with a GSI-16 audiometer using CD recordings prepared by Auditech of St. Louis. Standard

spondee lists were used to obtain speech recognition thresholds (SRT) for each ear, and the Northwestern University Auditory Test Number 6 (NU-6; Tillman & Carhart, 1966) lists were used to obtain word recognition scores for each ear. Stimuli for the word recognition tests were presented at 30 dB SL re: SRT for participant WS001; however, after the discovery of high frequency hearing loss in participant WS001, the presentation level was changed to 40 dB SL re: SRT, which is more efficient for both individuals with normal hearing and with hearing impairment (Gelfand, 2001).

Both NU-6 and the Phonetically Balanced Kindergarten word lists (PBK-50; Haskins, 1949) were used to obtain word recognition scores for WS001. However, after working with children with WS at the national conference, it was hypothesized that given the verbal memory and language skill strengths of individuals with WS, even the children would be able to respond to the NU-6 list. The intent was to begin with an NU-6 list and shift to a PBK-50 list if necessary. As will be noted later, problems with factors such as fatigue precluded assessment of word recognition in the children who participated in the study.

Otoacoustic emissions data

The goal of this task was to investigate outer hair cell function in individuals with WS. Distortion product otoacoustic emissions (DPOAEs) were obtained using an Intelligent Hearing System (IHS) SmartOAE instrument. Data were collected for each ear in response to primary tones (labeled f1 and f2), with the ratio of these frequencies (f2/f1) equal to 1.22. The frequency of f2 varied from 1,000 to 8,000 Hz in 1/8-octave steps. Twenty-five pairs of frequencies were presented and data from 16 sweeps were recorded and averaged for the response to each pair. Primary levels were set to f1 = 65 dB SPL and

f2 = 55 dB SPL. The criterion for an emission consistent with hearing loss was that it be below the 5th percentile for ears with normal hearing as reported by Gorga et al. (1997).

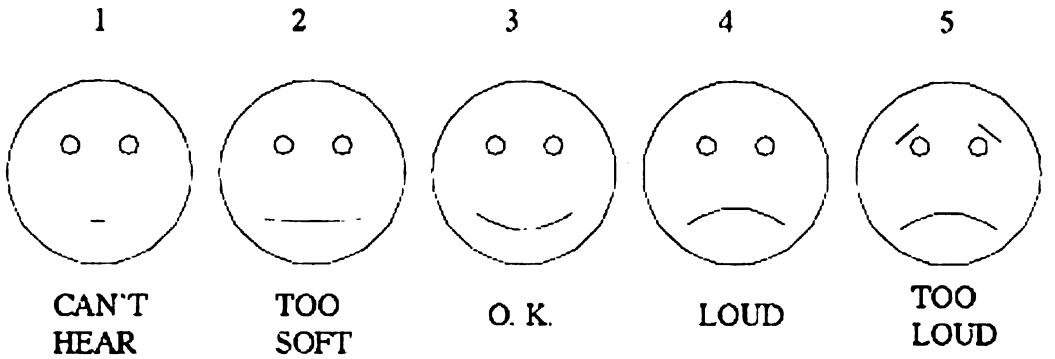
Loudness discomfort level (LDL) rating

The goal of this condition was to measure the loudness discomfort levels (LDLs) in individuals with WS and determine whether individuals with WS demonstrate lower LDLs for multi-talker babble and/or speech noise than would be expected in the general population. Speech noise generated by a GSI-16 audiometer was used for these measurements because for normally hearing individuals, speech-like signals yield lower thresholds of discomfort than do pure-tone signals (Dirks & Kamm, 1976). Thus, the impact of hyperacusis might be more readily observed with speech noise than with tones. In addition, the 12-talker multi-talker babble from the Revised Speech Perception in Noise Test (R-SPIN; Bilger, Nuetzel, & Rabinowitz, 1984) was used in this task to obtain the data to reflect real-world noisy communication situations.

In order to investigate a broad range of tolerance levels, the softest stimulus was presented at 5 dB HL, and the loudest stimulus was presented at 65 dB HL. Due to the concern that individuals with WS would not be able to tolerate loud stimuli, an upper level of 65 dB HL (approximately 85 dB SPL) was selected, lower than the loudness discomfort level for speech stimuli (100 dB SPL) obtained from a normal-hearing population reported by Dirks & Kamm (1976). Stimuli were presented binaurally at ascending levels of 5 to 65 dB HL in 10-dB steps, using TDH-49 earphones. Participants were asked to rate the signals on a five-point scale. Due to WS subjects' remarkable ability to process affect (Bellugi et al., 2000), and to recognize, discriminate, and remember unfamiliar and familiar faces (Rossen, Jones, Wang, & Klima, 1995), a picture

with five iconic faces was provided for this task (Figure 1). This picture was revised from Kawell, Kopun, and Stelmachowicz (1988) by changing the facial expressions and the wording of the rating scale. The surprised face with the word "hurts" used by Kawell et al. (1988) was removed to prevent upsetting individuals with WS who report oversensitivity to loud sounds. The facial expression "just right" in Kawell et al. (1998) was moved from the second lowest level to a neutral place (Number 3) in the current study and the wording was changed to "O.K." The instructions for this task were

Figure 1. Picture provided for LDL rating measure.



"You will hear different sounds. Point to the faces on this paper to show me how you feel. If you cannot hear it, just wait. If it is soft, point to this one (Number 2). If you feel comfortable, just like listening to music, point to this one (Number 3). If it is a little loud, but does not bother you, point to this one (Number 4). If it is too loud and you want me to turn it off, point to the last one (Number 5)." Signals were presented binaurally in a random order. Each signal level was presented twice. If the participant's response on the second trial was not consistent with the first, then the level was presented a third time. The responses rated "5 (Too Loud)" were recorded as the individuals' LDL to the stimulus. Participants' responses to both speech noise and multi-talker babble were

obtained using this task. The data for the speech noise were collected first, and then the multi-talker babble data were collected.

Hyperacusis questionnaire

A hyperacusis questionnaire that was an expansion of the form used by Klein et al. (1994) was provided to the parents of each participant. This questionnaire was constructed to obtain data on the prevalence of hyperacusis and otitis media, and was modified from Klein et al. by adding questions about the participant's past and present responses to sounds in order to investigate the relation between age and hyperacusis. A copy of the form is available in Appendix A. This questionnaire was either completed during the data-collection session or mailed back afterward. In order to be able to compare the results with the Klein et al. data, questions regarding the participant's behaviors covered hyperacusis and phonophobia together rather than as different responses to sounds.

CHAPTER 3

Results

Audiological assessment

Otoscopy

For participants WS001, WS002, WS003, WS004, WS018, and WS028, otoscopy revealed external canals within normal limits, no excessive cerumen and intact tympanic membranes. Otoscopy could not be performed on WS016 due to his unwillingness to allow the otoscope to be placed in his ears.

Immitance

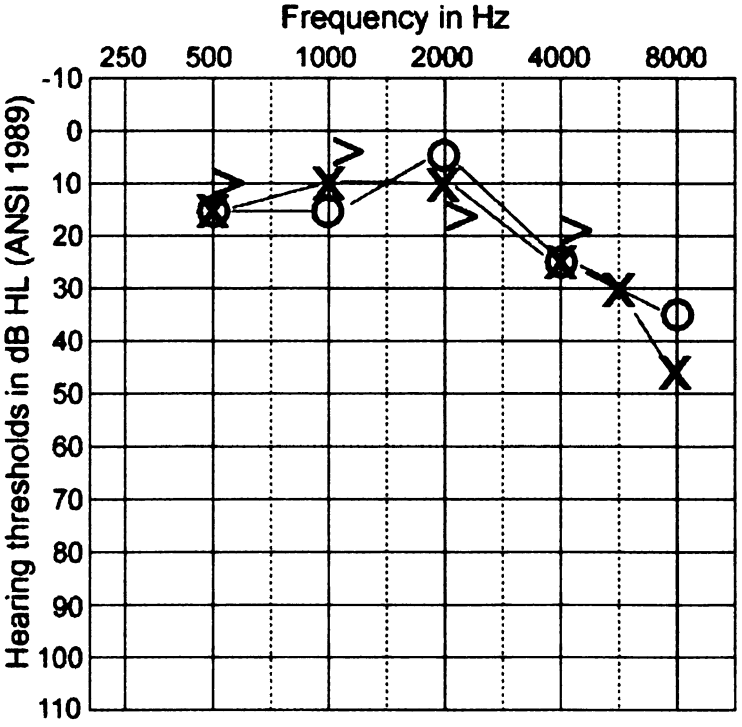
Tympanograms were obtained for both ears in five of the seven participants, and the results from four of five showed normal static admittance, equivalent volume, gradient and compliance. Participant WS001's had ear canal volumes greater than 90% of the ASHA (ASHA, 1990) data; however, his tympanograms suggested normal middle ear function. Tympanometry was accomplished only for the left ear in child participant WS016 and showed middle ear stiffness (flat tympanogram with ear canal volume 0.4 ml). Tympanometry for the right ear in WS016 could not be conducted due to the lack of a good seal and his unwillingness to continue this task. Tympanometry was attempted with child participant WS028, but could not be conducted because of her unwillingness to allow the probe to be placed in her ears. The individual data of Immitance measures are listed in the Appendix B.

Pure-tone audiometry

Air-conduction and bone-conduction thresholds for stimuli from 500 to 8,000 Hz for each participant are shown in Figure 2. Standard pure-tone thresholds were obtained

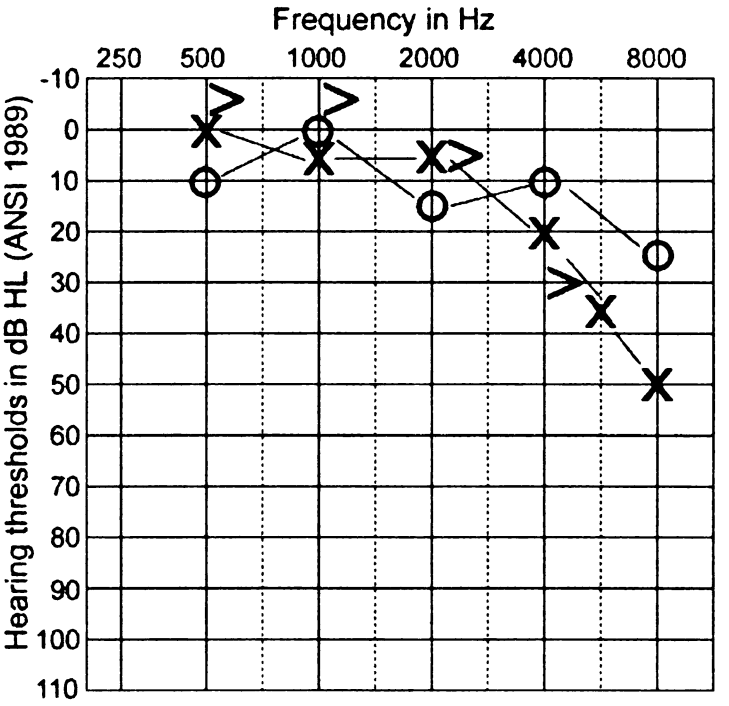
from all the participants, and extended high-frequency pure-tone thresholds were obtained from adult participant WS004. Thresholds ≥ 20 dB HL at any frequency in adults and 15 dB HL at any frequency in children were considered to indicate hearing loss. The criterion for identifying hearing loss as sensorineural was that there be no more than a single air-bone-gap ≥ 10 dB HL in the pure-tone thresholds. The degrees of hearing impairment were defined using the categories recommended by Jerger and Jerger (1980) for adult participants and by Northern and Downs (2002) for child participants. Five of the seven participants (WS001, WS002, WS004, WS018, and WS028) in the study demonstrated bilateral sensorineural hearing loss and three of those five participants (WS001, WS002, and WS018) showed a pattern of high frequency hearing loss.

Figure 2. Hearing thresholds for each participant (O: Right ear air-conduction thresholds; X: Left ear air-conduction thresholds; <: Unmasked right ear bone-conduction thresholds; >: Unmasked left bone-conduction thresholds; [: Masked right ear bone-conduction thresholds;]: Masked left ear bone-conduction thresholds).

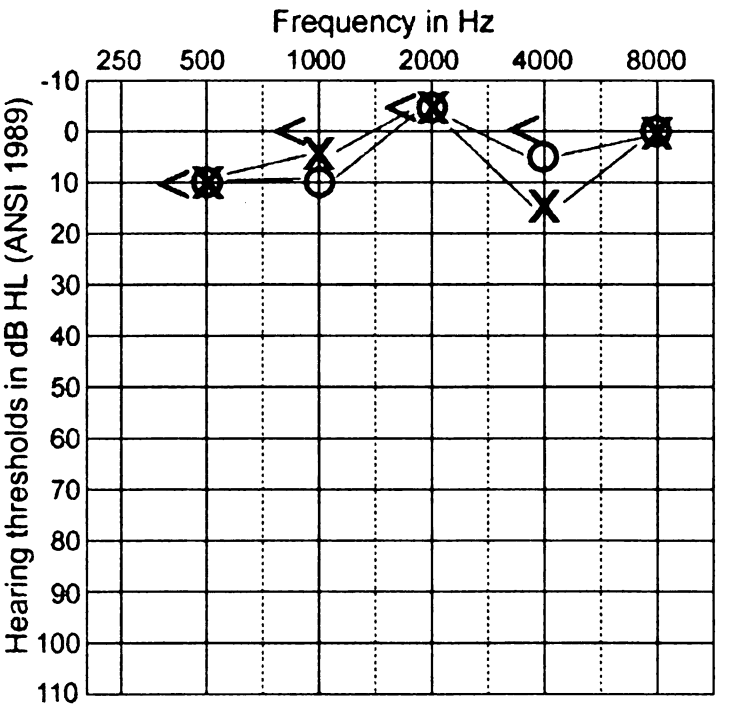


WS001

Figure 2 (continued). Hearing thresholds for each participant (O: Right ear air-conduction thresholds; X: Left ear air-conduction thresholds; <: Unmasked right ear bone-conduction thresholds; >: Unmasked left bone-conduction thresholds; [: Masked right ear bone-conduction thresholds;]: Masked left ear bone-conduction thresholds).

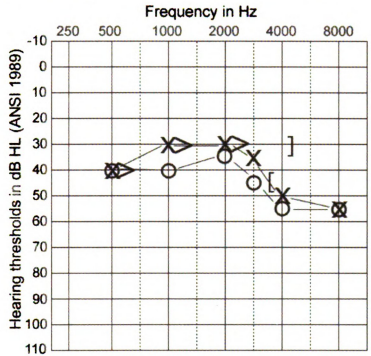


WS002

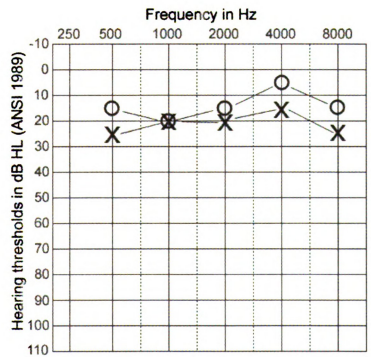


WS003

Figure 2 (continued). Hearing thresholds for each participant (O: Right ear air-conduction thresholds; X: Left ear air-conduction thresholds; <: Unmasked right ear bone-conduction thresholds; >: Unmasked left bone-conduction thresholds; [: Masked right ear bone-conduction thresholds;]: Masked left ear bone-conduction thresholds).

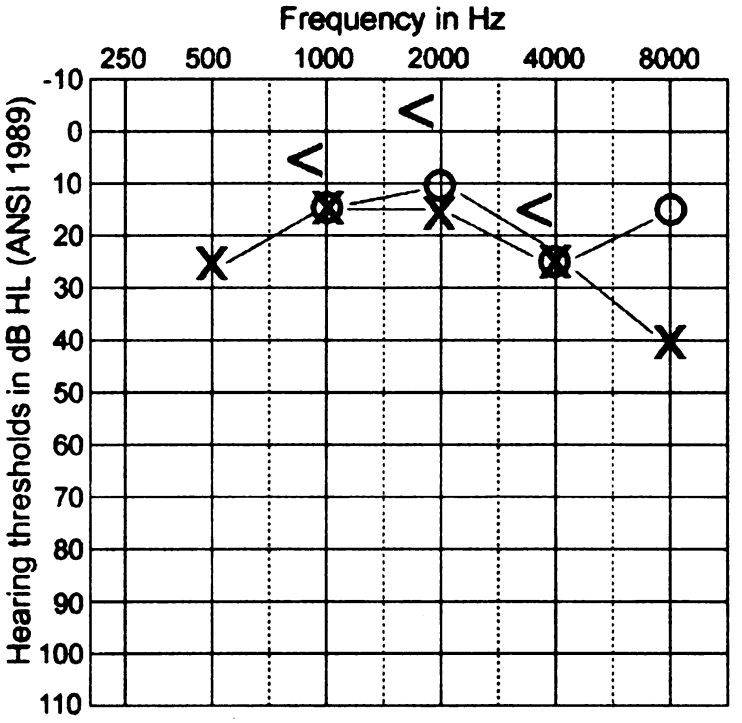


WS004

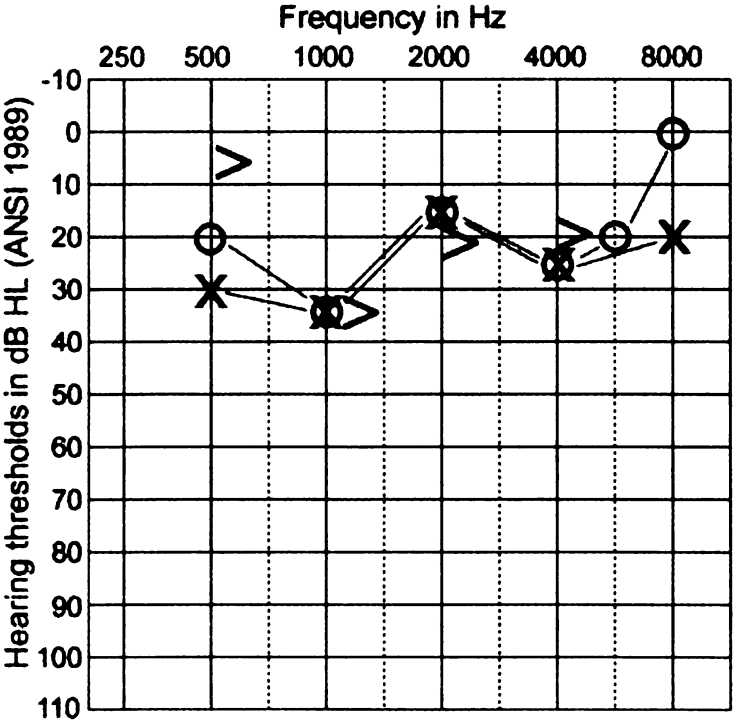


WS016

Figure 2 (continued). Hearing thresholds for each participant (O: Right ear air-conduction thresholds; X: Left ear air-conduction thresholds; <: Unmasked right ear bone-conduction thresholds; >: Unmasked left bone-conduction thresholds; [: Masked right ear bone-conduction thresholds;]: Masked left ear bone-conduction thresholds).



WS018



WS028

Participant WS001 demonstrated hearing within normal limits through 2,000 Hz in both ears. Thresholds above that point were indicative of a mild, sloping sensorineural hearing loss in the right ear and a mild-to-moderate, sloping, sensorineural hearing loss in the left ear. False-positive responses were only observed when high-frequency stimuli were presented and re-instruction was provided when they occurred. Participant WS001 was one of the two individuals in the current study who reported presence of tinnitus during the test. It is possible that participant WS001 had difficulty discriminating between the pure-tone stimuli and his tinnitus. The reliability of his pure-tone data was judged to be good.

Pure-tone thresholds for participant WS002's right ear were within normal limits through 4,000 Hz, and then indicated mild sensorineural hearing loss at 8,000 Hz. Left ear pure-tone thresholds were within normal limits through 2,000 Hz, and were indicative of mild-to-moderate, sloping, sensorineural hearing loss. The reliability of her pure-tone data was judged to be good. Participant WS002 showed a bone-conduction threshold poorer than air-conduction thresholds (i.e., a reverse air-bone-gap) at 4,000 Hz in both ears. The air-conduction and bone-conduction thresholds are within ±10 dB of each other; therefore, "it is possible and perfectly acceptable for a bone-conduction threshold to be a bit poorer than air-conduction thresholds at the same frequency, even if everything is done correctly" (Gelfand, 2001, p. 155-156). However, the 20 dB reverse air-bone-gap at 4,000 Hz in her right ear was not typical. The placement of the bone oscillator was checked and the thresholds at 4,000 Hz were re-tested during the data collection in order to rule out technical errors. It is important to note that participant WS002 also reported the presence of ringing in her ears during the test. It is possible that

participant WS002 had difficulty discriminating between the pure-tone stimuli and her tinnitus.

Participant WS003 was the only participant in the current study who showed normal pure-tone thresholds across the entire test frequency range in both ears. The reliability of his pure-tone data was judged to be good. Although the audiogram of WS003 showed a 15 dB air-bone gap at 4,000 Hz in the left ear, tympanograms for both of WS003's ears revealed normal middle ear function. Therefore, the air-bone-gap at 4,000 Hz in the left ear was not considered to indicate the presence of a conductive component to the hearing loss.

Participant WS004 demonstrated a bilateral mild (500 to 2,000 Hz) to moderate (above 2,000 Hz) sensorineural hearing loss. He showed some inconsistent responses to the stimuli during this task and re-instruction was provided when false-positive responses occurred. The reliability of his pure-tone data was judged to be only fair due to his inconsistent responses.

One child (WS016) showed a left unilateral hearing loss of an unidentified type. Tympanometric data for that ear were indicative of middle ear pathology, and mixed hearing loss could not be ruled out without bone-conduction data. Bone-conduction data were not collected because the child lost interest in the task and could not be coaxed to continue. The reliability of his pure-tone data was judged to be fair.

Pure-tone thresholds for participant WS018's left ear showed mild, rising, sensorineural hearing loss at 500 to 2,000 Hz, and then indicated mild-to-moderate, sloping, sensorineural hearing loss range in the high frequencies. Air-conduction thresholds for the right ear were obtained only from 1,000 to 8,000 Hz due to the

participant's loss of interest in the task. Right ear pure-tone thresholds showed a mild sensorineural hearing loss at 1,000 Hz, then a rise to the normal hearing range at 2,000 Hz, and finally a slope to mild hearing loss range at 4,000 to 8,000 Hz. The reliability of his pure-tone responses was judged to be good. No bone-conduction threshold was obtained at 500 Hz due to participant WS018's loss of interest in the task. Sensorineural hearing loss at 500 Hz in the left ear was confirmed by the normal immitance results for WS018.

Pure-tone thresholds for participant WS028's right ear showed mild-to-moderate, sloping, sensorineural hearing loss from 500 to 1,000 Hz, then a rise to mild, essentially flat, sensorineural hearing loss at 2,000 to 6,000 Hz, and finally a rise to normal-hearing range at 8,000 Hz. Left ear pure-tone thresholds showed mild to moderate, essentially flat, sensorineural hearing loss at 500 to 1,000 Hz, then a rise to mild, essentially flat, sensorineural hearing loss at 2,000 to 8,000 Hz. Due to WS028's fear of earphone placement and refusal to stay in the testing booth at the beginning of the audiological evaluation, the procedure was modified to help participant WS028 gradually overcome her fear. Play audiometry, using a Beltone 120 portable audiometer, was performed during the language evaluation session to help the participant get used to the earphone placement. WS028 was conditioned to put a block in a puzzle board when she heard pulsed tones through the earphones. VRA using FM signal (warble tones) was introduced after play audiometry to help her become comfortable with the testing booth. After this, participant WS028 was able to do play audiometry (using pulsed tones) in the testing booth, and her pure-tone data were collected using the GSI 61 audiometer. The reliability of her pure-tone data was judged to be good.

Extended high-frequency (EHF) data were obtained only for WS004, and they were converted to dB SPL by adding the reference equivalent threshold sound pressure levels (RETSPLs) recommended by Frank (2001). The test reliability was judged to be fair due to his false positive responses to stimuli. Table 3 shows the data comparison of EHF air-conduction thresholds of WS004 and the mean data of 32 individuals (16 male and 16 female) aged 20 to 30 years reported by Stelmachowicz, Beauchaine, Kalberer and Jesteadt (1989). These data were used for comparison because they were obtained from a population that included the age range of the adult participants in the current study (i.e., 240 individuals in the range of 10 to 60 years), the subgroups of each age range contained equal numbers of male and female individuals, and all of the individuals were reported to have normal middle ear function and a negative history of excessive noise exposure or ototoxic drug use. The data comparison shows that WS004's EHF thresholds were 56.6 dB or greater than the mean data for individuals aged 20 to 30 years reported by Stelmachowicz et al. (1989).

Table 3. Data comparison of extended high-frequency air-conduction thresholds obtained from WS004 and mean thresholds for 32 individuals age 20 to 30 years reported by Stelmachowicz et al. (1989).

	Right Ear (dB SPL)			Left ear (dB SPL)		
Frequency (kHz)	10	12.5	16	10	12.5	16
WS004	87.6	NR	NR	112.6	NR	NR
Stelmachowicz et al., 1989	31	38	72	31	38	72

NR= No response when stimuli reached the maximum audiometer output.

Speech audiometry

Speech recognition thresholds (SRT) and word recognition scores (WRS) were obtained from the four adult participants, ages 17 to 43 years. Pure-tone averages were calculated using the thresholds for 500, 1,000, and 2,000 Hz, the SRT data, and the WRS

data are shown in Table 4. Each of the adult participants demonstrated excellent word recognition as indicated by scores ≥ 92%. Speech audiometry could not be conducted with three child participants due to their loss of attention to the task.

Table 4. Speech audiometry and pure-tone-average data for the four adults who completed the speech audiometry tasks.

	Right Ear				Left Ear			
	PTA	SRT	WRS		PTA	SRT	WRS	
			Presentation level (dB HL)	% Correct			Presentation level (dB HL)	% Correct
WSA001	12 dB	15 dB	45 dB	98	12 dB	15 dB	45 dB	98
WSA002	8 dB	5 dB	45 dB	100	7 dB	0 dB	40 dB	100
WSA003	5 dB	10 dB	50 dB	96	3 dB	5 dB	45 dB	96
WSA004	38 dB	25 dB	65 dB	100	33 dB	25 dB	65 dB	92

Data for three of the four participants indicate consistency between pure-tone averages (PTA) and SRTs (i.e., a difference that is ≤ 6-12 dB; Brandy, 2002). The fourth participant's data were consistent for the left ear but differed by 13 dB in the right ear. Participant WS004 demonstrated some inconsistent responses to pure-tone stimuli and needed more re-instructions than the other participants during the pure-tone audiometric measurement. It is possible that this contributed to a slightly elevated pure-tone average. Any inconsistent responses (differing by more than 5 dB) can lead to a different result in terms of the consistency between PTA and SRT. This individual also had difficulty convincing his family members that he has hearing loss. Therefore, it is possible that WS004 exaggerated the severity of hearing loss in order to convince his family that he is hearing impaired.

Otoacoustic emissions data

DPOAE data were obtained from five of the seven individuals (WS001, WS002, WS003, WS004, and WS018). DPOAE measures were attempted with the two remaining child participants (WS016 and WS028), but they could not be conducted because of the children's unwillingness to allow the probe to be placed in their ears. DPOAE data for the participants in this study are shown in Figure 3.

The 95th percentile data for DPOAE amplitude for hearing-impaired ears and the 5th percentile data for DPOAE amplitude for normally hearing ears, as reported by Gorga et al. (1997) are plotted in Figure 3 for comparison. All points below the Gorga et al. data (i.e., at a lower dB SPL) have a significant probability of coming from ears with hearing loss. Data above the 95th percentile for hearing-impaired ears are considered indicative of hearing within normal limits. Data between the 5th percentile for normally hearing ears and the 95th percentile for hearing-impaired ears are considered difficult to interpret, and therefore ambiguous.

Of the five data sets shown in Figure 3, one was from an individual who had hearing within normal limits (WS003) and four were from individuals who demonstrated bilateral sensorineural hearing loss. The DPOAE data support the audiological finding of hearing loss in three (WS001, WS002, WS018) of the four participants who showed bilateral hearing loss.

Participant WS001 demonstrated pure-tone thresholds that sloped from within normal limits to the mild (right) or moderate (left) hearing loss range from 4,000 to 8,000 Hz. His DPOAE amplitudes in the frequency range of 1,500 to 3,000 Hz were indicative of normal hearing. DPOAE amplitudes fell below the 5th percentile of the Gorga et al.

data in the frequency range of 4,000 to 6,000 Hz, and thus were consistent with hearing-impaired ears. Thirty-eight percent of the DPOAE responses from the right ear and 33% of the responses from the left ear fell within the ambiguous range.

Participant WS002 also demonstrated thresholds that sloped from within normal limits to the mild (right) or moderate (left) hearing loss range at 8,000 and/or 4,000 to 6,000 Hz. Most of the DPOAE amplitudes above 2,000 Hz in her right ear were indicative of normal hearing. Her DPOAE amplitudes fell below the 5th percentile of Gorga et al. data in the frequency range of 6,000 to 7,000 Hz in the left ear, and thus were consistent with hearing-impaired ears. Thirty-three percent of the DPOAE responses from the right ear and 42% of the responses from the left ear fell within the ambiguous range.

Figure 3. Individual DPOAEs for participants WS001, WS002, WS003, WS004, and WS018.

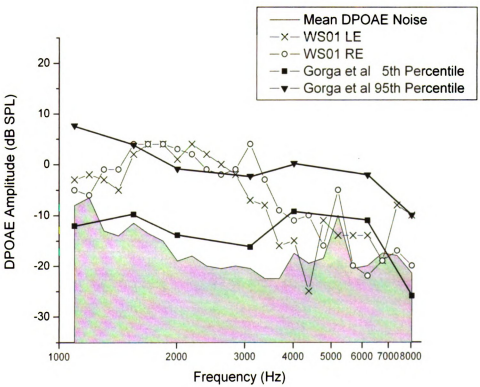
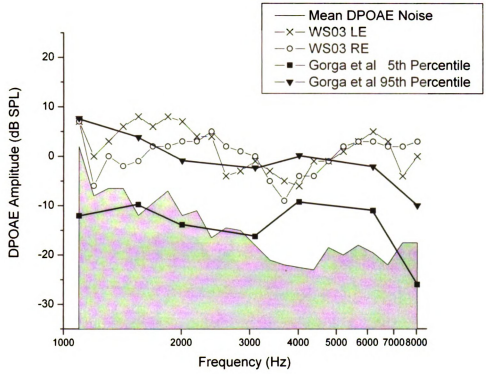


Figure 3 (continued). Individual DPOAEs for participants WS001, WS002, WS003, WS004, and WS018.



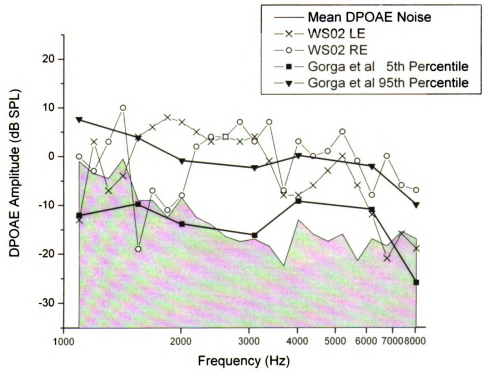
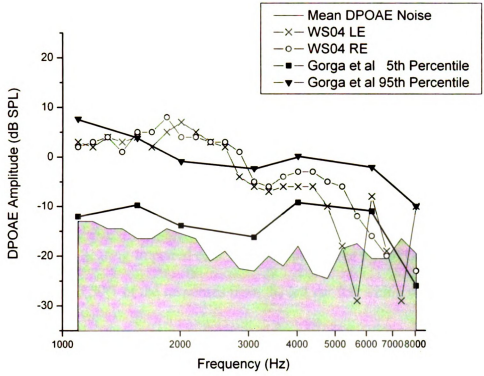
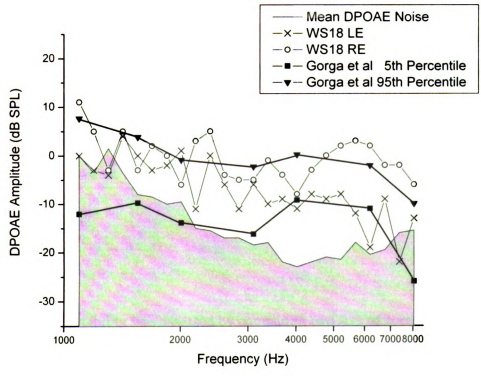


Figure 3 (continued). Individual DPOAEs for participants WS001, WS002, WS003, WS004, and WS018.





Participant WS003's pure-tone thresholds were within normal limits bilaterally. His DPOAE amplitudes in the frequency range of 1,500 to 3,000 Hz and above 5,000 Hz were indicative of normal hearing in both ears. Forty-six percent of the DPOAE responses from the right ear and 42% of the responses from the left ear fell within the ambiguous range.

Participant WS004's pure-tone thresholds indicated bilateral mild-to-moderate hearing loss across the stimulus frequency range. His DPOAE amplitudes between 1,500 and 3,000 Hz were indicative of normal hearing and DPOAE amplitudes above 5,000 Hz were consistent with hearing-impaired ears. Forty-six percent of the DPOAE responses from the right ear and 50% of the responses from the left ear fell within the ambiguous range.

Participant WS018 demonstrated thresholds that rose from mild hearing loss (at 1,000 Hz) to normal-hearing range (at 2,000 Hz), and then sloped to mild hearing loss in the right ear. She also demonstrated thresholds that sloped from mild-to-moderate hearing loss (above 4,000 Hz) in the left ear. The DPOAE amplitudes in the frequency range of 2,000 to 2,500 Hz and at frequencies above 4,500 Hz in her right ear were indicative of normal hearing. The DPOAE amplitudes in the frequency range of 5,500 to 6,500 Hz in her left ear were consistent with hearing impaired ears. Fifty-four percent of the DPOAE responses from the right ear and 75% of the responses from the left ear fell within the ambiguous range.

Test-retest reliability

Test-retest reliability was assessed by comparing two sets of pure-tone threshold data and DPOAE data for the youngest participant in the current study (WS018). Data

from the current study were compared to data obtained at the 2004 Williams Syndrome Association Conference two months earlier. It is important to note that the subjects for the study of Marler et al. (2004) and the subjects for this project do not overlap (i.e., because the subject had follow-up assessment for this project, she was not included in the conference screening data set reported).

Table 5 provides the pure-tone data comparison. Due to environmental constraints (i.e., ambient noise levels), the criterion for passing a hearing screening at the conference was 20 dB HL. A threshold search was conducted only for those frequencies at which the individual failed the screening. Differences between thresholds obtained at the conference and thresholds obtained at the clinic were within 5 dB HL. This is considered to be typical test-retest variation (Harrell, 2002).

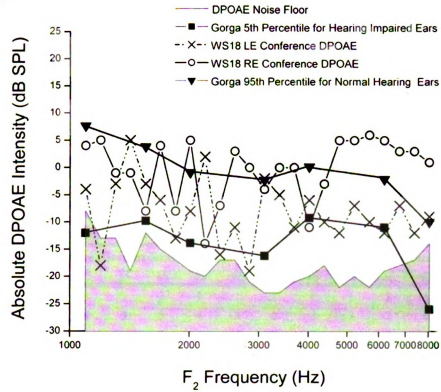
Table 5. Participant WS018's air-conduction thresholds obtained from hearing screening and diagnostic evaluation.

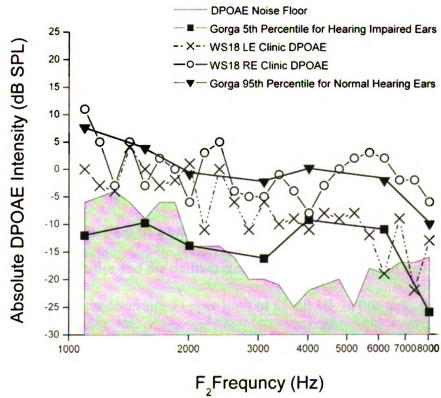
	Right ear (dB HL)						Left ear (dB HL)					
Freq. (Hz)	**500**	**1k**	**2k**	**4k**	**6k**	**8k**	**500**	**1k**	**2k**	**4k**	**6k**	**8k**
Conference data	Pass	Pass	Pass	30	25	Pass	25	Pass	Pass	30	45	40
Oyer Clinc data	20	15	10	25	25	15	25	15	15	25	40	40

"Pass" equals to a response to 20 dB HL stimuli.

Figure 4 provides the DPOAE data comparison. The DPOAE parameter settings in Marler et al. were the same as the current study. These two sets of data show a similar pattern and are consistent with audiometric findings. Sladen, Lamb and Hall (1996, as cited in Hall, J. W. III, 2000) reported that the intrasubject DPOAE amplitude variability was typically between 6 and 7 dB and as low as 3 dB SPL. The intrasubject amplitude variance for DPOAE by F2 frequency was within the dB range reported by Sladen et al. (1996) for 16 of 24 data points, including all of the points in the region between 4,000 and 6,000 Hz, the region in which data are consistent with hearing loss.

Figure 4. DPOAEs of participant WS018 obtained from hearing screening and diagnostic evaluation.





Loudness discomfort level (LDL) rating

LDL rating data were obtained from four adult participants (WS001, WS002, WS003, and WS004). The rating system was changed from a three-item scale to a five-item scale after the data of Participant WS001 were collected. Therefore, the LDL data from WS001 were excluded from the data analysis. The LDL data were not collected from any of the children due to the participants' fatigue and loss of attention to the task. The data for WS002, WS003 and WS004 are shown in Table 6.

Table 6. Loudness discomfort level rating scale data for the adult participants

	Speech noise stimulus level (dB HL)							Multi-talker babble stimulus level (dB HL)						
	5	**15**	**25**	**35**	**45**	**55**	**65**	**5**	**15**	**25**	**35**	**45**	**55**	**65**
WS002	2	2	3	3	3	5		1	2	3	3	5	5	
WS003	1	3	4	3	5	4	5	2	2	3	2	4	5	5
WS004	1	2	3	3	3	4	4	1	2	3	3	3	4	4

1=cannot hear; 2=too soft; 3=O.K.; 4=loud, but tolerable; 5=too loud.

Participant WS002, who showed a mild sensorineural hearing loss at 8,000 Hz in the right ear, and a mild-to-moderate sensorineural hearing loss above the 4,000 Hz range in the left ear, rated the speech noise stimuli as "too loud" at 55 dB HL and the multi-talker babble as "too loud" at 45 and 55 dB HL. Participant WS003, who showed hearing within normal limits in pure-tone audiometry, rated the multi-talker babble as "too loud" at 45 dB HL, but was inconsistent in identifying the discomfort level for the speech-noise stimuli (i.e., rated 45 dB HL as "too loud," but rated 55 dB HL as "loud, but tolerable"). Participant WS004, who showed a bilateral mild-to-moderate sensorineural hearing loss, rated both the speech noise and the multi-talker babble stimuli as "loud, but tolerable" at 65 dB HL. Due to the predetermined upper limit of 65 dB HL in this task, the investigator could not determine the level that participant WS004 would have rated "too loud."

Of the three individuals who performed the LDL tasks, two (WS002 and WS004) responded consistently to the stimuli and two (WS002 and WS003) demonstrated LDLs lower than the predetermined upper limit. Although limited, these findings were consistent with expectations prior to the initiation of the study.

The following unexpected findings were also observed. Participant WS002 showed bilateral high-frequency sensorineural hearing loss, and her LDL was lower than that of WS003, who showed pure-tone thresholds within normal limits. Participant WS003 demonstrated inconsistent ratings of the stimuli. Participant WS004's actual LDL was presumably higher than the upper limit allowed by the research design.

Hyperacusis questionnaire

Hypersensitivity to sounds

During or after data collection, all of the parents of the participants returned questionnaires regarding the individual's responses to loud sounds and history of hearing loss, and middle ear disease. All of them reported that their children were "bothered or frightened" by certain sounds during childhood and that all of them, including those now adults, were still "bothered or frightened" by these sounds when this survey was conducted. The following items were those most commonly selected by the parents as the sounds that cause the greatest discomfort: "firecracker (six out of seven)," "fire engine siren (five out of seven)," and "power saw (four out of seven)." Additional sounds selected by some parents as the sounds that caused the greatest discomfort to their children were "motorcycle," "loud auto muffler," "electric drill," "dog barking," "lawn mower," "squeaking toys," "hammering a nail," and "food blender."

The parents of three participants (WS001, WS003, and WS016) reported that the discomfort level associated with loud sounds became reduced as their children grew up; the parents of the remaining participants reported that the discomfort level remained the same. Only the father of individual WS003 reported that he had looked for professional help for his child's hyperacusis. That parent noted that his child liked to play music and did not complain about its intensity. The parent of participant WS004 reported that his child likes car races, so he wears ear protection and forces himself to tolerate the loud noises. When responding to a question focused only on "fear," the parents of participant WS001 and participant WS004 reported that these individuals continue to dislike the types of sounds that were reported earlier (e.g., firecracker, fire engine siren, power saw), but that these sounds no longer cause fear. The parents of the youngest participant (WS018) reported that their daughter still had fear of these sounds; however, during the experimental session, they stated that her hyperacusis had never been as disruptive as reported by other parents of children with WS. The parents of the remaining adult and child participants reported that fear of loud sounds was still an issue at the time this survey was conducted.

The following descriptions of the participants' most frequent responses to loud sounds were noted in the parent reports. Six out of seven sets of parents reported that the participant "covers ears with hands." Five out of seven reported "cries," five out of seven reported "cringes" and four out of seven reported "says something like I don't like it." All of the parents reported that the characteristics of sounds that most bothered the participants were how loud the sound is and a sudden sound.

Hearing

Six of seven sets of parents reported that no diagnosis of permanent hearing loss had been made before this study. The seventh, the parent of WS018 did not answer this question. During the evaluation session, the parent of WS018 reported that her child failed a hearing screening in kindergarten and that no follow-up diagnostic testing had been done prior to the current study. She was unable to provide any additional details regarding the hearing loss. The parents of WS018 also reported that hearing loss was not a noticeable problem in their daughter. Participant WS004 reported a history of hearing loss and rarely had middle ear infections; however, his parent reported that he had no diagnosis of permanent hearing loss and that he was "obsessive about being deaf." His parent also reported (as noted above) that participant WS004 likes to attend car races and forces himself to tolerate loud noise by using earplugs. Participant WS004 also reported that he understands the importance of using hearing protection when exposed to noise and that he uses earplugs to protect his hearing.

Middle ear infection

Two of seven sets of parents reported that their children (WS002 and WS003) never had ear infections, and three additional sets reported that their children (WS001, WS004, and WS018) rarely had ear infections. WS004's parents reported that he never had an ear infection in childhood, and rarely has one now. The remaining two sets of parents reported that their children (WS016 and WS028) had recurrent ear infections (several or constant) during childhood, but that they were less frequent as the participants grew up.

CHAPTER 4

Discussion

Audiological assessment

At the initiation of the current project, the prevalence of hearing loss in individuals with WS had not been widely studied. Due to the characteristic of hypersensitivity to sounds, most individuals with WS have been assumed to have normal or better than normal hearing. In fact, data reported by Ruangdaraganon et al. (1999) and Johnson et al. (2001) have shown that individuals with WS have a higher prevalence of hearing loss (33%) than is demonstrated by the children ages 3 to 10 years (9 in 10,000) (Van Naarden, Decoufle, and Caldwell, 1999). In addition, hearing screening and DPOAE data obtained by Marler et al. (2004) during a Williams Syndrome Association conference revealed that a surprising number of individuals with WS (74% of those examined) had poorer-than-expected auditory function. A report published by Cherniske et al. (2004) during the course of the current study confirms and expands previous reports of hearing loss in adults with WS. Cherniske et al. investigated multisystem function (i.e., cognitive, cardiovascular, musculoskeletal, gastrointestinal, endocrine, visual and auditory systems functioning) in 20 adults with WS over 30 years of age. Sixteen of these 20 adults had "standard audiological testing" (p. 256) in a hospital setting. Seventy-five percent had high-frequency sensorineural hearing loss. Half of these hearing losses were unilateral.

In the current study, five of seven participants had bilateral symmetrical sensorineural hearing loss. The configurations were either hearing loss only in the high

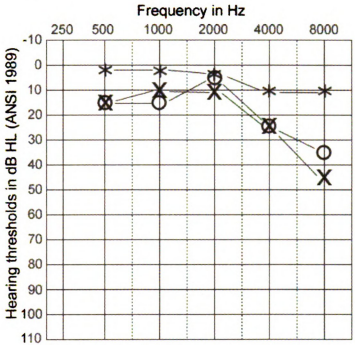
frequency range or hearing loss across the entire stimulus range, with thresholds greater (poorer) above 2,000 Hz than below 2,000 Hz.

Figure 5 compares the four adult participants' air-conduction thresholds to the median threshold data for the general population calculated using data and formulas provided by ISO 7029-1984. The ISO 7029-1984 data provides the audiometric information for the frequency range of 125 to 8,000 Hz from the otologically normal individuals aged range of 18 to 70 years. According to ISO 7029-1984, the criteria for identifying otologically normal was "a person in a normal state of health who at the time of testing is free from excess wax in the ear canals, is without known ear pathology and who has no history of undue exposure of noise" (p. 2). The report also provides formulas for calculating the expected value of the median hearing threshold shift relative to a group of persons aged 18 years. The median thresholds in Figure 5 were generated from data sets selected according to each participant's age and gender.

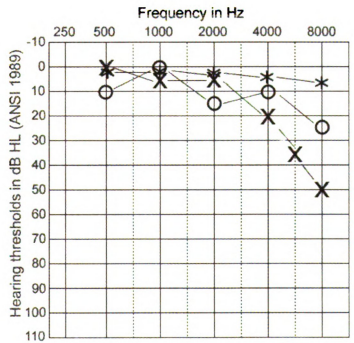
All of the air-conduction thresholds for individuals WS001 and WS004 were equal to or poorer than the median ISO 7029-1984 thresholds. The pure-tone thresholds of WS002 were equal to or poorer than the median thresholds with three exceptions. The left ear threshold for 500 Hz was 1.6 dB better than the median and the thresholds for both ears at 1,000 Hz were 1.83 dB better than the median. The data from WS003 were equal or poorer than median thresholds with just two exceptions. Thresholds from both ears at 2,000 Hz were 5 dB better than the median.

This data comparison shows that four of the adult participants had at least three thresholds in each ear that were 15 to 55 dB poorer than would be expected, given age and gender. For WS001 and WS002, these differences were greatest above 2,000 Hz.

Figure 5. Comparison of the four adult participants' air-conduction thresholds to expected median thresholds. The median was calculated from data and formulas provided by ISO 7029-1984 according to each individual's age and gender (O: Right ear air-conduction thresholds; X: Left ear air-conduction thresholds; *: Median threshold data calculated from ISO 7029-1984).

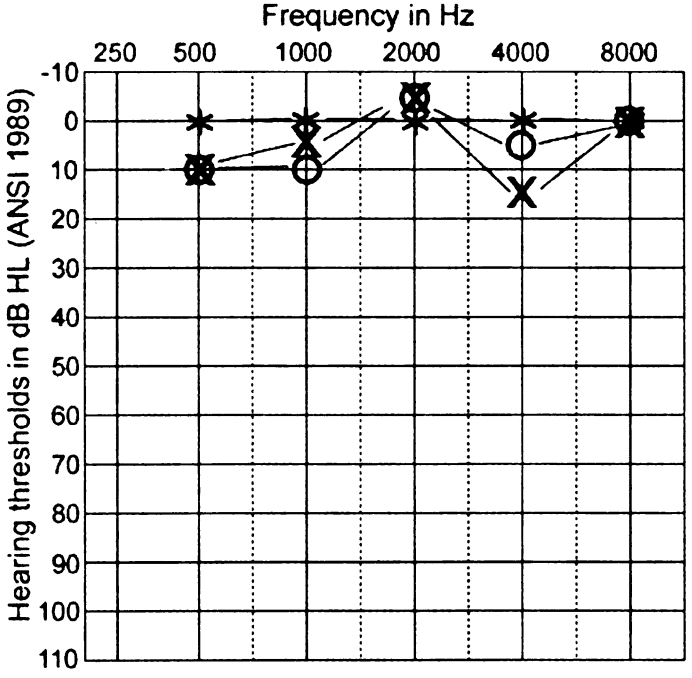


WS001 (37 y/o, male)

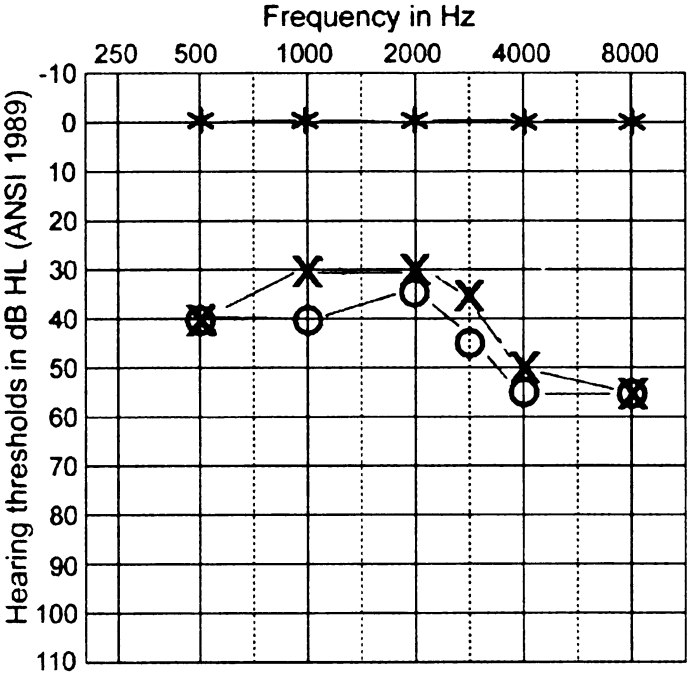


WS002 (29 y/o, female)

Figure 5 (continued). Comparison of the four adult participants' air-conduction thresholds to expected median thresholds. The median was calculated from data and formulas provided by ISO 7029-1984 according to each individual's age and gender (O: Right ear air-conduction thresholds; X: Left ear air-conduction thresholds; *: Median threshold data calculated from ISO 7029-1984).



WS003 (17.5 y/o, male)



WS004 (26 y/o, male)

Thresholds for WS004 were 30 to 55 dB poorer than would be expected given age and gender at all the test frequencies.

Figure 6 compares the three child participants' air-conduction thresholds to the median threshold data that was reported by Holmes, Niskar, Kieszak, Rubin, and Brody (1994). Holmes et al. obtained hearing thresholds in the frequency range of 500 to 8,000 Hz from 6,166 children ages 6 to 19 years in the Third National Health and Nutrition Examination Survey. In the study of Holmes et al., physical examinations and home interviews were performed to determine test exclusions (e.g., draining ears, colds, sinus problems, or exposure to loud noise over the past 24 hours). This study provides the mean and median thresholds by ear, gender, frequency and age group (6 to 11 years, 12 to 19 year). The median thresholds from the age group that matched each child participant's age were used for data comparison in Figure 6.

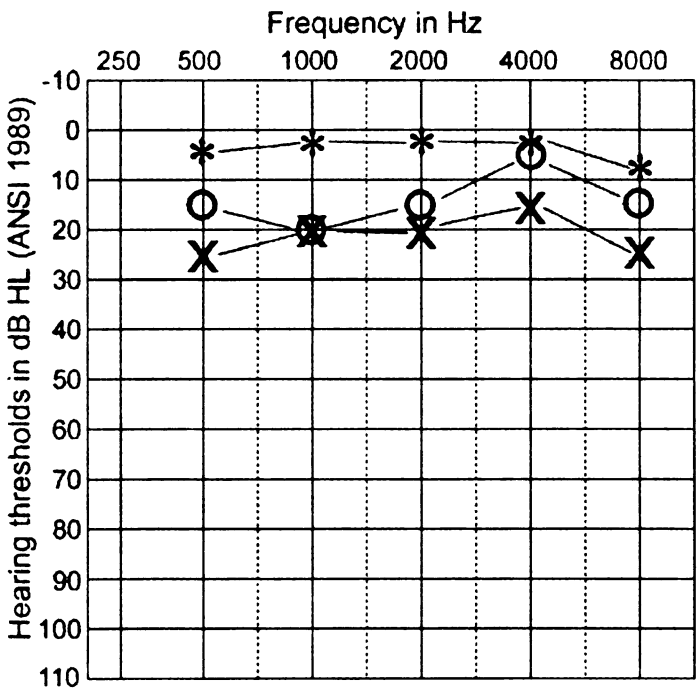
All of the air-conduction thresholds for individual WS016 and WS018 were poorer than the median thresholds reported by Holmes et al. The pure-tone threshold at 8,000 Hz in the right ear of participant WS028 was 8 dB better than the median thresholds for the female ages 7 to 11 years in Holmes et al. Only one pure-tone threshold at one frequency in one participant was observed to be better than those of normal hearing age-peers.

With the exception of isolated thresholds from three individuals (WS002, WS003, and WS028), all the participants in the current study showed pure-tone thresholds poorer than would be expected from age peers in the general population. Five of the seven participants in the current study showed bilateral sensorineural hearing loss. These

findings indicate that the prevalence of hearing loss in individuals with WS is greater than previously reported.

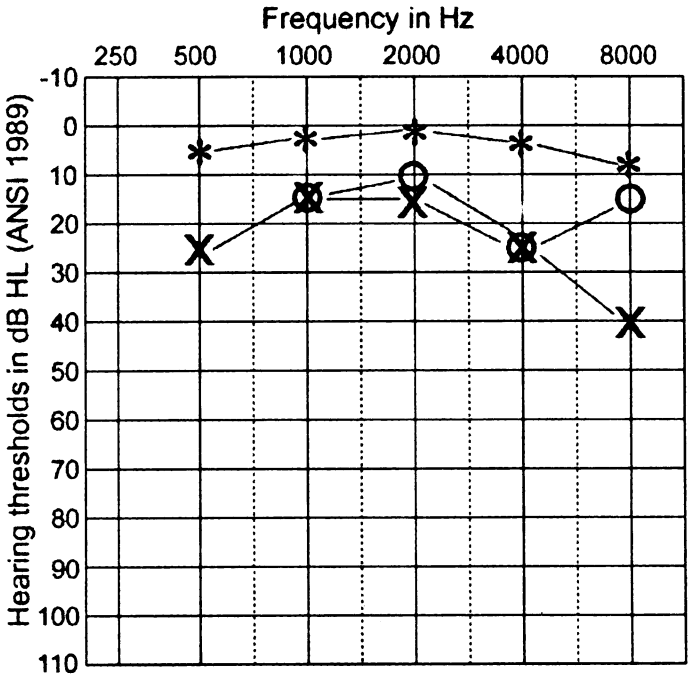
Although six of seven participants in the current study had no prior documented diagnosis of permanent hearing loss, four of these six participants showed sensorineural hearing loss. In addition, parental reports in the current study showed that most (six out of seven sets) of the participants' parents were not aware of the possibility of hearing loss in individuals with WS. They reported assumptions that their family members with WS would not have hearing impairment because of their oversensitive responses to sounds. These findings provide clear evidence that hearing impairment in individuals with WS has often been overlooked.

Figure 6. Comparison of the three child participants' air-conduction thresholds to the median thresholds reported by Holmes et al. (1994). The median was selected according to each participant's age and gender (O: Right ear air-conduction thresholds; X: Left ear air-conduction thresholds; *: Median threshold data reported by Holmes et al.).

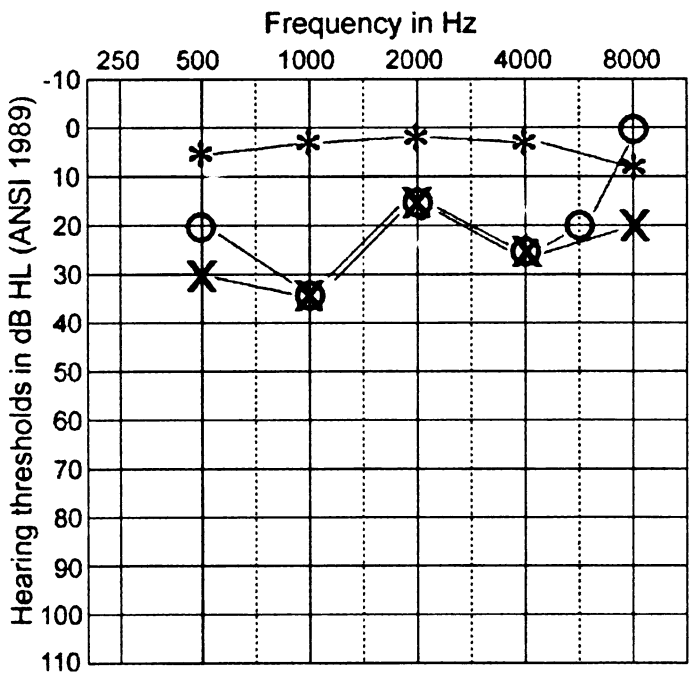


WS016 (7 y/o, male)

Figure 6 (continued). Comparison of the three child participants' air-conduction thresholds to the median thresholds reported by Holmes et al. (1994). The median was selected according to each participant's age and gender (O: Right ear air-conduction thresholds; X: Left ear air-conduction thresholds; *: Median threshold data reported by Holmes et al.).



WS018 (7.5 y/o, female)



WS028 (11.5 y/o, female)

Previous studies have shown the impact of hearing impairment on children's language development, academic achievements and social behaviors. Davis, Elfenbein, Schum and Bentler (1986) investigated the effects of degree of hearing impairment, age, and other factors on intellectual, social, academic, and language behaviors on 40 children with mild to moderately severe hearing impairment. According to Davis et al. (1986) "children with any degree of hearing loss appear to be at risk for delayed language development of verbal skills and reduced academic achievement" (p. 61). The researchers also reported that the two children with the mildest hearing loss in their study demonstrated some of the greatest vocabulary delays (i.e., 3 to 4 years on the Peabody Picture Vocabulary Test - Revised).

Bess, Dodd-Murphy and Parker (1998) defined minimal sensorineural hearing loss for three groups: unilateral sensorineural hearing loss (USHL, pure-tone average of 20 dB HL or greater in the impaired ear), bilateral sensorineural hearing loss (BSHL, pure-tone average between 20 dB and 40 dB HL bilaterally), and high-frequency sensorineural hearing loss (HFSHL, air-conduction thresholds greater than 25 dB HL at two or more frequencies above 2,000 Hz in one or both ears). They reported that 5.4 % of the children in the schools they worked with exhibited minimal sensorineural hearing loss (MSHL) and that 37% of children with MSHL failed at least one grade compared to a district rate of about 3%. Two of the three child participants (WS018 and WS028) in the current study met the Bess et al. definition of MSHL. Participant WS018 exhibited HFSHL in her left ear and participant WS028 demonstrated BSHL. Not only did these two child participants exhibit MSHL, but also two of the four adult participants (WS001 and WS002) demonstrated HFSHL. It is clear that because of MSHL, these two child

participants could be experiencing greater difficulties in school than do their normal-hearing age-peers. Both the Davis et al. and Bess et al. studies revealed the importance of identification of hearing loss in general populations of children; children with WS would be no different.

It is important to note that minimal sensorineural hearing loss could not be revealed by standard school hearing screenings recommended by the ASHA Audiology Panel (1997) which uses a 20 dB HL presentation level for pure-tone stimuli at 1,000, 2,000 and 4,000 Hz. Therefore, without a full hearing evaluation in a clinical setting, the possibility of ignorance of hearing loss is high.

The current study showed a high prevalence of hearing loss in the individuals with WS and low awareness of this hearing loss among their parents. Two of the three child participants in the current study demonstrated minimal sensorineural hearing loss. There is also evidence that minimal sensorineural hearing loss could have been missed by standard school hearing screenings. Thus, an annual hearing evaluation should be included in the routine test of the individuals with WS. This is especially important because we do not yet have evidence of age-of-onset patterns for this population.

Otoacoustic emission data

The DPOAE measures of two (WS001 and WS004) of the five participants with bilateral hearing loss showed significantly reduced DPOAE amplitudes relative to data provided by Gorga et al. (1997) for the high frequency range (i.e., from 4,000 to 6,000 Hz). Reduced DPOAE amplitudes, consistent with impaired outer hair cell function, were observed from 4,000 to 7,000 Hz in participant WS001 and above 5,000 Hz in participant WS004. Marler et al. (2004) recently reported that 74% of their participants showed

reduced DPOAE amplitude in the frequency range of 3,500-6,000 Hz. Impaired outer hair cell function results in sensorineural hearing loss. Therefore, the findings of both the current study and the Marler et al. study are consistent with the findings of sensorineural hearing loss (75%) reported by Cherniske et al. (2004).

Only one participant in the current study demonstrated hearing within normal limits. He did not demonstrate the absence of otoacoustic emission responses reported by Johnson et al. (2001). In the five participants who demonstrated thresholds within normal limits in the low and/or mid-frequency range, only a single isolated DPOAE in those ranges was consistent with hearing-impaired ears. None of these participants demonstrated the absence of OAE responses reported by Johnson et al. (2001).

Loudness discomfort level (LDL) rating

The current design of the LDL measure for this project did not allow for complete LDL measurements in the adult individuals with WS, and no data were collected for the children. The relation between hearing thresholds and LDL could not be investigated due to the small number of participants in this study. However, two out of three participants in this task assigned a rating of "too loud" at levels much lower than those expected from the general population.

The pictures and descriptions for LDL measures in the current study worked well in terms of instruction comprehension. The participants in this task were able to use these pictures adequately to express their feelings about the stimuli.

In a recent critical review of the most comfortable level (MCL) and LDL literature, Punch, Joseph, and Rakerd (2004) cited documentation that prior exposure to loud stimulus levels (i.e., use of a descending approach) tends to increase the MCL. This

literature also shows how an auditory referent influences the measures of MCL and LDL. A lack of an auditory referent in the current research design (i.e., random presentation of stimuli) may have contributed to the inconsistent responses that were observed from participant WS003. An ascending approach should be included in any future research design in order to provide an auditory referent and to help individuals with WS overcome the fear of loud stimuli.

The approach in the current study was flawed in that it did not include the option to continue increasing stimulus level to the point that yielded a rating of "5." In order to investigate whether individuals with WS have a lower tolerance for loud sounds than their normal age peers, future studies should emphasize conducting LDL measures on a large group of individuals with WS, the range of stimuli used should not be predetermined, and an ascending method should be used.

Hyperacusis questionnaire

All of the parents of the individuals with WS reported that their children were bothered by loud sounds in the past and were still sensitive to loud sounds at the time of data collection. All of the participants were reported to exhibit a fear response to loud sounds in childhood, and three of them (two adults and a child) have reportedly outgrown their fear to sounds.

Parental reports showed that the prevalence of recurrent otitis media in the current study (two of seven individuals) was lower than the rates for individuals with WS reported by Klein et al., 1990 (61%; ages 1 to 28 years) and Van Borsel et al., 1997 (54%; ages 1 to 45 years); however, it was close to the rate reported for normally developing children ages 1 to 17 years (26.9%, Lanphear, Byrd, Auinger, & Hall, 1997).

Thus the prevalence of recurrent otitis media in individuals with WS was not as high as reported in previous studies. However, it is possible that the data in this study may not represent the general population of individuals with WS due to the small number of the participants.

Although the prevalence of recurrent otitis media was not as high as reported in previous studies, it was consistent with the tympanometric findings in the current study. Five of the participants demonstrated normal middle ear function in tympanometry during data collection. The other two participants either exhibited middle ear stiffness or were not willing to be tested in this task. Although tympanometry could not be done in these participants, parental reports revealed a history of recurrent otitis media and a fear of probe placement resulting from poor experiences with middle ear disease treatment in the past. Recurrence of otitis media could have not only contributed to conductive hearing loss, but also led to minimal-to-mild unilateral or bilateral sensorineural hearing loss (Bess et al., 1996; Hunter, Margolis, Rykken, Le, Daly, and Giebink, 1996). Two of seven participants (WS016 and WS028) in the current study were reported as having recurrent otitis media; however, their parents also reported that they were not aware of the presence of hearing loss in the past. Therefore, it was not possible to confirm that recurrent otitis media contributed to their hearing loss. The relation between recurrent otitis media and high-frequency sensorineural hearing loss in individuals with WS has not yet been investigated. Thus future research into the cause of high-frequency hearing loss in individuals with WS should also include an investigation of the potential contribution of otitis media.

Evaluation procedures

The current study showed that it is possible to obtain reliable behavioral thresholds from individuals with WS. All of the adult participants in the current study were able to participate in conventional pure-tone audiometry by raising a hand to respond to the pure-tone stimuli. The modified audiological assessment recommended by Kile (1996) and Dille (2003) for testing individuals with Down syndrome can be used for testing children with WS (e.g., use of visual reinforcement to help children with WS pay attention to the task). Another approach that would be useful would be the gradual acclimatization to the assessment procedures that was used in the data collection of participant WS028. Modifications to standard procedures included letting the participant wear earphones in an environment where she felt comfortable, use of conditioned play audiometry outside the booth to help the participant practice how to respond to stimuli, use of VRA to help the participant become familiar with the testing booth, and finally, a shift from VRA to standard conditioned play audiometry in the booth. This process helped WS028 overcome the fear of earphone placement and led to reliable responses during hearing assessment. This approach would require more time than is typical in clinical settings; therefore, it will be necessary to plan ahead both for time in the clinic and in home activities. Perhaps practice at home with earphones (e.g., to listen to music) prior to hearing assessment can help individuals with WS to get used to earphone placement.

DPOAE measures should be included in the assessment of children with WS, especially when behavioral thresholds (pure-tone thresholds) cannot be obtained. Due to the frequent occurrence of otitis media in children with WS, tympanometry should be

also be included as a routine test in the audiological evaluation. Two child participants (WS016 and WS028) in the current study exhibited fear or dislike of the probe placement for tympanometry and DPOAE measures, or dislike of the placement of the earphones for pure-tone audiometry. Audiologists in the clinical setting or in future research should be cautious about this tendency and leave tympanometry and DPOAE measures to the end of the hearing evaluation in order to limit the impact of these fear responses on other aspects of hearing assessment. In addition, standard headphones should be used instead of insert phones during for pure-tone audiometry.

Summary and conclusion

Due to the small number of individuals evaluated, the data from the current study should be considered cautiously. Mild-to-moderate, sloping, sensorineural hearing loss in the high-frequency range (above 4,000 Hz) is the major finding of the current study. This finding supports ongoing reports of a high prevalence of hearing loss among children and adults with WS and strongly challenges the historical assumption that individuals with WS have better than normal hearing. The empirical evidence presented here demonstrates that reports of hypersensitivity to sounds do not justify a conclusion that overall hearing is intact or better than normal in individuals with WS. It is also clear that parents of individuals with WS in the current study had low awareness of hearing impairment in their children.

Bess et al. (1998) revealed the impact of sensorineural hearing impairment even at the minimal levels demonstrated by two of the three child participants in this study. An annual hearing evaluation including pure-tone audiometry, tympanometry, and DPOAE measure is strongly recommended for this population. Use of modified assessment

strategies can help children with WS overcome the fear of probe or earphone placement and enable them to pay attention to the task. Once the hearing impairment is diagnosed in individuals with WS, appropriate assistance (e.g., hearing aids, classroom amplification) should be provided. Individuals with mild-to-moderate, sloping, sensorineural hearing loss in the high-frequency range (e.g., WS001, WS002 and WS018) are not candidates for hearing aid fitting because hearing aids provide very limited benefit above 4,000 Hz frequency range. For these children, classroom amplification systems that increase the signal-to-noise ratio should be explored as a means to provide the teacher's voice at a level well above the classroom noise. Children with greater degrees of hearing impairment who are hearing aid users would also be expected to benefit from such systems. Although the cause and the mechanism of hearing loss in individuals with WS are still unknown, it is important to address fully and investigate the impact of hearing loss on these children's language development and academic achievement.

APPENDICES

APPENDIX A

Hyperacusis Questionnaire

Child's Name: _____ **Date of Birth:** _____
Gender: _____

Please answer the following questions about your child's past and present behavioral responses to loud sounds. In **Column A**, please answer according to the period (the child's age) when the hyperacusis was most severe (disruptive to his/her daily life and to the life of the family). In **Column B**, please answer according his/her present response to loud sounds.

Column A (past)	**Column B (present)**
1a. Was your child presently frightened or bothered by certain sounds? YES_____ NO_____	1b. Is your child presently frightened or bothered by certain sounds? YES_____ NO_____
2a. If your child was ever fearful of loud sounds, at what age did this begin, and was there an age when it was the most disruptive to his/her daily life? _____ _____ _____ _____	2b. If your child "outgrew" the fear of loud sounds, so what age did this become apparent? _____ _____ _____ _____
3a. How often had your child had ear infections? a. ____ Never b. ____ Rarely, one or two times c. ____ Several, over a few years treated with medication. d. ____ Constantly, requiring medication and eventually tubes	3b. How often has your child had ear infections? a. ____ Never b. ____ Rarely, one or two times c. ____ Several, over a few years treated with medication. d. ____ Constantly, requiring medication and eventually tubes
4. Had your child been diagnosed as having a permanent hearing loss? YES_____ NO_____	

IF THE ANSWERS TO QUESTIONS 1 AND 2 WERE "NO" (NO PROBLEMS WITH LOUD SOUNDS) – STOP HERE. OTHERWISE CONTINUE.

Column A (past)	Column B (present)
5a. On a scale of 1 to 5, in the right column, please rate the degree of discomfort each sound ***caused*** your child [where 1=least discomfort and 7=greatest discomfort]. If your child has never been exposed to the sound, mark N/E (left column for "no exposure").	5b. On a scale of 1 to 5, in the right column, please rate the degree of discomfort each sound ***causes*** your child [where 1=least discomfort and 7=greatest discomfort]. If your child has never been exposed to the sound, mark N/E (left column for "no exposure").
N/E Degree	**N/E Degree**
__ __ Airplane	__ __ Airplane
__ __ Motorcycle	__ __ Motorcycle
__ __ Loud auto muffler	__ __ Loud auto muffler
__ __ Garbage truck	__ __ Garbage truck
__ __ Fire engine siren	__ __ Fire engine siren
__ __ Hammering a nail	__ __ Hammering a nail
__ __ Power saw	__ __ Power saw
__ __ Electric drill	__ __ Electric drill
__ __ Telephone ringing	__ __ Telephone ringing
__ __ Firecracker	__ __ Firecracker
__ __ Squeaking toys	__ __ Squeaking toys
__ __ Dog barking	__ __ Dog barking
__ __ Lawn mower	__ __ Lawn mower
__ __ Playground noise	__ __ Playground noise
__ __ Loud music	__ __ Loud music
__ __ Vacuum cleaner	__ __ Vacuum cleaner
__ __ Food blender	__ __ Food blender
__ __ Train whistle	__ __ Train whistle
__ __ TV at normal volume level	__ __ TV at normal volume level
__ __ Other, please specify below	__ __ Other, please specify below
_____	_____
6a. How ***did*** your child react to above noises? (You can choose more than one type of reaction.)	6b. How ***does*** your child react to above noises? (You can choose more than one type of reaction.)
a. _____ Cover ears with hands	h. _____ Cover ears with hands
b. _____ Cries	i. _____ Cries
c. _____ Says something like "it hurts my ear"	j. _____ Says something like "it hurts my ear"
d. _____ Says something like "I don't like it"	k. _____ Says something like "I don't like it"

Column A (past)	Column B (present)
e. _____ Run away from sound f. _____ Cringes g. _____ Other, please specify below _____ _____	l. _____ Run away from sound m. _____ Cringes n. _____ Other, please specify below _____ _____
7a. In general, the most important characteristics of the sounds that ***bothered*** your child ***were*** (you can check more than one if necessary). a. _____ How loud the sound is b. _____ A low pitched sound like an air conditioner c. _____ A high pitched sound like a squeaky wheel d. _____ A sudden sound e. _____ Reverberated sounds f. _____ Other_____	7b. In general, the most important characteristics of the sounds that ***bother*** your child ***are*** (you can check more than one if necessary). a. _____ How loud the sound is b. _____ A low pitched sound like an air conditioner c. _____ A high pitched sound like a squeaky wheel d. _____ A sudden sound e. _____ Reverberated sounds f. _____ Other_____
8a. On a scale of 1 to 5 (where 1 is minimal discomfort and 5 is severe discomfort), how would you categorize your child's hyperacusis ***at its most prominent period in his/her life?*** Please circle the appropriate number. 1 2 3 4 5 **minimal discomfort** **severe discomfort**	8b. On a scale of 1 to 5 (where 1 is minimal discomfort and 5 is severe discomfort), how would you ***currently*** categorize your child's hyperacusis? Please circle the appropriate number. 1 2 3 4 5 **minimal discomfort** **severe discomfort**

9. Did you seek professional help for your child's hyperacusis?
   YES_____ NO_____

10. Are these any circumstance or behavior related to your child's progress to sounds, that you think important, but have not been checked in this questionnaire?

Completed by _____ Date _____
Relation _____
Questionnaire was adapted from Klein et al. (1990).

APPENDIX B

Individual Data from Immitance Measures

Table 7. Individual data from immitance measures.

	Static admittance (mmho)		Ear-canal volume (ml)		Gradient	
	Right	Left	Right	Left	Right	Left
WS001	0.5	0.4	2.1	1.8	0.4	0.5
WS002	1.1	1.0	0.9	1.0	0.7	0.7
WS003	0.3	0.4	0.8	0.9	0.5	0.5
WS004	0.7	0.6	1.8	1.7	0.5	0.5
WS016	NP	CNE	0.4	CNE	CNE	CNE
WS018	0.3	0.2	0.7	0.7	CNE	CNE
WS028	CNE	CNE	CNE	CNE	CNE	CNE

NP = No peak. CNE = Could not evaluate.

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