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Characteristics of auditory agnosia in a child with severe traumatic brain injury: A case report

Nina Hattiangadi,[a,*] Joseph P. Pillion,[b] Beth Slomine,[a] James Christensen,[c] Melissa K. Trovato,[c] and Lynn J. Speedie[a]

[a] *Department of Neuropsychology, Kennedy Krieger Institute, Johns Hopkins School of Medicine, USA*
[b] *Department of Audiology, Kennedy Krieger Institute, Johns Hopkins School of Medicine, USA*
[c] *Department of Physical Medicine and Rehabilitation, Kennedy Krieger Institute, Johns Hopkins School of Medicine, USA*

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Abstract

We present a case that is unusual in many respects from other documented incidences of auditory agnosia, including the mechanism of injury, age of the individual, and location of neurological insult. The clinical presentation is one of disturbance in the perception of spoken language, music, pitch, emotional prosody, and temporal auditory processing in the absence of significant deficits in the comprehension of written language, expressive language production, or peripheral auditory function. Furthermore, the patient demonstrates relatively preserved function in other aspects of audition such as sound localization, voice recognition, and perception of animal noises and environmental sounds. This case study demonstrates that auditory agnosia is possible following traumatic brain injury in a child, and illustrates the necessity of assessment with a wide variety of auditory stimuli to fully characterize auditory agnosia in a single individual.

Keywords: Auditory agnosia; Auditory perception; Auditory brain stem response; Verbal comprehension; Language processing; Complex sound processing; Pure word deafness; Temporal processing; Traumatic brain injury; Pediatric

1. Introduction

Early accounts of pure word deafness were often diagnosed solely based on the presence of impairments in auditory comprehension, repetition, and writing to dictation in the presence of spared pure tone audiometric findings. In most cases with the presentation of pure word deafness, however, exposure to a wider range of auditory stimuli has revealed additional impairments in other aspects of auditory processing. In a recent review of 63 cases of "pure word deafness" subjects, only five case studies reported normal nonverbal sound processing, with all others presenting with impairments in processing of music and/or environmental sounds (Pinard, Chertkow, Black, & Peretz, 2002). Auditory agnosia refers to a more generalized impairment in the recognition of sounds. While auditory agnosia is far from a new concept (Liepmann, 2001), the availability of a greater variety of auditory stimuli used in assessment of agnosia has led to greater appreciation of the complexity of the disorder. For example, several case studies describe a single individual who has demonstrated different agnosias at different times (Mendez & Geehan, 1988; Motomura, Yamadori, Mori, & Tamaru, 1986). These studies typically describe a patient presenting with an initial onset of "cortical deafness" but subsequently demonstrating gradual recovery for different auditory stimuli. In the Mendez and Geehan case study, for example, the initial presentation of cortical deafness was followed by inconsistent reactions to sound. Subsequently, improved pure tone thresholds, recognition of environmental noises, music recognition, and finally speech recognition returned in that order

* Corresponding author. Present address: Department of Psychology, The Children's Hospital of Philadelphia, 3405 Civic Center Boulevard, Philadelphia, PA 19104, USA. Fax: 1-215-590-5637.
E-mail address: hattiangadi@email.chop.edu (N. Hattiangadi).

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(Mendez & Geehan, 1988). Conversely, a degenerative course of progressively generalized auditory agnosias may be possible as well (Pinard et al., 2002).

Several lines of research implicate a disturbance of complex sound processing—particularly temporal processing—as the basis of word deafness and other auditory agnosias. In direct contrast to modularity theories, which state that different types of sounds are processed along neuroanatomically distinct pathways, temporal processing theories posit that spoken language can be conceptualized as a quantitatively more complex auditory signal along a continuum of speech and nonspeech sounds. More specifically, the degree of temporal processing required for appropriate perception of spoken language is significantly greater than is required for perception of nonspeech sounds (Fitch, Miller, & Tallal, 1997; Shannon, Zeng, Kamath, Wygonski, & Ekelid, 1995; Zatorre & Belin, 2001). Concurrently, the relative importance of temporal cues in the processing of speech sounds is greater than in the processing of nonspeech sounds (Griffiths, Rees, & Green, 1999). The net effect of these two characteristics renders spoken speech particularly vulnerable to the effects of a disruption in temporal auditory processing.

In most cases, the occurrence of pure word deafness or other auditory agnosias has been attributed to dominant unilateral lesions of Heschl's gyrus or bilateral lesions of the superior temporal lobe (Griffiths et al., 1999; Mesulam, 1985). However, auditory agnosias have occasionally been reported to result from damage sustained entirely at the subcortical or brainstem level, including bilateral lesions of the inferior colliculi (Johkura, Matsumoto, Hasegawa, & Kuroiwa, 1998; Vitte et al., 2002) or damage to thalamocortical auditory pathways (Shivashankar, Shashikala, Nagaraja, Jayakumar, & Ratnavalli, 2001; Taniwaki, Tagawa, Sato, & Iino, 2000).

We present a case of auditory agnosia in a child with non-penetrating traumatic brain injury. The case is relatively unique in terms of the mechanism of injury (non-penetrating traumatic brain injury) and location of identified neurological insult (primarily frontal, subcortical, and commissural). Furthermore, an extensive array of auditory stimuli was used in the assessment, yielding evaluation of aspects of sound processing such as perception of emotional tone and recognition of familiar voices that are rarely described in other case reports.

2. Case description

2.1. History

Prior to his injuries the patient, H.S., was a healthy young boy who was generally a low average student as indicated by standardized test scores and grades in core academic subjects. The patient had no documented learning disabilities, although given his low average academic performance it remains possible that mild undocumented language problems or other cognitive difficulties were present premorbidly. He had a history of frequent school changes due to family relocations and one grade retention in the 3rd grade. He was enrolled in a regular 6th grade program at the time of his injury.

H.S. sustained a non-penetrating traumatic brain injury shortly following his 12th birthday. At the time of his injury, the patient was reportedly riding on the front handlebars of a bicycle that was being propelled by another individual. The bicycle was hit from the side by an automobile. The patient, who was unhelmeted at the time of the accident, was thrown from the bicycle, striking his head first on the automobile, and again on the street. The circumstances of the accident are of particular interest, in that the structural brain injuries (described later) are primarily of the type produced by acceleration/deceleration associated with rotational forces. The child was reportedly unresponsive at the scene, with an initial Glasgow Coma Scale of 3. He was transported by air to a large pediatric trauma center, where he underwent a ventricular drain placement. He continued to require management and sedation for increased intracranial pressure following transfer to a Pediatric Intensive Care Unit. On Day #11, he was noted to demonstrate increased tone and a withdrawal to pain. Following several failed attempts at extubation, the patient underwent tracheostomy and placement of a gastrostomy tube, and was transferred to the Kennedy Krieger Institute's Brain Injury Unit on Day #28 for intensive inpatient rehabilitation.

At the time of his admission to the inpatient rehabilitation program, H.S. was demonstrating non-purposeful movements of all extremities, but with decreased movement on the left side. He was able to spontaneously open his eyes, but demonstrated a left gaze preference and only intermittent tracking from his left visual field to midline. Throughout the first month of his inpatient stay, H.S. made considerable gains in physical functioning; he was able to reach for objects, squeeze squeaky toys, imitate actions, and even ambulate with support. Despite these gains, he continued to demonstrate an inability to follow verbal commands. Early spoken language was characterized by echolalia and perseverative speech strings (e.g., "talk, talk, talk, talk."). Speech was hypophonic, breathy, and hoarse due to a right-sided vocal fold paralysis. Over time, this spontaneous speech evolved to include syntactically correct but perseverative statements (e.g., "What's your name?"). Approximately 2 months following his injury (Day #58), H.S. was first presented with a command in written form ("Write your name") and was able to comply immediately, indicating that he likely had been able to follow written commands for some time before this ability was first assessed. By three months post-injury, he was demonstrating significant amounts of appropriate speech. Although he remained unable to comprehend the majority of verbal speech presented to him, he was able to answer simple written questions regarding his name, age, and other personal data and was able to follow two-step written commands.

During the course of his inpatient hospitalization, H.S. demonstrated a greater ability to maintain sustained attention with less structure and reinforcement and to participate in reciprocal social exchanges using a combination of written and verbal language. Although considerable memory difficulties were still noted in H.S.'s daily functioning, as his rehabilitation progressed he demonstrated an ability to form new memories, learning the names of his therapists and navigating within portions of the building independently.

Approximately 5 months following his injury (Day #143), H.S. was discharged to home with close supervision and began attending the Specialized Transition Program (STP) at the Kennedy Krieger Institute, a transitional day-program providing educational and rehabilitation services. He was discharged from this outpatient program approximately 6 months following the date of his injury (Day #182), returning to a self-contained, highly structured classroom within his pre-injury school environment. At the time of his discharge from the STP program, H.S. was on a regular diet and was able to ambulate independently, with distant supervision required in community settings secondary to limited safety awareness.

2.2. Neuroimaging

Magnetic resonance imaging (MRI) of the brain was performed 10 days post-injury (see Fig. 1). The study included sagittal T1, axial T2, axial FLAIR, diffusion weighted images, axial gradient recalled echo, and magnetic resonance angiography. A table of neuroimaging findings is provided Table 1.

Table 1
Neuroimaging findings

White matter edema of the subcortical medial bifrontal regions, left > right
Cortical injury of the anterior temporal poles
White matter edema of the left parietal region
White matter edema of the left occipital region
Edema of the left splenium of the corpus callosum crossing the midline
Edema of the bilateral caudate nucleus
Edema of the right thalamus
Edema of the posterior limb of the right internal capsule
Edema of the left posterior thalamus extending in the lateral midbrain
Edema of the left cerebral peduncle

2.3. Neuropsychological functioning

Complete neuropsychological, educational, and speech and language evaluations were conducted approximately five months after date of injury, prior to H.S.'s discharge from the STP program. Because of his difficulties with comprehension of spoken language, an accommodated style of testing was employed, primarily consisting of written instructions used to supplement oral instructions, and simultaneous presentation of written stimuli to accompany all verbal stimuli. It is important to note, however, that H.S.'s performance on tests of written language comprehension was not age-appropriate, and therefore the use of written material to accommodate his speech perception deficits does not necessarily ensure adequate understanding of all task demands. Furthermore, normative data do not fully

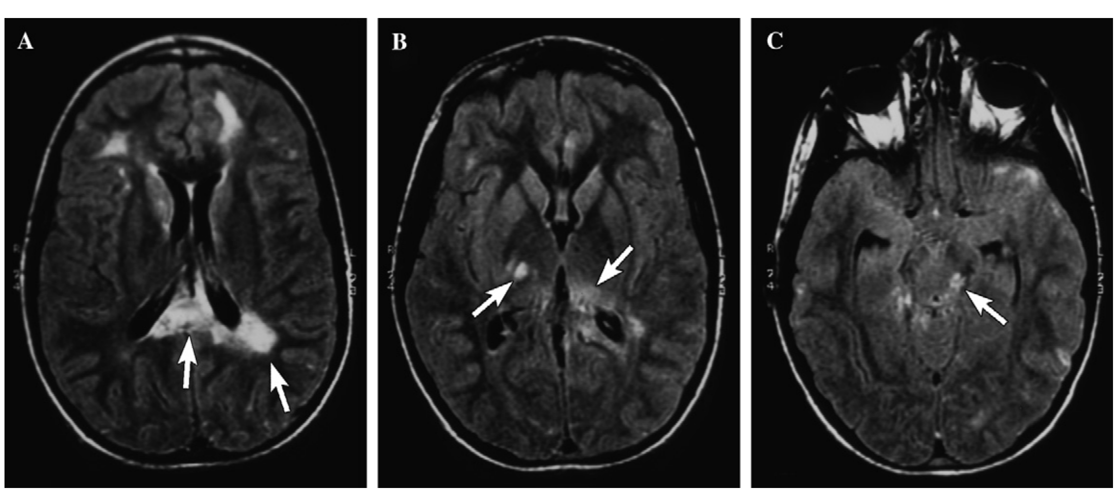


Fig. 1. T2-weighted images MRI images are shown, with increased signal consistent with edema. The arrows point to the following areas of edema: (A) left splenium of the corpus callosum crossing the midline, and left temporal–parietal white matter, (B) right and left thalamus, (C) left cerebral peduncle.

apply to such nonstandardized administration, although normative scores are provided as an estimate of the patient's performance in comparison to age-equivalent peers. Nonetheless, even with such accommodations, in comparison to his estimated low average premorbid functioning his performance clearly represents a global decline in virtually all areas of cognitive functioning.

Throughout the testing, attention, and concentration significantly impaired performance across tasks. Results revealed psychometric intelligence was in the deficient range. A relative strength in verbal comprehension was noted (when stimuli were presented in written form). Constructional tasks were attenuated due to a bilateral intention tremor and slow speed. Additionally, untimed academic skills were borderline to low average, whereas greater impairment was noted on speeded academic tasks. H.S. was also able to attend to and recall small amounts of organized information (brief stories) when presented in written form as indicated by average immediate story recall. Scores from this assessment period are provided in Table 2. Results are presented in scaled or standard scores unless otherwise noted.

Table 2
Cognitive evaluation

Wechsler Intelligence Scale for Children, Third Edition (WISC-III)[a]			
Information	4	Picture Completion	1
Similarities	5	Coding	1
Arithmetic	—	Picture Arrangement	2
Vocabulary	6	Block Design	1
Comprehension	1	Object Assembly	1
Digit Span	—	Symbol Search	1
Full Scale IQ[A]	53	Verbal IQ[A]	55
VIQ–PIQ = 19, ($p < .05$)		Performance IQ	47
		Verbal Comprehension	68
		Perceptual Organization	50
		Processing Speed	50
Delis–Kaplan Executive Function System (D-KEFS)[b]			
Number Sequencing	1	Letter Sequencing	1
NEPSY Developmental Neuropsychological Assessment[c]			
Comprehension of Instructions	1	Arrows	3
Wide Range Assessment of Memory and Learning (WRAML)[d]			
Verbal Learning	4	Story Memory[a]	9
Differential Ability Scales (DAS)[e]			
Recall of Objects—Immediate	T = 34	Recall of Objects—Delayed	T = 26
Woodcock Johnson Revised Test of Academic Achievement (WJ-R)[f]			
Letter Word Identification	78	Calculation	82
Passage Comprehension	78	Math Fluency	58
Reading Fluency	69	Writing Fluency	64
Broad Reading	71		
Math Calculation Skills	73		
Academic Fluency	64		
Clinical Evaluation of Language Functions—Third Edition (CELF-3)[g]			
Concepts and Directions	4	Formulating Sentences	3
Word Classes	4	Recalling Sentences	4
Semantic Relationships	4	Sentence Assembly	3
Listening to Paragraphs	3		
Global Language Comprehension	50		
Global Language Expression	50		
Total Language Score	50		

[A] Prorated score.
[a] Wechsler (1991).
[b] Delis, Kaplan, and Kramer (2001).
[c] Korkman, Kirk, and Kemp (1998).
[d] Sheslow and Adams (1990).
[e] Elliott (1990).
[f] Woodcock and Mather (1989, 1990).
[g] Semel, Wiig, and Secord (1995).

3. Tests of auditory processing

Tests of auditory processing were conducted during H.S.'s participation in the Kennedy Krieger Institute's inpatient and outpatient brain injury rehabilitation programs (approximately Day #28–Day #182). Standardized tests were administered when possible, but frequently tests had to be created to assess a specific auditory skill or to minimize cognitive demands inherent to some standardized tests. Tests that were created by the authors for the purpose of clinical assessment of this individual are indicated with an asterisk (*). The overall pattern of impaired and relatively spared abilities described here did not seem to be significantly affected by general improvements in cognitive functioning over the course of H.S.'s recovery. Although some skills that were initially above-chance improved even further as H.S. recovered, all tasks noted to be at chance levels early on remained at chance when reexamined at a later phase of H.S.'s course of recovery. Because normative information was not available for tests created by the authors and interest was primarily in the differentiation of intact from grossly impaired skills, a binomial distribution function was used to distinguish chance from above-chance performance. The probability of a successful performance on any given item was adjusted appropriately for each task. All skills falling at above-chance levels were significant at the $p < .001$ level.

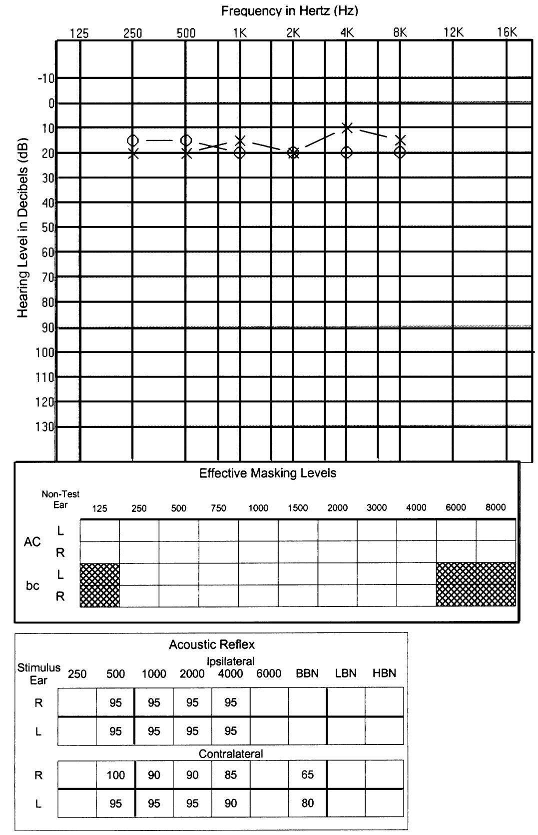


Fig. 2. Audiometric data depicting auditory sensitivity for the right ("O") and left ("X") ears. Ipsilateral and contralateral acoustic reflex thresholds are also shown. All data are in dB HL ANSI (1989).

3.1. Peripheral auditory function

1. Auditory sensitivity
   - *Materials and procedures:* Auditory sensitivity was assessed monaurally for speech and pure tones utilizing conditioned play audiometry.
   - *Results:* Results are shown in Fig. 2 and indicate the presence of normal peripheral auditory sensitivity bilaterally. Speech reception thresholds could not be established. However, speech detection thresholds were obtained at 5 dB HL for the right ear and −5 dB HL for the left ear.
2. Acoustic stapedial reflexes
   - *Materials and procedures:* Acoustic stapedial reflexes were measured utilizing a middle ear analyzer (Madsen Zodiac, model 901) for pure tones from 500 to 4000 Hz for ipsilateral and contralateral stimulation of both ears. Broad band noise signals were also utilized for contralateral stimulation of both ears.
   - *Results:* Acoustic reflexes were established at normal sensation levels for ipsilateral and contralateral simulation of both ears with pure tone stimuli (500–4000 Hz) and for broadband noise stimuli for contralateral stimulation of both ears, indicating that afferent and efferent elements of the acoustic reflex arc were intact.
3. Otoacoustic emissions
   - *Materials and procedures:* Distortion product otoacoustic emissions were obtained utilizing an ILO88DP otoacoustic emissions measurement unit. Primary tones (f_1 and f_2) were presented at 65 and 55 dB SPL with a frequency ratio between f_1 and f_2 of 1.2.11.
   - *Results:* Distortion product otoacoustic emissions (Fig. 3) were present for stimulation of both ears. Findings indicate the presence of normal outer hair cell function bilaterally.
4. Auditory evoked potentials—auditory brainstem responses (ABR)
   - *Materials and procedures:* Auditory brainstem responses (ABR) were recorded utilizing gold-cup biopotential electrodes affixed at the forehead and on each mastoid. Two-channel concurrent differential recording was employed with the high-forehead and low forehead electrodes serving as the active and ground electrodes, respectively. Each mastoid served as a reference for one of the two recording channels. Responses to 1024 clicks were averaged following filtering

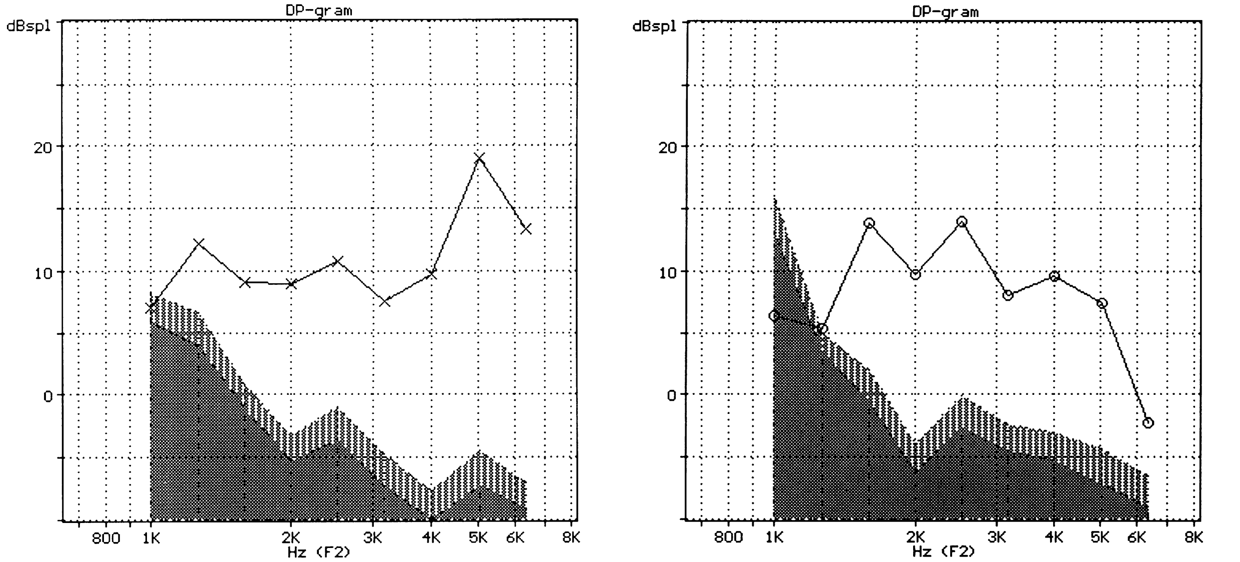
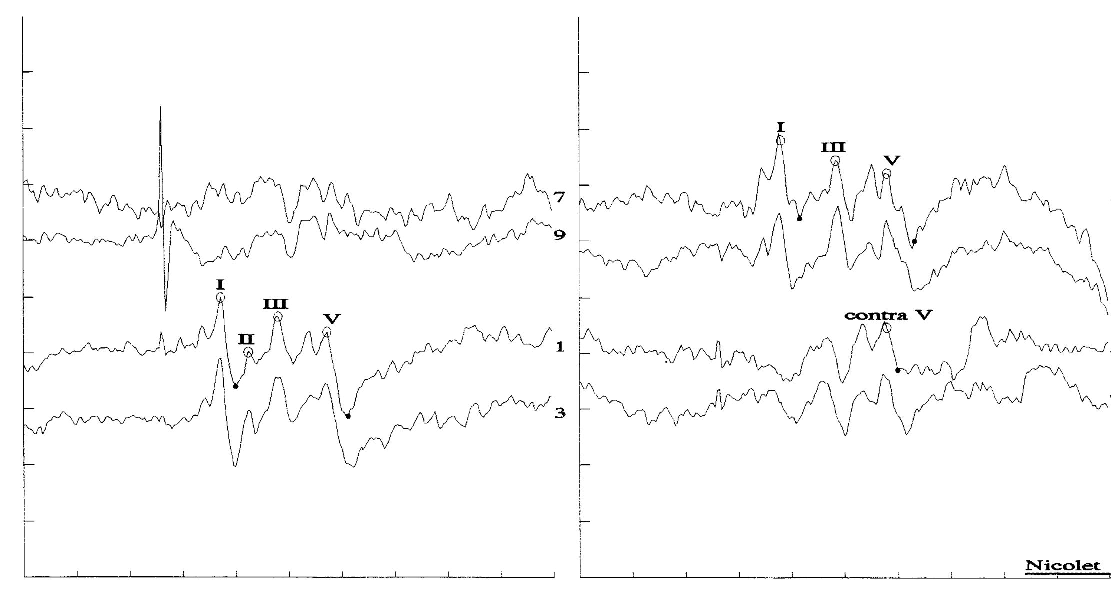


Fig. 3. Distortion product otoacoustic emissions (DPOAEs) for the left and the right ears. Responses for the left ear are shown on the left and responses for the right ear are shown on the right.

(100–3000 Hz), amplification, and rejection of artifacts. ABR testing was undertaken utilizing rarefaction clicks presented at 80 dB nHL at rates of 27.7 or 57.7 s.

- *Results:* The response waveforms are shown in Fig. 4 and reveal normal IPLIs (i.e., I–III, III–V, and I–V interpeak latency intervals) and waveform morphology for stimulation of both ears. For further evaluation for evidence of disruption in structure and/or function of the central auditory pathways, the ABR measurements were conducted using an increased click rate of 57.7/s. There was no significant degradation in waveform morphology or in the IPLIs following an increase in rate of click presentation to 57.7/s, suggesting the presence of normal structure and function of the auditory pathways at brainstem levels.

5. Auditory evoked potentials—middle latency response (MLR)



Sensitivity and Sweep Time Per Division

1	0.25 uV	2.0 msec	2	0.25 uV	2.0 msec	3	0.25 uV	2.0 msec	4	0.25 uV	2.0 msec
7	0.25 uV	2.0 msec	8	0.25 uV	2.0 msec	9	0.25 uV	2.0 msec	10	0.25 uV	2.0 msec

Fig. 4. Auditory brainstem responses (ABRs) to clicks presented at 80 dB nHL at a rate of 27.7/s. Responses to clicks presented to the left ear are on the left, and responses to clicks presented to the right ear are shown on the right. The responses for both ipsilateral and contralateral channels are shown.

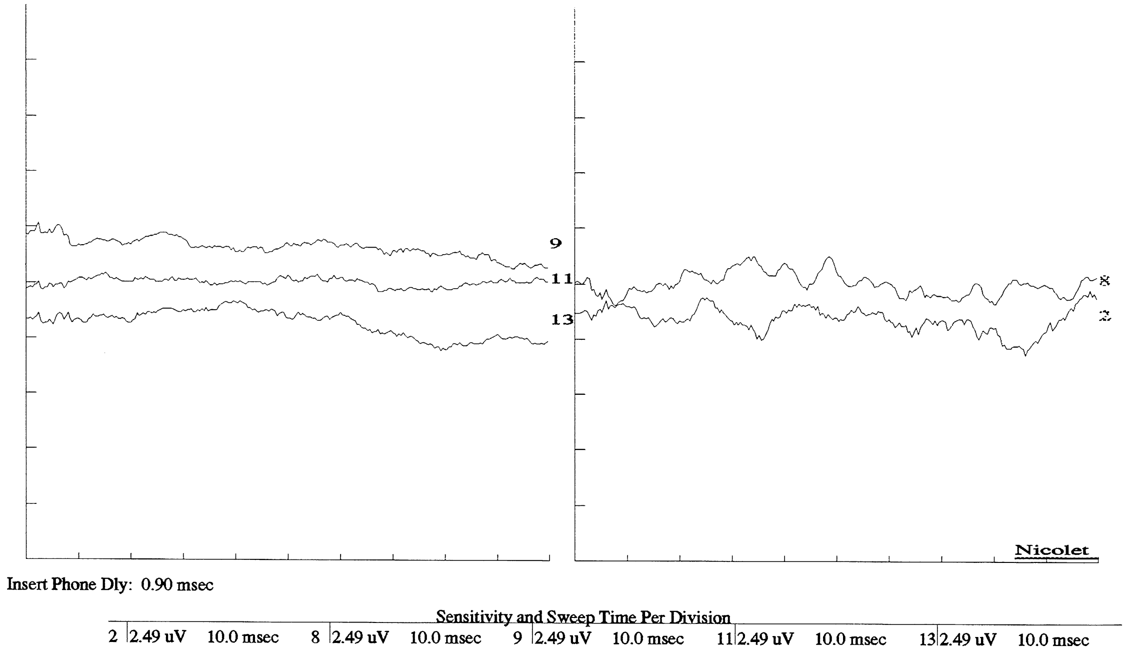


Fig. 5. Middle latency response (MLR) waveforms to clicks presented at 80 dB nHL at a rate of 2.5/s. Waveforms obtained to clicks presented to the left ear are shown on the left, with waveforms to clicks presented to the right ear shown on the right.

- *Materials and procedures:* Measurements of the middle latency response (MLR) were conducted utilizing rarefaction clicks presented at a level of 80 dB nHL at a rate of 2.5/s. Responses to 250 clicks were averaged following filtering (20–3000 Hz), amplification, and rejection of artifacts.
- *Results:* The MLR (Fig. 5) was absent for stimulation of both ears, which is consistent with bilateral lesions of auditory radiations and/or bilateral lesions in the auditory cortex (Scherg & von Cramon, 1986).

6. Sound localization
   - *Materials and procedures:* Sound localization abilities were assessed within a sound treated audiometric booth with speakers located at 45 and 315 degrees azimuth. Warble tones centered at 1000 Hz were utilized. The presentation level was 50 dB HL.
   - *Results:* Sound localization abilities were grossly intact, as only one error was noted in 22 trials (4.5% error rate) Table 3.

Table 3
Peripheral auditory function

Intact functions	Impaired functions
Auditory sensitivity	Middle latency response (MLR)
Acoustic stapedial reflexes	
Otoacoustic emissions	
Auditory brainstem responses (ABR)	
Sound localization	

3.2. Speech perception

1. Phoneme repetition
   - *Materials and procedures:* The Phoneme Recognition Test (Katz, 1998) was administered to assess the ability of H.S. to repeat speech phonemes spoken in isolation. The test consists of 34 items presented in an open set repetition task format.
   - *Results:* H.S. was able to repeat only 1/34 items accurately (3% accuracy rate), indicating severely impaired repetition of individual speech sounds.
2. Uncued word repetition
   - *Materials and procedures:* The Phonetically Balanced Kindergarten Word Lists test (PB-K) (Haskins, 1949) was utilized to assess H.S.'s word repetition abilities in an open set response format. The word lists consist of 25 single-syllable words presented at a level of 50 dB HL in quiet.
   - *Results:* Individuals typically perform at a level of 92% or better on the task. In contrast, H.S. correctly identified 1/25 words for the left ear (4% accuracy) and 0/25 for the right ear (0% accuracy), indicating a virtual absence of the ability to repeat even very simple spoken words.
3. Visually cued word recognition
   - *Materials and procedures:* The Northwestern University Children's Perception of Speech Test (NU-CHIPS) (Elliott & Katz, 1980) was also administered. NU-CHIPS is a closed-set picture-

pointing response task with four response alternatives. A monosyllabic noun is presented to the individual, with stimuli chosen from words that were found to be correctly identified by at least 22 of 25 children ranging in age from 3 to 3 1/2 years old. It would therefore be expected that a 12-year-old individual like H.S., despite his overall cognitive impairment, would do extremely well on this task in the absence of significant word recognition deficits. Items were selected so that each of the four response alternatives represented different vowel phonemes, with no other systematic phonological, orthographic, or semantic contrasts between the target and response foils.
- *Results:* Scores of 56% (14/25) were obtained for the right ear and 64% (16/25) for the left ear. Although this level of performance still indicates severely impaired levels of speech recognition compared to normal functioning, H.S. demonstrated dramatic benefit from pictoral cues relative to the open-format repetition task. Furthermore, it should be noted that the characteristic of this task of having a different vowel phoneme for each target and response foils renders it possible to perform well with perception of the vowel sound alone.

4. Visually cued verbal versus written word recognition
   - *Materials and procedures:* To directly compare spoken versus written single-word semantic association, H.S. also completed the first 24 items of the Peabody Picture Vocabulary Test, Third Edition (PPVT-IIIA) (Dunn & Dunn, 1997), with words presented first in verbal format and then in written format. This test uses an untimed format to allow an individual to pick from four pictures the one that most closely relates to the vocabulary word said by the examiner (or in our adapted written format, the written word shown to the individual by the examiner). The words are ordered to be progressively more difficult. The distractors (incorrect picture responses) are designed to be similar to the correct response in subject matter and complexity of illustration, but not to be similar in sound to the target word.
   - *Results:* H.S. was able to indicate comprehension of a few one- to three-syllable words from the pictoral vocabulary task (e.g., "hand," "helicopter," and "jumping") and benefited markedly from the context provided by a four-choice format. Nonetheless, when words were presented verbally with the examiner's mouth covered to eliminate visual cues, H.S. demonstrated chance performance on this vocabulary task (25%, 6 of 24 items correct). Items incorrectly answered in the verbal format were then administered in written format, by showing H.S. the written word and then having him choose a response. With this visual administration of the task, his performance jumped to 94% accuracy (17/18 correct), indicating that prior impairments were related to difficulties with auditory comprehension rather than limitations in vocabulary.
5. Verbal versus written sentence comprehension
   - *Materials and procedures:* The patient was tested informally using a variety of simple questions, (e.g., "What color is grass?"), simple and complex one- and two-step commands (e.g., "Touch your nose after pointing to the door."), and more complex questions (e.g., "Are you older than your father?") from the Complex Ideational Materials subtest of the Boston Diagnostic Aphasia Examination (Goodglass & Kaplan, 1978).
   - *Results:* When presented with the simple and complex questions and commands, the patient was unable to perform any of these when they were presented verbally in the absence of lipreading cues. However, he successfully completed virtually all of these when presented in written format. He was able to successfully complete 8 of 10 complex commands such as, "Point to the door after touching your nose." Occasional errors appeared to be related to impulsive or inattentive reading, in that H.S. appeared to be mistaking similar-appearing words (e.g., "head" for "hand").
6. Dichotic digit perception
   - *Materials and procedures:* The Dichotic Digit Task (Noffsinger, Martinez, & Wilson, 1994a) was administered at a level of 50 dB HL. The test consists of single pairs of 36 digits which are precisely aligned in onset and recorded onto an audio compact disc. H.S. demonstrated the ability to identify the words binaurally in a closed set response format preliminary to dichotic presentation. The response alternatives were shown to H.S. and were in front of him during the task, and he was instructed to repeat both digits.
   - *Results:* Normally hearing young adults typically identify both digits with 97% or greater accuracy during dichotic presentation (Wilson & Jaffe, 1996). H.S. identified 4/36 (11%) of digits presented to his right ear and 15/36 (42%) of digits presented to his left ear, indicating a significant left ear advantage for this task (Table 4).

Table 4
Speech perception

Above chance abilities	At chance abilities
Visually cued word recognition	Phoneme repetition
Written word recognition	Uncued word repetition
Written sentence comprehension	Verbal sentence comprehension
Left ear dichotic digit perception	Right ear dichotic digit perception

3.3. Perception of nonspeech sounds

1. Pitch perception*
   - *Materials and procedures:* Using SuperLab software, a computerized forced-choice pitch perception task was created. Two tones, each 500 ms in duration, were presented with an interstimulus interval of 500 ms. After each stimuli pair, the patient was asked to determine whether the tones had been the same or different. A visual analogue of this task, in which the patient responded whether two squares were the same or different color, was presented to ensure the patient's understanding of task demands. For the experimental pitch task, pitch stimuli were compared in each case to a 1000 Hz base tone, with counterbalancing of tone presentation across items. Tones were chosen to represent equal octave intervals above or below the 1000 Hz base tone, and were divided into three experiments requiring increasingly fine discriminations. "Same" and "different" items were counterbalanced to eliminate the effects of response bias. Tones compared to the base tone (1000 Hz) ranged from 125 to 8000 Hz, with discriminations as small as 5 Hz assessed.
   - *Results:* Results indicated random performance for discriminations less than 400 Hz, indicating severe impairment in discrimination of frequency. Normally hearing individuals typically discriminate frequency with differences of between 4 and 8 Hz at 1000 Hz (Wier, Jesteadt, & Green, 1977).
2. Melody perception*
   - *Materials and procedures:* Five songs commonly known to children were identified ("Itsy Bitsy Spider," "Happy Birthday," "Old McDonald," "Twinkle Twinkle Little Star," and "Mary Had a Little Lamb"), and the patient's father confirmed in advance of the task that the patient had been familiar with these melodies prior to his injury. To increase the patient's interest in the task and reduce the cognitive load of reading, five cards were prepared as stimuli, each containing the title of a song and a small illustrative color picture (e.g., a spider in a web for "Itsy Bitsy Spider," a birthday cake for "Happy Birthday"). Prior to beginning the task, the patient was presented with the title-and-picture stimuli. He affirmed his familiarity with the songs by singing each one, although singing output was notably atonal. For each trial of this task, the cards were presented in a random vertical arrangement at the patient's midline. The examiner then played the initial two phrases of a song (without words) on a keyboard hidden from the patient's view. Although the exact duration of the melodies was not controlled, melodies were approximately equivalent in length, as roughly indicated by the very small range (12–14) of the number of notes played in each melody. The patient was asked to indicate by pointing which song he had heard.
   - *Results:* Performance on this measure fell at chance, with 20% recognition of melodies (4/20 correct).
3. Emotional prosody
   - *Materials and procedures:* The Emotional Prosody Naming subtest of the Florida Affect Battery (Bowers, Blonder, & Heilman, 1992) was used to assess H.S.'s comprehension of emotional tone. This task consists of 20 audiotaped sentences of neutral semantic content (e.g., "The chairs are made of wood") spoken in either a neutral tone or one of four emotional tones (happy, sad, angry, or scared). To increase H.S.'s attention and to maximize accurate responding, he was asked to respond by referencing a vertical list of the emotions. To improve understanding and limit reading and working memory demands, each written emotional term (e.g., "happy") was accompanied by a cartoon face exhibiting that emotion. In addition, the term "neutral" was written as "neutral/nothing" to facilitate H.S.'s understanding of this term.
   - *Results:* This task was administered on two occasions. Although the patient appeared to listen attentively and responded following each item, recognition of affective intonation of speech was at chance, with correct responding to 10 of the 40 items. When administered the Facial Affect Naming task from the same battery, he was able to correctly identify 16 of 20 (80%) emotional facial expressions using the same word list without cartoon face cues, with errors typically related to underutilization of the "neutral" category. This ability to name facial emotion but not emotional prosody indicates that the deficit in processing emotional intonation does not represent a general impairment in emotional understanding, nor can it be explained by the complexity of task demands.
4. Environmental sounds
   - *Materials and procedures:* The Sound Effects Recognition Test (SERT) (Finitzo-Hieber, Matkin, Cherow-Skalka, & Gerling, 1977) was used to assess the patient's perception of environmental sounds. Three 10-item forms of this measure are available, resulting in a total of 30 environmental sounds (e.g., barking, hammering, dishes breaking). Each sound is presented on audiotape while the subject responds by pointing to one of four pictures presented in a 2×2 format. On each page, the target noise as well as three other possible noises are represented pictorially (e.g., a barking dog for the barking sound).

- *Results:* Perception of environmental sounds fell consistently above chance, despite significant levels of inattentive and uncooperative behavior. Performance across three forms of the SERT in a single session resulted in accurate perception of 17 of the 30 items (accuracy of 57% in comparison to a 25% chance performance). To reduce the impact of inattentive behaviors, the videotape of the testing session was independently reviewed by three members of the neuropsychology staff. Staff members were able to hear the item presentation but were not able to view the patient's responses, and were therefore blind to the correct or incorrect status of the patient's responding. On each item, the staff rated whether the patient demonstrated adequate attention to have responded to the stimuli appropriately. Elimination of the five items to which all staff agreed the patient had not demonstrated appropriate attention (e.g., by talking during item presentation or picking a response prior to presentation of the sound) still results in above-chance performance (15/25; or 60% in comparison to a 25% chance performance).

5. Animal noises*
   - *Materials and procedures:* A series of four-choice picture stimuli were prepared featuring different animals. Animals similar in appearance or species (e.g., wolf and dog, eagle and turkey) were not present on the same page. The patient was then presented with 17 different animal noises, culled from audio files made publicly available on the internet and chosen by the examiners for their similarity to the noise considered archetypal to each animal (e.g., "ribbit ribbit" for frogs). The noises were presented to the patient one by one from an audio compact disc, and he was asked to point to the picture of the animal making each noise.
   - *Results:* Perception of animal noises fell consistently above chance (71% in comparison to a 25% chance performance).
6. Cartoon voice recognition*
   - *Materials and procedures:* A series of four-picture stimuli were again created, this time consisting of cartoon characters reported by the patient's father as having been familiar to the patient prior to his injury. Characters felt by the examiners to be unusually similar in voice quality or other paralinguistic qualities (e.g., accent, stutter, and lisp) were not present on the same page. Brief sound clips of each character's voice were chosen from audio files made publicly available on the internet. Sound clips were chosen based on clarity, length, and absence of background noise such as sound effects, music, or other characters' voices. An attempt was made to avoid "catch phrases" that were known to be commonly associated with each character (e.g., "What's up Doc?" for Bugs Bunny), to prevent the possibility of matches based on linguistic content rather than voice characteristics. Sixteen sound clips, consisting of two different clips for each character, were chosen and presented to the patient in random order.
   - *Results*: Performance on this task was remarkably intact, with 88% accuracy for matching the sound clip to the cartoon character. Strikingly, even when the patient was able to correctly identify the character who had spoken, when asked he would completely misreport the nature of the utterance (e.g., stating that the character who said, "Be very quiet," said, "I'm getting closer") (Table 5).

Table 5
Perception of nonspeech sounds

Above chance abilities	At chance abilities
Frequency discriminations greater than 400 Hz	Frequency discriminations less than 400 Hz
Environmental noises recognition	Melody perception
Animal noises recognition	Emotional prosody recognition
Cartoon voice recognition	

3.4. Temporal and pattern auditory function

1. Gap detection
   - *Materials and procedures:* The Random Gap Detection Test (RGDT) (Keith, 2000) was administered on two occasions (4 months and 6 months post-injury) to assess H.S.'s temporal auditory processing abilities. The stimuli for this task consist of tone pairs of the same frequency, at octave frequencies ranging from 250 to 4000 Hz. Stimuli are 17 ms in total duration, with a 1 ms rise/fall time. Gap increments from 0 to 40 s are presented in random order, with 2 ms serving as the smallest non-zero gap. Visual cues consisting of a drawing of the two tones and written instructions were utilized in explaining the task to the child. The child was given the option of saying "one" or "two," or pointing to a pictoral representation of one or two to indicate how many tones were heard.
   - *Results:* Typical performance on gap detection tasks is in the range of 5.6 ms. for 5-year-old children and 5.2 ms. for adults (Trehub, Schneider, & Henderson, 1995). The normative data for children on the RGDT indicate that detection threshold should not exceed 20 ms. H.S. was unable to reliably identify gaps as large as 40 ms. (the maximum gap interval on the RGDT) during administration of the RGDT on both occasions, suggesting severely impaired temporal processing.

Table 6
Perception of temporal and duration cues

Above chance abilities	At chance abilities
None	Gap detection Durational pattern recognition

2. Durational pattern
   - *Materials and procedures:* The Durational Pattern Test (Musiek, 1994; Noffsinger, Wilson, & Musiek, 1994b) was utilized to further assess temporal auditory processing abilities. In this task, the participant is presented with a sequence of three long or short tones, and is asked to choose verbally or by humming which of six possible duration sequences (e.g. short, short, and long) matched the tones presented.
   - *Results:* Individuals typically perform at 80% accuracy on the task in comparison to chance levels of 17%. Utilizing a verbal response, H.S. correctly identified only 4/30 (13%) of the patterns correctly. For a hummed response, 2/30 items (7%) were correctly identified. Neither response style fell above chance levels.
3. Frequency pattern
   - *Materials and procedures:* The Frequency Pattern Test (Musiek, 1994; Noffsinger et al., 1994b) consists of a series of three tones in which one of the tones differs in frequency. The participant is asked to repeat the tonal sequence either verbally or by humming the temporal sequence (e.g., low, high, and high).
   - *Results:* Preliminary to administration of the task, H.S. was asked to identify whether pairs of tones differing in frequency were the same or different. He could not perform the screening tasks above chance levels. Consequently, the Frequency Pattern Test could not be administered (Table 6).

4. Discussion

Among published accounts of auditory agnosia, this case is atypical in many respects, including the mechanism of injury, age of the individual, and location of neurological insult. Furthermore, an extensive array of auditory stimuli was used in the assessment, yielding evaluation of aspects of sound processing such as perception of emotional tone and recognition of familiar voices that are rarely described in other cases. By presenting such an extensive range of stimuli, ranging in complexity from peripheral auditory function to high-level auditory perception and interpretation and spanning most known aspects of sound processing, we are able to present a more complete clinical picture.

The vast majority of auditory agnosias described have occurred in patients with vascular injuries, typically infarcts (Griffiths et al., 1999; Simons & Ralph, 1999). From recent reviews and case studies, we could identify four cases of auditory agnosia following traumatic brain injury: three following closed head injury (Franklin, 1989; Lambert, Eustache, Lechevalier, Rossa, & Viader, 1989; Vitte et al., 2002) and one following a gunshot wound to the head (von Stockert, 1982). Because vascular injuries are typically the causal event, very few cases of acquired auditory agnosia in the presence of otherwise normal language functioning have been identified in children (Buchman, Garron, Trost-Cardamone, Wichter, & Schwartz, 1986; Rapin, Mattis, Rowan, & Golden, 1977; Simons & Ralph, 1999). This case study is another demonstration that, although rare, auditory agnosia can occur following traumatic brain injury, and describes the first known occurrence of auditory agnosia following a traumatic brain injury in a child.

Another uncommon aspect of this case involves the location of identified lesions. Although the patient sustained a severe traumatic brain injury and therefore shearing and diffuse axonal injury are undoubtedly present, neuroradiological findings indicate that the temporal lobes are largely spared. Specifically, minimal focal injury appears to have occurred to primary and secondary auditory cortex such as the anterior transverse temporal (Heschl's) gyrus and adjacent areas of the superior temporal gyrus. Cortical injury appears to be largely limited to cortical contusions and edema to the temporal poles as well as aspects of the parietal and occipital lobes. Focal injuries occurred primarily at the level of the corpus callosum, caudate nucleus, internal capsule, thalamus, and cerebral peduncle.

Although subcortical or brainstem lesions are believed to have been causal in a few cases of auditory agnosia (Johkura et al., 1998; Shivashankar et al., 2001; Taniwaki et al., 2000; Vitte et al., 2002) these cases represent a very small proportion of published accounts. Ascending auditory tracts within the brainstem transmit input from both ears, indicating that a bilateral lesion would be needed to disrupt auditory perception at the brainstem level. Furthermore, the auditory tract shares close proximity with other vital motor and sensory pathways, and therefore specific disruption of auditory processing would be unlikely to occur in the absence of other profound neurological deficits (Griffiths, 2002). Nonetheless, cases of auditory agnosia have been described in the absence of cortical lesions, indicating that a disturbance of projections to the auditory cortex may be sufficient to create the syndrome. In the present case, the absence of a middle latency response during testing of auditory evoked potentials is consistent with impairment to thalamocortical pathways bilaterally

(Scherg & von Cramon, 1986). Furthermore, the suppression of input from the right ear on the dichotic speech task is also consistent with a disruption of access to functions subserved by the left hemisphere language areas. This case study presents further evidence that impairments in complex sound processing can occur following subcortical and brainstem injury despite the presence of intact auditory cortex bilaterally. Such a finding is consistent with recent research demonstrating that subcortical areas are not only key to temporal processing, but in fact show more response to fine temporal discriminations than cortical areas (Harms, Melcher, & Weisskoff, 1998). Processing of complex sounds such as speech is inextricably linked to perception of temporal cues, to the point where speech can be perceived on the basis of temporal cues alone (Shannon et al., 1995).

This case study indicates that auditory agnosia is possible following traumatic brain injury in a child with largely subcortical lesions, and provides a detailed description of such a child's pattern of performance. Nonetheless, several limitations of the methodology of this single-case study must be considered. The child's premorbid history does not rule out the presence of undiagnosed phonological or other language impairment prior to his injury, although clearly not to the degree present following his injury. Although the relative lack of injury to temporal lobes and clear subcortical lesions point toward damage of thalamocortical tracts as being causal, the child did experience widespread brain damage, and therefore any putative lesion site must be viewed with caution. Furthermore, limitations in the patient's general cognitive functioning and attention span often necessitated adapted administration of standardized tests. It was also necessary to create tests when standardized tests were too complex or did not exist for a specific category of auditory stimuli. Therefore, normative data are not available for most tasks, and it was not possible to equalize difficulty across tasks. Tests created by the authors were designed to have similar task demands and minimal cognitive and attentional demands, and were believed to be tests that would be "easy" for even a child with a significant brain injury. Furthermore, there was no direct relation between potential indicators of difficulty such as the duration of an auditory stimulus and H.S.'s rate of success on tasks. Nonetheless, unless normative information for each of these tasks could be collected on same-aged individuals with similar degrees of brain injury, task difficulty cannot be eliminated as a factor in this child's performance. Finally, the testing of auditory abilities for this patient was limited to factors that were felt to be clinically relevant. Additional testing of rhythm, meter, complex tones, or melodic-organization (e.g., scale-violated, countour-violated, or interval-violated melodies) would have been scientifically interesting (Sidtis, 1981; Vignolo, 2003), but was not felt to be clinically warranted given the child's limited cognitive and attentional resources and the stringent demands of his rehabilitation program.

Pure word deafness as a clinical condition has historically been used as evidence to support the qualitatively different nature of speech in comparison to nonspeech sounds. Recent challenges to this theory of modularity in auditory processing have suggested that pure word deafness, or strictly verbal agnosia, may be at least in part an illusory phenomenon arising from limitations in the range of auditory stimuli presented to subjects. With more detailed testing, deficits in the perception of music, environmental sounds, or other auditory stimuli are frequently observed to be present in an individual with verbal agnosia, either concurrently or at different points in the recovery process (Pinard et al., 2002). Newer studies have further eroded the boundary between traditional views of left-hemisphere semantic processing and right-hemisphere processing of nonverbal stimuli such as music and prosody. Recent experimentation has demonstrated not only the presence of impaired processing of nonverbal sounds in aphasics, but has further revealed that impairments in nonverbal and verbal processing are highly correlated, indicating shared neural resources (Saygin, Dick, Wilson, Dronkers, & Bates, 2003).

Deviation from the traditional view of modularity in auditory processing has led to a greater emphasis on the similarities and differences among acoustic features of various auditory stimuli. A deficit in processing music, for example, can result from impaired melody, impaired rhythm, or both, and deficits in these underlying characteristics of music may be more lateralized than amusia itself (Vignolo, 2003). Similarly, prosodic information is conveyed in frequency, intensity, durational features, articulation and voice quality, with each characteristic interacting with the others, and fluctuating in importance over the course of a single utterance in a single individual (Sidtis & Sidtis, 2003). By comparing H.S.'s performance on a variety of auditory processing tasks, we were able differentiate chance level performance from above-chance performance for individual tasks, while also identifying relative strengths and weaknesses across tasks. With such comparison, a pattern of relatively spared and relatively impaired abilities is delineated. Perception of speech, music, and prosody is markedly impaired, while perception of animal and environmental noises and familiar voice recognition are relatively spared. Nonetheless, the relative importance of various acoustic features in the processing of each of these types of auditory stimuli is largely unknown, and therefore an attempt to identify any disturbance in one or more underlying acoustic features must remain speculative.

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