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A Case Study of Pure Word Deafness: Modularity in Auditory Processing?

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Abstract

AL, a woman with an acquired disturbance of auditory processing beginning in the second decade, was originally diagnosed as having pure word deafness. Recent analysis with a wide range of stimuli suggests that her comprehension deficit also extends to a subset of musical and non-verbal environmental sounds. The perceptual demands of the different auditory stimuli appear to account for part of the apparent material specificity. Additionally, over the years, the presumed temporal lobe cortical pathology has been supplemented by a mild to moderate, peripheral low-frequency hearing loss and evidence of dysfunction in lower level auditory processing pathways. The current peripheral dysfunction closely resembles cases recently labeled as auditory neuropathy. The diagnosis of pure word deafness should not be based on a limited set of auditory stimuli; additionally, a careful assessment using modern audiological techniques should be performed to evaluate peripheral auditory functions.

Introduction

Pure word deafness is a syndrome of verbal auditory agnosia characterized by an inability to comprehend only spoken language despite intact hearing. It is a rare syndrome generally attributable to dominant unilateral lesions affecting Heschl's gyrus, although bilateral superior temporal lobe lesions have also been documented (Mesulam, 1985).

The classical 'behavioral neurology' literature on pure word deafness describes a syndrome in which auditory thresholds are preserved. There are problems with temporal resolution. Language processing is invariably affected, music processing is sometimes affected, heard speech cannot be repeated. Cortical evoked potentials are sometimes present, sometimes partially absent. Pathologically, there is claimed to be a disconnection between the secondary auditory cortical areas from Wernicke's area, unilaterally or bilaterally (Albert and Bear, 1957; Denes and Semenza, 1975; Saffran *et al.*, 1976; Auerbach *et al.*, 1982; Coslett *et al.*, 1984; Tanaka *et al.*, 1987; Yaqub *et al.*, 1988; Bauer, 1993; Adams *et al.*, 1997; Bauer and Zawacki, 1997).

In fact, there are four syndromes—auditory neuropathy, cortical deafness, auditory agnosia, and pure word deafness—which give rise to many overlapping symptoms. Part of the reason for this is that the structures involved often overlap; the other reason may be that the syndromes are not really distinct (Albert and Bear, 1957; Albert *et al.*, 1972; Adams *et al.*, 1977; Berlin *et al.*, 1998).

In this paper we present a detailed study of a subject, AL, clinically diagnosed with progressive pure word deafness. Our original evaluation of this woman was designed to address the question of whether the existence of pure word deafness provides evidence that speech is subserved by modular processes and hence is isolable from music appreciation, and the processing of environmental sounds.

Cases of pure word deafness are rare, but of considerable theoretical importance. The existence of the syndrome, it has been suggested, provides strong evidence for the claim that speech, music and environmental sound analyses are processed by distinct systems (Albert and Bear, 1957; Goldstein, 1974; Brick *et al.*, 1985). Alternatively, the absence of true pure word deafness would weaken such a claim. Unfortunately, the bulk of pure word deafness cases have consisted of clinical descriptions with a relatively limited set of test stimuli. Even so, many case studies have indicated that other non-verbal auditory stimuli were 'mildly' affected (Goldstein, 1974; Denes and Semenza, 1975; Goldstein *et al.*, 1975; Saffran *et al.*, 1976; Auerbach *et al.*, 1982; Coslett *et al.*, 1984; Metz-Lutz and Dahl, 1984; Buchman *et al.*, 1986; Tanaka *et al.*, 1987). In order to appreciate more fully

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the possibility of more extensive non-verbal impairment than previously appreciated in a fairly 'typical' pure word deafness case, clinical evaluation in our subject was supplemented by more detailed and controlled experimental testing of non-verbal processing. The results may call into question the concept of pure word deafness as a nosological entity.

Case report

Neurological history

Patient AL was first seen by a team of neurologists at McGill University and Harvard in 1974 at the age of 23 years. She had a normal developmental history with an uneventful full-term vaginal delivery and normal milestones. She reported that, at the age of 17 years, in her native India, she began experiencing difficulty with the comprehension of spoken language. There was no concomitant difficulty in hearing and her auditory thresholds were normal. Also there was no reported difficulty understanding music or environmental sounds. The hearing deficit progressed slowly over the next 4 years, and then stabilized. Clinical presentation and formal neuropsychological testing conducted 7 years after onset, led her neurologist to conclude that she had pure word deafness. The basis for this diagnosis was astute clinical observation in the office. According to the treating senior neurologist, 'she was unable to repeat even the simplest words aside from lip-reading. When I turned around, and rattled the keys in my pocket, she responded quickly: "those are your keys rattling". She was equally accurate in identifying coins jangling in my hand. I realized that this was Pure Word Deafness, as described in the textbooks' (Dr I. Libman, personal communication).

The patient was referred to Drs Norman Geschwind and M.-Marsel Mesulam at Harvard, who after a full evaluation concurred with this clinical diagnosis. She showed preserved auditory acuity and she was reliably able to recognize objects by their characteristic sounds. Perception of environmental sounds was judged to be normal, and she 'could recognize sounds such as crumpling of paper and jingling of keys' (Mesulam, 1982, p. 593). No mention was made regarding her processing of musical sounds. The results on both an examination of the ears and a neurological examination were normal, other than a tremulousness noted in her voice.

While the verbal comprehension deficit was her most prominent and profound deficit, AL also had less than adequate performance on visuospatial tasks, calculations, and reading of complex material. The patient was judged to be otherwise normal, if not superior, in other cognitive domains. The diagnosis made at that time was pure word deafness (specifically indicating a 'pure word deafness-like syndrome') of atrophic origin. No definite etiology was specified and no treatment was given. Milder deficits of left parietal function were also suspected.

AL was reported as one of the six initial individuals (patient 4) described by M.-M. Mesulam in his groundbreaking paper on primary progressive aphasia (PPA) syndrome (Mesulam, 1982). It is significant that of the 63 cases of PPA in the literature between 1982 and 1992, she remained the only one with onset below the age of 40 years, and the only one who primarily presented word deafness (Mesulam and Weintraub, 1992).

Table 1. Neuropsychological results for subject AL

Wechsler Memory Scale indices	
General memory	118
Verbal memory	106
Visual memory	136
Attention /concentration	107
Wide Range Achievement Test	
Reading	7th percentile
Spelling	34th percentile
Arithmetic	23rd percentile
Word fluency (FAS)	35 (56th percentile)
Trail making A	34 s
Trail making B	74 s
Multidimensional Aptitude Battery	
Verbal	92
Performance	88
Full scale	90

Over the following 10 years the patient remained stable. She successfully completed university training as a toxicologist (using written materials and functioning effectively as a deaf person). She married and continued to work in her profession. Both the patient and her spouse noted a minimal to slight gradual worsening of her auditory comprehension deficit over time. The subject was aware of difficulty understanding many spoken words, and could not use the telephone. She could recognize variation in speech loudness, but often confused a loud voice with an angry one. She compensated for her auditory deficit by lip reading and making skillful use of contextual cues. She reported that she rarely listened to music.

In 1992–1993, AL (now aged 38 years) was reevaluated by our research team. The main clinical features remained a tremulous voice and the obvious deficit in comprehending spoken (but not written) language. There were no apparent speech articulatory aberrations for phonemes whose acoustic characteristics mapped on to the frequency pattern of the hearing loss.

During her clinical evaluation she continued to be able to identify environmental sounds such as keys jangling and paper crumpling. It was subsequently noted that although limb coordination and gait were grossly normal, tandem gait deteriorated significantly when eyes were closed. The remainder of the neurological examination was normal.

In 1994 a complete neuropsychological examination was performed (see Table 1). She was tested in English, not her native language. On the Multidimensional Aptitude Battery [a paper and pencil analogue of the Wechsler Adult Intelligence Scale (WAIS)], she obtained verbal and performance scale values in the bottom 30% of the population at large. It should be noted, however, that this battery contains speeded subtests and requires English ability. The fact that her English reading speed was below average (she scored at about the seventh percentile in this area) may have affected her overall results. Her fine tactile perception, her finger tapping speed and her manual dexterity were all mildly impaired bilaterally. She had difficulty in articulation, and her performance was below average on oral word reading, written arithmetic and written spelling. In contrast, her immediate and delayed verbal recall were above average (the recall paragraphs were presented visually), and her immediate and delayed visual recall were superb. Her oral fluency was judged to be very good for a non-native speaker, and she had no problems copying complex geometric figures. The tests indicated no significant level of psychological stress. In general, the low scores on certain tests were felt to be due to the fact that these tests were speeded, and to the fact that English is not her native language.

The Face Emotion Test of Ekman (Ekman, 1976) was also administered in order to evaluate further her higher level, non-verbal processing ability. This test consists of 110 visual presentations of faces which appear individually and which express seven different emotions (i.e. disgust, sadness, surprise, anger, neutrality, happiness, fear). After each slide presentation, the subject is asked to indicate which of the seven emotions is being expressed. AL obtained 83.5 correct responses out of a total of 110 stimulus presentations, for a score of 76% correct (chance performance being 14.29% correct). The grand mean for the seven emotions judged by normal controls is 81.17%, so AL was slightly below normal (normally, subjects correctly recognize 86–93% of the emotional categories and score 19% correct on the neutral faces; Ekman, 1976). AL was judged to be unimpaired in the visual processing of emotional material.

Lesion localization

A series of tests was carried out between 1978 and 1994 to localize and delineate the responsible pathology.

In 1978 a computed tomography scan showed slightly more atrophy in the left hemisphere than that expected for someone her age and an increase in ventricular size. Auditory cortical evoked potentials were grossly abnormal to 90 decibel clicks and were markedly suppressed bilaterally to speech, especially when measured from the right ear. In contrast, in 1980, an audiogram (see Fig. 1) as well as brainstem evoked potentials were judged to be entirely normal. At the same time, the audiology report noted that the acoustic reflexes were absent on the right and elevated on the left (suggesting peripheral nerve involvement). These results were interpreted to mean that her pathology was primarily localized to the cortex, specifically to the left hemisphere.

In 1984 a follow-up brainstem auditory evoked response study with click stimuli demonstrated that waveforms were absent bilaterally (a single peak was recorded possibly from the cochlea). Repeat audiograms raised the possibility of bilateral partial sensorineural hearing loss.

In 1993–1994 an extensive audiological evaluation (including audiometry, pitch discrimination and pattern testing, gap detection, tone duration and decay, lip reading, auditory brainstem responses, auditory cortical evoked responses, and impedance testing) was undertaken. She also had a detailed otolaryngological evaluation, including electronystagmography. A full brain magnetic resonance imaging (MRI) scan and hexamethyl-propyleneamine oxime single photon emission computed tomography (Hm-PAO SPECT) were also performed.

At this point, the audiogram revealed a bilateral sensorineural loss—moderate to severe in the right ear and mild to moderate in the left ear (especially for low frequencies) (see Fig. 1). At this juncture on the brainstem auditory evoked response (BAER), wave 1 was clearly absent, as were all subsequent waveforms. The pattern of results on the audiograms deserves some explanation. With regards to the mild to moderate threshold loss, a comparable level of threshold loss resulting solely from a cochlear (sensory) lesion is usually associated with comparably poor speech discrimination only in the speech frequencies affected by the loss (speech sounds are usually 250–4000 Hz). So if there is, for example, a low-frequency loss, only low-frequency sounds (e.g. voiced sounds—b, d, g) would be affected. One due to an VIIIth nerve lesion is associated with speech discrimination and speech comprehension difficulties disproportionately worse than those expected from the threshold loss alone. So, in this case, if there is a low-frequency threshold loss, both low-frequency (e.g. voiced sounds) and high-frequency (e.g. fricatives—f, s) sounds may now be affected. Finally, one due to a central lesion is associated with speech discrimination and comprehension difficulties which are even further deteriorated. In this last case, the low-frequency threshold loss would now not only be associated with loss of both low- and high-frequency sounds. Additionally, the loss in the low-frequency sounds would be of greater severity than that predicted by the degree of threshold loss; thus a mild threshold loss could be associated with a moderately severe discrimination and comprehension loss in those frequencies (Katz, 1985; Sataloff and Sataloff, 1993; Becker *et al.*, 1994; Adams *et al.*, 1997). This pattern of audiogram results therefore supports lesion localization either at the VIIIth nerve level or more centrally.

With regard to the threshold loss especially in the low frequencies, a threshold loss resulting solely from a cochlear (sensory) lesion usually gives rise to an ascending air and bone conduction hearing level, indicating better performance at the higher frequencies. In sensorineural hearing loss, air and bone conduction are both reduced. The audiometric pattern most typical is that of a high-frequency hearing loss (downward sloping audiogram). That aspect of the audiogram results supports lesion localization more peripherally (Katz, 1985, Sataloff and Sataloff, 1993; Becker *et al.*, 1994; Adams *et al.*, 1997).

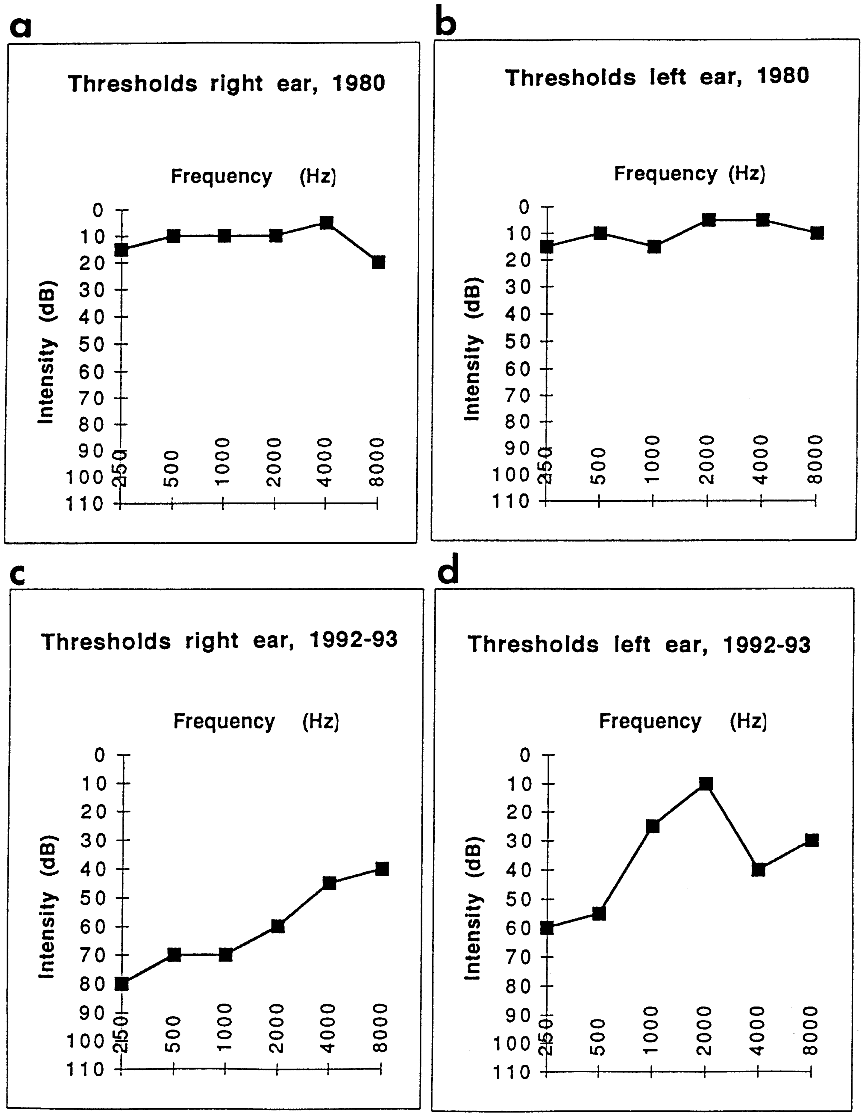

Fig. 1. Audiograms of patient AL in 1980 (a and b) and in 1992–1993 (c and d) for right and left ear thresholds. A deterioration in low-frequency thresholds is demonstrated over time.

The MRI scan of 1993 revealed evidence of small oval white matter hyperintensities in the left and right cerebellum on T2-weighted images, which were deemed non-specific (see Fig. 2). No cerebral hyperintensities were noted. A brain Hm-PAO SPECT scan revealed a focal perfusion defect in the left anterior temporal pole region (see Fig. 3).

The electronystagmogram in 1993 revealed near absent bilateral peripheral vestibular function. It was noted that the patient had never experienced oscillopsia (a visual sensation that stationary objects are swaying back and forth), thus leading to the conclusion that the vestibular disorder had been slowly progressive. The clinical findings of impairment of tandem gait with eyes closed and the loss of fine lines on the Snellen chart with head shaking, were, in the absence of oculomotor findings, consistent with peripheral vestibular dysfunction. The etiology of these changes remains unclear, but would be compatible with the involvement of both vestibular and auditory components of the VIIIth cranial nerve.

In 1995, electrocochleography (a measure of VIIIth nerve functioning) demonstrated no measurable action potentials in both ears. This result was consistent with auditory nerve damage outside the brainstem. Testing for otoacoustic emissions showed them to be strong in both ears, indicating that the cochlea itself was normal, and confirming the retrocochlear localization of her disorder. Nerve conduction studies performed in 1998 showed no evidence of peripheral neuropathy and a VDRL (venereal disease research laboratories) was negative.

In summary, there appears to have been a very slow progressive course over a 20-year period with respect to her auditory processing, but she has otherwise been neurologically stable. No multisystem degeneration or obvious cognitive deterioration has ensued. Anatomical and functional

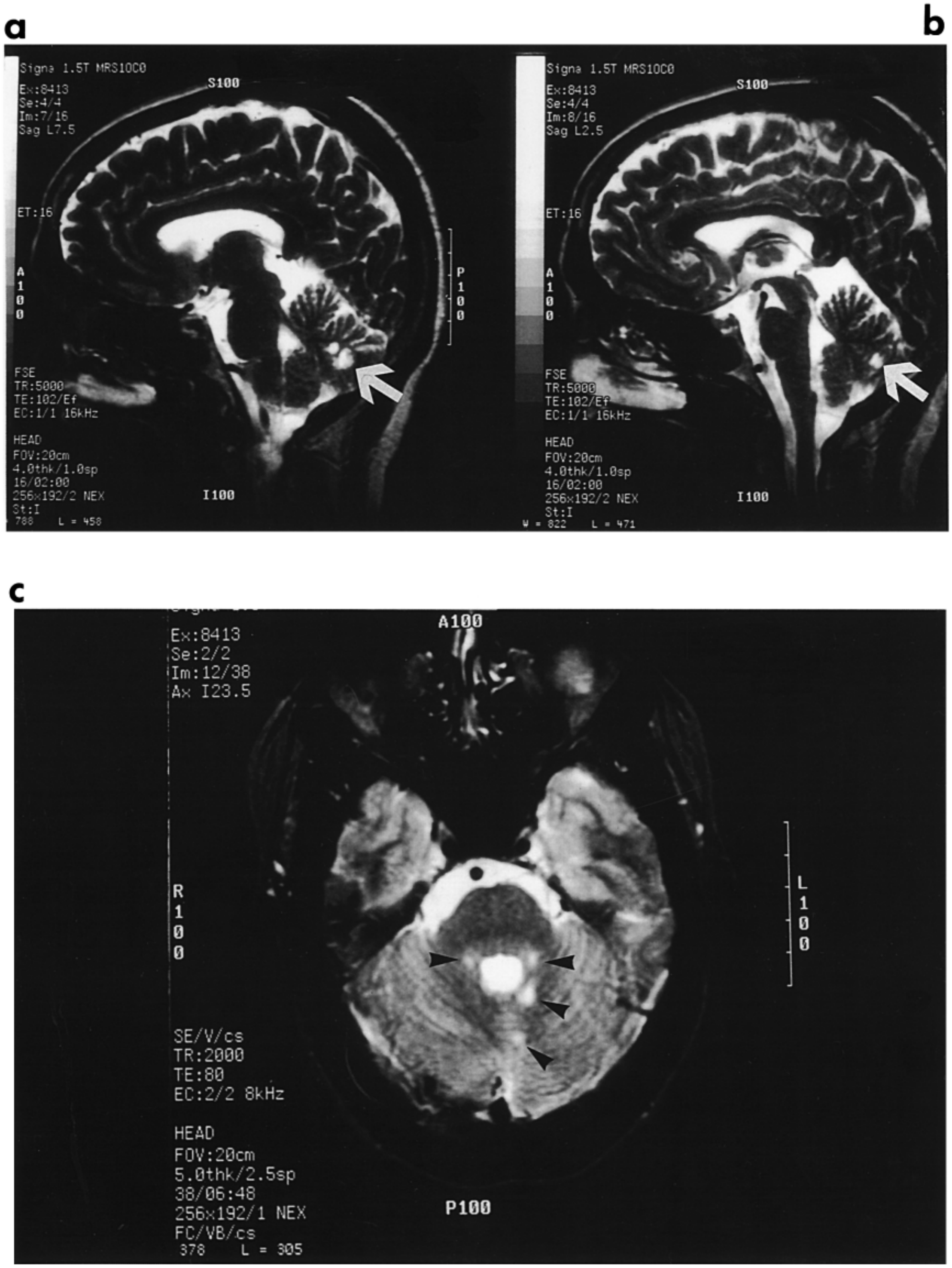

Fig. 2. Magnetic resonance imaging scan of AL (1993). (a) and (b) show T2-weighted (proton density) saggital images demonstrating small areas of increased signal in the left cerebellar vermis (white arrows). (c) shows the horizontal view (proton density) with no clear abnormality in the temporal cortices, but the deep cerebellar lesion is again visible (black arrows).

neuroimaging as well as neuroacoustic measures have demonstrated more extensive pathology compared with the initial (albeit less extensive) evaluation. Specifically, there has been more recent additional neurophysiological evidence of bilateral damage to the VIIIth nerve. Non-specific imaging abnormalities included white matter hyperintensities in the left and right cerebellum on MRI and a focal perfusion deficit in the anterior left temporal lobe on SPECT. Neurologically she remains normal except for the signs of vestibular dysfunction mentioned above.

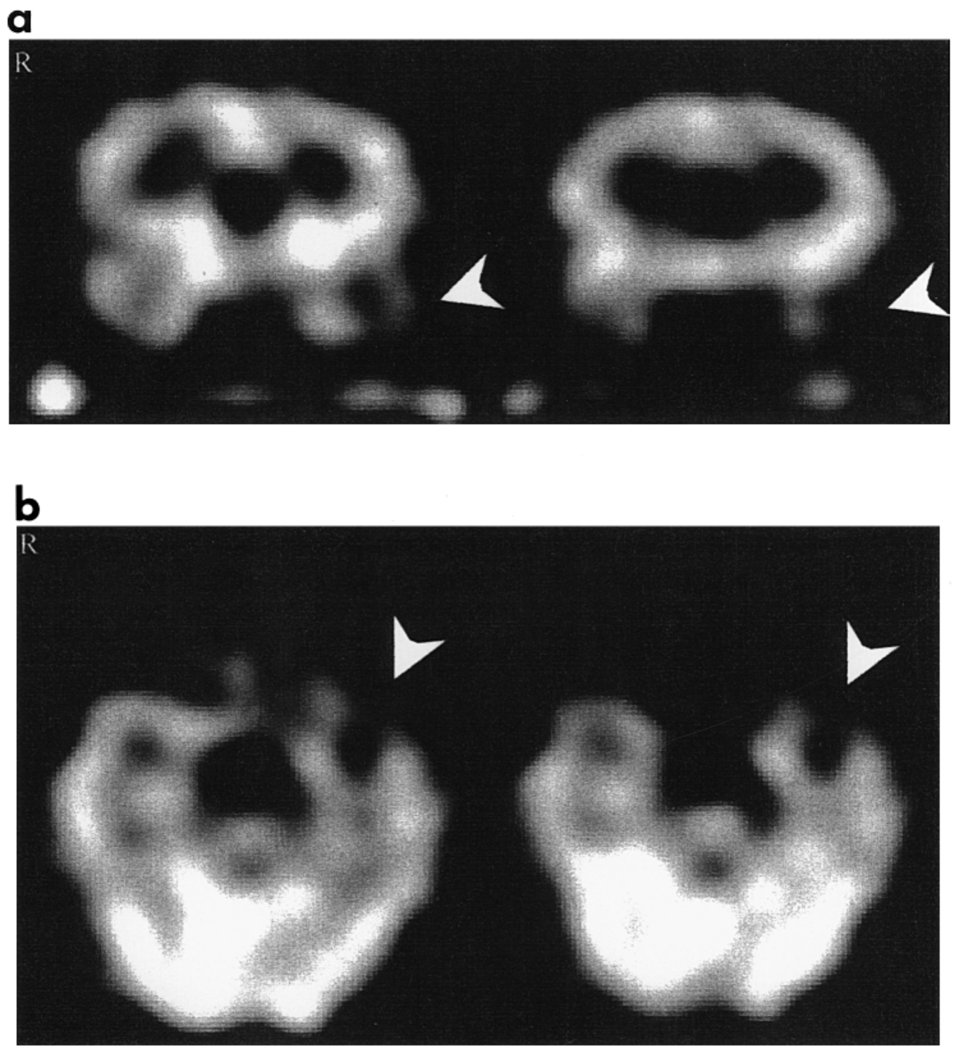

Fig. 3. SPECT scan with Hm-PAO (1992) demonstrating a mild focal decrease in cerebral blood flow at the left temporal pole (arrows). (a) shows two coronal sections through the temporal lobes, while (b) shows horizontal sections through the temporal lobes.

Experimental testing

General procedure

Subject AL was tested on a series of experimental cognitive tests between 1993 and 1995. All stimuli were presented on either a tape recorder, a visual screen, or a Macintosh Classic using Psychlab software (Bub and Gum, 1991). Instructions were written for all tests.

Processing of verbal non-auditory material

AL did not appear to have cognitive deficits outside the auditory modality, and this was substantiated by her performance on standard neuropsychological tests. There was no deficit of verbal or non-verbal memory when tested using the visual modality.

Materials and procedures. The written word to picture matching subtest from the Psycholinguistic Assessment of Language (PAL; Caplan and Bub, 1990; Caplan, 1992; Westbury, 1995) was presented to investigate AL's ability to carry out the processing of verbal material presented in the visual modality. In this test, 32 concrete nouns denoting vegetables, animals, fruits and tools were presented visually. The subject had to select one of two pictures as the correct match for the noun. The picture foils were semantically and visually similar to the nouns (e.g. deer as the target, moose as the foil). The noun targets were high- or low-frequency nouns, and were short (monosyllabic) or long (tri- or quadrisyllabic) (Caplan, 1992).

Results. Thirty of the 32 responses were correct. AL thus scored 94% correct (chance level performance being 50% correct), well within the normal range for her age. Normal controls tested on the PAL (aged 35–50 years) performed at 98.6% correct [standard deviation (SD) 3.2]. This gives AL a z-score of –1.44.

Processes for speech output: generating sound-based forms of words

We wished to assess whether AL's auditory language problems were limited to comprehension or additionally extended to a problem in generating sound-based forms of words for speech output.

Materials and procedures. Two homophone matching tasks were administered.

Homophone Matching A (from the PAL; Caplan and Bub, 1990; Caplan, 1992; Westbury, 1995)
In this test, 32 sets of paired pictures of common objects or actions were presented visually. The task of the subject was to indicate if the two pictured objects had the same verbal label. For instance, bat (the object) and bat (the animal) were presented as a pair of pictures. In this test, the orthographic labels denoting the pictured objects were identical. Sixteen homophones and 16 non-homophonic pairs were presented.

Homophone Matching B (Chertkow et al., 1994)
Eighty sets of common word pairs were presented written on a paper and the subject was asked to read them silently. All pairs had non-identical orthographic labels. Forty of the pairs were homophones and 40 were not. For instance, 'hair' and 'hare' share homophonic pronunciation, while 'cut' and 'cot' do not. The task of the subject was to indicate whether the two words had identical or non-identical pronunciations.

Results. On Homophone Matching A, AL obtained 100% (32/32) correct results (50% being chance). Normal controls (aged 35–50 years) performed at 96.2% correct (SD 8.4).

On Homophone Matching B, AL achieved 85% (68/80) correct results (chance performance being 50% correct). Normal controls performed at 96.25% overall. Given that AL is not a native speaker of English, these results were interpreted to be within the normal range.

It is generally accepted (Caplan, 1992) that homophone matching is carried out by generating a phonological form of each word which can then be compared. In the Homophone Matching A task, the homophonic words also shared orthography, and therefore subjects could conceivably match correctly on the task by generating orthography. In the second test, it is only on the basis of the stored phonological representation of the word that correct performance may be achieved. AL's normal performance on these two tests indicated intact phonological output processing when visual inputs were utilized. Having accessed the stored semantic representations of the word or object, AL had no difficulty generating the correct phonological output form.

Auditory input modality: word level processing (real words)

Five subtests of the PAL (Caplan and Bub, 1990; Caplan, 1992; Westbury, 1995) were used to test whole word auditory input processing for subject AL.

Auditory word to picture matching using semantic and visual distracters

Materials and procedures. In this first task, 32 words representing mainly animals, common fruits/vegetables, and common household items were presented. Half the items were one to two syllables long and the other half were three or more syllables long. Each recorded word was presented auditorily and was followed after a 2-s interval by two simultaneous pictures on the screen, one matching the previous word and the other denoting a distracter object. Foils were both semantically and visually similar to the targets (e.g. deer as the correct target and moose as the foil).

Results. On this task, performance was 72% correct (chance level being 50% correct). AL correctly matched 23 of 32 stimuli. Normal controls (aged 35–50 years) performed at 98% correct (SD 2.9). This gave AL a z-score of –8.97. AL's performance was thus poor, but above chance.

Auditory word to picture matching using phonological distracters (Caplan and Bub, 1990)

Materials and procedures. In this task, 17 words representing animals, common fruits/vegetables, and common household items were presented auditorily. As before, each recorded word was followed after an interval by two pictures, one matching the previous word, e.g. 'chair', and the other representing an object with a similar verbal label, e.g. 'pear'. These phonological distracters differed from the target by one phoneme in either the initial or the final position. The subject's task was, in both cases, to listen to the word on headphones and to indicate which picture best matched the heard word.

Results. Here, performance dropped dramatically: she obtained 53% correct (chance level being 50% correct), matching only nine items out of a possible 17. Normal subjects performed at 100% correct. She was, therefore, essentially at chance when distracters were phonologically similar.

This pattern suggests that partial phonological information was being extracted by AL from words presented auditorily. This partial information was sometimes sufficient to support correct matching against semantic distracters. Such a strategy collapsed in the face of phonologically similar distracters.

Auditory lexical decision

Materials and procedures. In this task, 80 words (40 meaningful and 40 specifically constructed nonsense words) were digitized on a Macintosh computer and presented auditorily. The meaningful words consisted of concrete nouns (common objects, animals, fruits or vegetables) and varied both in frequency (more than 40 per million to less than 5 per million) and length (either one syllable, or three or more syllables). Half the nonsense words had a change in one distinctive feature in a single phoneme in different syllabic positions (e.g. 'ponato'). The other 20 nonsense words were created by changing a syllable of the word so as to produce plausible alternatives (e.g. ‘harpsiform’ from ‘harpsichord’). A yes/no (word/non-word) response was required from the subject for each stimulus.

Results. AL’s performance was 56% correct (45/80; chance level performance being 50%) for words and non-words combined, with a total of 45 of 80 stimuli correct. She scored 57% correct for words (23/40), 55% correct for non-words (22/40) (chance 50% for both). Normal controls (aged 35–50 years) performed at 95.1% correct (SD 1.9). This gave AL a *z*-score of –20.58. She was thus markedly impaired on this task.

Repetition of oral meaningful and nonsense words

Materials and procedures. In this task, 40 words (20 meaningful and 20 nonsense words) were recorded for presentation through headphones. Half the words were short, the other half were long. The meaningful words represented common objects, animals, or fruits/vegetables. Nonsense words consisted of real words altered by substitution of two phonemes (e.g. ‘slazeny’). The subject was asked to repeat the word heard and the repetition was recorded. Items were scored both in terms of repetition being entirely correct and also in terms of single phonemes being correct, i.e. ‘partial’ repetition.

Results. Here performance was also very poor—0% for entirely correct words and 15% if part marks were attributed for single phonemes. By ‘part marks’ it is meant that if subject AL transcribed heard ‘precipice’ (prEsipis) as ‘express’ (EksprEs), part marks were attributed for ‘Eprss’, the phonemes repeated correctly.

Normal controls (aged 35–50 years) performed at 97.3% correct (SD 5.3). This gave AL a *z*-score of –18.36. Some examples of AL’s repetition errors are shown in Table 2. It can be seen that partial phonemic identification was occurring in a good number of cases, even though the whole word was incorrectly reproduced.

As can be seen in Table 3 (although we did not perform statistical tests), clearly, syllabic structure was perceived quite well. Also, if we examine performance by number of phonemes properly repeated, performance was markedly better than if the whole word was judged as correct or incorrect. Finally, vowels were markedly better perceived than consonants, especially in non-words. It is interesting to note here that performance seemed to be superior in non-words.

Writing to dictation

Materials and procedures. In this task, words and nonsense words were recorded for auditory presentation. Twenty-five were non-words (e.g. ‘snid’), and 20 were real words (e.g. ‘leather’). The real words denoted common concrete or simple abstract nouns. The subject was instructed to write down each word or nonsense word heard.

Table 2. Sample repetition errors to auditorily presented word stimuli for subject AL

Stimulus	Repeated form
b**ine**	d**ine**
n**i**d	l**i**st
bran**ch**	**br**i**dg**e (tch to dg)
w**eed**	tr**eed**
pensa**phon**	tele**phone**
b**i**p	s**i**x
precipi**ce**	**ex**p**ress**
pr**int**	w**itt**y
stetho**sc**ope	**sixt**y (sc to cs)
smurt	**swift** (clusters preserved)
job	**ja**w
d**ipe**	l**ike**
plant	**Frenc**h (cluster preserved)
far	**fr**yme
bro**sk**	a**x**e (sc to cs)
crocod**i**le	p**ork**f**ry** (cr to rk)
g**auz**e	h**ors**e (z to rs)

Letters shown in **bold** represent those preserved in whole or slightly transformed by the subject in the repeated form. Slightly transformed = inversions, cluster preservation, one distinctive feature difference.

Results. On this task, AL was virtually unable to transcribe heard words correctly (4% correct). Some details of her performance are shown in Table 4. There was again evidence of partial recognition of the stimuli. If part marks were given to correctly performed phonemes, she obtained 34% correct. By ‘part marks’, it is meant that if subject AL transcribed heard ‘precipice’ (prEsipis) as ‘express’ (EksprEs), part marks were attributed for ‘Eprss’, the phonemes transcribed correctly. Vowels were consistently more often identified than stops, and phonemes were more frequently identified within words than within non-words. AL succeeded in obtaining a 64.5% correct score for vowels in word stimuli (20 correct out of 31), and at the other extreme, obtained only 8.8% correct for stops in non-word stimuli (three correct out of 34). Normal controls (aged 35–50 years) performed at 99.1% correct (SD 2.0) overall. This gave AL a *z*-score of –47.35.

Overall, these results indicate severe disturbance in generating an intact phonological input from heard words. There was evidence in many instances of partial extraction of acoustic/phonological information, generally insufficient to support correct performance, unless the distracter items were easily differentiable in terms of their phonology.

Auditory input modality: word level processing (nonsense words)

Materials and procedures. Auditory input processing at a level more basic than (i.e. prior to) whole word representations was tested using an auditory nonsense words matching task (from PAL; Caplan and Bub, 1990; Caplan, 1992; Westbury, 1995). In this task, 40 pairs of pronounceable, monosyllabic,

Table 3. Repetition of oral meaningful and nonsense words by underlying units of analysis

	Word[a]	Phonemes[b]	Consonants	Vowels	Syllables
Non-words	0/20	43/95	21/63	22/32	41/45
	(0%)	(45.26%)	(33.3%)	(68.75%)	(91.1%)
Words	0/20	31/97	19/66	12/31	41/46
	(0%)	(32%)	(28.8%)	(38.7%)	(89.1%)
Both	0/40	74/192	40/129	34/63	82/91
	(0%)	(38.5%)	(31%)	(54%)	(90.1%)

Results presented as number correct/total number of stimuli.
[a]If not an exact replica of the stimulus then judged to be wrong, e.g. bif repeated as bik.
[b]Total sum of phonemes in stimulus, e.g. brosk = 5 phonemes b/r/o/s/k.

nonsense words were presented auditorily. Members of a pair were either identical ($n = 21$) or dissimilar ($n = 19$). Of the 19 dissimilar pairs, there were 11 stop consonant contrasts, five fricative pairs, one liquid pair, one nasal pair. There were no vowel contrasts. Also, there were seven voicing and six place contrasts. AL would probably have done much better had vowels been tested. The subject had to state whether the members of the pair were the same or different.

Results. Overall, AL correctly matched 63% of the pairs (25 correct out of 40). She was thus very impaired, chance performance being 50%. Normal controls (aged 35–50 years) performed at 94.6% correct (SD 8.7). This gave AL a z-score of –3.63. She was 86% correct in ‘same’ responses for matching pairs (18 correct out of 21), whereas chance was 52%. She was 37% correct for ‘different’ responses for dissimilar pairs (seven correct out of 19), whereas chance was 48%. She thus had an obvious bias towards responding ‘same’.

In summary, AL exhibited clear difficulties in the auditory processing of verbal material, with respect to words and nonsense words. She could neither match these to each other, nor reliably carry out lexical decisions, repeat the words, or write them to dictation. This deficit stood in contrast to her intact ability to use output phonological representations for homophone matching.

Auditory input modality: voice gender and emotional intonation judgments

Clinically, AL seemed able to judge tone of voice to some degree. This ability was, however, at times confounded with judgments of loudness of voice. For instance, the family reported that she stormed into the children’s room one day when they were talking loudly, certain that there were angry voices. We wished to evaluate whether her auditory deficits for spoken material extended to prosody. Two tests were administered: voice gender recognition and recognition of emotional intonation. The hearing impaired were not tested on any of these. We have no control data on these tests for hearing-impaired subjects. However, we may surmise indirectly, from what follows, on the effects of hearing loss on performance. The literature on the hearing impaired gives us indirect and few results on their performance. Grose and Hall (1996) report that patients with sensorineural hearing losses are poorer than normals at hearing a target melody from competing melodies (melodies are very similar to intonation). In terms of environmental sounds, Hedrick and Jesteadt (1996) found no significant differences between the hearing impaired and normals when amplitude was manipulated, but relative impairment for the hearing impaired when duration was manipulated (amplitude and duration are key features characterizing environmental sounds). Finally, Jerger *et al.* (1995) report that hearing impaired patients process gender (voice), but that for them gender is more independent of words; in other words, gender may be processed at a more elementary, acoustic level, less tied to its word context. Thus, we may surmise that the hearing impaired would be impaired with some aspects of environmental sounds, nevertheless, none of these data tell us the extent of the hearing impaired loss compared with someone like AL (Jerger *et al.*, 1995; Grose and Hall, 1996; Hedrick and Jesteadt, 1996).

Table 4. Writing to dictation: vowel versus stop consonant identification within real words and non-words (percentage correct)

	All stimuli	Word stimuli	Non-word stimuli
Whole word ($n = 45$)	4.4		
Speech sounds[a] ($n = 196$)	34.2	43.6	24.5
Vowels ($n = 57$)	52.6	***64.5***	40.0
Stops ($n = 67$)	23.9	36.4	***8.8***

[a]Vowel, stop, fricative, semi-vowel, nasal.

Voice gender recognition (Peretz et al., 1994)

Materials and procedures. Twenty sets of stimuli were prepared. Each stimulus consisted of a taping of either one of two French phrases: 10 of ‘C’est très bien’ (That’s fine); 10 of ‘ça va mieux’. (I’m feeling better). Half of each of the phrases were spoken by a young woman, the other half by a man. The subject’s task was to identify whether the voice heard was that of a man or a young woman.

Results. On the voice gender recognition task, AL’s performance was 55% correct (11 correct out of 20, with chance performance being 50% correct). She was equally impaired at recognizing the male and the female speaker's gender. Normal controls perform at ceiling on this task (Peretz *et al.*, 1994).

Recognition of emotions (intonation) (Bryden and MacRae, 1989)

Materials and procedures. For the second subtest, the stimulus set consisted of the four words 'power', 'bower', 'dower', and 'tower', each spoken by a male speaker in a tone of voice which was either happy, sad, angry, or neutral. An initial group of 20 subjects had rated each of the stimuli to ensure that the effect was perceived as intended (Bryden and MacRae, 1989). The 16 tokens were digitized on a modified PDP 11/40 computer, edited to a common length of 500 ms, equalized in loudness, and stored for playback. The stimuli were presented binaurally, with four sets of 18 trials (72 trials total). The subject's task was, after each word presented, to write down which of the four emotions was being expressed.

Results. For the recognition of emotions (intonation) task, AL was 25% correct (18 correct out of 72, which was equivalent to chance performance). The norms [cited in Bryden and MacRae (1989)] are between 65.54 and 78.82% correct.

Thus, AL was markedly impaired at recognizing either gender or tone of voice in auditory verbal material. This deficiency stood in striking contrast to her success in judging emotional content of facial expressions, for which she scored normally (see neuropsychology section).

Auditory input modality: processing of environmental and musical sounds

An essential feature of pure word deafness is that it is claimed to spare non-verbal auditory material. As noted, few clinicians have at hand a very rich store of such test materials. A formal test of non-verbal sound processing was therefore carried out, using a sound identification task and a sounds-to-picture matching task. Ten age-matched normal controls were also tested on the materials.

Sound identification

Materials and procedure. Twenty-seven sound clips, drawn from the *Sound Encyclopedia* (Lucasfilm Ltd and Sound Ideas, 1983) were digitized and recorded. These consisted of seven animals (e.g. lion, donkey), five musical instruments (e.g. violin, trumpet), five objects (e.g. tennis racket, hammer), and 10 vehicles (e.g. train, helicopter). The sounds were presented through headphones with a pseudo-randomized order of presentation in terms of categories. After each stimulus was heard twice, the subject was asked to identify the sound.

Table 5. Identification and recognition of environmental sounds (percentage correct)

	Identification (mean over two trials)		Recognition (picture matching)	
Category	Correct response	Same category	Correct response	Same category
Animals ($n = 7$)	43	14	86	14
Vehicles ($n = 10$)	10	30	50	40
Musical instruments ($n = 5$)	20	40	40	40
Objects ($n = 5$)	40	0	80	0
Overall ($n = 27$)	26	22	63	26

Results. Preliminary testing with four to six young controls in their mid-twenties showed that they scored a mean of 76% correct on the sound identification test. AL's performance for the sound identification task was only 26% correct overall (seven correct out of 27 sound clips). If same category responses were included as correct (e.g. the response 'car' for motorcycle), then her performance rose to 48% correct (13/27). There was some measure of variability in her identification success. She correctly identified three of seven animal sounds and two of five objects. In contrast, she was only able to identify correctly one of 10 vehicles and one of five musical instruments (see Table 5). For these latter two categories, there was a distinct tendency to produce same category or superordinate responses (e.g. 'it's a vehicle'). While some responses were inconsistent, a consistent pattern of response was seen in a subset of items (Table 6).

Sounds to picture matching

Materials and procedures. Forty-six sounds again drawn from the *Sound Encyclopedia* (Lucasfilm Ltd and Sound Ideas, 1983) were recorded. These consisted of 14 animals, four musical instruments, 11 objects, and 13 vehicles. For each sound, four pictures were constructed. One of the pictures corresponded to the concept whose sound was heard, while the three others were semantic distracters from the same semantic category. The order of presentation was randomized. The subject's task was to match (by pointing with her finger) the heard sound with the corresponding picture on the screen.

Results. Four to six young controls in their mid-twenties performed at a mean of 95% correct. Chance performance was 25% on this task. AL's performance was 63% correct overall. AL's performance was particularly good with animal and object sounds, and worse for musical instruments and vehicle sounds (see Table 5).

Instrument identification and instrument recognition (Peretz et al., 1994)

Materials and procedures. In this task, musical instruments were assessed with single instrumental pieces taken from commercial recordings, with stimuli administered at two

Table 6. Sample environmental sound identification

	Correct response	Same category response	Incorrect response
Animal sounds	***dog*** ***donkey*** ***cow***	**pig** **('cow', 'animal')**	
Vehicle sounds	***car*** motorcycle (correct, or 'car') **bus** (correct, or 'like a plane')	**wagon** ('car', 'airplane') **roller skate** ('car', 'airplane')	***sailboat*** ('animal talking', 'car') **truck** ('helicopter', 'noise') **airplane** ('water falling', helicopter')
Musical instruments	**violin** (correct, or 'musical instrument')	**trumpet** ('clarinet', 'violin')	***drum*** ('trumpet', 'vehicle')
Object sounds	***pitcher*** ***hammer***		***tennis racket*** ('animal', 'rolling pin')

sessions. The response mode was either (a) naming (i.e. identification), or (b) written multiple choice (i.e. recognition). Excerpts from 10 musical instruments (e.g. cello, harmonica, piano) were prepared on tape. Two blocks of randomized presentations of each of these 10 stimuli were presented (10 for the first block, nine for the second block). Stimulus presentation was identical for both tasks. For the identification portion, the subject had to write down the name of the instrument playing. For the recognition portion, the subject had to select the appropriate instrument from a written listing of all 10 instruments.

Results. On the identification portion of the task, AL's performance was 29% correct (5.5 correct out of 19, with chance performance being close to 10%. A half point was given for a violin response when the actual stimulus was a cello. The two instruments are auditorily close to one another, although still distinct). In contrast, nine to 10 controls (Peretz *et al*., 1994), produced scores of 83.3–100% correct. On the recognition portion of the task, AL scored only 11% correct (two correct out of 19), which is at chance.

The violin (an instrument of high pitch and long attacks) was correctly identified most often. The very poor performance on the recognition portion of the task was somewhat surprising, as recognition is usually easier than identification.

Thus, AL's performance was better than chance for environmental sounds, but remains markedly below what would be considered normal.

Auditory input modality: music comprehension

We wished to test formally whether AL's auditory deficit extended to music. A song recognition test was therefore devised.

Materials and procedures. A song recognition test was created and administered. This test was constructed in our laboratory, using a native Hindi speaker from the same region as the subject to select the songs likely to be familiar to AL from childhood. A set of excerpts from some very familiar, mostly popular, songs in AL's native language was selected as well as an equal number of not too familiar songs. Eighteen presentations were arranged. A native-born, healthy individual from AL's region decided upon the familiarity (or lack of it) of each item. The order of the excerpts was randomized and each excerpt appeared only once.

After each presentation the subject had to (a) decide whether the stimulus was a familiar or an unfamiliar one, (b) choose, from four possible written choices, which one was the correct song heard.

Results. AL's score on familiarity was 0% correct (0 of 18), with chance performance being 50% correct. Her score on recognition was 11% correct (two of 18) with chance performance being 25%. The slightly better performance on the recognition task was possibly an order effect; this task was administered after the familiarity task. She was thus clearly impaired on song recognition.

Discussion

The classical behavioral neurology literature views pure word deafness as a rare cortical syndrome whose existence demonstrates the modularity of auditory processing, presumably at a high linguistic level. Its existence in fact constitutes good evidence for the specificity of brain regions—the basis of the disease is claimed by some to be the anatomical disconnection of basic auditory processing regions from those (e.g. Wernicke's area) specialized for comprehension of language (Mesulam, 1985; Heilman and Valenstein, 1993).

Clinical evaluation of our subject AL as early as 1978 resulted in a clinical diagnosis of pure word deafness by all concerned, including leading figures in behavioral neurology. The current clinical reevaluation indicates that AL's deficit remains clearly limited to one cognitive domain; there appears to be no deficit in non-auditory cognition, memory, frontal lobe function, etc. Similarly, there appears to be no deficit in visual identification, recognition, and processing of stimuli—from written words to faces. Visual recognition of emotional expression in faces is intact. Furthermore, phonological output processing is intact, as evidenced by normal performance on the homophone matching tasks. AL's language problems have been exclusively limited to auditory comprehension.

For verbal material, it is clear that AL fails on all tasks that call for accurate activation of the phonological form of words for input and comprehension, while tending towards partially correct interpretation of certain phonemes within words. Furthermore, the impairment extends to comprehension of nonsense words as well. This pattern strongly suggests that the critical functional deficit precedes activation of the phonological word form, at either a basic psychoacoustic level of analysis, or at the level of phonemic analysis. The details of this functional deficit will be further explored in a subsequent study.

A claim was never made by either the initial or the subsequent treating clinicians for the absolute preservation of non-verbal processing in AL's case (it was at one point stated that AL's deficits were 'generally limited to language'). Nevertheless, the dissociation was considered striking and virtually complete. This initial clinical impression of 'pure word' impairment did not, however, bear up under our more detailed testing. On formal testing of environmental sounds, AL correctly identified only 26% of environmental stimuli and matched 63% correctly—far from normal. For the identification of music and voice gender, the deficit was even more striking. Detailed evaluation of patient AL, in other words, reveals her to be a case of auditory agnosia, rather than pure word deafness (or strictly verbal agnosia).

One can, of course, make the case that this performance on environmental stimuli was superior to AL's virtually complete inability to identify (e.g. repeat) words. It is more relevant, however, to explore why some environmental sounds might have been identified and others were not. There are a number of variables which might have influenced AL's success in identifying sounds. First, *repetitiveness* may have been an important factor. Dog sounds, cow sounds, the sound of a saw, are all repetitive (as were the sounds used initially by the neurologist who arrived at the diagnosis of pure word deafness). Second, some sounds are either much *louder* or much more *distinctive*; they manifest differences in frequencies, intensities, and durations that vary dramatically in either magnitude or scope. Car sounds and bus sounds are loud, and the sounds of a motorcycle and a trumpet are highly distinctive. Finally, the *duration* of the sounds might have been important. Both the tennis racket sound and the lion sound consist of short bursts—sounds that were, in fact, misidentified. In contrast, the sound of a violin is very elongated, and was identified correctly.

At a very simple level of analysis, consonant phonemes are non-repetitive, generally soft in volume, lack distinctiveness, and are of short duration. Thus, very few consonant phonemes would be recognized correctly by AL. In further experiments exploring the underlying processing deficit producing AL's errors, we have argued that these fundamental differences between word sounds and environmental sounds can easily explain the better performance for certain non-verbal material without recourse to any assumptions regarding modular processing differentiating the two sorts of material (Pinard *et al.*, 1993).

Is AL different in this pattern of performance from previously reported cases of pure word deafness? In fact, clear-cut cases of unadulterated pure word deafness (i.e. deficits affecting exclusively the linguistic component of complex auditory processing) are a rarity even among pure word deafness subjects. Indeed, of 63 cases reviewed, only five (Albert and Bear, 1957; Goldstein, 1974; Metz-Lutz and Dahl, 1984; Brick *et al.*, 1985; Yaqub *et al.,* 1988) reported what was considered to be normal non-verbal processing. In one additional case (Praamstra *et al.*, 1991), although non-verbal perception was claimed to be only mildly impaired, the evidence seems anecdotal and not too well documented. In all other 57 cases, deficits were in reality not strictly limited to verbal material (Goldstein, 1974; Denes and Semenza, 1975; Goldstein *et al.*, 1975; Saffran *et al.*, 1976; Auerbach *et al.*, 1982; Coslett *et al.*, 1984; Buchman *et al.*, 1986; Tanaka *et al.*, 1987). In all of these cases, although the main presenting manifestation may have been the verbal loss, some aspect of music and/or environmental sound perception was also documented as having been impaired.

Thus, it may be that 'apparent' pure word impairment is usually an epiphenomenon arising from the limited environmental stimuli hitherto available for testing. The picture that emerges from the present study calls into question the material specificity of the impairment which seemed evident on initial clinical testing. Where does this leave the question of material specificity in auditory comprehension? Certainly, we cannot ignore the five pure word deafness exception cases, nor can we ignore a small number of other case studies demonstrating the converse—impairment of music and environmental sound processing with sparing of speech (Spreen *et al.*, 1965). Peretz *et al.* (1994) even documented a functional impairment limited to perception of music and prosody. Our particular case, however, provides no evidence that speech, music and environmental sounds undergo analyses which are distinct, mutually exclusive, modular processes. It is not perfectly clear, however, from these data alone, whether this present lack of distinctiveness is due to (a) our more wide ranging and sensitive stimuli, (b) the fact that the lesion has progressed locally thus giving rise to more global symptoms, or (c) the fact that the lesion has progressed downward and thus gives rise to symptoms from different domains. These issues will be addressed more fully in a forthcoming article.

Regarding the underlying etiology in this case, the recent audiological investigations have raised important questions. Our initial impression was that the pathological locus in this individual originated at the temporal cortical level. Subsequently, we surmised, there was evidence of subtle spread which extended into the brainstem, cerebellar white matter, and peripherally to involve the vestibular and auditory VIIIth nerve components. The cerebellar auditory area is innervated from cells in different regions of the cochlear nuclear complex (Huang *et al.*, 1982; Brodal and Brodal, 1985; Nieuwenhuys *et al.*, 1988). There are extensive efferent fibers which descend from the auditory cortex, via the olivocochlear bundle and vestibular nerve, terminating in the organ of Corti (Warr, 1975; Adams, 1983). This efferent system serves to modulate sensory outflow from the cochlea (Nieuwenhuys *et al.*, 1988). Thus, this case could be argued to represent a 'system degeneration', with the lesions preferentially affecting the neural elements at various levels of the auditory processing system. If this formulation is correct, then these later changes in the brainstem and peripheral nerve reflect trans-synaptic retrograde as well as anterograde degeneration secondary to the initial cortical lesion (Mesulam, personal communication). We know of no other similar cases in the literature to date.

In terms of threshold losses, a typical threshold loss due to a cochlear lesion usually gives rise to an ascending air and bone conduction hearing level, indicating better performance at the higher frequencies; in sensorineural hearing loss, air and bone conduction are both reduced. The audiometric pattern most typical is that of a high-frequency hearing loss (downward sloping audiogram).

Thus, in terms of her performance on speech processing, as argued earlier, AL fits more neatly into a central type of loss, whereas in terms of threshold losses, she fits more into a cochlear—not VIIIth nerve—type of loss (Katz, 1985; Sataloff and Sataloff, 1993; Becker *et al.*, 1994; Adams *et al.*, 1997).

Regarding the SPECT results, the scan showed a focal perfusion defect seen in the left temporal lobe. This area seems localized to Brodmann areas 38 or also 20—and not the classical areas 22, 41, and 42. This might indicate focal brain degeneration or be the result of neural degeneration elsewhere (auditory cortex?) with inputs into the temporal pole. This area has recently been argued to have a role in language processing [recent functional neuroimaging studies on language processing indicate that quite anterior portions of the temporal lobe light up: in Price (1997) areas 38 and 20 as defined by Gilman and Newman (1992) activate during semantic decisions; in Mummery *et al.* (1999) areas 22 and 38 as defined by Gilman and Newman (1992) activate during speech perception tasks]. However, SPECT can also be unreliable and show perfusion defects even in normals, so interpretation must be guarded (Gilman and Newman, 1996; Price, 1997; Mummery *et al.*, 1999).

Recent audiological reports suggest an altogether different possibility, however—that AL presents as an unusual form of auditory neuropathy. This disorder, reported only within the last decade, is characterized by evidence of normal outer hair cell function (normal otoacoustic emissions or cochlear microphonics), in the presence of abnormal function in the auditory nerve (absent brainstem responses or abnormal electrocochleography). Cases are now being reported with onset any time from infancy to young adulthood (Starr *et al.*, 1996), and many (but by no means all) of these patients develop a more generalized neuropathy over time (Kaga *et al.*, 1996). One characteristic of the disorder is that language comprehension is more severely affected than non-language comprehension—exactly the pattern encountered in AL. Another feature is that the pure tone audiogram can be 'almost normal' or even normal at the beginning of the disorder, even when speech comprehension is impaired (Starr *et al.*, 1996; Kaga *et al.*, 1996). Recent work has indicated that auditory brainstem responses (ABR) can be falsely labeled as normal when cochlear microphonics are prominent, and this can result particularly in an apparently normal first wave form (Berlin *et al.*, 1993, 1994). Thus, the initial report of a normal ABR may have been misleading (and the stapedius reflex was not actually normal initially). AL's ABRs were, however, abnormal by 1984.

According to this latter formulation, the auditory neuropathy disorder was subtle initially, but has progressed and possibly may have spread centrally. This has also occurred in other cases of auditory neuropathy reported (Horoupian, 1989).

AL has persisted as an important 'outlier' or boundary case in the literature on primary progressive aphasia (PPA) (see Black, 1996). Her age of onset, symptomatology, and subsequent course have deviated substantially from the prototype of PPA, thus causing us to doubt that she be considered as a true case of PPA. Rather, we would suggest that she be reconsidered as an example of some different clinical entity. Either she represents an unusual centrifugal auditory system degeneration or an unusual case of auditory neuropathy. Either way, it should be noted that most pure word deafness cases in the literature share many features with AL, and if her initial clinical picture reflected early auditory neuropathy, then interpretation of evidence for central versus peripheral auditory system deficits will need to be revised.

The issue of modularity deserves some comments. Early on, only AL's language comprehension was selectively affected—not music or environmental sounds—and this corresponded with the cortical abnormality only. Only later as the lesion seemed to progress downward were music and environmental sounds more affected. This provides evidence for modularity at some high level. Even today, to the extent that environmental sounds are relatively preserved, this constitutes evidence for some modularity.

We suggest that pure word deafness, auditory agnosia, cortical deafness, and auditory neuropathy are in fact overlapping. Part of the reason for this is that initial cortical lesions give rise to retrograde degeneration, whereas peripheral lesions give rise to cortical dysfunction. AL's symptoms began (and have remained) with the presence of cortical dysfunction. There was a possibility that peripheral findings were being masked early on (a rather weak point). Clear evidence for peripheral dysfunction appeared only later. It does not seem reasonable to dismiss the persistent, early cortical findings (favoring a cortical origin). Although auditory neuropathy does cause disability in higher functions, these disturbances are qualitatively different from disturbances both peripherally and centrally.

This case underlines the necessity of detailed evaluation of patients with 'classical syndromes'. This also entails more adequate controls with lesions at different points of the auditory system.

The detailed neuropsychological and cognitive evaluation in fact suggested that the deficit was not as specific as initially believed, but extended to a subset of environmental sounds. Detailed electrophysiology suggested that a subtle auditory neuropathy is now present, and may have been present even initially. Finally, this case has been followed longitudinally for many years. It is only with the passage of time and the development of new technologies (e.g. to detect otoacoustic emissions and cochlear microphonics) that the diagnosis has been clarified (although it remains obscure even yet!). It is intriguing that the final diagnosis may involve an entity (auditory neuropathy) that had not even been discovered when this woman presented.

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A case study of pure word deafness: modularity in auditory processing?

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Abstract

AL, a woman with an acquired disturbance of auditory processing beginning in the second decade, was originally diagnosed as having pure word deafness. Recent analysis with a wide range of stimuli suggests that her comprehension deficit also extends to a subset of musical and non-verbal environmental sounds. The perceptual demands of the different auditory stimuli appear to account for part of the apparent material specificity. Additionally, over the years, the presumed temporal lobe cortical pathology has been supplemented by a mild to moderate, peripheral low-frequency hearing loss and evidence of dysfunction in lower level auditory processing pathways. The current peripheral dysfunction closely resembles cases recently labeled as auditory neuropathy. The diagnosis of pure word deafness should not be based on a limited set of auditory stimuli; additionally, a careful assessment using modern audiological techniques should be performed to evaluate peripheral auditory functions.

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Primary diagnosis of interest

Progressive pure word deafness

Author's designation of case

AL

Key theoretical issue

- Modularity in auditory processing: are speech, music and environmental sounds processed by distinct, mutually exclusive processes/structures

Key words: pure word deafness; auditory neuropathy; modularity of auditory processing; processing of speech, music and environmental sounds

Scan, EEG and related measures

1978–1984: Computed tomography scan, audiograms, brainstem auditory evoked response
1992–1998: Audiological evaluation: audiometry, pitch discrimination and pattern testing, gap detection, tone duration and decay, lip reading, auditory brainstem responses, auditory cortical evoked responses, impedance testing. Otolaryngological evaluation: electronystagmography, electrocochleography, otoacoustic emissions. Full brain magnetic resonance imaging, Hm-PAO SPECT, nerve conduction studies, VDRL

Standardized assessment

Neuropsychological assessment battery: Wechsler Memory Scale, Wide Range Achievement Test, word fluency, Trail making A and B, Multidimensional Aptitude Battery

Other assessment

Bub and Caplan Psycholinguistic Assessment of Language, voice gender recognition, recognition of emotions (intonation), sound identification and sounds to picture matching, instrument identification and instrument recognition, song recognition test

Lesion location

- A slowly evolving downwardly progressing lesion, over the course of 18 years
- 1978: Slightly more atrophy in the left hemisphere; increase in ventricular size
- 1984: Partially absent bilateral brainstem function

- 1993, 1994: Focal perfusion defect in left anterior temporal pole; totally absent bilateral brainstem function; near absent bilateral peripheral vestibular function (probably of slowly progressive origin); small oval white matter hyperintensities in left and right cerebellum
- 1995: Auditory nerve damage outside brainstem

Lesion type

Either the subject presents an unusual centrifugal auditory system degeneration (with trans-synaptic retrograde as well as anterograde degeneration secondary to an initial cortical lesion) or an unusual case of auditory neuropathy, subtle initially, which progressed and possibly spread centrally

Language

English