

PURE WORD DEAFNESS FOLLOWING BILATERAL LESIONS

A PSYCHOPHYSICAL ANALYSIS

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SUMMARY

A detailed clinical and psychophysical study of a woman who had developed pure word deafness associated with amusia after bilateral temporoparietal destructions is reported. The patient had a defect in temporal resolution encompassing auditory, visual and somatosensory modalities, but the clinical defect was limited to the auditory sphere. Auditory comprehension did not improve even if she was spoken to slowly, although marked improvement of temporal resolution was observed as the duration of a nonlinguistic sound was extended. Also, she exhibited a supramodal defect in the perception and reproduction of rhythm, which was rate-dependent. These and other findings led to the following conclusions: (1) the auditory modality is much more dependent on temporal resolution than other sensory modalities; (2) for a full understanding of the mechanism of pure word deafness, not only the defect of temporal resolution but also many other factors, for example, defective discrimination of loudness, pitch or tone duration have to be taken into consideration; and (3) rhythm sense strongly depends on a supramodal capacity of temporal resolution.

INTRODUCTION

Pure word deafness is defined as the inability to comprehend and discriminate spoken language while spontaneous speech, reading, reading comprehension and writing are preserved, and comprehension of nonverbal sounds is relatively spared. Recent experimental studies have implicated defects in auditory processing (Jerger *et al.*, 1969; Kanshepolksy *et al.*, 1973; Albert and Bear, 1974; Auerbach *et al.*, 1982), temporal resolution (Albert and Bear, 1974), phonemic discrimination (Chocholle *et al.*, 1975; Denes and Semenza, 1975; Saffran *et al.*, 1976; von Stockert, 1982) or prephonemic discrimination (Auerbach *et al.*, 1982).

We recently had the opportunity of studying a patient with pure word deafness associated with amusia and mild auditory agnosia for environmental sounds. We have analysed temporoparietal lobe functions by several psychophysical and musical studies on this patient. The relationship between temporal resolution and clinical manifestations is discussed.

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CASE REPORT

The patient, a 26-year-old right-handed Japanese woman, began to take lessons in playing the organ at the age of 6 years and the piano at the age of 15. She graduated from a junior college at the age of 20. Her past medical history was unremarkable, and she denied taking contraceptive pills.

On January 9, 1982, while watching television, she suddenly developed a right hemiparesis and her speech became garbled and incomprehensible. She was admitted to a local hospital, where initially she could produce only a few words. The number of words later gradually increased. Her spontaneous speech contained a moderate number of verbal and literal paraphasias. Verbal comprehension, repetition and reading aloud were moderately impaired; however, she was able to sing a nursery rhyme. The general impression of the admitting physician was a predominantly posterior type of aphasia. Computed tomography (CT) revealed a low-density area in the left temporoparietal region. Left carotid angiograms showed no abnormal findings. The CSF was normal. She was thought to have had a cerebral infarct and supportive treatment was given. Her hemiparesis disappeared within a few days and her aphasia improved gradually.

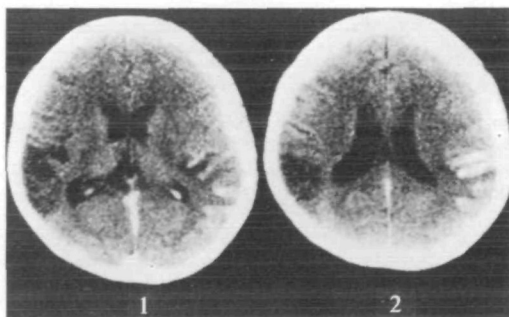


FIG. 1. CT scan of the brain after the second stroke demonstrating an enhancing lesion in the right posterior temporoparietal region and an older non-enhancing lesion in the left posterior temporoparietal region including a part of Wernicke's area.

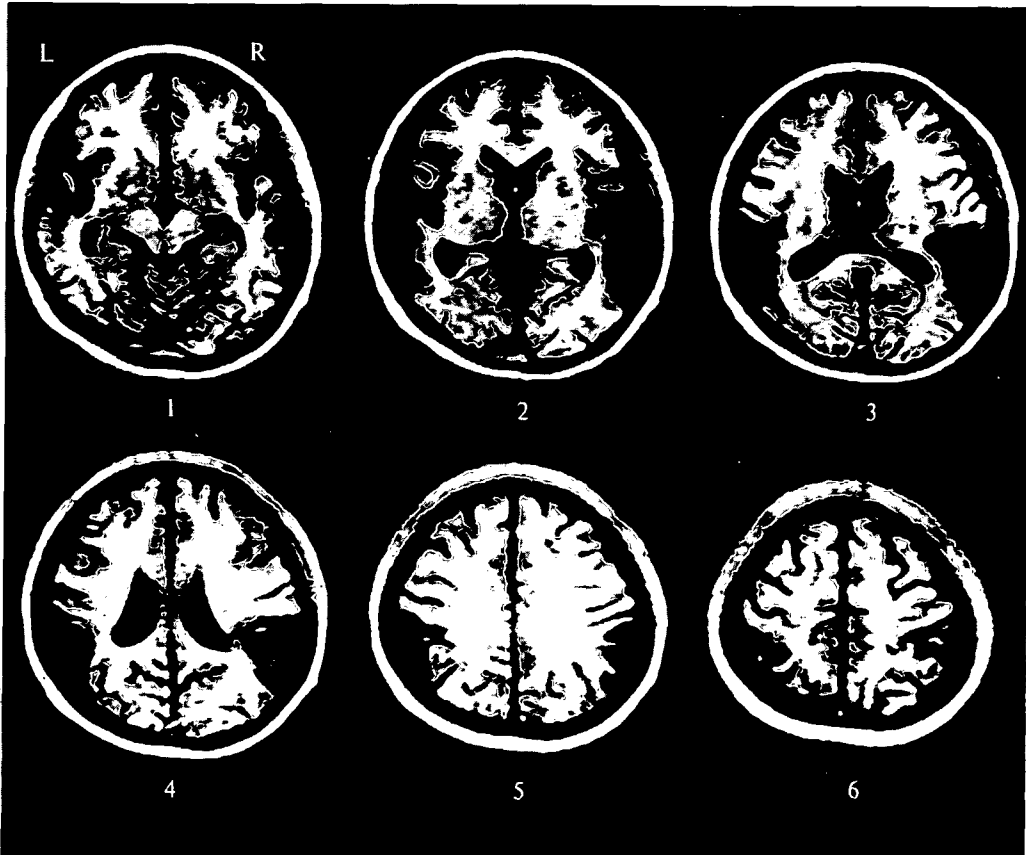
On January 5, 1985, she suddenly had a generalized convulsion. She was at once admitted to another hospital, where she was drowsy for ten days. Thereafter, she became unable to understand spoken words, although she was able to follow written commands. The remainder of the neurological examination was normal. A CT scan demonstrated an enhancing lesion in the right posterior temporoparietal region and an older nonenhancing lesion in the left posterior temporoparietal region including a part of Wernicke's area (fig. 1). Bilateral carotid and vertebral arteriograms were normal. An EEG showed persistent theta activity in the right parieto-occipital region. She was transferred to the Neurology Service of Hyogo Brain and Heart Center at Himeji on May 1, 1985, for further examination.

On admission she was attentive, cooperative and orientated in time and place with a good insight into her illness. General physical examination revealed a normal heart rate and rhythm. Blood pressure was 120/80 mmHg in both arms. There were no carotid bruits. Remote and recent memory was intact. Calculations were done moderately well. She could identify most nonverbal sounds such as a telephone ringing or water running from a tap, but could not understand spoken words, which sounded like an echo. Visual fields were normal to confrontation, and the cranial nerves intact except for auditory function, which will be described in detail below. Findings of sensory and cerebellar tests were unremarkable. Motor examination revealed normal muscle tone without weakness. The tendon reflexes were normal bilaterally without pathological reflexes. Right-left orientation, two-point discrimination, skin graphaesthesia, point localization, finger naming, optokinetic nystagmus, visually-guided reaching, stereognosia and spatial ability were all intact. Double simultaneous stimulation (auditory, tactile and visual) was correctly perceived on both sides.

Laboratory findings were all normal, including immunology, coagulation and cardiac surveys

(echocardiography and ambulatory Holter electrocardiography). An EEG, performed on May 9, 1985, was within normal limits.

Magnetic resonance imaging (MRI) with 10 mm slices, performed on May 27, 1985, revealed that the left lesion was situated in the middle and posterior portion of the superior temporal gyrus involving most of Heschl's gyrus and the geniculotemporal auditory radiation. There was another lesion in the left parietal lobe involving the supramarginal and angular gyri. The right lesion, more extensive than the left, was in the middle and posterior portion of the superior temporal gyrus and in the parietal lobe involving the supramarginal and angular gyri. Involvement of Heschl's gyrus and the auditory radiation was also more extensive on the right (figs 2, 3, 4).



FIGS 2, 3, 4. MRI performed on May 27, 1985 showing bilateral posterior temporoparietal lesions including the primary auditory area and the auditory radiation on each side, greater in the right hemisphere. Figs 2, 3, and 4 show horizontal, coronal and sagittal sections, respectively (in fig. 4, the upper figures show the left hemisphere and the lower show the right).

RESULTS

Neuropsychological Examination

Abnormally loud and dysprosodic spontaneous speech was fluent with very rare phonemic paraphasic errors. Visual confrontation naming, reading aloud, reading

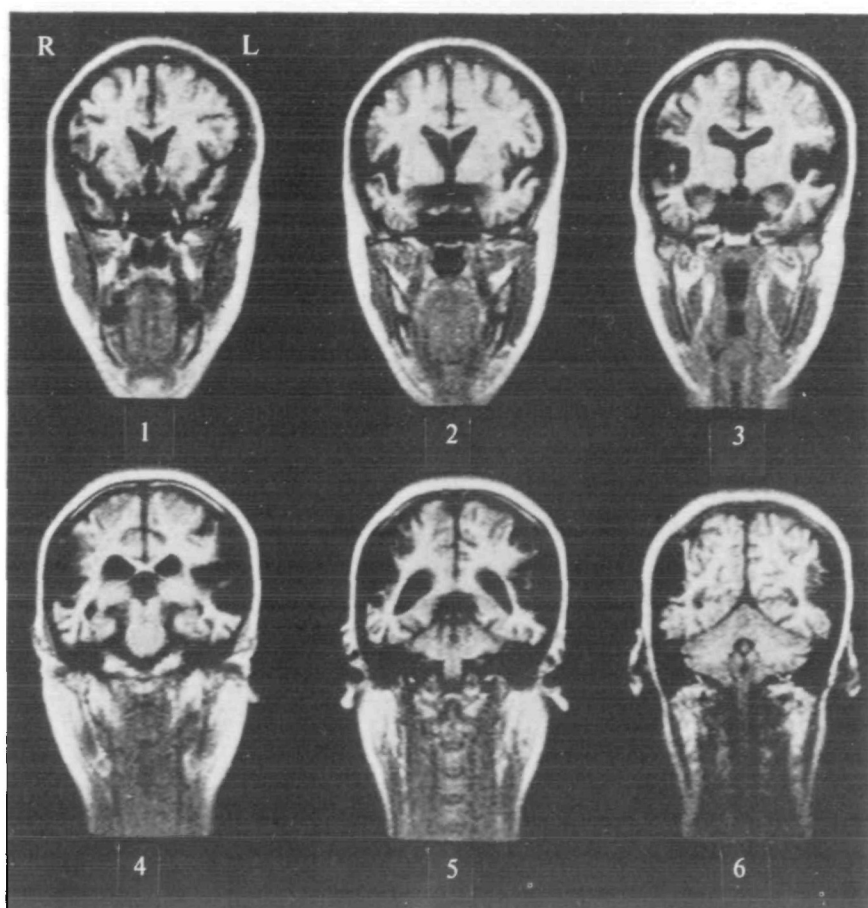


FIG. 3

comprehension and spontaneous writing were normally performed. She could correctly execute four-step written commands. Her score of the Token Test presented as written commands, performed on May 10, 1985, was 92% correct. Verbal comprehension was markedly impaired and she was unable to identify even a single word. Slowing the rate of speech by increasing the interval between syllables, or lip-reading, did not improve her comprehension. Repetition of a single word and a phrase was severely impaired. The results of the standard language test of aphasia (SLTA) (Takeda, 1977), performed on May 10, 1985, are summarized in Table 1. Praxis was normal. Copying of a cube was done well. Colour recognition and visual object recognition were normal. Her intelligence level according to WAIS, performed on May 14, 1985, was as follows: Verbal IQ = 84, Performance IQ = 91 and Whole Scale IQ = 86.

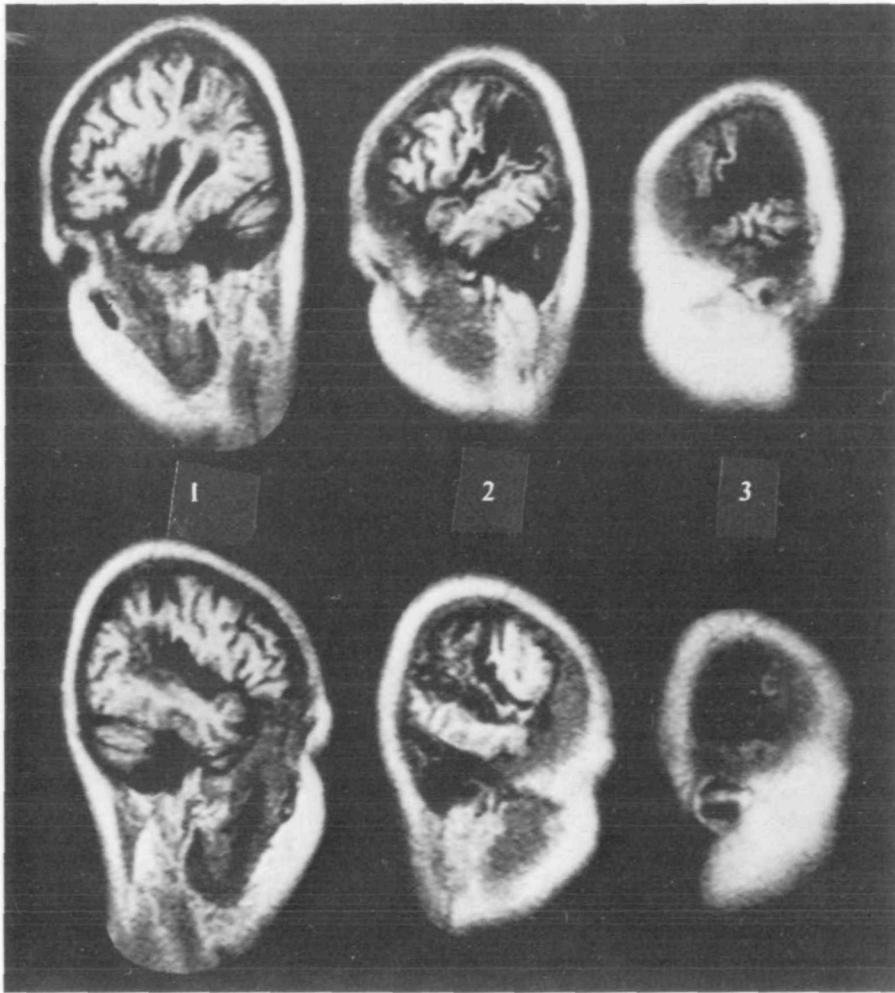


FIG. 4

Audiological Examination (May 16, 1985)

The external auditory meatus and ear drums were normal. Impedance audiometry revealed normal tympanic membrane mobility bilaterally. Both ipsilateral and contralateral acoustic reflexes were elicited at normal reflex thresholds. Standard pure tone threshold audiometry showed a mild bilateral sensorineural loss at the lower and higher frequencies. Thresholds for the higher frequencies were more elevated bilaterally, especially on the left (fig. 5). In the range of speech frequencies (500, 1000, 2000 Hz), an average of 15 dB hearing loss was noted in the right ear and 8 dB loss in the left ear. Speech reception threshold could not be determined because the patient was unable to recognize and repeat even a single word. However, she

reported that she could hear some sound. The discrimination ability was zero, although the hearing for pure tones was near-normal at the speech frequencies.

TABLE 1. RESULTS OF THE STANDARD LANGUAGE TEST OF APHASIA (SLTA)

<i>Subtest</i>	<i>Score</i>
Auditory comprehension (words, sentences, commands)	0 correct
Speech	
Naming	100% correct
Explanation of action in a picture	100% correct
Explanation of cartoon	100% correct
Generation of word list	18 animal names/min
Repetition (words, sentences)	0 correct
Reading aloud (Kanji, Kana, sentences)	100% correct
Reading comprehension (Kanji, Kana, sentences, written commands)	100% correct
Writing	
Spontaneous writing (Kanji, Kana, explanation of cartoon)	100% correct
Dictation (Kanji, Kana, sentences)	0 correct
Calculation	100% correct

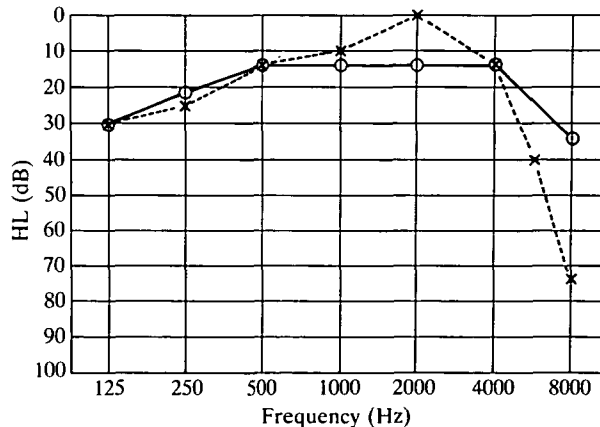


FIG. 5. Pure tone threshold audiometry. × = left ear; ○ = right ear; HL = hearing level.

Sound localization. The patient was blindfolded and then exposed to a canon sound of about 3 s duration through a tape recorder which was placed at 50 cm from her. The sound was randomly delivered from six directions (right, left, front, behind,

above and below). Ten trials were performed in each test. She was correct in 90% of the trials from the right and in 80% of the trials from the left. Accuracy in localizing the sounds from other directions was from zero to 40%. Normals reached 100% correct detection of the sound from all directions. These findings indicate that the patient has a moderate impairment of sound localization.

Nonverbal sound discrimination. The test consists of 25 familiar sounds presented through a tape recorder, including sound-producing objects, animals or events (e.g., a miaowing cat, an ambulance siren). She correctly named 19 sounds.

Distinguishing categories of sounds. The patient was told that she would hear through a tape recorder a sound of one of the following three categories; music (vocal or instrumental), noise and language (English, French, German, Chinese or Japanese), and was asked to categorize the sound. She performed this categorization task almost perfectly; however, she had some difficulty in identifying the gender of a recorded voice and types of musical instruments. Also, she was totally unable to discriminate the languages.

Linguistic Studies

These and the following other special investigations were performed over an eight-week period after admission. All the linguistic tasks were enunciated by the same native Japanese male according to the method of Auerbach *et al.* (1982).

Vowel identification. The patient was asked to identify long vowels (about 1 s duration), a, e, i, o, u; 150 randomly-ordered vowels were presented verbally. Special care was taken to avoid cues from lip-reading or facial gestures. Her average correct response was 47%.

Stop consonant-vowel discrimination. All items were a combination of a consonant and a vowel /a/. The initial stop consonants differed according to a place of articulation (labial /b, p/, alveolar /d, t/, or velar /g, k/) or voicing (voiced /b, d, g/, voiceless /p, t, k/).

1. Binary decision with single distinctive feature. The patient was asked to point out one of two possible choices given in written form in each test. Nine tests of 100 items each were presented. The results are given in Table 2. The average correct response across all the nine tests was 49%.

2. Binary decision with two distinctive features. These tests of 100 items each

TABLE 2. RESULTS FOR STOP CONSONANT-VOWEL DISCRIMINATION: BINARY DECISION WITH SINGLE DISTINCTIVE FEATURES

	<i>Condition</i>			<i>Nature of distinction</i>
Distinction	/ba/-/pa/	/da/-/ta/	/ga/-/ka/	Voiced/voiceless
% correct	52	50	50	
Distinction	/ka/-/ta/	/pa/-/ta/	/pa/-/ka/	Place of articulation for voiceless stops
% correct	47	52	54	
Distinction	/da/-/ga/	/da/-/ba/	/ba/-/ga/	Place of articulation for voiced stops
% correct	45	47	44	

were presented in the same way as the tests above. The only difference was that the forced choice was between two phonemes that differed by two features instead of one. The following results were obtained: /ka/-/da/ distinction, 50% correct; /ga/-/pa/ distinction 49% correct; /ba/-/ta/ distinction 53% correct. The average correct response across all the 3 tests was 51%.

These linguistic studies indicate that she was able to identify long vowels at a better than chance level but failed in stop consonant-vowel discrimination, with no particular pattern of error, even if the number of distinguishing features increased. From these observations, the patient's 'pure word deafness' seems largely or entirely to be caused by a phonetic decoding defect.

Tests of Musical Capacity

Expressive musical ability. The patient was asked to sing familiar nursery rhymes with the help of written lyrics. Both pitches and rhythms were severely impaired.

Instrumental musical ability. The patient was able to play familiar nursery rhymes on an electric organ with the help of music, but often hesitated during the performance.

Receptive musical ability. 1. *Recognition of melodies.* Free field presentation of 18 nursery rhymes played on an electric organ yielded the following results: verbal identification, 28% correct; a five-way multiple-choice of corresponding pictures, 89% correct.

These results suggest that recognition of melodies was relatively preserved. The poor verbal identification would reflect a defect in retrieving the verbal components.

2. *Tonal pattern discrimination.* The patient was asked to determine whether a sequence of either two or four tones played on an electric organ was ascending or descending. The duration of each tone was about 1 s; 50 items each were administered. The results are shown in Table 3. The patient thus displayed a profound impairment of perception of tonal pattern. In a sequence of four tones, only one octave interval was identified correctly above chance level.

Her failure to distinguish tonal pattern agrees well with the results of experiments

TABLE 3. RESULTS FOR TONAL PATTERN DISCRIMINATION

<i>Two tones</i>		<i>Four tones</i>	
Interval*	% correct	Interval*	% correct
1/2 tone	50	1/2 tone	48
Whole	54	Whole	40
Third	34	Third	56
Fifth	52	Octave	76
Sixth	48		
Octave	46		

* Interval between neighbouring tones.

by Diamond and Neff (1957) and Neff *et al.* (1975) who showed that bilateral ablation of auditory cortex in the cat or monkey destroys auditory pattern discrimination.

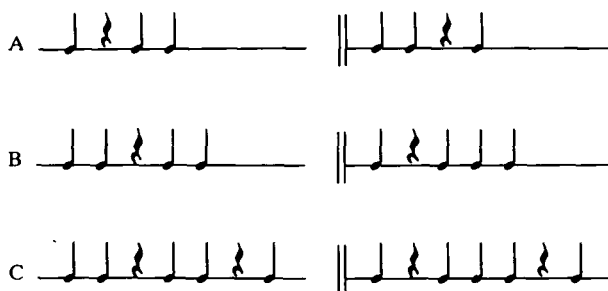


FIG. 6. Examples of pairs of rhythm patterns. A, B and C show pairs of 3, 4 and 5 beats, respectively.

3. *Rhythm discrimination.* Following the method of Mavlov (1980), rhythm perception was investigated using three modalities: auditory (electric organ), visual (penlight flashes) and tactile (taps on the patient's palm). Simple rhythm patterns consisting of 2 to 5 stimuli variously interspaced by 1 or 2 empty stimuli were used (fig. 6). Fifty pairs were presented, and the patient was asked to determine whether two rhythm patterns were the same or different. In all three modalities, they were presented at a 'fast' (200 ms interval) or a 'slow' (1000 ms interval) rate in a random fashion.

Disturbances were seen across all the three modalities at the fast rate but, at the slow rate, the response improved significantly in all modalities; by audition from 52 to 88% correct, by vision from 62 to 94% and by touch from 62 to 88%.

4. *Rhythm reproduction.* For rhythm reproduction, 10 simple rhythm patterns composed of 2 to 5 elements were presented through three modalities in the same way as the previous test. The patient was asked to reproduce the rhythm orally. Each of the above patterns was presented three times for each modality.

Similar results to the previous test were obtained; by audition from 33 to 83% correct, by vision from 57 to 90% and by touch from 40 to 87%.

Neurophysiological Investigation

An MEB-5100 (Nihon Koden, Tokyo) was used in recording auditory evoked potentials (brainstem and late latency auditory evoked potentials). These investigations were performed on May 19, 1985.

Brainstem auditory evoked potentials (BAEPs). Clicks (0.1 ms) of 90 dB Hearing Level (HL) were presented at 10 Hz monaurally. Recordings were derived from Cz to the ear lobe of the stimulated side. The contralateral ear was masked for all trials. Each trial represented 2048 responses. Responses obtained in each ear are presented in fig. 7. All BAEP components (waves I–VI) were recorded with normal amplitude and latency, indicating integrity of the cochlear mechanism and brainstem auditory pathways.

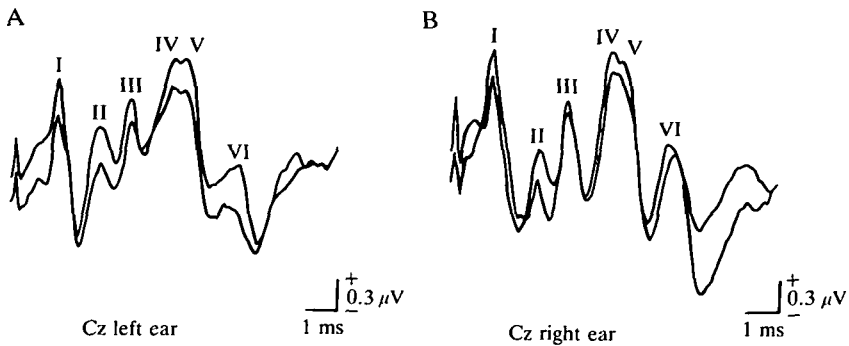


FIG. 7. Brainstem auditory evoked potentials. Recordings are from Cz to the left (A) and to the right (B) ear, respectively; 2048 signals are averaged. Waves I-VI are intact in both tracings.

Late latency auditory evoked potentials (LLAEPs). The click was given to both ears randomly one per 2 s, 100 dB HL. Recordings were derived from Nz to T3 and T4. The low filters were set at 0.5 Hz, and the high filters at 30 Hz. Each trial represented 128 responses in random order. P₁, N₁, P₂ and N₂ were easily identified with latencies at about 55, 140, 185 and 225 ms, respectively. The peak latencies were all within normal limits except for N₁, which was slightly prolonged. Also, overall late components showed low amplitudes (fig. 8).

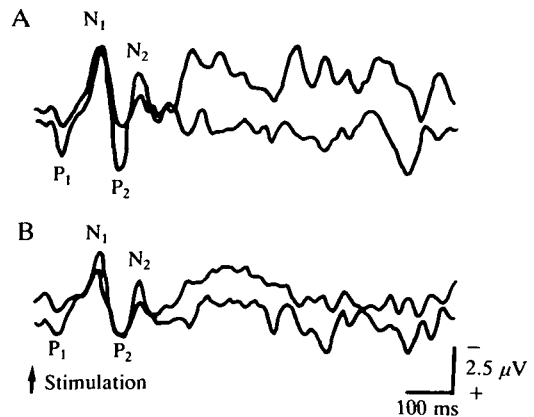


FIG. 8. Late latency auditory evoked potentials. Recordings are from Nz to T3 (A) and to T4 (B), respectively; 128 signals are averaged randomly. Filters are set at 0.5 Hz and 30 Hz.

Neuroacoustic Studies

Threshold-duration function. The patient was asked to tell only whether she heard a sound or not as the duration of the sound was varied over the range from 5 to 1000 ms. Tone bursts generated by an electric stimulator (SEN-7103, Nihon Koden, Tokyo) coupled with an acoustic stimulator (SSS-3100, Nihon Koden) were delivered monaurally. Fig. 9 shows the relationship between duration and intensity of the

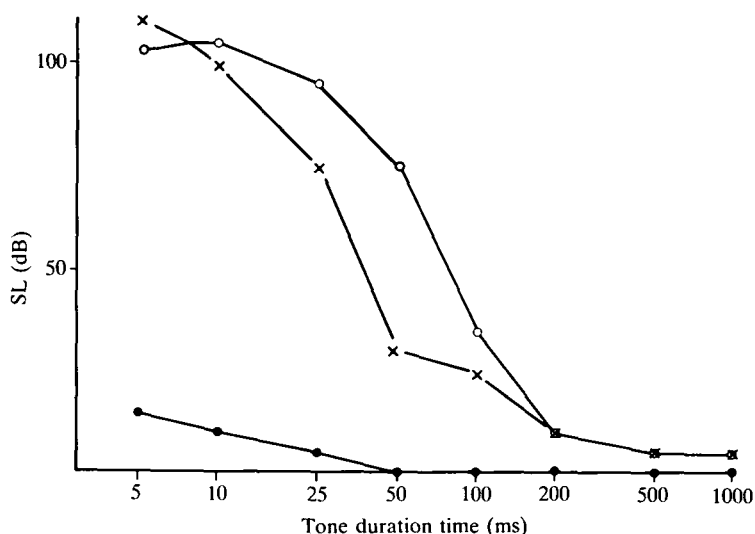


FIG. 9. Threshold-duration at 1000 Hz. Note that the sound intensity decreases as the duration of sounds increases. SL = sensation level. o = patient (L ear); x = patient (R ear); ● = normal subject (both ears).

stimuli to obtain threshold response for both right and left ears at 1000 Hz. As the duration became short, sound intensity had to be raised considerably (100–105 dB Sensation Level, SL) to maintain the threshold response in our patient. In normal subjects the effect was less than 15 dB SL.

Loudness discrimination. Loudness discrimination was tested by the quantal psychophysical method (Jerger *et al.*, 1969). Short-intensity increments were added to a steady-state, pure-tone signal at 5 s intervals. The patient was instructed to indicate verbally whenever she detected a change in the loudness of the steady tone. HL of the steady-state signal was based at 80 dB for 1000 Hz. The patient reached 70% correct detection in the right ear and 50% in the left ear for intensity increment of 3 dB. A 100% correct detection was not achieved until the increment size was 6 dB in both ears. Normal subjects in our laboratory reached 100% correct detection in each ear for an intensity increment of only 2 dB.

Temporal auditory acuity. To examine the temporal resolution of the auditory system, clicks of 0.1 ms duration generated by an MEB-5100 coupled with an SEN-7103 electric stimulator were delivered binaurally through earphones at 100 dB HL. Two tests of click fusion and click counting were performed following the method of Albert and Bear (1974).

1. *Click fusion test.* The interval between two brief binaural pulses was varied, the patient being asked to report whether she heard one or two clicks. Normals can distinguish two clicks at 1 to 3 ms separations (Hirsch, 1975). Our patient, by the method of ascending and descending limits, fused at intervals of 300 ms.

2. *Click counting test.* The patient was asked to count the number of clicks given

in 1 s. The counting in normals ranges from 9 to 11 clicks/s (Auerbach *et al.*, 1982). Our patient was inaccurate for rates greater than 2/s.

The results of both click fusion and click counting tests suggest that our patient had a severe defect in temporal auditory resolution.

3. *Other studies using click sounds.* The perceptual disorder of click sounds was further explored by two methods: (a) threshold-number of clicks and (b) threshold-interval of clicks.

(a) *Threshold-number of clicks.* The patient was asked to tell only whether she heard clicks or not as the number of clicks was varied over a range from 1 to 128. The interval between clicks was fixed at 10 ms. Threshold was estimated by the method of ascending and descending limits.

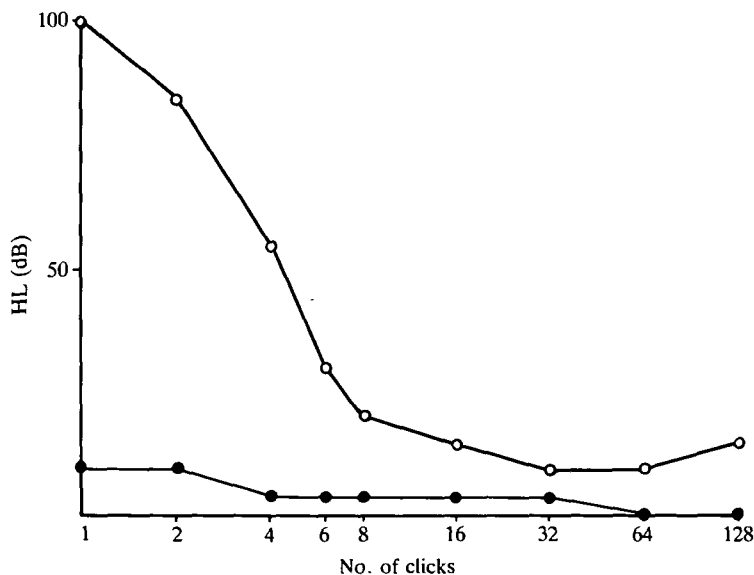


FIG. 10. Threshold-number of clicks. Interval of clicks was fixed at 10 ms. Note that the sound intensity decreases as the number of clicks increases. HL = hearing level. ○ = patient; ● = normal subject.

Our patient could detect a single click only at 100 dB HL. However, when the number of clicks was increased, the threshold decreased progressively. In normal subjects in our laboratory, the effect was less than 10 dB HL (fig. 10). This phenomenon seems to be due to the effect of temporal summation, which occurs at a cellular level when two or more subthreshold stimuli are applied in close succession (Brinley, 1980).

(b) *Threshold-interval of clicks.* The threshold with varying intervals of clicks was first examined. The patient was asked to tell only whether she heard clicks or not as the interval of clicks was varied over a range from 1 to 500 ms. The number of clicks was fixed at 2, 4 and 8, respectively. Threshold was estimated in the same way

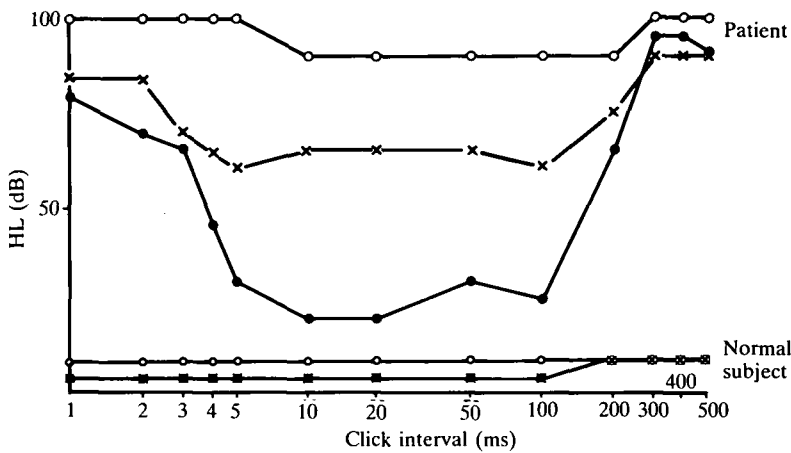


FIG. 11. Threshold with varying intervals of clicks. The number of clicks was fixed at 2, 4 and 8, respectively. As the interval between clicks became shorter from 300 to 100 ms, the threshold decreased but increased again when the intervals of clicks were less than 5 ms. HL = hearing level. \circ = 2 clicks; \times = 4 clicks; \bullet = 8 clicks.

as the preceding test. As the intervals between clicks were shortened from 300 to 100 ms, the threshold decreased. It remained constant with intervals between 100 and 10 ms and then increased again as the interval shortened. In normals in our laboratory, the effect was less than 10 dB HL (fig. 11).

These results can possibly be interpreted as follows. The lower threshold at click intervals between 300 and 10 ms appears to be due to the effect of temporal summation and the higher threshold at intervals of less than 5 ms may be related to the influence of the relative refractory period. During the initial period of an action potential, the membrane cannot respond to any stimuli, regardless of their intensity; this interval, the absolute refractory period, is approximately 0.4 to 1 ms in mammalian nerve fibres. Following the absolute refractory period, the membrane becomes responsive to stimuli of greater intensity than the normal threshold, that is, the threshold for excitation has been increased. The threshold then declines towards the normal level, this interval representing the relative refractory period (about 3 to 4 ms in mammalian nerve fibres) (Mashima, 1975; Brinley, 1980). From these considerations, the higher threshold at intervals of less than 5 ms can be attributed to the influence of the relative refractory period. However, even within these periods, as the click number was increased, the threshold decreased in our patient, suggesting that temporal summation is also operative during these periods.

To elucidate the relationship between the threshold and the number and interval of the clicks, we administered a further test, the assessment of threshold with varying intervals of clicks within the fixed duration of click burst. The duration of click burst was fixed at 200 ms and within that interval, the number of clicks was increased from 2 to 201, that is, interval of clicks was shortened from 200 to 1 ms. Other methods were exactly the same as the immediately preceding test. At an interval of 200 ms,

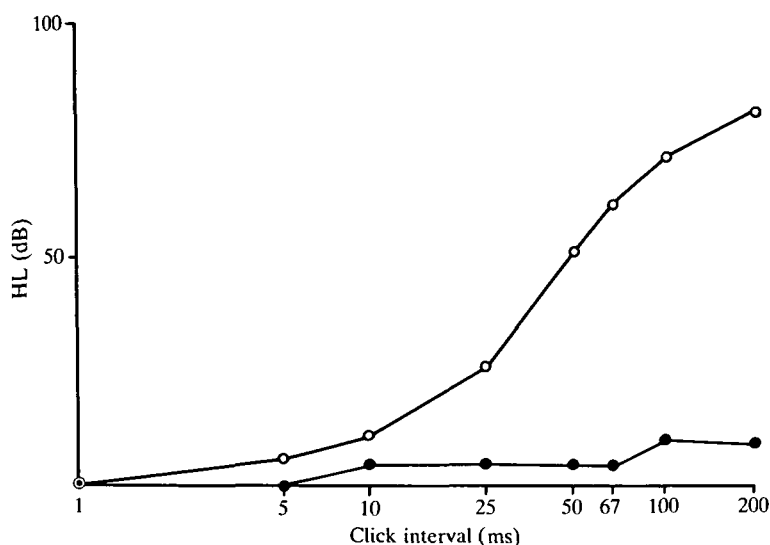


FIG. 12. Threshold with varying intervals of clicks within 200 ms intervals. Duration of click burst was fixed at 200 ms. Note that the threshold decreases as the intervals of clicks are made shorter. HL = hearing level. \circ = patient; \bullet = normal subject.

our patient could detect a sound only at 80 dB HL. As the interval between clicks was made shorter, however, the threshold for the detection of the sounds decreased progressively. The results at fast click rates agreed nicely with those of fig. 9 for a 200 ms tone burst. In normals, a similar trend was noted with a threshold of less than 10 dB HL throughout (fig. 12). This result confirms that the effect of temporal summation is present in this patient.

Discrimination Test in Various Sensory Modalities

The fact that rhythm perception in this patient improved at a slow rate in all modalities led us to further examination of the relationship between the duration and the interval of various sensory modalities. We designed the following tests for this purpose.

Auditory system. The SEN-7103 electric stimulator coupled with the SSS-3100 acoustic stimulator was used in the following examination.

1. *Duration discrimination test of tones.* A small increment in duration was added successively to a steady-state tone signal at periodic intervals monaurally. The subject was instructed to indicate verbally whenever she detected a change in the duration of the steady tone. The duration of the steady-state signal was 200 ms, and SL was based at 80 dB for 1000 Hz. In the present case, a duration increment of 380 ms was required to reach 100% correct detection in both ears. Normal subjects in our laboratory reached 100% correct detection by each ear with a duration increment of 20 to 30 ms.

2. *Tone fusion test.* Two tones with the same duration and intensity were presented monaurally, and the patient was asked to report whether she heard one or two tones as the interval between the two tones was varied. The duration of a tone was varied over the range from 20 to 500 ms. For each duration SL was set at 20 dB above the threshold which had been measured in the threshold-duration function test (fig. 8) except for the 20 ms duration, where SL was 100 dB. Normals in our laboratory could distinguish two tones with an interval greater than 1.0 ms in both ears through all durations. In our patient, the interval necessary to discriminate two tones decreased as the duration of a tone increased (fig. 13).

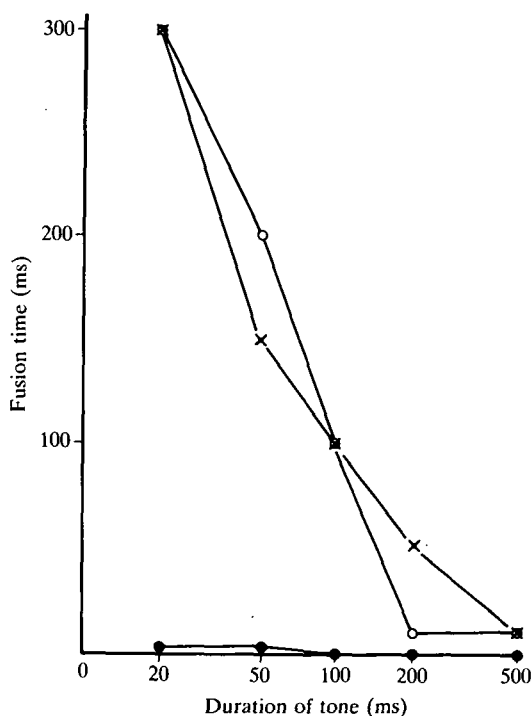


FIG. 13. Tone fusion test. Two tones of the same duration and intensity were presented. Sensation level was 20 dB above the threshold of the duration. Ability to discriminate two tones improved as the duration of a tone was prolonged. ○ = patient (L ear); × = patient (R ear); ● = normal subject (both ears).

Visual system. 1. *Duration discrimination test of lights.* The ability to detect a small change in light duration was measured with tachistoscopic presentation. Two lights, each being a quadrilateral figure of 15 × 10 cm with a green filter, were projected to the centre of a screen placed in front of the patient at an eye level of 120 cm. Two lights were presented in the same way as the duration discrimination test of tones. The duration of the steady-state signal was 200 ms. In the present case, a duration increment of 300 ms was required to reach 100% correct detection. Normals in our laboratory reached 100% correct detection for duration increment of 40 ms.

2. *Light fusion test.* Two lights of the same figure as in the test above were

presented with a tachistoscope coupled with a two-pulse generator (electric stimulator SEN-7103). Two lights of the same duration were presented and the patient was asked to report whether she saw one or two lights as the interval between two lights was varied. The duration of a light was varied over the range from 5 to 500 ms. A result quite similar to that of tone fusion test was observed, that is, the interval necessary to discriminate two lights decreased as the duration of a light increased (fig. 14). Normals in our laboratory could distinguish two lights for intervals greater than 30 ms through all durations.

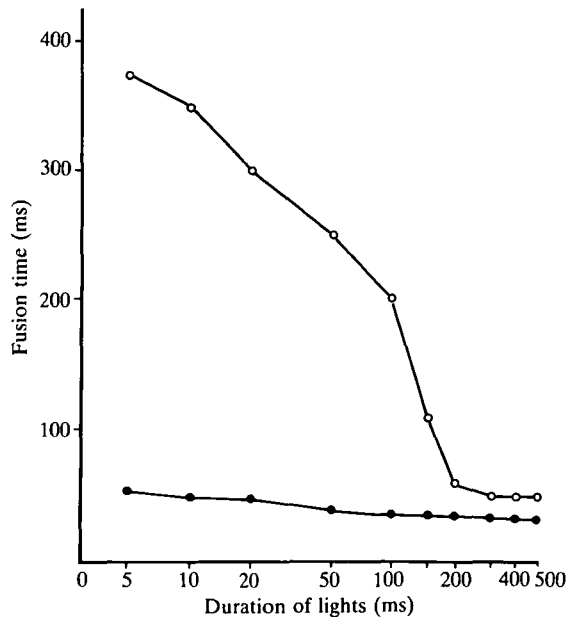


FIG. 14. Light fusion test. Two lights of the same duration were presented. A similar result to that of tone fusion test (fig. 13) was seen. ○ = patient; ● = normal subject.

Somatosensory system (pain fusion test). Two electric stimulations (0.5 ms, 100 V) were presented in each palm through the general evoked response stimulator SMP-3100 (Nihon Koden, Tokyo) coupled with a two-pulse generator. The interval between two brief pulses was varied and the patient was asked whether she felt one stimulation or two. Normals in our laboratory could distinguish two stimulations with a 20 to 30 ms interval in both palms. This patient fused these stimulations at intervals of 240 ms in the right palm and at 220 ms in the left.

Summary of Major Findings

We have presented a 26-year-old woman with pure word deafness who had cerebral infarcts bilaterally in the temporoparietal lobes. A series of studies revealed

the following features. For auditory processing, (1) pure tone threshold audiometry was within normal limits in the frequency range of speech sounds; (2) BAEPs were also within normal limits, and LLAEP components, although low in amplitude, were relatively preserved; (3) sound localization was moderately disturbed; (4) temporal auditory processing as determined by click fusion and click counting was extremely abnormal; (5) ability to discriminate loudness and duration of tones was abnormal; (6) the threshold for detection of a tone decreased as the duration of a tone was prolonged; (7) auditory temporal summation was observed when the number of clicks was increased or the interval of clicks was shortened; (8) ability to discriminate two tones improved as the duration of a tone was prolonged.

For visual processing, (1) ability to discriminate duration of two lights was abnormal; (2) temporal visual processing as determined by light fusion was extremely abnormal; (3) ability to discriminate two lights improved as the duration of light was prolonged. Temporal somatosensory processing as determined by pain fusion was extremely abnormal.

Linguistic studies with natural speech indicated that she had great difficulty in discriminating stop consonant-vowel combinations with no particular pattern of error, but identification of long vowels was better than chance level.

The results of musical ability were as follows: (1) she could sing familiar nursery rhymes, but explosively and monotonously; (2) instrumental musical ability was relatively preserved; (3) ability of melody recognition was also relatively preserved; (4) tonal pattern discrimination was extremely abnormal; (5) disturbance of rhythm perception and reproduction was seen at a fast rate in all modalities, but at a slow rate this supramodal defect improved markedly in all modalities.

Further neuropsychological testing demonstrated some difficulty in discriminating the gender of a recorded voice and types of musical instruments, and great difficulty in discriminating the language of a speaker. Identification of broad categories of sounds was relatively intact and only a mild auditory sound agnosia was demonstrated.

DISCUSSION

The clinical picture in our patient was compatible with pure word deafness. She was unable to comprehend spoken words with intact speech production, reading and writing abilities. Comprehension of nonverbal sounds was also relatively spared. Other features included expressive and receptive amusia. In the following paragraphs, neurophysiological and psychophysical aspects, musical disturbance and the relationship between temporal resolution and auditory comprehension will be discussed.

Neurophysiological and Psychophysical Aspects

Except for only mild hearing loss, our patient had no audiometric defect, and her BAEPs were normal. Hence her poor recognition of linguistic sounds represents

a disturbance of central auditory processing. Although low in amplitude, relative preservation of LLAEP components in our patient may be due to residual auditory cortical structures. In the literature which describes cortical auditory evoked potentials in pure word deafness, electrophysiological findings vary from normal to abnormal responses over the left, right or both sides (Albert and Bear, 1974; Shoumaker *et al.*, 1977; Michel *et al.*, 1980; Auerbach *et al.*, 1982). These findings suggest the variability in pathology of patients with pure word deafness.

The moderate impairment of sound localization is consistent with the findings in cats or monkeys after experimental bilateral ablations of the auditory cortex (Neff, 1968; Neff *et al.*, 1975). However, in man this seems to be a variable concomitant (Jerger *et al.*, 1972). As indicated by Jerger *et al.* (1972), this impairment seems to be related to an interaural imbalance in the relation between signal duration and loudness, which was also present in our patient.

In the examination of nonlinguistic auditory stimuli, the present case showed that the threshold for the detection of a sound markedly decreased as the number of clicks was increased or the interval of clicks was shortened. This finding suggests that auditory temporal summation described by Motomura *et al.* (1986) was also present in our case. But their case, unlike ours, had auditory agnosia with bilateral subcortical lesions. Temporal summation appears to exist regardless of symptoms when the bilateral auditory radiations are partially destroyed.

In the present case, a disturbance of temporal resolution was found in all the three sensory modalities tested. This finding has led us to speculate that there is a 'time-organizing system' which is both independent of and central to the sensory mechanism. Efron (1963) found that, compared with right-brain-damaged nonaphasics, aphasics with left hemisphere lesions were severely impaired in determining the order when pairs of tones or coloured lights were presented and that the aphasics with more posteriorly located lesions had more severe visual sequencing difficulties. The subsequent studies of Edwards and Auger (1965), Van Allen *et al.* (1966), Goldman *et al.* (1968), Goodglass *et al.* (1970), Carmon and Nachshon (1971), Albert (1972), Swisher and Hirsh (1972) and Carmon (1978) using sequential auditory and/or visual or audiovisual stimuli, supported this view, although the performance of right-brain-damaged subjects was more impaired than that of normal subjects. Also, in monkeys, the inferior parietal lobule and the banks of the superior temporal sulcus are considered to be a polymodal area (Pandya and Kuypers, 1969; Jones and Powell, 1970; Mesulam *et al.*, 1977; Seltzer and Pandya, 1978, 1980; Bruce *et al.*, 1981; Hyvärinen, 1981). These observations indicate that the 'time-organizing system' might be in the temporoparietal region, predominantly on the left side. However, in the present case, it was only after bilateral temporoparietal lobe infarctions that a marked defect of auditory perception was observed. Hence damage to the temporoparietal regions bilaterally might be necessary for production of marked disturbance of the 'time-organizing system'.

Amusia

In the reported cases of pure word deafness with musical impairment, the disturbance seems to be mainly receptive (Liepmann and Storch, 1902; Bonvicini, 1905; Stertz, 1912; Schuster and Taterka, 1926; Klein, 1927; Reinhold, 1950; Ziegler, 1952; Denes and Semenza, 1975; Auerbach *et al.*, 1982; Coslett *et al.*, 1984). But in our case, musical expression was much more severely involved than musical perception, as in the case of Klein and Harper (1956). Since our patient was unable intelligibly to hear her own words, this may account for her difficulty in musical expression.

This patient had several characteristic disturbances in musical perception. Generally, basic components of music are considered to be composed of the pitch and the rhythm (Wertheim, 1969; Gordon, 1970, 1978; Gates and Bradshaw, 1977). Recognition of pitches was markedly impaired in our case. As for rhythm perception and reproduction, a supramodal defect was seen at a fast rate, supporting the view of Mavlov (1980). However, this condition significantly improved in all modalities at a slow rate, indicating rhythm sense is essentially preserved. Since perception of rhythm requires registration of time intervals, this rate-dependent supramodal defect can be attributed to a supramodal defect in temporal resolution. There is some evidence that rhythm patterns are preferentially processed by the left hemisphere (Halperin *et al.*, 1973; Papçun *et al.*, 1974; Robinson and Solomon, 1974; Gordon, 1978). Also, Mavlov (1980) reported a case of a professional musician who developed a supramodal defect only in the recognition and reproduction of rhythm patterns following a vascular stroke in the left posterior parietal or parietotemporal region. In the light of our own and these findings, we can conclude that rhythm sense probably reflects one aspect of a 'time-organizing system' situated in the temporo-parietal region, predominantly in the left.

Thus we believe that the expressive and receptive musical disabilities in this patient are secondary to central hearing loss.

Our patient was able to discriminate broad categories of sounds: voices, music and environmental sounds; however, she was unable to distinguish foreign languages. Also, she had some difficulty in identifying types of musical instruments and the gender of a recorded voice. From these observations, it seems that she had difficulty in distinguishing sounds belonging to the same 'timbre category'. It has been reported that the right hemisphere is responsible for perception of the intonation contours of speech (Blumstein and Cooper, 1974) and for timbre perception (Milner, 1962; Kallman and Corballis, 1975; Mazzucchi *et al.*, 1982). Hence her difficulty may reflect the larger right hemisphere lesion.

With regard to melody recognition, her ability was relatively preserved. Bever and Chiarello (1974) called attention to the possibility that different nervous structures may be engaged in the processing of musical stimuli, depending on the subject's contact with music and that a melody may be perceived either as its overall melodic contour or as an organized arrangement of components. Bever and Chiarello (1974) concluded that naive listeners do in fact perceive melodies in a gestalt fashion and

that the processing is predominantly in the right hemisphere, while musically experienced listeners perceive melodies in an analytic fashion and that the processing is predominantly in the left hemisphere. Hence, in the light of her experience in music, her relatively preserved melody recognition in the presence of a large right hemisphere lesion may be due to an interaction of the right and left hemisphere functions.

Relationship between Temporal Resolution and Auditory Comprehension

In the present case, the clinical defect was limited only to auditory modality, although psychophysical data revealed a supramodal defect in the temporal resolution of these modalities. This finding suggests that the auditory modality is much more dependent upon a temporal resolution than other sensory modalities. Somatosensory and visual perception demands more spatially-orientated data processing. Therefore, these capacities of hers would have not been affected. Also, she had an extremely abnormal click fusion and click counting ability. Especially, the threshold of click fusion (300 ms) was much higher than those reported by others: 15 ms in Albert and Bear (1974) and 30 ms in Auerbach *et al.* (1982). These findings support the view of Albert and Bear (1974) who emphasized that the defect in auditory temporal resolution was an important factor for the development of pure word deafness. However, there still remains a problem which cannot be explained by this hypothesis. Her auditory comprehension did not improve at all, even if she was spoken to slowly enough, despite the marked improvement in temporal resolution as the duration of a tone was prolonged. This indicates that there are other factors besides a defect in auditory temporal resolution for the poor recognition of spoken words in this case. This patient had marked impairment in discrimination of loudness, pitch and tone duration. In addition, she often complained that a human voice sounded like an echo. These factors and/or other unknown factors may be responsible for her poor recognition of spoken words.

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