Cortical Deafness, Auditory Agnosia, and Word-Deafness: How Distinct are They?

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Word-deafness (verbal auditory agnosia, agnosia for speech, agnosia for acoustic language) is the most devastating of all the disorders of the central auditory pathway in children, whether the disorder is present from infancy (dysphasia) or is acquired (aphasia). Typically, word-deaf children are mute and understand little or nothing of what is said to them; they resemble deaf children in this respect, except that it is their brains that are abnormal rather than their ears. Judging from personal experience and from the literature, there are many more word-deaf children than adults. The literature on adults consists entirely of isolated case reports. While there are now over a hundred descriptions of children with so-called epileptic aphasia, these have been summarized in several recent reports (e.g., Holmes, McKeever, & Saunders, 1981; Dugas et al., 1982; Dulac, Billard, & Arthuis, 1983; Aicardi, in press). Epileptic aphasia produces the most dramatic form of word-deafness in the pediatric age-group, yet it may not be the most frequent since there are no data on the prevalence of word-deafness among autistic and congenitally dysphasic children.

The ability to hear, process, and comprehend speech is indispensable for normal language acquisition. Sustained speech input is also required for the maintenance of speech during the preschool years. Most congenitally deaf children fail to acquire speech (and language) unless educated through the visual channel; the same is true of word-deaf infants. Preschool children who are deafened suddenly by meningitis become mute immediately or within days; so do preschool children with acute epileptic aphasia. Abrupt onset of deafness or word-deafness may result in a dramatic decrease in verbal output, even in older children, despite the advantage provided by better practiced speech and by a sparing of the ability to process language through the visual modality (reading and writing). Adolescents and adults with acquired deafness do not lose their speech, although its acoustic properties will eventually deteriorate somewhat through lack of feedback. Word-deaf adults speak but complain that they don't know what they are saying.

There is controversy as to whether word-deafness, in both its developmental and acquired forms, should be considered an auditory disorder or a linguistic disorder specific to the decoding of acoustic language (phonology). This controversy exists not only for word-deafness in children but also for the acquired syndrome in adults; pure syndromes of "cortical" deafness, auditory agnosia, and verbal auditory agnosia are exceedingly rare, if they exist at all. The reason for this lack of "pure" cases is that the cerebral insults responsible for these syndromes are not selective (damage may be cortical or subcortical in either hemisphere or both). Pathology often involves areas adjacent to the auditory pathway, so that most patients have associated deficits, especially during the acute phase of the illness. These concomitant signs may complicate the interpretation of the language deficit. Occasional word-deaf adults may be "cortically" deaf, most have an auditory agnosia, and they may have a hemiparesis or a transient aphasia as well.

Children with congenital or acquired word-deafness almost never have a demonstrable structural lesion on computerized tomography (CT); at best, evidence for localization of dysfunction is an abnormal EEG. Word-deaf children are difficult to test behaviorally because of their age, lack of comprehension of speech, and frequent behavior disorders. In order to gain an understanding of the children's deficits, it has been necessary to consider evidence from adults in whom word-deafness was the result of a stroke and in whom the extent of cerebral infarction was demonstrable on CT or at autopsy. Adults with word-deafness can be alert and cooperative, making detailed testing possible. Furthermore, they can describe what they are experiencing.

Few word-deaf adults and even fewer word-deaf children have undergone detailed behavioral evaluation of their auditory and language abilities, and none seem to have undergone electrophysiologic recordings with state-of-the-art paradigms (Novick, Kovrich, & Vaughan, 1985). Whether the elementary auditory perceptual deficits found in word-deaf persons explain their inability to understand speech is an open question and raises the more basic issue of whether decoding phonology calls for abilities fundamentally different (i.e., linguistic rather than acoustic) from those required for other auditory perceptual tasks (Liberman et al., 1967; Albert et al., 1972; Liberman, 1974).

Definitions

Sensorineural hearing loss denotes loss of auditory sensitivity due to pathology in the ear or VIIIth nerve. Central auditory deficits are behaviorally defined deficits in auditory perception and processing whose anatomic substrate is virtually always undefined (see later). "Corti-

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cal" deafness, which is the most severe form of central auditory deficit, refers to unresponsiveness to all sounds because of bilateral pathology in the cerebral hemispheres (not necessarily in the auditory cortex, as will be seen). Auditory agnosia refers to the inability to identify the source of environmental sounds, to discriminate among them and between these sounds and speech, despite the ability to hear sounds. It denotes bilateral hemispheric pathology in all but exceptional cases.

Verbal auditory agnosia or word-deafness refers to the inability to comprehend speech despite preserved ability to process language visually and to perceive and recognize environmental sounds. Adult patients with acquired word-deafness not only speak; typically, they can calculate, read, write spontaneously, and copy, but they cannot repeat what is said to them or write to dictation. Most word-deaf adults, notably those with bilateral temporal lobe pathology, have auditory agnosia, and occasionally, some are cortically deaf, usually transiently. Young word-deaf children are mute, and few acquire language adequately unless: 1) it was already well developed before they became word-deaf; 2) their word-deafness is relatively short-lived; or 3) they are educated as though they are deaf.

Central deafness is exceedingly rare because each ear projects to both sides of the brain stem, thalamus, and cortex. Occasional patients have severe loss of auditory sensitivity due to lesions, such as a pontine glioma (Makishima & Ogata, 1975) or demyelinating plaques (Dayal, Tarantino, & Swisher, 1966), which affect the brain stem auditory pathway bilaterally. Kernicterus due to neonatal hyperbilirubinemia is often invoked as an example of loss of hearing due to pathology in the auditory relays of the brain stem. In fact, hyperbilirubinemia may produce pathology in the cochlea or VIIIth nerve as well as in the brain stem nuclei (Kaga, Kitazumi, & Kodama, 1979) and may not qualify as an example of central hearing loss. Neonatal asphyxia in newborn monkeys produces cell loss in the inferior colliculus (Ranck & Windle, 1959); the behavioral consequences of this pathology in man have been reported by, for example, Kileny & Berry (1983).

Epileptic aphasia is a term applied with increasing frequency to children with acquired verbal auditory agnosia as though it were synonymous with it. In fact, there are a few children with epileptic aphasia whose deficit is predominantly an expressive one, with better comprehension than expression (e.g., Sato & Dreifuss, 1973; Deonna, Fletcher, & Voumard, 1982; van de Sandt-Koenderman et al., 1984). There are children with verbal auditory agnosia who do not have seizures or an abnormal EEG, especially among those in whom the deficit is developmental rather than acquired. Epileptic aphasia is a chronic disorder that must be differentiated from transient loss of speech (Todd's aphasia) during or following a partial seizure that involves the language areas of the dominant hemisphere (Hécaen & Piercy, 1956; Deonna et al., 1977; de Negri, Doria, & Veneselli, 1983).

Organization of the Central Auditory Pathways

Each of the ears projects to both hemispheres. Studies in animals and dichotic studies in man, in which competing stimuli are presented simultaneously to both ears (Darwin, 1974), have demonstrated conclusively that the projection from each ear to the contralateral temporal lobe dominates the projection to the ipsilateral side (Sparks, Goodglass, & Nickel, 1970). The left hemisphere is known to be dominant for most aspects of language, namely the processing of consonant sounds (Studdert-Kennedy & Shankweiler, 1970), syntax (Liberman, 1974; Rankin, Aram, & Horwitz, 1981), and the meaning of verbal messages (semantics) (Boller, Kim, & Mack, 1977). It is even more strongly dominant for the production of speech than for its comprehension. In fact, the isolated right hemisphere is virtually mute (Gazzaniga & LeDoux, 1978). The right hemisphere is superior to the left for processing the prosody of speech (intonation, rhythm, and melody, characteristics that convey its affective tone) (Heilman, Scholes, & Watson, 1975) as well as for music (chords and melodies) and other functions including visuo-spatial skills, facial recognition, emotion, and attention-arousal (Heilman & Valeenstein, 1979; Weintraub & Mesulam, 1983). Hemispheric asymmetry for the processing of stop consonants and music appears to be innate (Eimas et al., 1971; Morse, 1979; Molfese, 1983).

Lesions of the primary auditory cortex do not impair the perception of pitch, loudness, or the onset or cessation of auditory stimuli, but they do prevent animals from judging duration, order, and pattern of stimuli and the location of sound in space (Ravizza & Belmore, 1978). Decoding of more complex aspects of auditory signals is presumed to engage auditory association areas, with phonologic decoding of speech occurring predominantly on the left which is anatomically larger than the right (Geschwind & Levitsky, 1968). Comprehension of the meaning of speech is impaired, despite preserved ability to decode phonology, in patients with lesions in themore posterior Wernicke's area of the left temporal lobe (Boller, Kim, & Mack, 1977). Dichotic studies illustrate the importance for the decoding of speech of interhemispheric transmission via commissural fibers which connect the auditory association areas on the two sides. Left ear extinction (inability to report stimuli presented to the left ear during dichotic tasks), despite the ability to report words presented to either ear alone, occurs in patients with right temporal lesions and in those with commissurotomies for the control of epilepsy (Sparks & Geschwind, 1968; Sparks, Goodglass, & Nickel, 1970). Information conveyed via the ipsilateral pathway from the left ear to the left temporal lobe is suppressed when it is competing with information from the crossed pathway coming from the right ear. Subcortical loss of commissural fibers coming from the right auditory association areas, together with destruction of the acoustic radiations on the left, explains the occasional case of word-deafness caused by a single leson on the left (Geschwind, 1965).

Cortical Deafness

A number of case reports of word-deafness associated with auditory agnosia state that, when the patient suddenly lost the ability to comprehend speech, he complained he could not hear and was unresponsive to environmental sounds, music, and speech (e.g., Klein & Harper, 1956; Lhermitte et al., 1971; Jerger, Lovering, & Wertz, 1969; 1972; Chocholle et al., 1975; Goldstein, Brown, & Hollander, 1975; Leicester, 1980; Özdamar, Kraus, & Curry, 1982). Some case reports include a behavioural pure-tone audiogram showing a moderate or severe flat hearing loss; others show a high tone loss. These audiograms are often dismissed by authors on the grounds that they are unreliable and reflect the patient's auditory inattention or represent a presbyacusis consonant with the patient's age, and that, in any case, the loss of auditory sensitivity was transient and not severe enough to account for the patient's word-deafness. Such an interpretation must be accepted skeptically, especially since a premorbid audiogram is usually lacking. The severe sensorineural hearing loss of the 21-year-old man studied in great detail by Jerger et al. (1969) cannot be disregarded. It evolved later into a persistent (for at least 10 weeks) asymmetric high tone loss, at which time cortical evoked potentials to clicks and tones were absent. Hearing loss was severe and permanent in patients described by Clark & Russel (1938) and Leicester (1980). A 8-year-old boy with word-deafness and epileptic aphasia observed by this author complained at first that he could not hear and his pure-tone audiogram showed a threshold at 70-80 dB, which returned to normal on retest two months later. Unfortunately, no electrophysiological testing was available to help evaluate these observations. The 6-year-old boy with congenital word-deafness studied at post-mortem by Landau, Goldstein, & Kleffner (1960) had cystic infarcts of the auditory and auditory association cortex bilaterally, together with severe degeneration of the medial geniculate bodies, had a normal audiogram (deviations from normal on two other audiograms were considered unreliable). No doubt these divergent observations reflect differences between individuals both in terms of extent of pathology and of brain organization.

As already stated, bilateral ablation of the auditory cortex in animals does not produce deafness, that is, complete and permanent unresponsiveness to sound. The Landau, Goldstein, & Kleffner (1960) case, as well as most cases of adults with bilateral infarction of the auditory cortex, confirm this in man (e.g., Coslett, Brashear, & Heilman, 1984). The word "cortical" is put in quotes since in at least one case (Clark & Russell, 1938) the cause of "cortical" deafness was reported to be entirely subcortical. In patients in whom the hearing loss is transient, edema of the white matter at the time of the stroke or diaschisis in lower relays of the afferent auditory pathway may be at fault. Inattention to auditory stimuli may play a role in explaining the lack of startle to loud sound and unreliable audiograms reported in some patients (e.g., Albert et al., 1972; Chocholle et al., 1975), since the right hemisphere is thought to play an important role in the control of arousal and attention (Heilman & Valenstein, 1979).

In order to document cortical deafness, one would need to have not only behavioral data and pure-tone audiograms indicating complete lack of responsivity to sound, but also the electrophysiologic evidence provided by normal threshold of waves I-V (or I-VIII) of the brainstem evoked response, and absent cortical potentials, including perhaps the so-called "middle" potentials that signal activation of geniculate neurons and primary auditory cortex (Picton et al., 1974). A cortically deaf patient (described by Özdamar, Kraus, & Curry, 1982) fulfilled these criteria and his documented cortical deafness later evolved into an auditory agnosia, as is regularly the case. A patient with auditory agnosia rather than cortical deafness had normal brainstem and middle potentials (Parving et al., 1980), while a word-deaf patient described by Michel & Peronnet (1980) had no cortical evoked potentials to tones. Ludlow et al. (1985) reported that a 23-year-old patient with cortical deafness following presumed herpes simplex encephalitis had behavioral pure-tone thresholds between 65 and 95 dB and normal thresholds for brain stem evoked responses. They did not describe his cortical potentials. Moreover, their patient was not deaf but had selective deficits in auditory processing.

Auditory Agnosia

Auditory agnosia is usually taken to mean inability to identify environmental auditory stimuli despite normal hearing sensitivity to pure tones. Vignolo (1969) pointed out that this definition is ambiguous: one needs to determine whether the patient can discriminate among sounds, which is a function for which the right hemisphere is dominant, or whether the patient is unable to identify the source of the sound, an ability most likely to be affected by left-sided pathology. Findings may depend on the manner in which one has tested the patient: 1) is he to match a sound he has just heard with one in a series of several sounds presented successively; 2) is he to match it with one of several pictures of animals or objects producing different sounds, or 3) is he to name it (Spreen, Benton, & Fincham, 1965; Vignolo, 1969)? The diagnosis of auditory agnosia based on the last technique would be shaky in aphasic patients or those with anomia for stimuli presented to the auditory channel (Denes & Semenza, 1975).

Most patients with auditory agnosia not only cannot name but they cannot recognize environmental sounds, reflecting the bilaterality of their pathology. They often report that sound is unpleasant and they may not be able to distinguish speech from other sounds. Some of them have difficulty localizing a sound-source in space (Jerger et al., 1969; Lhermitte et al., 1971; Chocholle et al., 1975; Michel & Peronnet, 1980; Coslett, Brashear, & Heilmann, 1984).

Many patients with auditory agnosia also have amusia, that is an inability to perceive music as such, to identify melodies, or the timbre of different instruments.

Many of them cannot sing or whistle a well known tune, play a musical instrument, or reproduce a pattern of taps (e.g., Klein & Harper, 1956; Lhermitte et al., 1971; Albert et al., 1982; Chocholle et al., 1975; Denes & Semenza, 1975; Oppenheimer & Newcombe, 1978). A few have had systematic evaluations, for example, using subtests of the Seashore Measures of Musical Talent. Amusia, like auditory agnosia without word-deafness, suggests right-sided pathology (Spreen, Benton, & Fincham, 1965; Roeser & Daly, 1974; Critchley & Henson, 1977).

Because right-sided pathology is more likely to interfere with the appreciation of melodies than the left does not imply that music is localized only to the right hemisphere. Based on dichotic studies, Bever & Chiarello (1974) found left ear preference for music in non-musicians and right-ear preference in musicians whose technical knowledge of music evidently engaged the activity of the left hemisphere as well as the right. That the superiority of the right hemisphere for the perception of music and for the prosody of speech rests on common physiological mechanisms is plausible, but unproven.

One way to document auditory agnosia is through use of the odd-ball paradigm during evoked potential recordings (Simson, Vaughan, & Ritter, 1977; Novick, Lovrich, & Vaughan, 1985). A late positivity peaking after 300 ms (P300) occurs in the evoked response to one of two stimuli if it is presented rarely and unexpectedly in a train of the other one. This is enhanced if the patient is asked to respond selectively to the rare stimulus. First, one must determine that the patient has late cortical evoked responses to each of the two auditory stimuli; lack of a P300 would be an electrophysiological correlate of inability to discriminate between them.

Verbal Auditory Agnosia Word-Deafness

Word-deafness is an agnosia for speech sounds or, more technically, an inability to decode phonology. Strictly speaking, one should not diagnose word-deafness in a patient who also has an auditory agnosia because word-deafness implies lack of comprehension of speech despite preserved ability to hear, discriminate, and recognize environmental sounds and music. In reality, most word-deaf adults also have an auditory agnosia (Goldstein, 1974). It is uncertain if the auditory agnosia is sufficient to explain the word-deafness or, more plausibly, whether bilateral pathology is required to produce both. Word-deaf children may be less likely than adults with structural lesions to have a severe agnosia for non-speech sounds, but this impression may be unreliable since so few have been examined.

Some word-deaf adults state that they cannot "hear" themselves speak, may speak too loudly, and may state that they know what they want to say but do not know whether they succeeded in doing so (Wohlfart, Lindgren, & Jernelius, 1952; Klein & Harper, 1956). This inability to monitor speech can be demonstrated by delayed auditory feedback. A delay of approximately 200 ms severely disrupts the speech of most normal persons (Chase et al., 1959; Burke, 1975). Several word-deaf adults have been found to be totally insensitive to delayed auditory feedback (Lhermitte et al., 1971; Gazzaniga et al., 1973; Chocholle et al., 1975; Michel & Peronnet, 1980) and these patients do not increase the intensity of their voices like normals when their speech is masked by loud noise.

Word-deaf adults are not aphasic since they are able to speak, name objects to visual confrontation or to touch, read aloud and silently with comprehension, and write spontaneously. In other words, their "inner language" is preserved, and they can continue to program speech despite the lack of feedback. They may make some paraphasic errors in both their spontaneous utterances and their writings. These are more likely to be phonemic paraphasias than semantic paraphasias, which may be a function of their difficulty decoding and monitoring phonology (Coslett, Brashear, & Heilman, 1984; van de Sandt-Koenderman et al., 1984). Some word-deaf patients may have other more or less mild aphasic symptoms as residua of a left-sided stroke.

Word-deafness is a deficit that is specific to the processing of language presented to the acoustic channel. Children with epileptic aphasia and word-deafness acquired in mid-childhood continue to read and write, but their ability to express oral language varies. Preschool children with word-deafness are mute, and may remain mute or severely dysfluent for years. Like adults, word-deaf children are able to process language visually and they acquire language through the visual channel in the form of reading or signing (e.g., Stein & Curry, 1968). This may be because brain dysfunction affects the parieto-temporal cortex, notably the angular gyrus (a late developing cortical area critical to higher-order modality non-specific linguistic processing) or perhaps because brain dysfunction impairs subcortical areas, such as the thalamus (an area which may be as critical for language as the cortex) (Ojemann, 1976; Crosson, 1984). In some children, inadequate learning of signs may be due to cognitive limitations, apraxia of the hands, lack of motivation to communicate, or impaired attention, which are all concomitant signs of brain dysfunction. Inadequate learning of visually presented language is often a function of inadequate exposure to fluent signing or to the use of an inappropriate method for teaching reading which stresses a phonic rather than an iconic (whole-word) approach.

A number of tests of elementary auditory functions have been carried out in word-deaf adults, most of whom were able to discriminate pitch and loudness. Some had difficulty judging the duration of sounds, integrating duration and intensity of near-threshold short sounds (Zwislocki, 1960), judging the relative loudness of sounds presented to the two ears; and some had a tendency to fuse two successive stimuli not separated by an unusually prolonged interval (Jerger et al., 1969; Jerger, Lovering, & Wertz, 1972; Albert & Bear, 1974; Chocholle et al., 1975; Saffran, Marin, & Yeni-Komshian, 1976). A few patients' comprehension was reported to improve when the rate of speech presented to them was slowed (Albert

& Bear, 1974; Oppenheimer & Newcombe, 1978). This seems to emphasize the importance of the temporal aspects of speech for its processing, although this finding was not universal (Coslett, Brashear, & Heilman, 1984).

In those patients in whom this type of study was carried out, it was consonant rather than vowel discrimination which was affected (Klein & Harper, 1956; Chocholle et al., 1975; Denes & Semenza, 1975; Saffran, Marin, & Yeni-Komshian, 1976; Oppenheimer & Newcombe, 1978). Consonants are known to carry more of the linguistic message than vowels and to be processed more efficiently in the left than the right side of the brain (Studdert-Kennedy & Shankweiler, 1970). Consonants are coarticulated with vowels into syllabic acoustic signals lasting up to 250 ms. The part of the signal that encodes consonants is only a brief portion of this signal. For example, stop consonants differ by a brief (40 ms) segment characterized by a rapid change in the frequency of the acoustic signal (formant transition), or by a brief pause between the opening of the vocal tract and the onset of the vibration of the vocal cords (Liberman et al., 1967). Difficulty discriminating among stop consonants is found in some dysphasic children. Their performance improves if formant transitions are artificially lengthened (Tallal & Piercy, 1978; Tallal & Stark, 1981). Children with word-deafness seem to be particularly deficient for this task (Frumkin & Rapin, 1980). Whether one is justified in concluding that dysphasic children with predominantly expressive deficits and word-deaf children differ only in the severity of this common phonologic decoding deficit seems doubtful, but it is an unanswered question.

Saffran, Marin, & Yeni-Komshian (1976) suggested that their word-deaf adult patient without auditory agnosia or amusia, who had impaired phonetic analysis skills and right ear suppression on dichotic tasks, processed language through the right hemisphere because of pathology on the left. Unfortunately, documentation of pathology strictly limited to the left hemisphere and mapping of brain activity during linguistic tasks were not available to document this hypothesis. Whether inability to discriminate consonants is prerequisite and sufficient cause for word-deafness remains to be determined.

If few satisfactorily detailed studies of auditory and linguistic function have been reported in word-deaf adults, fewer still seem to have been performed in young children, no doubt in part because of the difficulties involved. Stein & Curry (1968) reported that their patient had an auditory agnosia as well as word-deafness. Five of 14 patients described by Worster-Drought (1971) and at least one of four studied by Cromer (1978) seem to have had some degree of auditory agnosia as well as word-deafness. As stated earlier, Frumkin & Rapin's (1980) four patients with word-deafness were unable to discriminate stop consonants. Deonna, Fletcher, & Voumard (1982) described the verbal productions of a 2½-year-old boy with a few days episode of epileptic aphasia associated with left-sided paroxysmal EEG discharges. The child regressed transiently to saying only single words. Comprehension appeared less impaired than production, and phonology was not disproportionately distorted, so that he probably was not word-deaf. Van de Sandt-Koenderman et al. (1984) made monthly recordings over a period of almost two years of the speech of a 6½-year-old girl with two episodes of epileptic aphasia affecting comprehension and production. They counted the number of words she produced in three minutes and the mean length of her utterances; both were dramatically decreased while she was symptomatic. Her articulation was very defective but syntax was not affected. At the height of her illness she made many paraphasic errors and produced neologisms. Defective phonologic production is characteristic of word-deaf children (e.g., McGinnis, 1982; Worster-Drought, 1971; Gascon et al., 1973) and is probably a reflection of their defective perception of phonology.

Cromer (1978) studied the written syntax of 6 children with severe receptive and expressive dysphasia and compared it to those of 6 congenitally deaf children. The deaf children made more grammatical errors, but they attempted more sophisticated syntactical constructions. Bishop (1982) compared the knowledge or oral, written, and signed English grammar of 9 children with epileptic aphasia and word-deafness, 25 children with predominantly expressive dysphasia, and 31 congenitally deaf children. She found that the children with word-deafness were deficient in comprehension of written and signed, as well as oral, syntax, and that their performance was similar to that of deaf children. The knowledge of the predominantly expressive dysphasics was far superior, although not quite up to norms. She suggested that the deficit of the deaf and word-deaf children was attributable to their learning of language by eye rather than by ear, and that "signed exact English" is not well adapted for conveying the syntax of oral language.

None of these studies answers the basic question of whether word-deafness is an auditory defect affecting linguistic and non-linguistic stimuli equally, or whether it is specific to the linguistic task of decoding phonology (Liberman et al., 1967). This question was considered with two chronically word-deaf adolescents who did not have a general auditory agnosia and who had acquired some ability to comprehend speech and to express themselves orally, although their phonology and syntax remained quite deficient. They had normal brainstem auditory evoked responses to clicks and normal N100 P200 waves of the cortical evoked response to verbal stimuli. They had no difficulty making a pitch discrimination as reflected by a P300 wave triggered by use of the odd-ball paradigm. They were unable to make a phonetic discrimination until they were trained using a visual cue. Even then, their P300 waves were markedly attenuated. They could not be trained to make a semantic discrimination for which training with a visual cue was not possible. Their performance and late potentials to visual analogs of these tasks were normal. Although more detailed behavioural testing of the ability to discriminate other nonverbal auditory stimuli will be required before confidently stating that

their deficit is specific for phonological decoding, the data are suggestive, and a methodology for approaching this question is at hand.

Electrophysiology, using the Brain Electrical Activity Mapping approach of Duffy (1982), can provide data on the localization of cortical activity during verbal and nonverbal tasks. Imaging of the brain's metabolism during such tasks by measuring blood flow, glucose utilization, or the local concentration of neurotransmitters are powerful techniques. However, because they employ radioactive compounds, they will have limited application in children who are not severely ill. Further, their use is not completely without question. For example, a single boy with word-deafness was found not to have the expected increase in blood flow over fronto-temporal regions during a verbal naming task (Lou, Brandt, & Bruhn, 1984).

Epileptic Aphasia and Word-Deafness in Children

There is a large literature on the subject of epileptic aphasia, starting with the report of Landau and Kleffner in 1957. It comprises several large reviews of published cases (Fejerman & Medina, 1980; Holmes, McKeever, & Saunders, 1981; Toso et al., 1981; Dugas et al., 1982). More than 100 cases have been described. Most of these papers focus on the medical and electroencephalographic aspects of the illness, on its prognosis, and on how to manage it.

The EEG abnormalities may be much more common in sleep than awake (Dulac, Billard, & Arthuis, 1983), and in fact may be almost continuous during sleep in some children (Kellerman, 1978). The discharges consist of spike and spike-wave complexes; they may be unilateral or bilaterally focal over temporal or parieto-temporal regions, or be generalized (Rodriguez & Niedermeyer, 1982). In general, the abnormalities may vary in location and severity over time and are only rarely correlated with the severity of the language disturbance (e.g., Rapin et al., 1977; Holmes, McKeever, & Saunders, 1981; Aicardi, in press). Although many children have generalized, focal motor absence, or partial complex seizures, some children have no clinically detectable seizures despite their abnormal EEGs. Whether such children should be treated with anticonvulsants in a debated issue, especially since control of clinical seizures in children who have them does not often improve the language disorder dramatically (Deuel & Lenn, 1977).

The language disorder may precede or follow the onset of the seizures, in some cases with an interval of many months (Forster, Braun, & Weidner, 1983). It may start acutely or insidiously, and may persist long after the seizures have disappeared and the EEG has returned to normal, which happens regularly in due course. This suggests that both the language disorder and the abnormal EEG are the manifestations of a transient brain dysfunction that may leave permanent sequelae, rather than the seizures being responsible for the word-deafness. Epileptic aphasia resembles the benign epilepsy of childhood with Rolandic spikes by the character of its EEG discharges and their exacerbation during sleep, by the disappearance of both the seizures and the EEG abnormality by adolescence, and in most cases, by the lack of overt structural pathology. It is all but benign considering its duration and drastic consequences for language and behavior. The course of the illness is extremely variable, with abrupt loss of language in some children and insidious progression over many months in others. In some children language skills fluctuate with transient remissions. The illness may extend over many years. In some cases, recovery may be incomplete; other children recover entirely (Mantovani & Landau, 1980).

Prognosis appears to be better in school-age children who develop epileptic aphasia after language is well established (Forster, Braun, & Weidner, 1983; Bishop, in press) than in toddlers who become mute and understand virtually nothing of what is said to them. These young children are in essentially the same situation as those in whom word-deafness is present since infancy (Worster-Drought & Allen, 1930); McGinnis, 1963; Cromer, 1978; Maccario et al., 1982). Some congenitally dysphasic word-deaf children have an abnormal EEG, with or without seizures; some do not, and therefore cannot be classified as having epileptic aphasia.

Word-deafness may be associated with a number of behavior disorders (Petersen et al., 1978). Some appear to be secondary to loss of comprehension and lack of a vehicle for expression since they improve when the children learn to communicate through the visual channel. They usually use pantomine, signs, reading, or a communication board. In contrast with older children and adults, lip-reading is rarely effective for younger children, as it is an inefficient channel for learning language. In other children, behavior disorders — which may include aggression, temper tantrums, inattentiveness, or withdrawal — appear to be primary and represent but another manifestation of the child's brain dysfuntion (Rapin & Allen, 1983). Some word-deaf children are frankly autistic. Their prognosis for languge learning and development is guarded unless particular pains are taken to foster socialization. Their gaze avoidance, stereotypes, and cognitive deficits are not conducive to communication through any channel. On the other hand, there are word-deaf children who have no behavior disorder and who invent gestures and communicate very effectively by pointing, pantomine, or pictures. They are the children who will do well with Total Communication (sign language together with oral speech) and with reading.

The pathology of word-deafness associated with epileptic aphasia is unknown. The EEG abnormalities and auditory deficit point to the temporal lobes. Almost all children have normal CT scans and spinal fluid, although the boy described by Landau, Goldstein, & Kleffner (1960) had bilateral porencephalies in his temporal lobes. Rarely word-deafness may be the result of an encephalitis (Lou, Brandt, & Bruhn, 1977; Gupta et al., 1984; Ludlow

et al., 1985). The pathology is presumed to be bilateral in most cases, since unilateral damage sustained in early life does not preclude the development of language, whether it be on the right or the left (Annett, 1973; Dennis & Kohn, 1975; Rankin, Aram, & Horwitz, 1981). Perhaps dysfunction is unilateral in some children who happen to be particularly strongly lateralized for language. Boys are more likely to suffer from word-deafness than girls. There are also a few cases in siblings (Landau & Kleffner, 1957) which raises the possibility that genetic factors may be at play in some cases.

In short, word-deafness in children, whether it occurs in the context of epileptic aphasia or is congenital, is a devastating disorder that does not have a single etiology. It is very difficult to give a prognosis at its start. Anti-convulsants are needed for children with clinical seizures and are worth trying in those with an abnormal EEG and no seizures. The role of ACTH or steroids, if any, is controversial (Aicardi, in press). Remedial education based on supplementation of the defective auditory channel by the visual one is in order. Research is needed in order to develop better prognostic criteria, define the etiology of epileptic aphasia, and delineate the language disorder in more detail so as to gain better understanding of its pathophysiology.

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