Deafness and Cerebral Plasticity

Deafness refers to the inability to perceive auditory stimuli. The condition may be congenital or acquired. Numerous causes for acquired deafness have been identified, including bacterial and viral infections leading to meningitis or encephalitis. In addition, children aged 2 years or older may significantly improve communication skills. Cochlear implants can substitute for the missing auditory receptor functions and may help to improve impaired hearing function. In adults with severe or profound deafness due to inner ear impairment, cochlear implantation may significantly improve communication skills. Children aged 2 years or older may also be potential candidates for this type of hearing prosthesis.

Deafness can cause severe disability in children and adults. The auditory system conveys information about environmental acoustics, and, in humans, learning and use of language for communication are inherent functions of audition. Deafness in children affects the development of speech, language, and cognitive abilities. In both children and adults, inability to hear may severely impair social capabilities. Thus, efforts to restore auditory perception are crucial.

Cochlear implants can substitute for the missing auditory receptor functions and may help to improve impaired hearing function. In adults with severe or profound deafness due to inner ear impairment, cochlear implantation may significantly improve communication skills. Children aged 2 years or older may also be potential candidates for this type of hearing prosthesis. However, before a cochlear implant is considered, various tests of the auditory system need to be performed to ensure that benefits to the patient will be sufficient to warrant this invasive procedure.

Deafness occurring before the acquisition of language skills is described as prelingual, whereas hearing loss developing during or after language acquisition is termed perilingual or postlingual deafness. In adults with postlingual deafness, language performance appears to be best when the period from the loss of hearing to the introduction of the cochlear implant is relatively short. Individuals with prelingual deafness may achieve a limited ability to differentiate sounds with the aid of cochlear implants, even when they receive the implant as an adult. However, with prelingual deafness, a quasinatural language development may be achieved only when cochlear implants are introduced at a relatively early age.

The success of cochlear implants in the treatment of total deafness depends on anatomically and functionally intact neuronal transmission from the cochlea to the auditory cortex. Similar to other sensory systems, the auditory system is able to reorganize its structure and function after partial or total loss of its receptor function. This ability is termed “plasticity.” When metabolism in the auditory cortex of deaf patients has been restored by the phenomenon called “cross-modal plasticity,” the auditory cortex can no longer respond to signals from a cochlear implant installed afterward.

AUDITORY SYSTEM AND FUNCTIONAL STUDIES

Several neurophysiologic tests, such as those measuring acoustic emissions or auditory evoked potentials, have been developed to assess the auditory neuronal pathway. There is accumulating evidence that functional imaging procedures such as functional MRI (fMRI), SPECT, and PET give reliable information about the neurologic status of the primary and associative cortices.

Friston et al. (4) performed basic methodologic research to compare fMRI and PET with respect to levels of statistical inference and power in detection of neuronal activation. From their work, it is possible to conclude that set-level inference (which infers that a given number of clusters comprising an observed activation profile is unlikely to have occurred by chance) is generally more powerful than cluster-level or voxel-level inference. Therefore, for set-level inferences, if the signal (relative to resolution) is strong, as it is in fMRI, then the optimum extent threshold should be greater than the expected number of voxels for each cluster. If the signal is weak, as in PET, the extent threshold should be small.

Investigations of auditory cortex activation with fMRI, SPECT, and PET are described below. For example, fMRI can detect increases in regional cerebral blood flow during cochlear electrical stimulation in deaf patients. In 2 binaural deaf patients studied by Berthezene et al. (5), fMRI showed that electrical stimulation of the left ear induced an increase in maximum signal intensity of approximately 8% in the right and 6% in the left auditory cortex.

Suarez et al. (6) used SPECT with 99mTc-ethylcysteinate dimer as a tracer to examine 5 postlingually deaf patients with multichannel cochlear implants and assessed changes in regional cerebral blood flow through a stimulation paradigm before and after the operation. They demonstrated a significant increase in blood flow in the primary auditory area after surgery. They also observed a significant asymmetric increase in blood flow in the frontal lobes when the patient listened to sequential language sequences.
Evidence of phonological and orthographic processing deficits associated with dyslexia is emerging. In fMRI studies in children, Temple et al. (7) observed significant differences between dyslexic children and control subjects with normal reading ability in the activation of cortical regions during phonological and orthographic tasks. After participation in a remediation program, dyslexic children showed increased activity in the area of the left prefrontal cortex that was normally activated in children without reading impairment.

Recently, PET studies indicated a central origin for several types of tinnitus. In patients who could alter tinnitus loudness by performing voluntary oral facial movements, these changes affected cerebral blood flow in the auditory cortex contralateral to the ear in which tinnitus was perceived, whereas unilateral cochlear stimulation caused bilateral effects (8).

The neural mechanisms involved in listening to sentences and then detecting and verbalizing a specific word are poorly understood. PET activation studies have been performed to identify the areas of the human brain that are involved in hearing speech in quiet and in background noise environments (9). This study revealed that word repetitions activated mainly auditory and motor areas of the brain, whereas more difficult tasks, such as speech in noise or multitalker noise activated linguistic, attentional, cognitive, working memory, and motor planning areas.

Recent PET examinations compared glucose metabolism in the auditory cortex of human subjects with early-onset deafness and healthy control subjects with ears plugged (10). Interestingly, the rate of glucose metabolism in the primary and associative auditory cortices was higher in deaf patients than in healthy subjects.

The biologic constraints and effects of experience on the cerebral organization of language have been studied by fMRI in deaf and hearing subjects (11). The effects of deafness, age of language acquisition, and bilingualism were assessed by comparing results from normally hearing, monolingual native speakers; congenitally deaf signers of American Sign Language (ASL); and normally hearing, bilingual speakers who were native signers of ASL and speakers of English. In this comprehensive examination, a strong and repeated activation of the classical language areas of the left hemisphere was observed in hearing and deaf subjects, while processing their native language, English or ASL. However, deaf subjects reading English did not display activation in these regions. These results suggest that the early acquisition of a natural language is important in the expression of the strong bias for these areas to mediate language, independently of the form of language. In addition, native signers—hearing or deaf—displayed extensive activation of homologous areas within the right hemisphere, indicating that the specific processing requirements of the language may also, in part, determine the organization of the language systems of the brain.

Deafness and Cerebral Plasticity

As reported in this issue of the Journal of Nuclear Medicine, Lee et al. (12) found that the glucose metabolism of 9 postlingually deaf patients was significantly lower than that of age- and sex-matched control subjects in both anterior cingulate gyri and superior temporal cortices. Glucose metabolism in these cortical regions, which represent Brodmann areas 24 (BA24), BA41 (primary auditory cortex), and BA42 (auditory association cortex), showed a significant positive correlation with duration of total deprivation of hearing capability. Six of the 9 patients received cochlear implantation after the PET scan, and speech perception performance testing was done thereafter. Interestingly, there was a tendency toward a poorer prognosis for cochlear implantation in patients with a longer duration of deafness (r = 0.73).

These results are in accordance with those of a previously reported study by Lee et al. (13). In this investigation, the glucose metabolism of 15 prelingually deaf patients was preoperatively reduced in the superior temporal cortices (BA41, BA42, and BA22) as well as in the inferior frontal cortices (BA44 and BA45). After cochlear implantation, the size of the hypometabolic areas correlated positively with a hearing capability score (r = 0.81).

The results of these studies support the notion that when a cochlear implant is installed after restoration of metabolism in the auditory cortex of deaf patients by cross-modal plasticity, the auditory cortex can no longer respond to signals from the implant.

In contrast, Ito et al. (14) found that in 9 totally deaf patients the duration of deafness correlated with the reduction of glucose metabolism in the auditory cortex—that is, the longer the duration of deafness, the lower the glucose metabolism as measured by 18F-FDG PET. However, in this study the PET examinations were not assessed quantitatively. Furthermore, the PET results were not compared with healthy age- and sex-matched control subjects. Thus, from a current perspective, the study has substantial methodologic limitations. The discrepant results of the studies by Lee et al. (12) and Ito et al. (14) may thus be due to methodologic rather than physiologic differences.

In an earlier PET study, Ito et al. (15) reported remarkably low metabolic rates for glucose detected in the auditory cortex before cochlear implantation. However, these metabolic rates returned to normal levels after cochlear implantation. Thus, these findings suggest that the activation of the speech comprehension mechanism of the higher brain system can be initiated by sound signals from the implanted device.

Herzog et al. (16) examined 2 postlingually and 2 prelingually deaf patients by H215O PET with respect to cortical responses to the stimulation of cochlear implants. Relative elevations of regional H215O activity were observed in the primary and secondary auditory cortices. Thus, this method demonstrated objective responses of the central auditory system during electrical stimulation in profoundly deaf patients.
deaf patients, even in those who had never been able to hear. Naito et al. (17), using H15O PET, also investigated 3 prelingually and 6 postlingually deaf patients who had received multichannel cochlear implants. They demonstrated that speech stimulation may cause an increase of regional cerebral blood flow in the auditory cortex of postlingually deaf patients. The increase was smaller in prelingually than in postlingually deaf patients.

Using 18F-FDG PET, Kang et al. (18) studied 5 prelingually and 3 postlingually deaf patients before and after cochlear implantation. Two of the 3 postlingually deaf patients showed disappearance of the hypometabolic area in the contralateral auditory cortex and good speech perception after cochlear implantation. The other patient, who had no hypometabolism in the auditory cortex before cochlear implantation, did not show any change in hypometabolism or improvement in poor speech perception after cochlear implantation.

These studies clearly demonstrated that improvement in speech perception after cochlear implantation cannot be expected in deaf patients who have had no hypometabolism in the auditory cortex before receiving the implant. A normalized glucose metabolism in the auditory cortex before cochlear implantation may thus be the sequela of cross-modal cerebral plasticity—that is, reorganization of neuronal structure and function.

The clinical importance of the study by Lee et al. (12) in this issue of The Journal of Nuclear Medicine lies in the potential for the use of 18F-FDG PET to identify those deaf patients who may benefit from cochlear implantation. According to the National Institutes of Health (NIH) Consensus Statement (1), the limited prediction of implant efficacy in a specific individual remains a pressing issue. 18F-FDG PET may provide improved identification of potential candidates for successful cochlear implantation.

Several studies have suggested that a diminished neuronal inhibition could be involved in cerebral plasticity mechanisms. The inferior colliculus neurons have been shown to display age-related changes, including a decreased number of GABAergic neurons, decreased basal concentrations of GABA, decreased GABA release, decreased glutamate decarboxylase levels, decreased GABA receptor binding, and subtle GABA_A receptor-binding changes (19–21). Changes in the central auditory system after cochlear ablation were also observed in transmitter release and uptake of glycine and N-methyl-D-aspartate receptors in animal experiments (22). These observations may stimulate PET investigations using appropriate tracers in humans for better understanding of pathophysiology of cerebral plasticity.

The directions for future research on cochlear implantation are summarized by the NIH Consensus Statement (1): Research must attempt to explain the wide variation in performance across individual cochlear implant users. New tools, such as functional imaging of the brain, might be applied to unexplored variables—for example, the ability of the implant to activate the central auditory system. Investigations of the role of higher level cognitive processes in cochlear implant performance are needed.

ACKNOWLEDGMENTS

This commentary is dedicated to Dr. Roland Felix on the occasion of his 65th birthday. The authors thank Petra Rautenberg for excellent assistance in preparing this manuscript.

Michael Cordes, MD
St. Theresien-Krankenhaus
Nuremberg, Germany

Zbigniew K. Wszolek, MD
Mayo Clinic
Jacksonville, Florida

REFERENCES


1442 The Journal of Nuclear Medicine • Vol. 44 • No. 9 • September 2003