Strokes and Behavior: Disorders of Higher Cortical Functions Following Cerebral Disease

Disorders of Language and Related Functions

François Boller, M.D.

IN ADDITION to motor and sensory deficits, strokes may produce impairment of language, of visual-spatial perception, and of other aspects of cognitive behavior known collectively as "higher cortical functions." These disorders are of interest to the physician because they are helpful in localizing the site of a cerebral lesion. Knowledge of these disorders is necessary for evaluation of a patient's level of functioning, for selection of appropriate rehabilitation, and for determining prognosis. Furthermore, the cognitive disorders produced by strokes may be considered "experiments of nature" which offer fascinating insight into the organization of the central nervous system and so-called "brain and behavior relationships." The cerebral lesions induced by tumors, infections, and trauma are often less clearly localized than some vascular lesions. Strokes, therefore, represent the best pathological model from which one can infer the organization of behavior.

The various aspects of higher cortical functions affected by strokes include 1) attention and consciousness, 2) language and related functions, 3) visual-spatial perception and constructional abilities, 4) memory, 5) intelligence, and 6) emotional behavior and personality. These are also the steps usually followed by a mental status examination.1 This article will deal with aphasia, apraxia, and related disorders.

Aphasia

Broca's observation 120 years ago that strokes affecting the left hemisphere may produce aphasia marked the birth of a new branch of science, known today as neuropsychology. The hemisphere dominant for speech is the left hemisphere, not only in almost every right-handed person, but also in the majority of left-handed persons. The language areas of that hemisphere are supplied by the middle cerebral artery. It is estimated that up to 40% of all stroke victims suffer aphasia, which includes disorders of writing, reading, calculating, performing, and understanding gestures. Recent reviews4,5 consider these features more comprehensively than is appropriate here.

Aphasia may be defined as a disorder of previously intact language occurring as a result of brain damage. It must be emphasized that all aspects of symbolic communication tend to be affected in aphasia and that aphasic patients have trouble expressing themselves not only orally but also in writing and even with gestures. Similarly, both auditory comprehension and reading are usually impaired.

The best way of diagnosing aphasia is by listening to the patient while taking the history and performing simple tests of naming, repetition, comprehension, writing, and reading.2 Many formal tests have been developed to diagnose the presence of aphasia and quantify its severity. The token test4 is widely used because it is easy to administer and to score and appears to be highly reliable.

Since stroke victims usually show more impairment in some channels of communication than in others, aphasic patients differ considerably from one another. The variety of clinical pictures has led to classifications of aphasic syndromes that are sometimes confusing.

It has been known for many years that aphasia can be divided into nonfluent and fluent types just by listening to the patient's speech output. This distinction remains valid today and is important for several reasons. Ordinarily, nonfluent aphasics tend to have lesions located in the anterior, or pre-rolandic part of the brain, while fluent aphasics suffer lesions in the posterior, or retro-rolandic area. In addition, patients with nonfluent aphasia often seem to be unable or unwilling to talk, write, or communicate in any way and are therefore referred to as "inhibited," while patients with fluent aphasia may talk more than normal and appear to be "disinhibited." This distinction is important for understanding some aspects of the functional organization of the brain. In addition, the rehabilitative approach to an inhibited patient is quite different from that to a disinhibited patient (see the section on Therapy). In practical terms the distinction between nonfluent and fluent aphasia offers a basis for a relatively simple classification of aphasic syndromes.

The differentiation between nonfluent and fluent...
aphasia is based on several characteristics of conversational speech. Patients with nonfluent aphasia have a decreased rate of output, altered rhythm, and difficulty with articulation (dysarthria). Phrase length (number of words between pauses) is decreased, and utterances are spoken with obvious effort, although the few words produced may convey a considerable amount of information. In contrast, fluent aphasics have a normal or sometimes even an increased rate of output. Their effortless speech occasionally comes out in inappropriate abundance ("press of speech") but with normal rhythm and articulation. Phrase length is normal or increased. Inappropriate substitutions of one word for another ("verbal paraphasias") are common and at times "made-up words" (neologisms) may appear. In contrast to the nonfluent, fluent aphasics may convey very little information despite their considerable verbal output.

Nonfluent Aphasias

Broca’s (Motor) Aphasia. This form of aphasia is characterized by reduced (non-fluent) verbal output in both speech and writing. Comprehension often appears intact or nearly intact on informal examination although it is rarely perfect, as shown by the poor performance of these patients when asked to carry out complex commands requiring a succession of responses. Written questions and commands are particularly likely to evoke defective responses. In some cases comprehension is more severely impaired, so there may be considerable overlap between this form of aphasia and global aphasia. Not infrequently, patients who initially present with global aphasia later improve in their ability to understand and to talk and, therefore, evolve into a typical Broca’s aphasia.

In addition to right hemiparesis, patients with Broca’s aphasia frequently also have difficulty with gestural behavior, particularly that involving oral and facial muscles (buccofacial apraxia). “Broca’s area” classically corresponds to the pars opercularis of the third frontal convolution (Brodmann’s area 44). It has been shown, however, that many patients with Broca’s aphasia have a lesion extending well beyond the limits of “Broca’s area.” The extent and location of the lesions are important from the point of view of prognosis: it has been stated that patients with lesions limited to Broca’s area have a rapid recovery. About 20% of all patients with aphasia have Broca’s aphasia.

Global Aphasia. This aphasia produces the most severe deficit. Patients with global aphasia show severe impairment of all linguistic functions. They say little, understand only a few questions and commands, and repeat only the simplest sounds. Reading, writing, and gestural behavior are usually impaired. A right hemiplegia is the rule. It is not unusual to observe this condition initially, followed by a residual picture of another type of aphasia (as, for example, Broca’s aphasia).

The lesion usually involves a considerable portion of the territory of the middle cerebral artery, particularly the perisylvian area. Global aphasia is the most common type of aphasia (20 to 25%).

Transcortical Motor Aphasia. Patients with this syndrome have markedly reduced output with relatively intact comprehension. Naming is also quite good; repetition is practically perfect. Reading is preserved, but writing is nearly always impaired. In the patient whose reduced output is linguistically correct and who can name, understand, repeat, and read quite well, it is doubtful that the disorder is an aphasia. The lesion is in fact often located outside the classic “language area” and may involve the supplementary motor cortex or the dominant hemisphere. Although uncommon, it may be seen in patients with strokes affecting the territory of the anterior with greater weakness in the lower limb and shoulder than in the forearm and hand.

Fluent Aphasias

Wernicke’s (Sensory) Aphasia. In Wernicke’s aphasia, output is typically fluent but at times is incomprehensible (“jargon”). Comprehension, as a rule, is severely impaired, but some patients with severe jargon aphasia can show good performance on tests of auditory comprehension. The typical patient, when questioned or given commands, usually stops talking and appears to listen to what he is told, only to fall back into a jargon that often bears no apparent relationship to the examiner’s words.

These patients may have no motor deficit or only a mild right hemiparesis. They are usually unable to read either aloud or for comprehension. Wernicke’s area is considered to be located in the posterior part of the left superior temporal gyrus (Brodmann’s area 22). Here again, it is the rule rather than the exception for lesions to extend beyond this area. Wernicke’s aphasia comprises about 15% of the aphasic population. Recent observations suggest that the incidence of Wernicke’s aphasia increases steadily with age, while the incidence of all other clinical types of aphasia increases to a peak between ages 52 and 57 and then declines steadily with increasing age.*

Conduction Aphasia. Patients with conduction aphasia understand almost everything but, when attempting to talk, have striking difficulty in uttering the right sounds, as if they were groping for them through successive approximations. In addition, these patients repeat phrases or sentences poorly. Their writing is usually quite impaired while reading ability varies. Buccofacial apraxia (see below) is frequent. These patients are not usually hemiplegic but may have a right homonymous hemianopsia. The lesion may be located in the paths connecting Wernicke’s and Broca’s area. Diagnosis is often based on a discrepancy between relatively preserved comprehension and clearly impaired repetition. Conduction aphasia constitutes up to 10% of aphasic patients.

Anomic Aphasia. In this syndrome, also called amnestic aphasia, the main disturbance consists of difficulties in naming on confrontation and in word finding in spontaneous speech. The naming and word-
finding problems occur with a wide variety of word types and must be distinguished from problems in naming specific types of stimuli, such as colors or other visual or tactile stimuli. Nonaphasic misnaming may occur with diffuse or generalized neurological involvement as well. True anomia is present in about 5% of the total number of cases of aphasia.

Transcortical Aphasia. In this syndrome repetition is good, but comprehension is impaired. In extreme cases repetition is the only remaining language function, and there may be "echolalia" where patients meaninglessly repeat the words addressed to them. This severe form has also been called "isolation of the speech area." The name transcortical sensory aphasia indicates a subtype of fluent aphasia in which comprehension is clearly worse than repetition. This syndrome is found in about 2% of aphasic patients.

Other Syndromes

Alexia Without Agraphia. Reading difficulties are frequently found in aphasia and may take different forms according to the site of the hemispheric lesion. True alexia without agraphia, or pure word blindness, is a rare but particularly interesting form of reading disorder. In the classic form patients have no disorder of spoken language and can write normally but are unable to read even the material they may have just written. Difficulties in object naming are often found in the initial stage. These patients usually have a right homonymous hemianopsia. Lesions are found in the left occipital lobe and in the splenium of the corpus callosum.

Word Deafness. Also rare is pure word deafness, in which patients present with a striking inability to understand spoken language despite normal hearing ability. Oral expression, writing, reading aloud, and reading comprehension show little or no abnormality. The lesion may be found only in the left temporal lobe, although usually both temporal lobes show disease.

Therapy For Aphasia

There is considerable controversy about the efficacy of aphasia therapy, which includes sessions between therapist and patient, programmed instruction, and use of non-verbal symbols. Prognosis is affected by the nature of lesion, age of onset, tendency to left-handedness, and "attitude" of the patient. In non-fluent patients efforts are made to stimulate the patients and encourage them to speak more. Fluent patients instead are encouraged to be more critical of their own verbal production. Recent studies have shown that aphasics who receive specific language therapy show a significantly greater improvement than controls.

Apraxia

Apraxia may be defined as an inability to carry out motor activities in the presence of an intact motor and sensory system and normal comprehension, attention, and cooperation. Patients with ideomotor apraxia are unable to carry out such actions as making a fist, or brushing their teeth on command, although they have no trouble carrying out the same tasks spontaneously. Patients in whom movements of the oral and perioral muscles, such as sticking out the tongue or pretending to sip from a straw, are specifically affected are said to have buccofacial apraxia. Apraxia is thought by Geschwind to be another example of a disconnection syndrome that interrupts the pathways connecting receptive with motor areas. Apraxia is frequent in aphasic patients but may occur as an isolated phenomenon.

Other disorders directly or indirectly related to aphasia may be found in stroke patients but are not discussed here for lack of space. These include disorders of calculation ability, the Gerstmann syndrome (consisting of finger agnosia, right-left disorientation, agraphia, and acalculia), and auditory and visual agnosia.

Acknowledgment

This project was supported in part by the General Medical Research Service of the Veterans Administration.

References


Strokes and behavior: disorders of higher cortical functions following cerebral disease.
Disorders of language and related function.
F Boller

doi: 10.1161/01.STR.12.4.532

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://stroke.ahajournals.org/content/12/4/532.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Stroke can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Stroke is online at:
http://stroke.ahajournals.org/subscriptions/