Introduction to disorders of higher cortical function

We have seen that relatively restricted focal lesions can produce discrete changes in sensory and motor system functioning. We will now begin to illustrate that focal lesions also can produce discrete changes in cognitive-intellectual functions as well. Disorders of higher cortical function can be quite difficult to localize and the terminology employed to describe these conditions can be quite confusing. You need only be familiar with a small number of conditions. We will discuss disorders of language (aphasia), recognition (agnosia), and complex movement (apraxia). You will see that diagnosing a focal disorder of higher cognitive function requires considerable effort to insure that the symptoms are not arising on another basis. This suggests that our ability to measure these functions is much less precise and direct than our ability to study clinical phenomena involving sensory and motor systems.

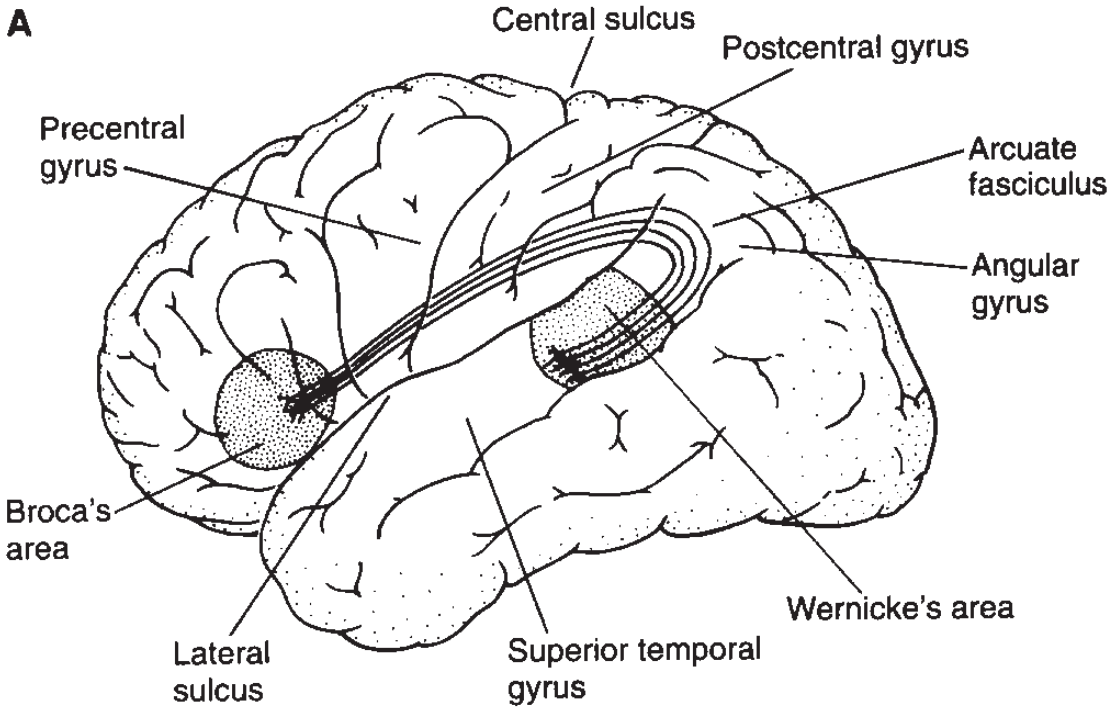
While it is beyond the scope of this course to discuss the cerebral organization of cognitive-intellectual faculties in depth, several points are worth noting. As will shortly become evident, the anatomy of higher cortical faculties is only modestly understood such that the pathways and networks subserving these functions cannot be specified with the same degree of detail and precision that you have encountered in sensory and motor systems. What is known is largely learned through the study of individuals with lesions (e.g. stroke, tumor, trauma). These lesions seldom are discrete, and therefore the effects they produce tend to be diffuse such that multiple cognitive abilities are often disrupted or impaired at once. The diffuse effects of most cortical lesions makes it very challenging to study the localization of specific mental abilities.

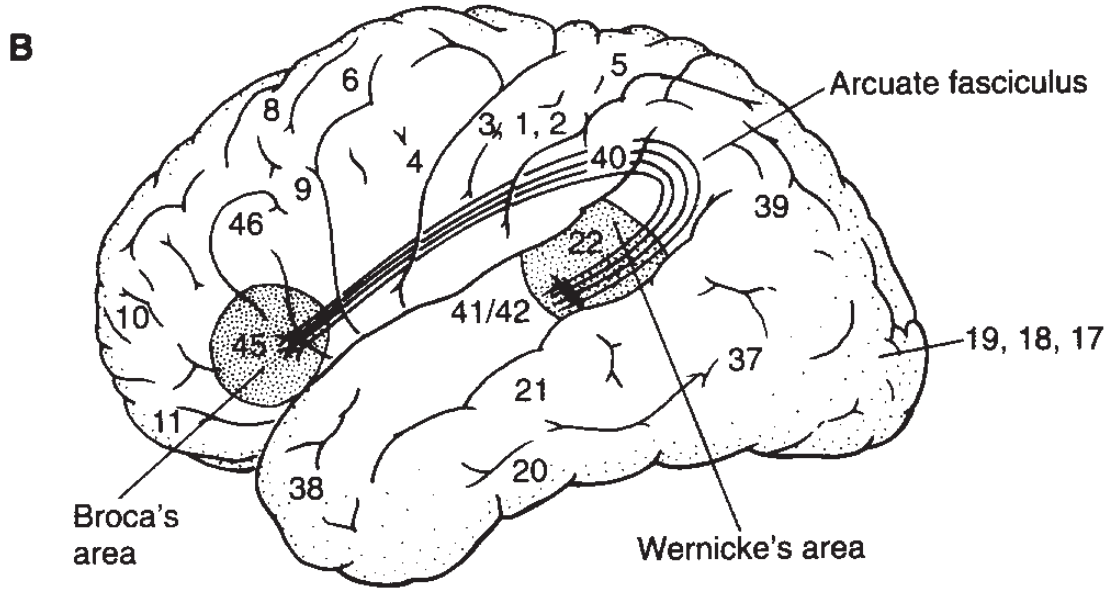
APHASIA. Aphasia refers to a disturbance in one or more of the many facets of language produced by injury to certain parts of the brain specialized for these functions. Disturbances of language usage that are due to paralysis or incoordination of the musculature of speech or writing, or to poor vision or hearing, or to severe generalized intellectual impairment are not, of themselves, aphasia. Patients with aphasia may or may not have additional sensory or motor deficits, or other cognitive-intellectual deficits. The modalities of language include written and spoken language, comprehension, and reading.

There are many nomenclature systems for classifying the aphasias. No one system is universally accepted. For purposes of this course, we will adopt the most simple distinction. Aphasias are often categorized as either FLUENT or NONFLUENT contingent upon the ease in which words are articulated.

NONFLUENT APHASIA. In 1861, Paul Broca, a French surgeon, observed a patient suffering from right hemiplegia and loss of speech and writing. The patient was able to understand speech, but he could articulate only one word “tan.” This patient died, and postmortem inspection of the brain revealed that there was a cavity filled with fluid on the lateral aspect of the left hemisphere. When the fluid was drained, a large left hemisphere lesion could be seen which included the first temporal gyrus, the insula and corpus striatum, and the frontal lobe, including the second and third frontal convolutions as well as the inferior portion of the transverse convolution.

Today, the condition this patient demonstrated is known as BROCA'S APHASIA, OR NONFLUENT APHASIA. The essential characteristics include: awkward articulation, restricted vocabulary, restriction of grammar to the simplest, most over-learned forms, and relative preservation of auditory comprehension. Utterances often are restricted to short telegraphic phrases consisting of nouns and verbs only, or even to single words. Reading is affected only mildly, while written language is usually impaired at least as severely as spoken language. Nonspeech oral movements often are affected. Often the patient has awareness of the deficit.





This form of aphasia arises from a lesion involving the third frontal convolution of the left hemisphere, the subcortical white matter, and extending posteriorly to the inferior portion of the motor strip (prerolandic gyrus). The left cerebral hemisphere is dominant for language in the vast majority of left and right handed individuals. The right hemisphere is estimated to be dominant for language in only 1 to 2% of the non-neurologically compromised population.

FLUENT APHASIA. In 1874, Karl Wernicke noted that lesions of the posterior portion of the left superior temporal region produced an aphasia in which comprehension was poor. The critical features of this syndrome included impaired auditory comprehension and fluent, easily articulated but paraphasic speech. Paraphasia refers to the production of unintended syllables, words or phrases during effort to speak. The two most frequently occurring paraphasias are termed LITERAL and SEMANTIC paraphasias. Literal paraphasias consist of pronunciation of the syllables of a word in the wrong order or with additional, unintended sounds. For example, “pipe” may become “Hike…no, pike…pipe!” Semantic paraphasias arise when a related but unintended word is inadvertently substituted for the intended word (e.g. “my boss” in place of “my teacher”). Because these patients’ speech generally is fluent, Wernicke’s aphasia often is referred to as FLUENT APHASIA. The impairment in auditory comprehension may be evident even at the level of one word. The patient may repeat the examiner’s words uncomprehendingly, or with paraphasic distortions. Reading generally is poor as is writing. Disorders of reading are called DYSLEXIA, while disorders of writing are termed DYSGRAPHIA. The individual may have little awareness of the condition, or may make light of the condition and evidence little or no frustration. Hemiparesis is unlikely because the lesion is in the posterior portion of the superior temporal gyrus.

Characteristics of Fluent & Nonfluent Aphasia

	Nonfluent (Broca’s)	Fluent (Wernicke’s)
Speech	awkward	easily articulated
Fluency	nonfluent	fluent paraphasic
Grammar	restricted	normal
Auditory comprehension	preserved	impaired
Reading impairment	mild	minimal
Written language	impaired	impaired
Awareness of deficit	present	limited
Hemiplegia	usually	rare

Cases of pure aphasia without sensory or motor loss, and without dysfunction in other cognitive-intellectual areas are rare because lesions usually do not respect anatomical boundaries. Thus, there is considerable variability in presentation across patients. Like other medical conditions, these cognitive disorders also evolve across time and occasionally largely resolve.

AGNOSIA Aphasia is a quite common neurological symptom, while agnosia is relatively rare. Classically, agnosia is defined as a disorder of recognition that cannot be attributed to elementary sensory defects, general mental deterioration, attentional disturbances, aphasic disturbance, or unfamiliarity with the presented stimuli. Agnosias most often are modality specific; the patient fails to recognize material presented in a specific sensory channel (e.g. vision) but is successful when allowed to experience the stimuli in another channel (e.g. tactile, auditory). Thus, a patient who does not recognize a hammer by sight may correctly label it when allowed to handle it. Visual, auditory and tactile agnosias have received the most attention

AGNOSIA

	Functional deficit	Anatomy
Visual agnosia		
a) visual form agnosia	May include inability to recognize objects and body parts but can recognize same when touched	case reported after accidental carbon monoxide intoxication that produced bilateral damage to visual association cortex (Areas 18 and 19)
b) prosopagnosia	limited to inability to recognize familiar faces yet know they are seeing a face	Bilateral occipitotemporal gyrus
Auditory agnosia		
pure word deafness / auditory agnosia for speech	unable to comprehend spoken language but can read, write and speak normally and comprehension of nonoral sound is intact	Bilateral symmetric cortical-subcortical lesions of anterior portion of superior temporal gyri, sparing Heschl's gyrus
Somatosensory agnosia		
asteroagnosia / tactile form agnosia	inability to recognize an object by touch alone, but can identify with sight	Temporal-parietal junction?

The neuroanatomy of agnosias are not well understood and the nomenclature is exceedingly lengthy and complex. One form of visual agnosia is termed prosopagnosia. Patients with this disorder may perform normally on face discrimination and face matching tasks, but are completely unable to identify whose face they are viewing when the face previously was well known to them. The lesions causing this condition typically are bilateral and involve the cortex and the white matter in the occipitotemporal gyrus. Like other agnosias, in rare cases in which the lesion for this disorder is unilateral, it tends to be restricted to the right hemisphere.

APRAXIA This condition arises when there is an inability to perform a complex motor activity even though underlying essential motor and sensory functions are intact and the patient is able to understand the presented request. Essentially, apraxias are disorders in which there is a loss of ability to perform an earlier learned skilled movement such as cutting with a knife, turning a door knob, buttoning a shirt, blowing out a candle, or sealing an envelope. Most of these patients also have aphasia. Apraxia is common in patients with focal lesions and in patients with degenerative dementias, but patients rarely spontaneously complain of this disturbance and they often appear unaware of their deficit. Testing involves requesting patients to perform actions using pretend or actual objects, or through imitation, again with or without the object present.

APRAXIAS

	Performance impairment	Anatomy
Ideational	reflects a conceptual deficit in which incorrect object is used to perform correct action	likely dysfunction in region of left temporal-parietal junction is most responsible
Ideomotor	inability to execute motor sequence such as inserting key and turning to unlock door due to sequencing errors or perseveratively performing prior action	lesion in circuit that includes inferior left parietal lobe and supplementary motor cortex

Patients with ideomotor apraxia have normal dexterity but have great difficulty demonstrating how to use a tool or instrument, particularly when the instrument is not present. The deficit may be elicited by asking a patient to show you how to use a real or imaginary hammer. Patients with this form of apraxia tend to make perseverative and/or sequencing errors. Perseverative errors refer to the patient continuing to perform a previously requested or executed movement. Sequencing errors arise when more than one movement is necessary to execute a command. For example, using a key to unlock a door requires inserting the key in the lock and then turning the key. The patient making sequencing errors will reverse this sequence. Other errors include using a body part as an object. For example, rather than pretending to hold an object to perform an action, such as using a key to open a door, the patient acts as if his or her hand is the key. The lesions underlying this disorder are not well understood. Again this is because the performance of skilled movements to command actually involves a multiplicity of activities, and performance impairments may arise from disruption anywhere in the often extensive circuits. The network may include the left inferior parietal lobe that contains the representations of skilled movements, the supplementary motor area and basal ganglia that are important in transcoding these representations into a motor program, and the motor cortex, which implements these programs. Lesions of any of these areas, except the motor cortex, may produce ideomotor apraxia, as may lesions which disconnect these areas from one another and from the motor cortex that controls limb movement.

The terms ideational apraxia and conceptual apraxia sometimes are used interchangeably to refer to the conceptual errors that some patients make when performing complex actions. An example is that a patient may use a brush to perform the actions of a fork, yet be able to tell you when asked that the tool being held is indeed a brush. A more complex and traditional illustration of an ideational apraxia is the patient who in making coffee using a drip coffee maker cannot understand that it is necessary to put the coffee grounds in a filter rather than directly in the water. A distinction between ideational/conceptual apraxia and ideomotor apraxia is that the performance of a patient with ideomotor apraxia improves when instead of imitating an action the patient uses an actual object, whereas in conceptual/ideational apraxia, the use of real objects does not enhance performance. Another difference is that some patients with ideational/conceptual apraxia perform the correct motor behavior but they simply use the wrong tool whereas the deficit for those with ideomotor apraxia is in the performance of the motor sequence.