simplest sentence because the visual information from the written words (presented to the intact left visual hemifield) cannot reach the language network. Objects in the left hemifield may be named accurately because they activate nonvisual associations in the right hemisphere, which in turn can access the language network through transcallosal pathways anterior to the splenium. Patients with this syndrome also may lose the ability to name colors, although they can match colors. This is known as a *color anomia*. The most common etiology of pure alexia is a vascular lesion in the territory of the posterior cerebral artery or an infiltrating neoplasm in the left occipital cortex that involves the optic radiations as well as the crossing fibers of the splenium. Because the posterior cerebral artery also supplies medial temporal components of the limbic system, a patient with pure alexia also may experience an amnesia, but this is usually transient because the limbic lesion is unilateral.

Apraxia and Aphemia *Apraxia* designates a complex motor deficit that cannot be attributed to pyramidal, extrapyramidal, cerebellar, or sensory dysfunction and that does not arise from the patient's failure to understand the nature of the task. *Apraxia of speech* is used to designate articulatory abnormalities in the duration, fluidity, and stress of syllables that make up words. Intoning the words may improve articulation. It can arise with CVAs in the posterior part of Broca's area or in the course of frontotemporal lobar degeneration (FTLD) with tauopathy. *Aphemia* is a severe form of acute speech apraxia that presents with severely impaired fluency (often mutism). Recovery is the rule and involves an intermediate stage of hoarse whispering. Writing, reading, and comprehension are intact, and so this is not a true aphasic syndrome. CVAs in parts of Broca's area or subcortical lesions that undercut its connections with other parts of the brain may be present. Occasionally, the lesion site is on the medial aspects of the frontal lobes and may involve the supplementary motor cortex of the left hemisphere. *Ideomotor apraxia* is diagnosed when commands to perform a specific motor act ("cough," "blow out a match") or pantomime the use of a common tool (a comb, hammer, straw, or toothbrush) in the absence of the real object cannot be followed. The patient's ability to comprehend the command is ascertained by demonstrating multiple movements and establishing that the correct one can be recognized. Some patients with this type of apraxia can imitate the appropriate movement (when it is demonstrated by the examiner) and show no impairment when handed the real object, indicating that the sensorimotor mechanisms necessary for the movement are intact. Some forms of ideomotor apraxia represent a disconnection of the language network from pyramidal motor systems so that commands to execute complex movements are understood but cannot be conveyed to the appropriate motor areas. *Buccofacial apraxia* involves apraxic deficits in movements of the face and mouth. *Limb apraxia* encompasses apraxic deficits in movements of the arms and legs. Ideomotor apraxia almost always is caused by lesions in the left hemisphere and is commonly associated with aphasic syndromes, especially Broca's aphasia and conduction aphasia. Because the handling of real objects is not impaired, ideomotor apraxia by itself causes no major limitation of daily living activities. Patients with lesions of the anterior corpus callosum can display ideomotor apraxia confined to the left side of the body, a sign known as *sympathetic dyspraxia*. A severe form of sympathetic dyspraxia, known as the *alien hand* syndrome, is characterized by additional features of motor disinhibition on the left hand. *Ideational apraxia* refers to a deficit in the sequencing of goal-directed movements in patients who have no difficulty executing the individual components of the sequence. For example, when the patient is asked to pick up a pen and write, the sequence of uncapping the pen, placing the cap at the opposite end, turning the point toward the writing surface, and writing may be disrupted, and the patient may be seen trying to write with the wrong end of the pen or even with the removed cap. These motor sequencing problems usually are seen in the context of confusional states and dementias rather than focal lesions associated with aphasic conditions. *Limb-kinetic apraxia* involves clumsiness in the use of tools or objects that cannot be attributed to sensory, pyramidal, extrapyramidal, or cerebellar dysfunction. This condition can emerge in the context of focal premotor cortex lesions or *corticobasal degeneration*.

Gerstmann's Syndrome The combination of *acalculia* (impairment of simple arithmetic), *dysgraphia* (impaired writing), *finger anomia* (an inability to name individual fingers such as the index and thumb), and *right-left confusion* (an inability to tell whether a hand, foot, or arm of the patient or examiner is on the right or left side of the body) is known as Gerstmann's syndrome. In making this diagnosis, it is important to establish that the finger and left-right naming deficits are not part of a more generalized anomia and that the patient is not otherwise aphasic. When Gerstmann's syndrome arises acutely and in isolation, it is commonly associated with damage to the inferior parietal lobule (especially the angular gyrus) in the left hemisphere.

Pragmatics and Prosody *Pragmatics* refers to aspects of language that communicate attitude, affect, and the figurative rather than literal aspects of a message (e.g., "green thumb" does not refer to the actual color of the finger). One component of pragmatics, *prosody,* refers to variations of melodic stress and intonation that influence attitude and the inferential aspect of verbal messages. For example, the two statements "He *is* clever." and "He is *clever*?" contain an identical word choice and syntax but convey vastly different messages because of differences in the intonation with which the statements are uttered. Damage to right hemisphere regions corresponding to Broca's area impairs the ability to introduce meaning-appropriate prosody into spoken language. The patient produces grammatically correct language with accurate word choice, but the statements are uttered in a monotone that interferes with the ability to convey the intended stress and affect. Patients with this type of *aprosodia* give the mistaken impression of being depressed or indifferent. Other aspects of pragmatics, especially the ability to infer the figurative aspect of a message, become impaired by damage to the right hemisphere or frontal lobes.

Subcortical Aphasia Damage to subcortical components of the language network (e.g., the striatum and thalamus of the left hemisphere) also can lead to aphasia. The resulting syndromes contain combinations of deficits in the various aspects of language but rarely fit the specific patterns described in Table 36-1. In a patient with a CVA, an anomic aphasia accompanied by dysarthria or a fluent aphasia with hemiparesis should raise the suspicion of a subcortical lesion site.

Progressive Aphasias Aphasias caused by major cerebrovascular accidents start suddenly and display maximal deficits at the onset. These are the "classic" aphasias described above. Aphasias caused by neurodegenerative diseases have an insidious onset and relentless progression. The neuropathology can be selective not only for gray matter but also for specific layers and cell types. The clinico-anatomic patterns are therefore different from those described in Table 36-1.

CLINICAL PRESENTATION AND DIAGNOSIS OF PRIMARY PROGRESSIVE APHASIA (PPA) Several neurodegenerative syndromes, such as typical Alzheimer-type (amnestic) and frontal-type (behavioral) dementias, can also undermine language as the disease progresses. In these cases, the aphasia is an ancillary component of the overall syndrome. When a neurodegenerative language disorder arises in relative isolation and becomes the primary concern that brings the patient to medical attention, a diagnosis of PPA is made.

LANGUAGE IN PPA The impairments of language in PPA have slightly different patterns from those seen in CVA-caused aphasias. Three major subtypes of PPA can be recognized. The *agrammatic variant* is characterized by consistently low fluency and impaired grammar but intact word comprehension. It most closely resembles Broca's aphasia or anterior transcortical aphasia but usually lacks the right hemiparesis or dysarthria and has more profound impairments of grammar. Peak sites of neuronal loss (gray matter atrophy) include the left inferior frontal gyrus where Broca's area is located. The neuropathology is usually an FTLD with tauopathy but can also be an atypical form of Alzheimer's disease (AD) pathology. The *semantic variant* is characterized by preserved fluency and syntax but poor single-word comprehension and profound two-way naming impairments. This