Extrasylvian aphasias

Alfredo Ardila

Thumbnail
Language disturbances observed in cases of damage outside of the “brain language area” are known as extrasylvian or transcortical aphasia. Preserved language repetition is its distinguishing characteristic. Two major subtypes of extrasylvian aphasia have been traditionally distinguished: extrasylvian motor aphasia and extrasylvian sensory aphasia, each one presenting rather different clinical manifestations. Additionally, striatocapsular and thalamic damage can result not only in speech but also in some language defects. More recently, it has been hypothesized that the cerebellum contributes to cognitive processing, particularly the processing of linguistic information because the cerebellum has reciprocal connections not only with the left inferior frontal gyrus, but also the left lateral temporal cortex.

Key points
• Aphasia observed in cases of damage outside the “brain language area” (perisylvian area of the left hemisphere) is known as extrasylvian aphasia.
• Usually an extrasylvian motor aphasia and extrasylvian sensory aphasia are distinguished.
• Sometimes aphasia can be found associated with striatocapsular and thalamic pathology.
• Right cerebellar damage may lead to some verbal deficits, such as impaired grammar and decreased verbal fluency.

Historical note and nomenclature
Wernicke and Lichtheim introduced the term "transcortical aphasia" (Wernicke 1881; Lichtheim 1885). Wernicke initially described 2 distinct types of aphasia, motor and sensory, as well as the anatomical demonstration to support this division (Wernicke 1874). Wernicke's original proposal was expanded during the following years. Lichtheim, in his paper "On Aphasia," presented a simple diagrammatic illustration of Wernicke’s model of language functions (Lichtheim 1885). This model, usually known as the Wernicke-Lichtheim aphasia model, includes 7 different types of language disturbances associated with brain pathology. According to the model, aphasia can be subcortical, cortical, and transcortical. Each one includes 2 subtypes: motor and sensory. Finally, conduction aphasia represents a relatively isolated defect in language repetition, observed when the sensory language area becomes disconnected from the motor language area. Preserved language repetition represents the distinguishing characteristic of transcortical aphasias. The term "transcortical" represents one area of the Wernicke-Lichtheim aphasia model of the cortical networks involved in language and ideation, in which the exact phonological reproduction could be accomplished by the aphasic patient, but the meaning (idea) could not be extracted and reproduced.

Goldstein analyzed several varieties of aphasia gathered under the title of transcortical aphasia. The term, as originally proposed, draws a distinction between aphasias with or without repetition defects. Goldstein, in his book Die Transkortikalen Aphasien, noted that the term "transcortical" was archaic and misleading (Goldstein 1917). "Transcortical aphasia," however, remained the most widely used designation for a distinctive group of aphasic syndromes (Berthier 1999). The term "extrasylvian," instead of "transcortical," has been proposed to designate this group of language disorders (Benson and Ardila 1996). For the
Purpose of this summary, the term "extrasylvian" will also refer to aphasias that arise from injury to either subcortical areas or areas outside of the cerebral hemispheres (eg, the cerebellum). A description of perisylvian aphasias may be found elsewhere.

Frequency of transcortical aphasia is relatively low. In the Copenhagen aphasia study (Pedersen et al 2004), 270 acute stroke patients with aphasia (203 with first strokes) were included consecutively and prospectively from 3 hospitals in Copenhagen, Denmark. The assessment was repeated 1 year after stroke. The frequencies of the different types of aphasia in acute first stroke were as follows: global 32%, Broca 12%, isolation 2%, transcortical motor 2%, Wernicke 16%, transcortical sensory 7%, conduction 5%, and anomic 25%. One year after stroke, the following frequencies were found: global 7%, Broca 13%, isolation 0%, transcortical motor 1%, Wernicke 5%, transcortical sensory 0%, conduction 6%, and anomic 29%.

Clinical manifestations

Two major subtypes of extrasylvian (transcortical) aphasia have been traditionally distinguished: extrasylvian motor aphasia and extrasylvian sensory aphasia, each one presenting rather different clinical manifestations. In both major subtypes, however, language repetition is preserved. Occasionally, both subtypes appear simultaneously (mixed extrasylvian aphasia).

Extrasylvian motor aphasia. Different names have been applied to nonfluent aphasia with preserved repetition, including dynamic aphasia (Luria 1980), anterior isolation syndrome (Benson and Geschwind 1971), and transcortical motor aphasia. Goldstein noted that transcortical motor aphasia could be observed in 2 different circumstances: during recovery from Broca aphasia at a time when repetition had recovered better than spontaneous language and as a primary difficulty in the initiation of articulation (Goldstein 1948). The term transcortical motor aphasia has been used to refer to 2 different language disorders: lack of verbal initiative associated with left prefrontal pathology (Luria dynamic aphasia) and defects in language initiation observed in cases of damage in the left supplementary motor area (Ardila and Lopez 1984). Language and speech disturbances associated with frontal lobe pathology include: aphemia, Broca area aphasia, transcortical motor aphasia, mutism, and impoverished discourse. The initial mutism observed in cases of medial frontal pathology can be followed by language initiation disturbances associated with nearly normal repetition. This language defect corresponds to the aphasia of the left supplementary motor area (Alexander et al 1989). Two subtypes of extrasylvian motor aphasia have been proposed: subtype I (dynamic aphasia associated with left dorsolateral prefrontal pathology) and subtype II (supplementary motor area aphasia) (Benson and Ardila 1996).

Extrasylvian motor aphasia-type I. Extrasylvian motor aphasia-type I corresponds to Luria dynamic aphasia (Luria 1976). It is characterized by nonfluent verbal output, lack of verbal initiative, good comprehension, and good repetition of spoken language. Patients with this subtype of aphasia use as few words as possible, answer questions by reiterating many of the words and grammatical structures presented in the question (echolalia), and on occasion produce perseverative responses. Sentences tend to be started but not finished. Poor verbal fluency, impoverished narrative production, reduced use of complex and precise syntax, and poor inhibition of high-association responses have been described following left prefrontal damage (Luria 1976; Kertesz 1999). Patients with extrasylvian motor aphasia-type I perform series speech well once the series has been initiated. Recitation of nursery rhymes and naming the days of the week are often performed successfully if initiated by the examiner. Open-ended phrases are easily completed by these patients. Comprehension of spoken language is good, at least for conversational language; however, many patients have difficulty handling sequences of complex material and some show defects interpreting
relational words. Interestingly, despite preserved language understanding, patients with this type of aphasia have difficulties following verbal commands.

The difficulty of these patients to initiate a response is complicated by significant apathy, and behavioral withdrawal is usually observed. The patients seem distant and not interested in engaging in social conversation. Luria proposed that in dynamic aphasia, the patient’s behavior is not controlled by language, and the dissociation between language and overt behavior represents an executive control disorder impairing language at the pragmatic level. He further proposed that in dynamic aphasia there exists an inability to form a linear schema of the sentence (Luria 1980). Some authors have supposed that in dynamic aphasia a selective impairment of verbal planning occurs, particularly at the level of “macroplanning,” i.e., generating sequences of novel thoughts and ideas (Bormann et al 2008). Impairment in the hierarchical organization of knowledge specific to the task at hand has been pointed out (Gold et al 1997). Alexander suggested that this type of aphasia could be more accurately defined as an executive function disorder than an aphasia (Alexander 2006). He proposed that the progression of clinical disorders from aphasia to discourse impairments can be interpreted as a sequence of procedural impairments from basic morpho-syntax to elaborated grammar to narrative language, correlated with a progression of the focus of the damage from posterior frontal to polar, or lateral frontal to medial frontal, or both.

The ability of these aphasic patients to repeat utterances is unexpectedly good in dramatic contrast to their nonfluent spontaneous output. Although the patients often echo a word or phrase, they are usually not fully echolalic. The ability to name on confrontation is often limited. Both contextual and phonemic cues can help. Three types of errors are found in confrontation naming:

1. Perseveration: the patient continues giving a past response for a new stimulus.
2. Fragmentation: the patient responds to a single feature of the stimulus, not to the whole stimulus.
3. Extravagant paraphasias: instead of the target name, the patient presents a free-association answer that becomes an extravagant deviation.

Writing is almost always defective. The tendency is so marked that the term "agraphia without alexia" is an appropriate description. Sentences are incomplete, and the patients with extrasylvian motor aphasia type-II must be continuously encouraged to continue writing. Reading, on the other hand, is significantly preserved. This dissociation between reading and writing is understandable considering that reading, to a significant extent, represents a visual-perceptual ability, whereas writing is a motor production skill.

Patients with this type of aphasia usually have a good language repetition, and even echolalia can be observed. Nonetheless, under certain circumstances, the patients introduce some changes to the word or sentence to be repeated. If the word contains a phonological error (and, hence, it is a pseudo-word), then the patient will correct the error when repeating. If a low probability word is presented, then the patient may replace it for a similar but more probable word. If a sentence containing a grammatical error is presented to the patient, then the patient will correct the error when repeating it. If the sentence is absurd or it has an extremely low probability, then the patient will make the sentence more probable and correct it (Ardila and Rosselli 1992). In patients with frontal pathology, this tendency to correct errors is observed not only in spoken but also in written language. Written pseudo-words are frequently read as real words.

Neurologic findings in this type of extrasylvian motor aphasia are variable. Hemiparesis is uncommon. Pathological reflexes involving the dominant limb are often present. Both conjugate deviation of the eyes and unilateral inattention have been recorded in the initial stages in some cases of dynamic aphasia. Damage is expected to involve Brodmann area 45, which is situated in front of Broca’s area, and adjacent brain areas (Rapcsak and Rubens 1994).
Extrasylvian motor aphasia-type II. This type of aphasia is characterized by a nonfluent output but good comprehension and repetition of spoken language. It has been referred to as transcortical motor aphasia (Kertesz 1979), the syndrome of the anterior cerebral artery (Rubens 1975), and supplementary motor area aphasia (Ardila and Lopez 1984; Alexander et al 1989). These patients present a disordered speech and language that is characterized by delayed articulatory initiation and disturbed clarity of speech, in contrast to their nearly normal output in repetition and essentially normal comprehension and naming. They show neither echolalia nor the forced-completion phenomenon. Reading aloud may be near-normal, but reading comprehension is usually limited (Rubens 1975). Inability to read letter-by-letter in meaningful words has been described (Ardila and Lopez 1984). Writing is slow, laborious, and incomplete, and shows occasional paragraphias.

Three major differences separate this type of extrasylvian motor aphasia from the first subtype: (1) associated neurologic findings, (2) locus of brain pathology, and (3) the patient’s behavior during verbal communication.

The associated neurologic findings are more evident in the lower extremities. Weakness, hyperreflexia, contralateral extensor toe signs, and sensory loss involve the lower extremities, but basically normal arm, hand, and face strength is characteristic. If midline frontal damage is extensive, the patient may show incontinence and a right grasp reflex.

The locus of the damage is located in the left supplementary motor area. The supplementary motor area is located outside the classical language area (left perisylvian area), and aphasia seems unexpected in cases of supplementary motor area damage. However, some vague mention of language disturbances and even mutism associated with pathology in the supplementary motor area is found in the neurologic literature since the beginning of the century. Nonetheless, only after demonstrating that electrical stimulation of this area produced speech arrest was a role in language ascribed to the supplementary motor area (Penfield and Roberts 1959). This brain area is known to receive considerable input from the basal ganglia and anterior thalamus. It has efferents to the caudate and dorsolateral frontal convexity. The adjacent cingulate gyrus appears to play a role in motivation and motor initiation tasks. The initial mutism followed by delayed and impoverished responses during recovery may be interpreted on the basis of disturbances to a key motor activation center.

The patient’s behavior during verbal communication is strikingly different from the behavior observed in dynamic aphasia. Although apathy and lack of interest in verbal communication is found in dynamic aphasia, patients with this second type of extrasylvian motor aphasia make a tremendous effort to speak. Nonetheless, they fail in initiating language, but once language is initiated or prompted by the examiner, they can complete the word or the sentence. When attempting to speak, a long delay may take place, but during the delay some movements of the lips and even panting can be observed.

Usually, recovery in this type of extrasylvian motor aphasia is surprisingly good. Long-lasting impaired performance in the verbal fluency tasks, however, has been observed (Deblieck et al 2003). It has been suggested that this language disorder is a pure motor disturbance that does not involve language function; therefore, it does not deserve inclusion as a type of aphasia. Nonetheless, it involves a significant disturbance in the use of language (pragmatic language function), and most authors include it among the aphasic syndromes.

Extrasylvian sensory aphasia. In the Lichtheim-Wernicke aphasia model, extrasylvian (transcortical) sensory aphasia was interpreted as a disconnection of the sensory (auditory) language area from a "concept area." Repetition and echolalia as the cardinal features of this aphasia have been emphasized (Goldstein 1917; Berthier 1999). Conversational language in extrasylvian sensory aphasia is fluent, often featuring paraphasias (especially neologic and semantic substitutions) and emptiness in content. Preserved ability to repeat is surprising in this context. Repetition sometimes is a true
echolalia. Frequently, these patients will incorporate words and phrases uttered by the examiner into the ongoing output when apparently failing to understand the meaning of the words (Berthier 1999). Unlike the echolalia observed in dynamic aphasia, these patients will echo incorrect syntactical structures, nonsense words, and even foreign phrases. The verbal output is abundant. Once started, extrasylvian sensory aphasia patients can count, recite the days of the week, and show a strong completion phenomenon for poems and statements. Although their serial speech is good, the patient cannot perform these tasks on command (Goodglass 1993).

Language comprehension is severely abnormal, often to the point of total incomprehension, and stands in evident contrast to the ease with which they reiterate the examiner’s statements. Naming is also seriously defective in these patients. Characteristically, extrasylvian sensory aphasia patients neither name an object nor identify it when the name is presented by the examiner. Surprisingly, however, the ability to read aloud may be preserved, even though paralexic errors are often observed. Reading comprehension, on the other hand, is severely defective. Writing is usually defective and, in general, resembles the disturbance in written language observed in Wernicke aphasia patients.

Extrasylvian sensory aphasia patients usually have no associated motor deficits. Sensory abnormalities are often present. Some disorder of the right visual field is frequent, being either an inferior quadrantanopia or a complete hemianopia. Inattention is sometimes observed. Damage is found in the temporal-occipital and parietal-occipital areas (Rapcsak and Rubens 1994). It should be emphasized that, as many patients have no obvious elementary neurologic deficit, the potential for misdiagnosis is considerable. Patient language may be misinterpreted as a psychotic language.

It has been proposed that 2 subtypes of extrasylvian sensory aphasia can be distinguished. The postulated distinction has been referred to as extrasylvian sensory aphasia-type I and extrasylvian sensory aphasia-type II (Benson and Ardila 1996).

**Extrasylvian sensory aphasia-type I.** The symptom cluster of this first type of extrasylvian sensory aphasia resembles the language features described as amnesic aphasia and the first subtype of transcortical sensory aphasia (Kertesz 2006). Spontaneous language is fluent, speech comprehension is poor, and repetition is normal. Conversational language tends to be contaminated with semantic paraphasias and sometimes neologicistic substitutions. Echolalia is common. Naming is strongly impaired, and nouns are produced at the most non-specific levels (eg, “animal” instead of “bird” or “robin”) (Martensson et al 2013). Reading aloud may be preserved, but reading comprehension is defective. Writing is defective to a variable degree.

The presence of semantic paraphasias and the helpfulness of phonemic prompting during naming tests suggest that the word itself may not be lost to the patient, but it cannot be retrieved in confrontation naming tasks. For these patients, a visually presented object does not trigger the appropriate word response and, conversely, a spoken word does not evoke a visual image (eg, when asking the patient to draw the meaning of a word, such as a "house" or "dog"). The patient is unable to retrieve meanings for words he or she can easily repeat. For these patients, the visual percept is disconnected from its lexical representation. Visual agnosia and right visual-field impairments may be present. The basic word-finding defects observed in this form of aphasia are identical to the word selection anomia suggested by some authors (Benson 1988), but they tend to be severe and accompanied by considerable comprehension problems.

The brain damage is usually located at the junction of the temporal, parietal, and occipital lobes (roughly, the lower angular gyrus and Brodmann area 37).

**Extrasylvian sensory aphasia-type II.** Extrasylvian sensory aphasia-type II has the language features described by Kertesz as his second type of transcortical sensory aphasia
Damage to the angular gyrus is known to produce a verbal amnesic defect characterized by fluent output with few semantic paraphasias, variable ability to comprehend spoken language, and excellent ability to repeat words and sentences coupled with a notable deficiency in word finding. Patients with this second subtype of extrasylvian sensory aphasia may produce empty speech due to the lack of significant words and remarkable number of circumlocutions. Reading and writing tend to be seriously impaired, and some or all components of the Gerstmann syndrome (right and left disorientation, finger agnosia, acalculia, and agraphia) will be present.

Extrasylvian sensory aphasia-type II resembles semantic aphasia. Semantic aphasia has been defined as an inability to simultaneously recognize the elements within a sentence (Head 1926). It has been proposed that patients with semantic aphasia have difficulties in the following types of language constructs (Luria 1976):

• Sentences with a complex system of successive subordinate clauses (e.g., "The person who came with the man who had the book is the doctor. Who is the doctor?").
• Reversible constructions, particularly of the temporal and spatial type (e.g., "The circle is above the triangle." may be indistinguishable from "The triangle is above the circle." due to the plausible reversibility of the objects in the sentences).
• Constructions with double negatives.
• Comparative sentences (e.g., "Elephants are bigger than dogs.").
• Passive constructions (e.g., "The earth is illuminated by the sun.").
• Constructions with attributive relations (e.g., "My brother's father" and "my father's brother" may erroneously be considered the same person).

Luria also stated that these spatial disorders not only incidentally accompany semantic aphasia but that semantic aphasia itself was a defect in the perception of simultaneous structures transferred to a higher symbolic level. Patients with semantic aphasia have difficulties integrating the elements of a sentence into a whole and fail to grasp the meaning of the relationship. They also have difficulties in naming and in understanding language, particularly if language includes elements of spatial nature. Although conversational language may be adequate for social purposes in these patients, their lexicon is notably limited.

It appears reasonably certain that left posterior parietal damage is associated with right and left disorientation, finger agnosia, agraphia, acalculia, anomia, and some degree of visuospatial language disturbance. If the damage extends anteriorly toward the temporal lobe, the finding of alexia with agraphia and a broader, more profound comprehension disturbance will be observed. If damage involves both of these areas plus the anterior-lateral occipital lobe, a clinical picture of classic extrasylvian (transcortical) sensory aphasia occurs. Boatman and colleagues induced transcortical sensory aphasia transiently by electrical interference during routine cortical function mapping in 6 adults. Transcortical sensory aphasia was associated with multiple posterior cortical sites, including the posterior superior and middle temporal gyri (Boatman et al 2000).

**Mixed extrasylvian aphasia.** A significant decrease of cerebrovascular perfusion due to hypoxia, carbon monoxide poisoning, acute hypotensive shock, cardiac arrest, or other serious illnesses can produce ischemic infarction that mildly or robustly involves the cerebral border zone, the area between 2 major vascular territories (watershed area). If the infarction includes the border zone areas (i.e., between the middle cerebral artery territory and that of both the anterior cerebral and the posterior cerebral arteries), a combined or mixed extrasylvian aphasia can occur (Maeshima et al 1996). The language disorder has sometimes been called "isolation of the speech area" or simply "isolation syndrome." In this type of language disturbance, the most dramatic clinical characteristic is the preserved ability to repeat spoken language despite severe impairment of all language functions (Berthier..."
The mixed extrasylvian aphasia has all the characteristics of global aphasia except for the preservation of repetition.

Patients with mixed extrasylvian aphasia have little or no spontaneous language. Their verbal output consists almost entirely of what has just been spoken by others (echolalia). These patients, however, may present elements of the completion phenomenon. When prompted with the beginning of a common phrase, the patient may repeat what has been said and then continue the phrase to completion. The patient’s articulation is good, and series speech (days of the week, nursery rhymes, etc.) is comparatively good once the task is started. Their verbal output, however, appears involuntary, automatic, and without understanding.

Language comprehension is usually severely impaired. Language repetition, even though dramatically preserved compared to other language functions, remains limited and is often below normal. The length of the phrase repeated is limited to a few words. These patients may or may not correct grammatically improper phrases when repeating; they will repeat pseudo-words and foreign words accurately. Confrontation naming is abnormal, sometimes producing neologisms or semantic paraphasias, but more often giving no response at all. The ability to read aloud, the ability to comprehend written language, and the ability to write are severely (usually totally) impaired. Extrasylvian mixed aphasia has been usually interpreted as an isolation of the perisylvian speech areas due to disconnection from surrounding brain areas (Berthier 1999). Mixed extrasylvian aphasia has been reported as a postictal epileptic phenomenon (Yankovsky and Treves 2002).

The neurologic findings can vary considerably. Some patients show bilateral upper motor neuron paralysis, a severe spastic quadripareisis that indicates bihemispheric damage. Other patients have right hemiplegia and sensory loss. A visual field defect, usually a right hemianopsia, is present in some cases. Some patients with mixed extrasylvian aphasia have few basic neurologic difficulties, showing only weakness that is most marked in the shoulder and hip musculature and variable degrees of cortical sensory loss. Most often, however, mixed extrasylvian aphasia is found in patients with severe brain damage, and a whole host of additional neurologic and neurobehavioral disorders are present.

Subcortical aphasia. Since Wernicke, it has been accepted that aphasia can represent the product of damage to neural networks made up of both cortical and subcortical structures. Nonetheless, whether true aphasia results from isolated subcortical brain damage, or due to a cortical extension or cortical deactivation, remains unanswered. Subcortical pathology frequently includes altered speech, often beginning with total mutism followed by hypophonic, slow, sparse output, and poorly differentiated, amelodic speech. Subcortical aphasias with similar speech and language defects have also been reported in children (Gout et al 2005). Contemporary neuroimaging techniques have permitted far better understanding of subcortical pathology involved in aphasia. Two neuroanatomical areas are most frequently discussed in subcortical aphasias: the striatocapsular region and the thalamus.

Patients with striatocapsular damage show significant articulation defects. Their language output appears truncated, but it is not agrammatic. Speech mechanisms are generally impaired with defects in articulation and prosody. Comprehension is intact for casual conversation but breaks down when complex syntax is presented. Word-finding problems may be noted. Early mutism in progressive nonfluent aphasia is associated with insula and inferior frontal atrophy extended into the left basal ganglia (Gorno-Tempini et al 2006). Alexander and colleagues have proposed 6 subtypes of verbal output impairment dependent on the specific neuroanatomical locus of striatocapsular damage, demonstrating that considerable variation in speech and language impairment can follow this type of pathology (Alexander et al 1987). Frequently, extension that involves the cortex is present in these cases. Extensive subcortical damage is required to produce pure striatocapsular...
aphasia, but prognosis is worse when the posterior limb of the internal capsule is involved (Liang et al 2001). When no cortical abnormalities are found on MRI, the severity of the left striatocapsular aphasia is associated with the extent and severity of the left cerebral cortical hypoperfusion on brain perfusion SPECT, particularly in the left temporal cortex (Choi et al 2007).

It has been speculated that this heterogeneity in the aphasia pattern in cases of basal ganglia pathology may reflect variations in cortical hypoperfusion resulting from large vessel stenosis. To test this hypothesis, Hillis and colleagues analyzed a consecutive series of 24 patients with left caudate infarct identified with diffusion-weighted imaging that underwent language testing and perfusion-weighted imaging less than 24 hours from onset of symptoms (Hillis et al 2004). Specific regions in perisylvian cortex were rated for the percentage of the region that was hypoperfused. Aphasia type was determined on the basis of speech fluency, comprehension, and repetition performance on the language tests. Results demonstrated that in patients with acute left caudate infarct, the presence and type of aphasia reflected regions of hypoperfusion and generally followed predictions based on chronic lesion studies regarding anatomical lesions associated with classic aphasia types. Subcortical aphasia is generally associated with diffuse hypometabolism in the ipsilateral cerebrum (frontal, parietal, temporal, occipital, putamen, thalamus) and in the contralateral cerebellum; and diffuse hypermetabolism in the contralateral cerebrum and ipsilateral cerebellum (Kim et al 2012).

Subcortical stimulation has been used in mapping the different subcortical pathways involved in language. Duffau and colleagues selected 115 patients with grade II gliomas located in language areas (Duffau et al 2008). Patients underwent operation after induction of local anesthesia; direct electrical stimulation to perform online cortical and subcortical language mapping was used. One or several of the following subcortical language pathways were identified in all the patients: (1) arcuate fasciculus (eliciting phonemic paraphasia when stimulated); (2) inferior frontooccipital fasciculus (eliciting semantic paraphasia when stimulated); (3) subcallosal fasciculus (resulting in extra-sylvian motor aphasia during stimulation); (4) frontoparietal fasciculus (resulting in speech apraxia during stimulation); and finally (5) fibers coming from the ventral premotor cortex (inducing anarthria when stimulated).

Murray analyzed spoken language production in Huntington and Parkinson diseases. It was found that in terms of syntax, patients with Huntington disease produced shorter utterances, a smaller proportion of grammatical utterances, a larger proportion of simple sentences, and fewer embeddings per utterance than their non-brain-damaged peers. The Huntington disease group also produced utterances that were shorter and syntactically simpler than those of the Parkinson disease group, despite similar performances on cognitive and motor tests. The only syntactic difference between the Parkinson disease group and their control group was that patients with Parkinson disease produced a smaller proportion of grammatical sentences (Murray 2000). In corticobasal degeneration and progressive supranuclear palsy, primary progressive aphasia has been reported to be particularly common (Kertesz and McMonagle 2010).

Thalamic pathology associated with aphasia usually produces an acute, catastrophic clinical picture with hemiplegia, hemisensory loss, and alterations in the level of consciousness (Benabdeljlil et al 2001). The initial language abnormality is mutism, which typically improves to a verbose, paraphasic, but hypophonic jargon output. Anomia is often severe. Although thalamic aphasia resembles other fluent paraphasic aphasias, patients with thalamic aphasia show decreased comprehension. When they attempt to repeat a word or phrase, their verbal output is far better than their conversational speech. A similarity to extrasylvian sensory aphasia has been noted, even though syntactic impairments have also been reported (De Witte et al 2006). Language disorder syndromes noted following thalamic damage can be categorized into 3 subtypes: (1) medial (left paramedial thalamic area,
involving dorsomedial and centromedian nuclei), (2) anterior (left anterolateral nucleus), and (3) lateral (left lateral thalamus). Lesions in the mediodorsal and the centromedian-parafascicular nuclei of the thalamus have been related to semantic retrieval deficits, probably because of their involvement in the activation of phonological representations (Pergola et al 2013). It has been suggested that thalamic nuclei and systems are involved in multiple processes that directly or indirectly support cortical language functions: lexical-semantic functions, working memory, visual processing in reading, and category-specific naming (Crosson 1999). Infarcts in the left posterolateral territory have also been associated with aphasia (Carrera et al 2004). On the other hand, patients may present with classical symptoms suggesting aphasia following thalamotomy (repetition, comprehension, fluency, and naming abnormalities). They may also present with “freezing of speech” (Bruce et al 2004). The left thalamus seems to bring online the cortical network involved in language processing (Metz-Lutz et al 2000).

Acute aphasia has been sometimes observed in multiple sclerosis (Devere et al 2000). Progressive nonfluent aphasia has been occasionally reported in progressive supranuclear palsy (Josephs et al 2005).

**Cerebellar language disorders.** It has been hypothesized that the cerebellum contributes to cognitive processing, particularly the processing of linguistic information (Leiner et al 1991; 1993; Hernandez-Muela et al 2005). The cerebellum has anatomical connections to the cerebral cortex, through which it can affect language function (Ackermann et al 2007). The neodentate cerebellar area projects via the thalamus to the frontal lobe, especially the prefrontal areas and Broca area. The frontal lobe reaches the neodentate in humans via several pathways. This neural cerebellar-frontal loop can affect cognitive, especially linguistic, functions. Cerebellar damage has been associated with impaired grammar and decreased verbal fluency (Akshoomoff and Courchesne 1992; Daum et al 1993; Silveri et al 1994), and also language understanding, naming, and repetition (Karaci et al 2008). It has been further argued that the cerebellum has reciprocal connections not only with the left inferior frontal gyrus but also the left lateral temporal cortex (Booth et al 2007).

Abe and colleagues selected 15 patients with chronic Broca aphasia (Abe et al 1997). Using the results of SPECT, they divided them into patients with (group 1) and without (group 2) crossed cerebellar diaschisis. Language function of the 2 groups was compared. Patients in group 1 showed classical Broca aphasia, whereas patients in group 2 showed mainly word-finding difficulty. Patients with crossed cerebellar diaschisis had infarcts involving the lower part of the frontal gyrus, but patients without crossed cerebellar diaschisis did not, which suggests that this region may have functional and anatomical connections with the cerebellum.

Schatz and colleagues found slowing in cognitive processing, particularly linguistic processing, associated with cerebellar pathology (Schatz et al 1998). Hubrich-Ungureanu and colleagues used MRI in 1 left- and 1 right-handed subject in a conventional block design (Hubrich-Ungureanu et al 2002). Regions of activation were detected after performance of a silent verbal fluency task. In the right-handed volunteer, they found an activation of the left frontoparietal cortex and the right cerebellar hemisphere, whereas in the left-handed volunteer the activation was seen in the right frontoparieto-temporal cortex and the left cerebellar hemisphere. The authors assumed that their results demonstrate that cerebellar activation is contralateral to the activation of the frontal cortex even under conditions of different language dominance. It is noteworthy to mention that verb generation is associated with activation of the right cerebellum, whereas reading of verbs results in both left and right activation (Frings et al 2006). Wollmann and colleagues demonstrated in Friedreich ataxia decreased motor and mental reaction times, reduced verbal span, deficits in letter fluency, impaired acquisition and consolidation of verbal information, proactive interference effect, and alterations in complex visuoperceptual and visuoconstructive abilities (Wollmann et al 2000).
2002). Nonetheless, it has been emphasized that although some minor defects in language can be found, aphasia is unusual in cerebellar pathology (Frank et al 2010).

Gottwald and colleagues examined 21 patients with cerebellar lesions due to tumor or hematoma, and 21 matched controls (Gottwald et al 2004). They concluded that lesions of the right cerebellar hemisphere lead to verbal deficits, whereas those of the left lead to nonverbal deficits. The generally greater impairment of those patients with a right-sided lesion was interpreted as resulting from the connection of the right cerebellum to the left cerebral hemisphere, which is dominant for language functions and crucial for right hand movements. It has been suggested further that the cerebellum is involved both in speech and singing; Callan and associates hypothesized that the left cerebellum differentially processes prosodic properties (melodic information used for singing) whereas the right cerebellum differentially processes segmental properties (phonological information used for speech) (Callan et al 2007). Jansen and colleagues, using fMRI with healthy subjects, corroborated that crossed cerebral and cerebellar language dominance is a typical characteristic of brain organization (Jansen et al 2005). Mariën and colleagues reported a 58-year-old right-handed man who presented with mild transcortical sensory aphasia associated with some alexia and agraphia following an ischemic infarction in the vascular territory of the right superior cerebellar artery (Mariën et al 2009). However, language defects secondary to left cerebellar pathology have also been reported (Galiano Blancart et al 2011).

Clinical vignette

A 55-year-old right-handed woman suffered a sudden inability to speak yet had preserved comprehension. Her right hemiparesis was mild. MRI suggested a left anterior cerebral artery vascular accident. She was mute for nearly 1 month. Two months later, scarce and effortful spontaneous language was observed. She cried easily when attempting to speak. She was depressed, sad, and frustrated with her condition. The patient mentioned frequent urinary urgency and loss of sphincter control on a few occasions.

At language evaluation, she had halted and effortful speech. She placed particular effort on getting started with a sentence or word. Consequently, intonation and sentence length were impaired. Together with her effort to initiate language, she frequently became tearful. No agrammatism, paragrammatism, or dysarthria were evident. Voice volume was normal. Oral description of a picture was characterized by effortful, halted speech, and there was no suggestion of neglect or simultanagnosia. Comprehension of orally presented words (nouns, verbs, numbers, colors, geometric shapes) was normal. Execution of simple and complex commands was intact. Identification of body parts was in the normal range. There were no indications of right-left confusion. No phoneme discrimination defects were found. Word retrieval skill in naming tests was moderately impaired. Her performance was characterized by occasional semantic paraphasias and, again, difficulty starting a word. Once she said a word, she would repeat it 2 or 3 times. Semantic and phonemic cueing were usually helpful. Repetition of syllables, words, pseudo-words, and phrases was normal. Her performance was flawless for short and medium length sentences and high and low frequency sentences (ie, sentences constructed from common versus uncommon words). Reading aloud was effortful but less halted than spoken language. No paralexias were observed. Despite being able to read relatively fluently both complex words and paragraphs, she had notable difficulty with spelling or reading letter-by-letter. Her reading comprehension was normal. She could write spontaneously and to dictation. No paragraphias were observed. A diagnosis of an extrasylvian motor aphasia-type II was proposed.

Etiology

Aphasia without repetition disturbances frequently indicates pathology located outside the perisylvian cortical region. Cerebrovascular infarcts represent a common etiology.
Extrasylvian motor aphasia-type II has been usually described associated with vascular disorders of the left anterior cerebral artery (Rubens 1975). Extrasylvian motor aphasia-type II, however, has also been observed with other etiologies (eg, interhemispheric tumors).

Decreased cerebrovascular perfusion for diverse reasons can result in ischemic infarction involving the cerebral border-zone between 2 major vascular territories and an extrasylvian aphasia of variable severity (Cauquil-Michon et al 2011; D’Amore and Paciaroni 2012; Joinlambert et al 2012). Extrasylvian aphasias can occur with either intracerebral or extracerebral tumors (Bizzi et al 2012) and arteriovenous malformations. Head injury, abscess, or any pathology involving some critical brain areas can also cause extrasylvian aphasia. A case of extrasylvian sensory aphasia in a child associated with subacute sclerosing panencephalitis was reported (Korkmaz et al 2007). Neologistic jargon aphasia and agraphia has been reported in primary progressive aphasia (Röhrer et al 2009).

The language impairment observed in the dementia of the Alzheimer type has been compared with extrasylvian sensory aphasia (Cummings et al 1985), and progressive jargon aphasia can be the initial manifestation of Alzheimer disease (Deramecourt et al 2010).

**Pathogenesis and pathophysiology**

Extrasylvian aphasias are associated with left hemisphere pathology. Much of the involved brain region corresponds to cerebral tissue lying at the junction between the middle cerebral artery and the vascular beds of the anterior or posterior cerebral arteries. Maeshima and colleagues observed that transcortical mixed aphasia may be caused by the isolation of perisylvian speech areas, even if a lesion is located in the inferior frontal gyrus, due to disconnection from surrounding areas (Maeshima et al 2002).

Preserved repetition represents the hallmark of extrasylvian aphasias. Language repetition defects in perisylvian aphasia are due to a diversity of factors (eg, phonemic discrimination errors, verbal memory defects, and verbal apraxia). Patients with extrasylvian aphasia do not present these defects responsible for the language repetition impairments observed in perisylvian aphasias. Hemorrhage is the most common subcortical pathology associated with aphasia.

Blank and colleagues investigated the brain systems engaged during propositional speech and 2 forms of nonpropositional speech: counting and reciting overlearned nursery rhymes (Blank et al 2002). Bilateral cerebral and cerebellar regions were involved in the motor act of articulation, irrespective of the type of speech. Three additional, left-lateralized regions, adjacent to the Sylvian sulcus, were activated in common: the most posterior part of the supratemporal plane, the lateral part of the pars opercularis in the posterior inferior frontal gyrus, and the anterior insula. Therefore, both nonpropositional speech and propositional speech were dependent on the same discrete subregions of the anatomically ill-defined areas of Wernicke and Broca. Propositional speech was also dependent on a predominantly left-lateralized neural system distributed between multimodal and amodal regions in posterior inferior parietal, anterolateral, and medial temporal and medial prefrontal cortex. The evidence from this study indicates that normal communicative speech is dependent on a number of left hemisphere regions remote from the classic language areas of Wernicke and Broca. Destruction or disconnection of discrete left extrasylvian and perisylvian cortical regions, rather than the total extent of damage to perisylvian cortex, will account for the qualitative and quantitative differences in the impaired speech production observed in aphasic stroke patients.

**Epidemiology**

No information was provided by the author.

**Differential diagnosis**
Extrasylvian motor aphasia has to be differentiated from depression and dementia. The general patient’s behavior (apathy and withdrawal) can resemble depression. Depression and dementia, however, may coexist with aphasia. The articulatory initiation defect observed in cases of damage of the supplementary motor area often results in anxiety, frustration, and depression. In frontal dementias, including Pick disease, a decrease in verbal output and echolalia are often observed. It can be proposed that extrasylvian motor aphasia represents a frequent element in frontal dementia. Some subtypes of schizophrenic psychosis (eg, catatonic schizophrenia) also have to be considered as a differential diagnosis in the first subtype of the extrasylvian motor aphasia associated with left prefrontal pathology.

Differential diagnosis between progressive nonfluent aphasia and Alzheimer disease represents an important diagnostic question. It has been observed, however, that pronounced literal paraphasic errors distinguish progressive aphasia patients from Alzheimer disease patients. Progressive aphasia patients also have anomia, decreased letter fluency, neologisms, difficulty on phrase repetition, decreased phrase length, and a decreased rate of verbal output (Mendez et al 2003).

In extrasylvian sensory aphasia, the greatest diagnostic confusion occurs with dementia. However, Alzheimer disease can include a language disturbance that has been equated with an extrasylvian sensory aphasia. In consequence, the differential diagnosis will be between a focal language defect and a global cognitive impairment. It is likely that some cases of extrasylvian sensory aphasia go improperly diagnosed because of its frequent lack of other neurologic abnormalities. Psychotic language represents a probable confusion.

**Diagnostic workup**

Language repetition represents the most crucial test in extrasylvian aphasias. Conservation of language repetition as compared with impairments in some other language functions is expected in extrasylvian aphasias. Several comprehensive test batteries have been developed in testing for aphasia: Boston Diagnostic Aphasia Examination (Goodglass et al 2000), Multilingual Aphasia Examination (Benton and Hamsher 1994), and Western Aphasia Battery (Kertesz 2006). In diagnosing for aphasia, the use of comprehensive language test batteries can be advisable. Some language subtests are typically included in these batteries:

- Spontaneous language: telling a story, describing a picture, etc.
- Language comprehension: phoneme discrimination, pointing (objects, body parts, colors, actions), following commands, etc.
- Naming: objects, body parts, colors, actions.
- Repetition: syllables, easy and difficult words, short and long sentences.
- Reading ability and comprehension: letters, pseudo-words, words, sentences, paragraphs.
- Writing: spontaneous, by dictation, by copy (letters, pseudo-words, words, sentences, paragraphs).

**Prognosis and complications**

The severity of aphasia and the pattern of recovery are variable. Recovery may be related to the size of the lesion and the etiology of the damage. Additional factors influencing recovery include age and handedness. Extrasylvian aphasias associated with degenerative diseases obviously have poor prognosis. In vascular disorders, some improvement is expected. Recovery in extrasylvian motor aphasia-type II is expected to be good (Kertesz 1999). Recovery in extrasylvian sensory aphasia is variable, but at least some language improvement is expected. In general, aphasias associated with border-zone infarcts have a good long-term prognosis (Flamand-Roze et al 2011).
Management

During the past few decades, formal language rehabilitation programs have become widely available. Most individuals with language disturbances receive formal diagnostic evaluations, and many attempt remedial therapy programs. Single case reports and group studies have demonstrated the potential effectiveness of aphasia therapy (Basso et al 1979; Kertesz 1988; Basso 2003). Mild or significant residual language defects usually remain. Anomia represents the most frequent residual defect, and anomia treatment is a major question in aphasia therapy (Cornelissen et al 2003). Different approaches to rehabilitation of anomia have been described. Some are restitutional and attempt to reactivate lexical-semantic or phonological representations to improve word retrieval. Others are intended to reorganize language functions by engaging alternative cognitive systems to mediate word retrieval or by exploiting residual abilities to circumvent the impairment (Maher and Raymer 2004). Other treatments have approached the short-term memory as a strategy to improve language understanding (Salis 2012).

In general, aphasia therapy is directed to (1) maintain the patient's verbal activity, (2) gradually increase the level of difficulty in retraining and relearning language, (3) provide the patient with successful strategies to communicate, and (4) encourage the patient to continue rehabilitation efforts outside the professional program (Benson and Ardila 1996). Language therapy also provides psychological support and may play an important psychotherapeutic role. The influence of pharmacological treatment with bromocriptine, a dopaminergic agonist, has been considered for its effects on extrasylvian motor aphasias. Albert and colleagues attempted to restore speech fluency in a patient with longstanding extrasylvian motor aphasia by treating his symptoms of hesitancy and impaired initiation of speech with bromocriptine (Albert et al 1988). Unfortunately, treatment response to bromocriptine is highly variable (Gupta 1995; Bragoni et al 2000). Individuals with impairments of the intentional aspects of language are more likely to benefit than those with other forms of nonfluent aphasia, suggesting an influence of bromocriptine on circuits necessary to activate spontaneous language (Raymer 2003). Administration of galantamine, on the other hand, has been observed to have a beneficial effect on chronic post-stroke aphasia patients, particularly in subcortical aphasias (Hong et al 2012).

Although significant progress in aphasia rehabilitation has been achieved, many questions remain unsolved regarding how neuroscience principles can potentially be used to maximize aphasia recovery (Raymer et al 2008).

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