Aphasia Handbook

aphasia
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失語症
실어증

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To my professor and friend

Alexander R. Luria

With inmense gratitude

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Preface

The study of the language disturbance associated with brain pathology (aphasia) represents the real beginning of cognitive and behavioral neurosciences; and throughout the sciences' history, aphasia has continued as one of the most significant and extensively analyzed brain syndromes.

The importance of the study of aphasia is extensive. (1) Aphasia has an enormous clinical significance. The most common etiology of aphasia is a stroke (close to 80% of the aphasia cases), and in about one third of stroke cases language difficulties are observed. This results in approximately 1 in 272 or 0.37% or 1 million people in USA that present aphasia (National Institute of Neurological Disorders and Stroke, 2013; www.rightdiagnosis. com/ artic/ninds_aphasia_information_page_ninds.htm). (2) From a neurological point of view, aphasia has significantly contributed to the understanding of the human cerebral organization and the role of different brain areas and systems. (3) From the cognitive perspective, aphasia has advanced our understanding of human cognition, including the evolution of human cognition. And, (4) from the linguistic point of view, aphasia has furthered the analysis about the organization of the human language. As a matter of fact, aphasia is a central issue in different clinical and fundamental areas, including speech-language pathology, neuropsychology, neurology, psychology, and linguistics.

This textbook attempts to integrate the most basic information on aphasia. It has been divided into 12 chapters because it is the optimal length for a textbook. Initially an introduction to the topic is presented, starting with the history of aphasia, the brain conditions potentially resulting in aphasia, and the language abnormalities that can be found in aphasia. The second section analyzes the clinical manifestation of the oral and written language disturbances; in other words: the aphasia, alexia, and agraphia syndromes. The third section reviews the associated disorders and the aphasia manifestations in some special population. The last section is devoted to assessment and rehabilitation issues in aphasia.

This book has been written for the purpose of having some easy-to-use, basic information on aphasia. It attempts to cover the fundamental issues in aphasia and can be used as a textbook in basic aphasia courses. Potentially, this book can also be useful for different professional clinicians working with aphasia patients. Please, feel free to download, copy, print, and in general, use it as you consider most convenient.

I want to express my most sincere gratitude to all my colleagues that encouraged me to write this textbook. My special thanks go to Dr. Monica Hough and Felipe Ardila for their invaluable support and editorial help.

I. BASIC CONSIDERATIONS

Chapter 1

History of aphasia

Introduction

Aphasia can be defined as the **loss or impairment of language caused by brain damage** (Benson & Ardila, 1996). The modern conception of aphasia began in 1861 when Paul Broca presented the case of an individual who suffered a loss of language associated with brain pathology at the Anthropological Society of Paris. However, before Broca some older reports described language impairments observed after a pathological brain condition.

In this book, the aphasia history will be divided in four epochs: **Pre-classical** (until Broca's report in 1861), **Classical** (until WWII), **Modern** (until the 1970s, when the CT scan was introduced), and **Contemporary** (since the 1970s).

Pre-classical Period (until 1861)

It is usually assumed that the first known references to a language disturbance associated with brain pathology appears in Egypt in the so-called **Edwin Smith Papyrus** (Figure 1.1) about 1,500 BC, although probably it was copied from an older papyrus written between 3,000 and 2,200 BC (Tesak & Code, 2008). The Edwin Smith Papyrus is a medical text and surgical treatise, including 48 case histories, beginning with the injuries to the head. In at least five cases, some mention is made to loss of speech due to a head fracture.

Figure 1.1. Edwin Smith Papyrus (ca. 1500 BC)

However, the first explicit reference to the role of the brain in language disturbances is found in the Hippocrates' Corpus (ca. 400 BC) (Figure 1.2). Hippocrates clearly referred to two different types of language disturbances: **aphonos** ("without voice") and **anaudos** ("without hearing") corresponding to the two major aphasia syndromes. Hippocrates may be regarded as the first direct antecessor of contemporary aphasiology.
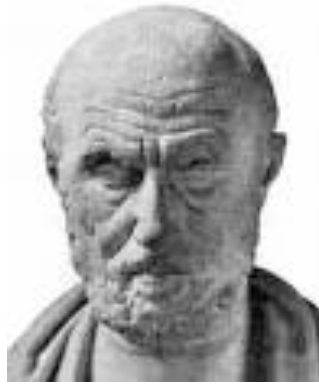
Figure 1.2. Hippocrates of Cos (ca 460 - 370 BC)

During the Roman Empire Valerius Maximus (ca. 20 AD) described the first case of **traumatic alexia** (acquired inability to read as a result of a head trauma). However, during this historical epoch, cognition was related to the cerebral ventricles and not really with brain tissue (Benton, 1981).

During the XV--XIX centuries, several papers describing languages pathologies were published. Antonio Guaneiro during the XV century reported two aphasic patients, one with a fluent paraphasic speech and the other one with a non fluent speech. The first description of **alexia without agraphia** (disturbed ability to read with preserved writing) is found by Girolamo Mercuriale, an Italian philologist and physician, most famous for his work *De Arte Gymnastica* (Figure 1.3)

Figure 1.3. Girolamo Mercuriale (1530-1606)

During the XVII century*,* Johann Schmitt and Peter Schmitt reported several aphasic patients with different symptomatology, including difficulties for naming and repetition impairments. During the XVIII century, new reports of diverse language disturbances are published, including anomia and jargon (Gesner), agraphia (Linné), preserved ability to sing (Dalin), and even dissociation for reading in different languages (Gesner) (Benton, 2000).

The XIX century is most crucial in the history of aphasia. Bouillaud (Figure 1.4) was a French physician who in 1825 distinguished two different types of language pathologies. One pathology had an articulatory basis, and the other pathology was amnesic in nature. These language profiles roughly corresponded to the two basic aphasic variants, mentioned before him by several authors beginning with Hippocrates.

Figure 1.4. Jean-Baptiste Bouillaud (1796 – 1881)

In 1843*,* Jacques Lordat, a professor of anatomy and physiology at Montpellier in France proposed a similar dichotomy. He described the inability to produce words, referred to as ***verbal asynergy***, and a disturbance in the ability to recall words, referred to as ***verbal amnesia***. The term, ***agraphia,*** was introduced by Ogle in 1867, to describe the acquired loss in the ability to write. Ogle found that although aphasia and agraphia usually occur together, they also can be dissociated.

At the beginning of the XIX century, Franz Josef Gall, a neuroanatomist and physiologist, developed the so-called "**cranioscopy**", a method directed to determine the mental and psychological characteristics of an individual based on the analysis of the external shape of the skull. Johann Spurzheim, his follower, renamed it as "**phrenology**" (*phren* = mind; *logos* = study) (Figure 1.5). Phrenology assumed that the brain is the organ of the mind, and that certain brain areas have localized, specific functions or modules. Language, for instance, depends on the orbital aspects of the frontal lobes; this area was supposed to be unusually increased resulting in a protrusion of the eyes ("oxen eyes") in people with excellent verbal memory, and also with language skills and literature ability. In recent times, phrenology has frequently been ridiculed because the concepts seem extremely naïve; however, phrenology historically played a significant role in the study of brain organization of cognition, explicitly stating that any type of cognition is the result of some brain activity.
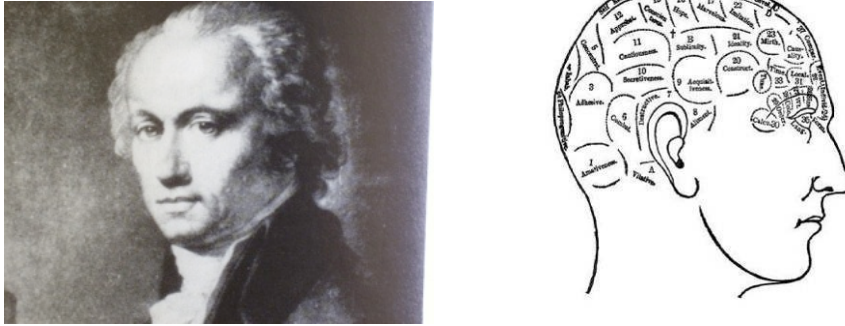
Figure 1.5. Franz Josef Gall (1758 – 1828) and the phrenology map.

Classical Period (1861-1945)

During early 1861, the Anthropological Society of Paris was discussing the potential relationship between intellectual capacity and volume of the brain. On April 4th, Auburtin presented the case of a patient who had lost his speech but was able to understand language. Paul Broca (Figure 1.6), French physician, anatomist, and anthropologist heard of a patient, named Leborgne who was transferred to the Bicêtre Hospital where Broca was working; the patient had lost the ability to speak, and could only produce the syllable "tan" (later, in the history of science, he has been known as **Tan**). Mr. Leborgne died on April 17, and his case was presented the next day in the meeting of the Anthropological Society of Paris. Broca named Tan's problem **aphemia** (loss of articulated speech). Broca determined that Leborgne's lesion was situated in the left posterior frontal lobe (Figure 1.7). Broca presented a second case a few months later and for the next two years he was able to collect 12 more cases in support of the localization of articulated language. He proceeded to directly propose that when an individual has lost the ability to speak, the pathology is situated in the left hemisphere, whereas lesions in homologous areas in the right hemisphere did not result in language deficits. In 1865, Broca stated: "*Aphemia is related with lesions of the third frontal gyrus in the left hemisphere*". Currently, it is recognized that about 25 years before, Mark Dax had already referred to this asymmetry in the brain organization of language, but his paper seemingly was not ever published.

Figure 1.6. Pierre Paul Broca (1824 – 1880)
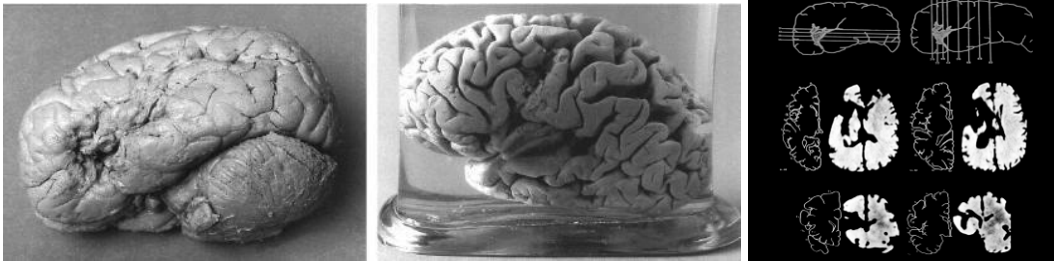
Figure 1.7. Leborgne's brain is preserved at the Museum of Man in Paris. At the right, sample of the first neuroradiological images of his brain.

A lively and heated discussion emerged about the name of the language disorder reported by Broca, initially referred to as aphemia. Previously, Lordat had used the name **alalia**, and Trousseau (1865) (Figure 1.7), a French physician, disapproved the name aphemia. According to Trousseau, aphemia was synonymous of infamy; he proposed the name **aphasia** (from ancient Greek ἀφασία ἀφασία (ἄφατος, ἀ- + φημί), "speechlessness"). Broca then proposed an alternative name, "**aphrasia**". But finally, **aphasia became the accepted label for this language disturbance**

Figure 1.7. ArmandTrousseau (1801–1867)

The second major advance in the study of the aphasia was represented by the doctoral dissertation of a German student, Karl Wernicke, in 1874 (Figure 1.8). Wernicke proposed **two different types of aphasia, motor and sensory**. Later, he proposed a third type of aphasia, named "**conduction" aphasia**, based in the diagrammatic descriptions of the brain areas involved in language. Later in 1885, he proposed with Lichtheim, a model for the interpretation and classification of aphasia, usually known as the **Lichtheim--Wernicke model**, or the **classical model of aphasias**. This model includes two major types of aphasia (motor and sensory) each one with three variants (cortical, subcortical, and transcortical). Conduction

aphasia, characterized by language repetition defects, is due to a disconnection between the sensory and motor areas of the language. This classification of aphasias has become the most influential model of aphasia, significantly guiding research in the area over the last century.
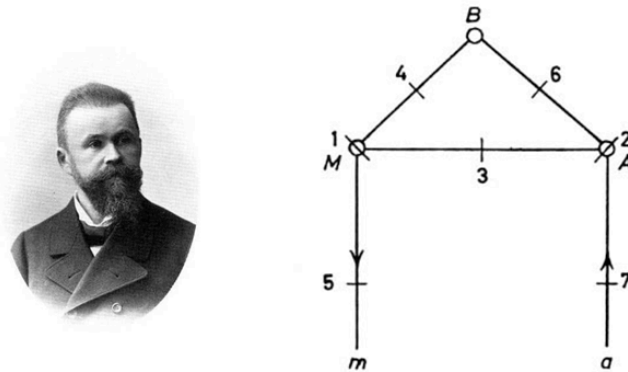
Figure 1.8. Carl Wernicke (1848-1905) distinguished seven variants in the language disturbances associated with brain pathology: (1) cortical motor; (2) cortical sensory; (3) conduction; (4) transcortical motor; (5) subcortical motor; (6) transcortical sensory; (7) subcortical sensory.

In his book about aphasia published in 1891, Freud criticized the "diagram makers" and the strict localization of language functions in the brain. In 1906, Pierre Marie (Figure 1.9) overtly rejected the **localizationist** approach of language in his critical paper *"The Third Frontal Gyrus does not Play Any Special Role in Language Functions"*. During the late XIX century and early XX century, many authors maintained a holistic viewpoint with regard to the brain organization of language. Indeed, the influence of this holistic perspective in interpreting brain organization of psychological processes significantly advanced during the first decades of the XX century and many researchers partially or totally supported this interpretation, including Head (1926), Wilson (1926), Pick (1931), Weisenburg & McBride (1935), Wepman (1951), and Bay (1962).

Figure 1.9. Pierre Marie (1853 –1940)

The opposite or localizationist viewpoint was upheld by many researchers, but in particular by Joseph Jules Dejerine (Figure 1.10) who developed the idea of the "**language area**" in the brain. This idea has been supported by most authors in the aphasia area. There is a general agreement that **language is related to activity in the perisylvian areas of the left hemisphere.**
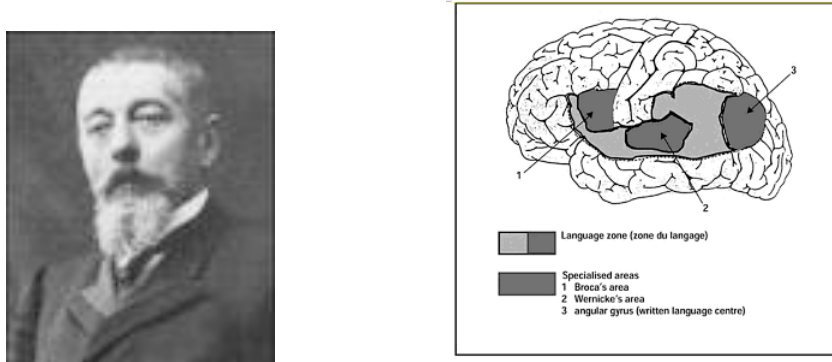
Figure 1.10. Joseph Jules Dejerine (1849 – 1917) supposed that there is a brain area involved in language, situated around the Sylvian fissure in the left hemisphere

Henry Head (Figure 1.11) presented a clinical/psychological approach to aphasia. He also introduced an original fourfold classification of disturbance of function in aphasia including verbal defects (verbal aphasia), syntactic defects (syntactic aphasia), nominal defects (nominal aphasia), and semantic defects (semantic aphasia). In his classical book, *Aphasia and Kindred Disorders of Speech,* published in 1926, he also presented some general guidelines for aphasia testing and insisted on the need to use comparable procedures for aphasia diagnosis.

Figure 1.11. Henry Head (1861 – 1940)

Gestalt psychologists such as Goldstein (1948) (Figure 1.12) and Conrad (1949) proposed that brain damage interferes with the basic function (*gestalten*), resulting in existing variations in observed symptomatology due to differences in organization of the whole brain. Goldstein

referred to his holistic approach as **organismic aphasiology**. Scientific support for the holistic approach was partially derived from experiments with animals conducted by Lashley (1929); his research suggested that brain function is not the result of a specific neuroanatomical structure, but the result of the integrated participation of an extensive volume of brain tissue,

Figure 1.12. Kurt Goldstein (1878 – 1965)

Modern Period (until the 1970s)

During WWII, the significant number of patients with brain injuries associated with language and other cognitive disorders increased the need for developing reliable diagnostic and rehabilitation procedures for aphasia. In different countries, special sections devoted to the diagnosis and rehabilitation of aphasia patients were created. The first important post-war result of this increased interest in aphasia was the book "*Traumatic Aphasia*" written by Alexander Romanovich Luria (1.13), published in Russian in 1947 and in English in 1970. An original classification and interpretation of aphasia was presented, assuming that in each type of aphasia, a specific level of language processing is impaired (Table 1.1). Luria's influence in aphasia interpretation has been very significant. His aphasia interpretations were further developed in later books, *Higher Cortical Functions in Man (1962), The Working Brain (1973),* and *Basic Problems of Neurolinguistics* (1976).

Figure 1.13Alexander Romanovich Luria (1902 – 1977)

Type of aphasia	Impaired factor
Acoustic-agnosic	Phoneme discrimination
Acoustic-amnesic	Verbal memory
Amnesic	Semantic structure of the words
Semantic	Understanding logic-grammatical quasi-spatial relations
Afferent motor	Articuleme discrimination
Efferent motor	Kinetic melody of speech, inner speech
Dynamic	Verbal initiative

Table 1.1. According to Luria, aphasia is due to the impairment of a specific level of language processing, and hence, a specific factor (or level of language processing) is impaired.

Luria took a midway stance between the localizationist and holistic approaches, acknowledging that both perspectives had remarkable merit. He considered language to be a complex functional system, requiring many different steps to achieve both comprehension and production; simultaneous participation of multiple cortical areas are required for language processing. Although each cortical area performs a specific process, it also participates in different **functional systems**. Thus, the first temporal gyrus participates in phoneme discrimination, and its damage causes difficulty in all functional systems requiring phoneme discrimination; language represents a complex functional system and different types of language impairments are associated with damage in specific brain areas (Figure 1.14)
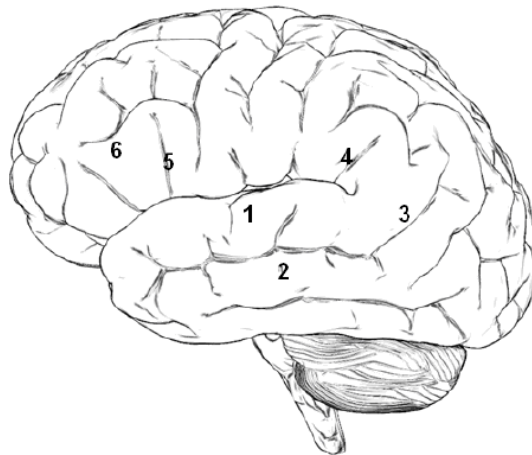
Figure 1.14. According to Luria, language is a complex functional system including different factors. Diverse language impairments are associated with left hemisphere lesions: (1) disturbances in phonemic discrimination; (2) impairments in verbal-acoustic memory; (3) word-retrieval difficulties (semantic associations); (4) language repetition defects; (5) agrammatism and speech apraxia; (6) adynamia of verbal processes.

Since the mid 1960s, clinical and theoretical interpretation of aphasia in the US (and also in a significant part of the western world) have been guided by Norman Geschwind (Figure 1.15) and the so-called Boston Group (Goodglass, Kaplan, Kertesz, Benson, Alexander, etc.). Geschwind interpreted cortical syndromes as disconnection syndromes; in 1962, he published his most classical paper in this area, with the title "*Disconnection Syndromes in Animals and Man*". In 1965, he organized a center for aphasia research at the Boston Veterans Administration Hospital, currently known as Harold Goodglass Aphasia Research Center. Geschwind further developed Wernicke's classical ideas and aphasia interpretations. These interpretations are known as the **Wernicke**-**Geschwind model** of language processing; this model proposes serial language processing, distinguishing seven different components of language: primary auditory cortex, Wernicke's area, arcuate fasciculus, primary visual cortex, angular gyrus, Broca's area, and primary motor cortex. These seven areas interact to form working language network in the left hemisphere. Conduction aphasia represents the best example of a linguistic disconnection syndrome (Figure 1.16)

Figure 1.15. Norman Geschwind (1926– 1984)
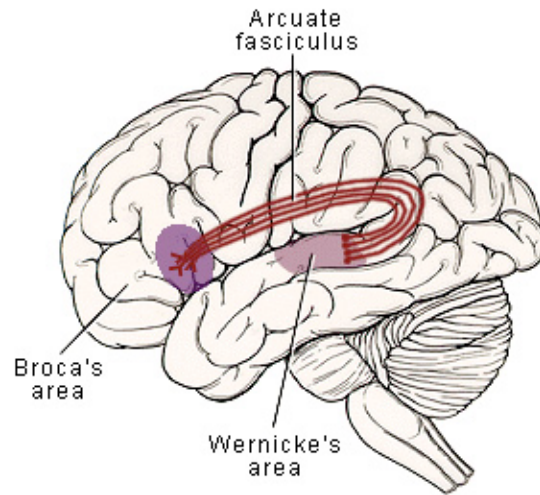
Figure 1.16. Following Wernicke's interpretation of conduction aphasia, Geschwind proposed that the impairment in the arcuate fasciculus (pathway supposedly connecting the Wernicke's and Broca's areas) was responsible for the repetition defects observed in conduction aphasia.

The Boston Group considers that the three major language parameters in aphasia classification are fluency, repetition, and understanding. Any aphasia syndrome can be classified simultaneously using these three parameters (Table 1.2)

⌘ non fluent	• Repetition poor	understanding poor	GLOBAL
		understanding good	BROCA
	• repetition good	understanding poor	MIXED TRANSC
		understanding good	MOTOR TRANSC
⌘ fluent	• repetition poor	understanding poor	WERNICKE
		understanding good	CONDUCTION
	• repetition good	understanding poor	SENSORY TRANSC
		understanding good	ANOMIC

Table 1.2. Aphasia syndromes can be classified using three language parameters: fluency, repetition and understanding.

During the second half of the XX century, significant research on aphasia was observed in different countries world-wide. In France, Henri Hécaen and François Lhermitte intensively analyzed language lateralization, childhood aphasia, reading and writing disturbances, and other aphasia related questions. In Italy, De Renzi, Vignolo, and Gainotti researched different aphasia issues. In Germany, Poeck made significant contributions to aphasia understanding. In England, Weigl, Warrington, and Newcombe also intensively researched aphasia. In Canada, André Roch Lecours and colleagues published research studies on different aspects of brain organization of language. In Latin America, Fernando Dalmas (Uruguay), Archibaldo Donoso (Chile), and others developed influential research programs on language and speech disturbances associated with brain pathology. In North America, aphasia research has been guided by D. Frank Benson, Edith Kaplan, Harold Goodglass, and Arthur L Benton among others (Figure 1.17). Different classification and interpretations of aphasia have been proposed (Table 1.2), but as a matter of fact these interpretations have become progressively more similar (Figure 1.18). .

Figure 1.17. Some major aphasia researchers during the second half of the XX century. From left to right: Henri Hécaen (1912 -1983), Harold Goodglass (1920-2002), André Roch Lecours (1936-2005), Edith F. Kaplan (1924 - 2009), and D. Frank Benson (1928-1996).

Luria, 1966	Benson & Geschwind, 1971	Hécaen & Albert, 1978	Kertesz, 1979	Benson, 1979	Lecours et al., 1983
Efferent motor	Broca's	Agrammatic	Broca's	Broca's	Broca's
Sensory	Wernicke's	Sensory	Wernicke's	Wernicke's	Wernicke's type I
Afferent motor	Conduction	Conduction	Conduction	Conduction	Conduction
Dynamic	Transcortical motor	Transcortical motor	Transcortical motor	Transcortical motor	Aspontaneity
—	Transcortical Sensory	Transcortical Sensory	Transcortical Sensory	Transcortical Sensory	Wernicke's type II
—	Isolation language area	—	Isolation	Transcortical mixed	—
Semantic Amnesic	Anomic	Amnesic	Anomic	Anomic	Amnesic
—	Global	—	Global	Global	—
—	Aphemia	Pure motor	—	Aphemia	Pure anarthria

Table 1.2. Some aphasia classifications. Except for Luria's proposal, the rest of the classifications are quite similar.

global aphasia   Broca's aphasia   Wernicke's aphasia

conduction aphasia   anomic aphasia   transcortical sensory aphasia

transcortical motor aphasia
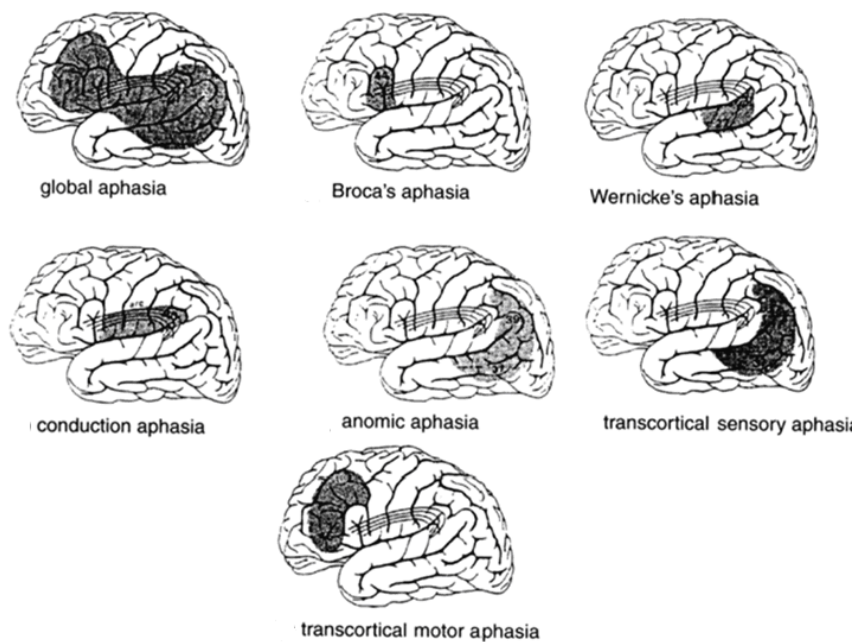
Figure 1.18. It has been observed that damage in some brain areas result in specific aphasia syndromes (from: www.studyblue.com/notes /note/ n/a phasia-typologies/ deck/231350.)

Contemporary Period (since the 1970s)

The introduction of computerized tomography (CT scan) during the 1970s represented a major revolution in different neuroscience areas, including aphasia. More reliable clinical/anatomical correlations became available. It was observed that other anatomical areas beyond the perisylvian area of the left hemisphere ("language area") could be impaired in cases of aphasia; for instance, it was observed that aphasia was frequently associated with subcortical pathology, and discussion and interpretation of subcortical aphasias re-emerged.

Three major advances can be identified during this contemporary period: first, the development and diffusion of neuroimaging techniques, initially (during the 1970s and 1980s) the anatomical techniques (**CAT** and **MRI**), and further (during the 1990s and later) the functional techniques (particularly **fMRI** and **PET**). These advances led to a new interpretation of brain organization of cognition in general and language in particular, resulting in the so-called "**functional model**" of brain organization of cognition. In general, it has been observed that the brain areas involved in language processing can be broader than the perisylvian area of the left hemisphere (classical "language area"). For instance, the supplementary motor area frequently is found to be activated during the performance of diverse verbal tasks.

A second major advance involves the progressively extended use of standardized procedures for aphasia assessment. Some aphasia tests and test batteries have become especially popular and widely used by speech language pathologists, neurologists, and neuropsychologists world-wide. These tests and protocols include the **Boston Diagnostic Aphasia Examination** (Goodglass & Kaplan, 1972, 1983, 2001), the **Multilingual Aphasia Examination** (Benton, Hamsher &, Sivan, 1994), the **Western Aphasia Battery** (Kertesz, 1982, 2006), the **Boston Naming Test** (Kaplan, Goodglass, & Weintraub,, 1983, 2001), the **Token Test** (De Renzi & Vignolo, 1962, 1978) and many others.

A third major advance in aphasia during the last few decades is the development and extension of diverse rehabilitation techniques and strategies (Basso, 2003; Paradis, 1993). Melodic Intonation Therapy is a good example of an aphasia rehabilitation technique that has become significantly extended and has proven to be successful in non-fluent aphasias (Sparks, Helm & Alberto, 1974).

By the same token, some new interpretations and classifications of aphasia disorders have been recently proposed (Table 1.3 and 1.4). These classifications attempted to integrate contemporary knowledge about brain organization of language in normal and abnormal conditions.

	Pre-Rolandic	*Post-Rolandic*
Peri-Sylvian	Broca's Type I (triangular syndrome)	Conduction (parietal-insular syndrome)
	Broca's Type II (triangular-opercular-syndrome)	Wernicke's Type I (posterior insular-temporal isthmus syndrome)
		Wernicke's Type II (superior and middle temporal gyrus syndrome)
Extra-Sylvian	Extra-Sylvian Motor Type I (left prefrontal dorsolateral syndrome)	Extra-Sylvian Sensory Type I (temporal-occipital syndrome)
	Extra-Sylvian Motor Type II (supplementary motor area syndrome)	Extra-Sylvian Sensory Type II (parieto-occipital angular syndrome)

Table 1.3. Some recent proposals of aphasia classification. The proposal suggested by Benson and Ardila (1996) is presented. According to this proposal, two major dimensions for aphasia classification can be used: aphasia can be peri-Sylvian or Extra-Sylvian; aphasia, on the other hand, can be pre-Rolandic or Post-Rolandic. Sub-types for some aphasia are distinguished, and aphasia syndromes are related to anatomical syndromes.

Type	*Impairment*
Primary (central) aphasias	*Language system impaired*
Wernicke-type aphasia (fluent aphasia)	Phonological level Lexical level Semantic level
Broca-type aphasia(non-fluent aphasia)	Sequencing expressive elements at syntactic and phonetic level
Secondary (peripheral) aphasias	*Mechanisms of production impaired*
Conduction aphasia	Disconnection (or segmentary ideomotora verbal apraxia)
SMA aphasia	To initiate and maintain voluntary speech production
Dysexecutive aphasia	*Language executive control impaired*
Extra-Sylvian (transcortical) motor aphasia	Executive control of language

Table 1.4. The proposal suggested by Ardila (2010) is presented. A major distinction is established between primary aphasias (Wernicke's with three subtypes, and Broca's) and secondary aphasias (conduction aphasia and aphasia of the supplementary motor area); finally a "dysexecutive aphasia" is distinguished.

Summary

Initial reports of oral language disturbances associated with brain damage were presented during the Egyptian Empire. The first report of a disturbance in written language was found during the Roman Empire period. Hippocrates was the first to distinguish that there are two different types of language impairments associated with brain pathology. During the XV- to XIX centuries, diverse observations about language disturbances in cases of brain damage were observed and presented to the scientific community. However, modern aphasia history (and in general cognitive neurosciences history) typically begins with Broca's case report of a loss of language in 1861. Later, Wernicke proposed a classification and interpretation of aphasia that has become the most influential framework on current thinking. Dejerine identified the so-called "language area" in the brain corresponding to the perisylvian area of the left hemisphere.

Controversy ensued relative to a holistic versus localizationist interpretation of language organization during the late XIX century and early XX century. After WWII, various researchers in different countries continued the clinical and theoretical study of aphasia; however, it appears that the two most influential approaches to aphasia have been Luria's interpretation of language as a complex functional system and the Wernicke-Geschwind model of language processing.

With the advent of neuroimaging techniques, it has been possible to obtain more accurate clinical/anatomical correlation of diverse language impairments. Furthermore, it has been observed that the critical areas of the brain initially identified relative to their involvement in language processing are more extensive than previously assumed. During recent decades, progressively extended use of standardized procedures for aphasia assessment has been observed, with some test batteries becoming particularly popular in the evaluation of aphasia. Additionally, this has led to development of various rehabilitation techniques as well as extension of new and diverse therapeutic strategies.

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Chapter 2

Aphasia etiologies

Introduction

Any abnormal condition affecting the brain areas involved in language (Figure 2.1) can result in aphasia. However, the specific symptoms of the language impairment depend upon the particular brain area that is affected. Anterior damage results in language productions impairments (possibly a Broca's type of aphasia), while posterior pathology is associated with language understanding difficulties and disturbances in the phonological, lexical and semantic language systems (often Wernicke's type of aphasia). Damage in the surrounding areas is associated with so-called perisylvian (transcortical) aphasias in the anterior and posterior regions, respectively.*.*
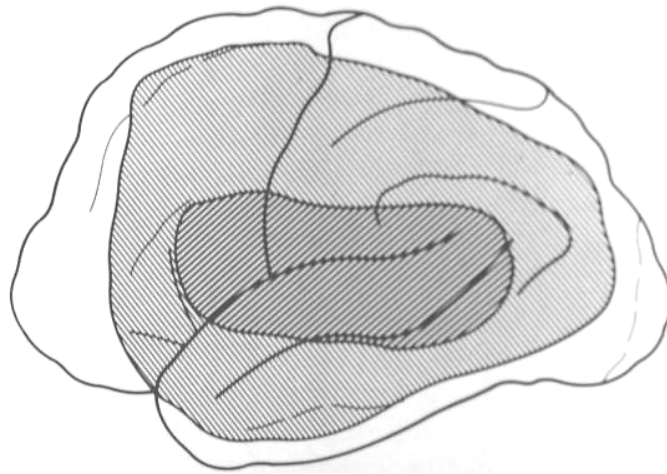
Figure 2.1. Damage in the perisylvian area (language area; darker area) of the left hemisphere can result in Broca's aphasia (frontal), Wernicke's aphasia (temporal) or conduction aphasia (parietal). Damage in the surrounding area (light grey area) is associated with so-called perisylvian (transcortical) aphasias.

Diverse etiologies of central nervous disturbances are recognized (Table 2.1). Some of them are frequently associated with aphasia, particularly vascular disorders and traumatic head injury; but there are other abnormal brain conditions also potentially associated with aphasia, such as neoplasms (tumors), infections, and degenerative conditions. Although other brain abnormalities (developmental, metabolic, and nutritional diseases; and disorders due to drugs

and chemical agents) can be associated with speech and language abnormalities, they do not represent specific aphasia etiologies.

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Vascular disorders
Traumatic brain injury
Neoplasms
Infections
Degenerative conditions
Developmental diseases
Metabolic diseases
Nutritional diseases
Disorders due to drugs and chemical agents

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Table 2.1. Etiologies of nervous system pathologies. The first five can result in aphasia. Although the last four can be associated with speech and language abnormalities, they do not represent specific aphasia etiologies.

Vascular disorders

Aphasia is observed in about one-third of the patients with so-called **cerebrovascular disorders** or **cerebrovascular accidents** (**CVA**) (stroke). However, aphasia subtype is variable and can change over time. In the acute stage of recovery, the most frequent aphasia is global aphasia; however, aphasia profile often changes during the stroke evolution with the most frequent aphasia subtype one year later being anomic aphasia (Table 2.2). As a matter of fact, anomia represents the most important aphasia symptom manifestation and long-term aphasia sequelae.

A CVA refers to a disruption in normal brain function due to any pathological condition of the blood vessels: walls of the vessels themselves, accumulation of materials, changes in permeability, or rupture. Stroke can be caused either by a clot obstructing the flow of blood to the brain or by a blood vessel rupturing and preventing blood flow to the brain. Consequently, there are two major types of strokes: **obstructive (ischemic**) and **hemorrhagic**.

At the onset of the CVA, a sudden neurological deficit (e.g., hemiplegia, aphasia, etc.) is often observed. In severe cases, CVA can be associated with coma. The development of the neurological deficit may take seconds, minutes, hours and occasionally even days, depending upon the specific type of CVA. Loss of consciousness is frequent in hemorrhagic CVAs, but infrequent in ischemic CVAs. Recovery is observed during the following hours, days, or weeks after the accident. As the results of decreases in edema (swelling) and diaschisis (extended impairment effect due to the broad connectivity of each brain area with the rest of the brain), symptomatology is progressively reduced to focal sequelae. The neurological or

neuropsychological residual deficit typically reflects the site and the size of the lesion (Figure 2.2).

	Acute first stroke	One year after stroke
global	32%	7%
Broca	12%	13%
Wernicke	16%	5%
conduction	5%	6%
anomic	25%	29%
transcortical motor	2%	1%
transcortical sensory	7%	0%
isolation	2%	0%
	100%	61%

Table 2.2. Copenhagen aphasia study including 270 stroke patients (Pedersen et al., 2004)
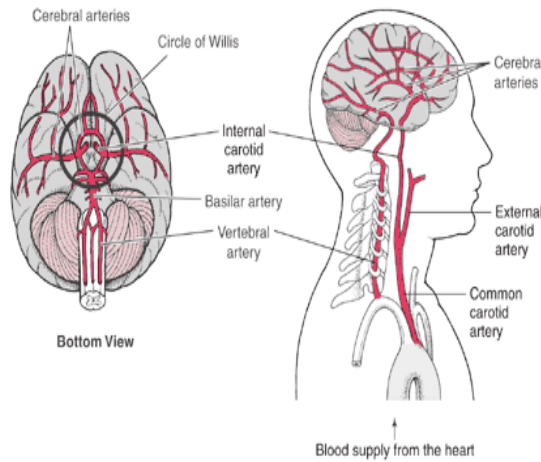
Figure 2.2. Blood supply to the brain. Blood goes to the brain through two different systems: the carotid system and the vertebrobasilar system. The first one originates the anterior and middle cerebral arteries, while the second one originates the posterior cerebral artery. The interconnection between both systems is known as the circle of Willis.

The incidence (new cases in one year) and prevalence (cases in the population at a certain moment) of CVA is very high (Table 2.3). Incidence has been estimated as about 80-150/100,000 and the prevalence in over 500/100,000. Mortality is close to 10% (Ropper & Samuels, 2009).

Indeed, stroke is the third leading cause of death in many countries. More than 140,000 people die each year from stroke in the United States. Stroke also is the leading cause of serious, long-term disability in many countries. About 75% of all strokes occur in people over the age of 65 and the risk of having a stroke more than doubles each decade after the age of 55. About 25% of the cases result in severe disability.

There are some well-established risk factors that increase the probability of having a CVA. These include: hypertension, age, cardiac disease, diabetes, obesity, and smoking; the three initial ones could be regarded as the major risk factors for CVA; the last three also are important risk factors, but not as significant as hypertension, age, and cardiac disease.

Characteristic	Total no. of respondents*	Prevalence of stroke (%)[†]	(95% CI[§])	Estimated no. of U.S. residents with a history of stroke
Age group (yrs)				
18–44	128,328	(0.8)	(0.7–0.9)	852,000
45–64	137,738	(2.7)	(2.5–2.9)	1,926,000
≥65	87,351	(8.1)	(7.7–8.5)	3,036,000
Sex[¶]				
Men	136,201	(2.7)	(2.5–2.8)	2,694,000
Women	219,911	(2.5)	(2.4–2.7)	3,145,000
Race/Ethnicity[¶]				
White, non-Hispanic	279,419	(2.3)	(2.3–2.4)	4,017,000
Black, non-Hispanic	27,925	(4.0)	(3.6–4.5)	772,000
Asian/Pacific Islander	5,974	(1.6)**	(1.0–2.7)	60,000
Hispanic[††]	25,539	(2.6)	(2.1–3.3)	616,000
American Indian/Alaska Native	5,535	(6.0)	(4.5–7.8)	126,000
Multiracial	6,519	(4.6)	(3.7–5.6)	136,000
Education[¶]				
Less than 12 years	38,202	(4.4)	(4.0–4.9)	1,365,000
High school graduate	109,830	(2.6)	(2.5–2.8)	1,863,000
Some college	93,228	(2.7)	(2.5–2.9)	1,474,000
College graduate	113,944	(1.8)	(1.6–1.9)	1,108,000
Total	356,112	(2.6)	(2.5–2.7)	5,839,000

* The sums of the sample sizes in each category might not add up to the total number of respondents because of unknown or missing information.
[†] Weighted percentage of respondents who reported a history of stroke.
[§] Confidence interval.
[¶] Weighted percentages are age adjusted to the 2000 U.S. standard population.
** The relative standard error of this estimate is 20%–30% and should be interpreted with caution.
[††] Might be of any race.

Tabla 2.3. Percentage of respondents reporting a history of stroke (according to the Behavioral Risk Factor Surveillance System, United States, 2010).

Aphasia is associated with CVAs involving the left middle cerebral artery (Figure 2.3). As a matter of fact, there is a significant correspondence between the territory of the middle cerebral artery and the surrounding brain area relative to language. CVAs involving the anterior cerebral artery, however, can be associated with the so-called "aphasia of the supplementary motor area", and extrasylvian (transcortical) motor (or dysexecutive) aphasia. CVAs involving the territory of the left posterior cerebral artery are frequently associated with alexia without agraphia (pure alexia) but not with aphasia.
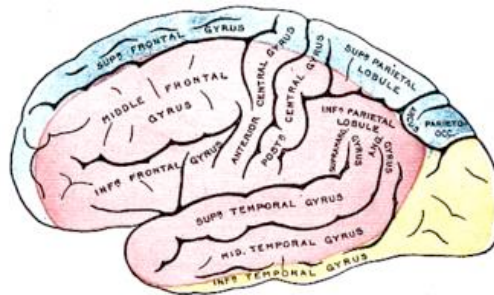
Figure 2.3. Cortical territory irrigated by the anterior (blue), middle (red) and posterior (yellow) cerebral arteries.

Furthermore, the specific aphasia subtype depends upon the particular branch of the middle cerebral artery that is involved (Table 2.4). When the main trunk of the left middle cerebral artery is involved, a global aphasia is found; when some specific branches are impaired, more diverse types of language disturbances may be observed.

Vascular territory	Type of aphasia
Main trunk of the left middle cerebral artery	Global aphasia
Orbitofrontal, pre-rolandic	Broca aphasia
Rolandic	Dysarthria
Parietal anterior	Conduction aphasia
Posterior parietal, angular	Sensory extrasylvian
Temporal	Wernicke aphasia
Lenticulostriate	Dysarthria, subcortical aphasia
Anterior cerebral artery	Aphasia of the supplementary motor area
Posterior cerebral artery	Alexia w/o agraphia

Table 2.4. Type of aphasia observed in cases of CVAs of different vascular territories

Types of CVA

As mentioned, the two major types of strokes are distinguished: **Occlusive (ischemic**) and **hemorrhagic** (Figure 2.4).

Occlusive (ischemic)

Two different conditions can be found relative to ischemic stroke: (1) **Embolism**: it is the occlusion of a vessel by material floating in arterial system. The emboli are usually formed from blood clots but are occasionally comprised of air, fat, or tumor tissue. Embolic events can be multiple and small, or single and massive; (2) **Thrombosis**: is the formation of a blood clot (thrombus) inside a blood vessel, obstructing the flow of blood through the circulatory system.
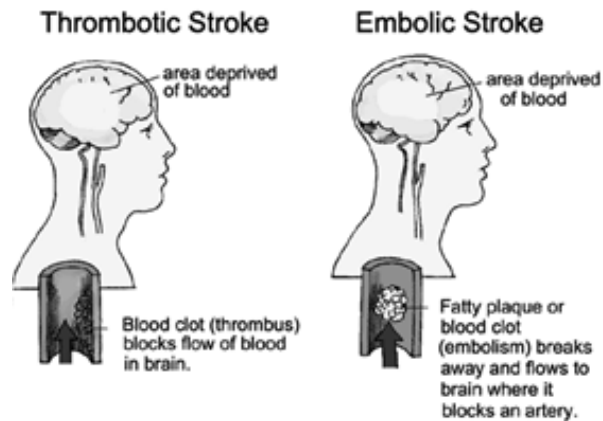
Figure 2.4. Thrombotic and embolic stroke

Hemorrhagic

Brain hemorrhage is another type of stroke. It is caused by an artery in the brain bursting and causing localized bleeding in the surrounding tissues. The pooled blood collects into a mass called a **hematoma.** These conditions increase pressure on nearby brain tissue.

Two major types of brain hemorrhage are distinguished (Figure 2.5):

(1) **Subarachnoid hemorrhage**: There can be bleeding into the **subarachnoid**, the space between the arachnoid and the pia mater, the innermost membrane surrounding the central nervous system. Most frequently, it is caused by bleeding from a cerebral aneurysm, but also can be due to bleeding from an arteriovenous malformation or head injury; Injury-related subarachnoid hemorrhage is often seen in the elderly who have fallen and hit their head. Among the young, the most common injury leading to subarachnoid hemorrhage is motor vehicle crashes.

(2) **Intracerebral hemorrhage**: is a type of stroke caused by bleeding within the brain tissue itself. It is most commonly caused by hypertension, arteriovenous malformations, or head trauma.
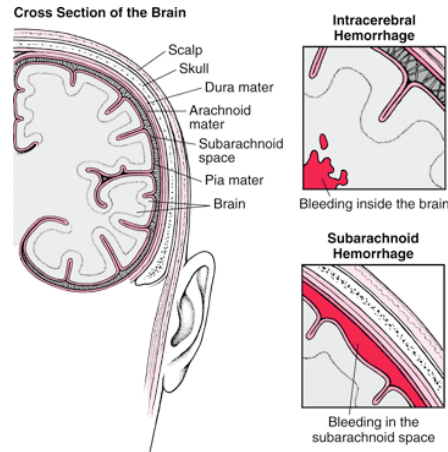
Figure 2.5. Two major types of hemorrhages: Intracerebral hemorrhage and subarachnoid hemorrhage

Traumatic brain injury

Traumatic brain injury (**TBI**), also called acquired brain injury, closed head injury, or simply head injury occurs when a sudden trauma causes damage to the brain. TBI can result when the head suddenly and violently hits an object, or when an object pierces the skull and enters brain tissue. Symptoms of a TBI can be mild, moderate, or severe, depending on the extent of the damage to the brain.

A person with a mild TBI may remain conscious or may experience a loss of consciousness for a few seconds or minutes. Other symptoms of mild TBI include headache, confusion, lightheadedness, dizziness, blurred vision or tired eyes, ringing in the ears, bad taste in the mouth, fatigue or lethargy, a change in sleep patterns, behavioral or mood changes, and trouble with memory, concentration, attention, or thinking. A person with a moderate or severe TBI may show these same symptoms, but may also have a headache that gets worse or does not go away, repeated vomiting or nausea, convulsions or seizures, an inability to awaken from sleep, dilation of one or both pupils of the eyes, slurred speech, weakness or numbness in the extremities, loss of coordination, and increased confusion, restlessness, or agitation.

TBIs may have different effects, including: (1) direct damage in the brain; (2) creation of a blood supply impairment interrupting the normal cerebral blood flow, (3) hemorrhages and hematomas; (4) brain edema (swelling), (5) subsequent possible infection; and finally, (6) origination of an epileptic focus.

Thus, the consequences of TBI can be diverse, and they include cognitive and behavioral sequelae such as impairments in attention and memory, impulsivity, irritability, and aphasia in addition to potential motor (paresis, dysarthria, etc.) and sensory defects (visual field defects, etc.).

One important criterion tool used to assess the severity of a TBI is the **Glasgow Coma Scale** that evaluates three types of responses: best eye opening, best motor response, and best verbal response (Table 2.5). The highest score is 15 and the lowest score is 3. It is assumed that a score over 13 is found in a mild TBI and below 8 suggests a more severe TBI. A score between 9 and 13 is indicative of a moderate TBI.

Response	Score
Eye opening	
Opens eyes spontaneously	4
Opens eyes in response to speech	3
Open eyes in response to painful stimulation (eg, endotracheal suctioning)	2
Does not open eyes in response to any stimulation	1
Motor response	
Follows commands	6
Makes localized movement in response to painful stimulation	5
Makes nonpurposeful movement in response to noxious stimulation	4
Flexes upper extremities/extends lower extremities in response to pain	3
Extends all extremities in response to pain	2
Makes no response to noxious stimuli	1
Verbal response	
Is oriented to person, place, and time	5
Converses, may be confused	4
Replies with inappropriate words	3
Makes incomprehensible sounds	2
Makes no response	1

Table 2.5. Glasgow Coma Scale.

In TBI, two different effects can be distinguished: (1) **Primary damage**: resulting from the impact to the brain; (2) **Secondary damage**: response to the injury (edema, hypoxia, hypotension, vasospasm, etc.)

Types of TBI

Two major types of TBI can be distinguished: closed and open (or penetrating)

<u>Closed.</u>

In closed head injury two different possibilities are separated: concussion and contusion.

Concussion. A concussion is a significant blow to the head that temporarily affects normal brain functions and may result in unconsciousness. A concussion may result from a fall in which the head strikes against an object or a moving object strikes the head. Significant jarring in any direction can produce unconsciousness. It is thought that there may be microscopic shearing of nerve fibers in the brain from the sudden acceleration or deceleration resulting from the injury to the head. The length of unconsciousness may relate to the severity of the concussion. Often victims have no memory of events preceding the injury or immediately after regaining consciousness with worse injuries causing longer periods of amnesia

Contusion. A contusion is a bruise of the brain. It appears as softening with punctate and linear hemorrhages in crowns of the gyri and can extend into the white matter in a triangular fashion with the apex in the white matter. Old contusions appear as brownish stained triangular defects in the cortex and underlying white matter. They occur on the orbital frontal surfaces and temporal poles in most instances (Figure 2.6).

Figure 2.6. The impact of a traumatic head injury is transmitted to the anterior and orbital frontal lobe and to the anterior and mesial temporal lobe.

<u>Open (penetrating).</u>

In open head injury there is a fracture of the skull, rupture of meninges, and the brain is penetrated (for instance, a gunshot wound).

Speech and language characteristics

Motor deficits including dysarthria are frequently found in the acute stage of TBI but tend to improve with evolution. Speech defects are found in about 60% of the cases acutely and 10% in long term follow-up. Most often the speech defect corresponds to a mixed dysarthria because of the nature of the brain-damage.

Traumatic aphasia depends upon the brain zone(s) that is impacted in the trauma. If the language areas are damaged, aphasia will be observed. Furthermore, the specific aphasia characteristics depend on the specific location of the damage: left posterior frontal damage can result in a Broca´s type of aphasia; left temporal impairment in Wernicke´s type of aphasia, etc. Aphasia is more frequently found in open head injury because of the focal nature of the injury. For instance, a gunshot in the left temporal lobe most likely will result in a fluent aphasia.

In cases of closed TBI, it is not unusual to find memory deficits and attention difficulties in addition to some word-finding defects and general difficulties with complex language. Although an overt language defect may not be recognized in a routine clinical examination, specific language testing may show some mild language difficulties; the term **sub-clinical aphasia** has been used to refer to this mild language impairment that is not overtly observed, but found only with specific language testing.

Neoplasms

A neoplasm (tumor) is any growth of abnormal cells, or the uncontrolled growth of cells. **Primary brain tumors** start in the brain, rather than spreading to the brain from another part of the body. A **metastatic brain tumor** is a mass of cancerous cells in the brain that have spread from another part of the body (Figure 2.7).
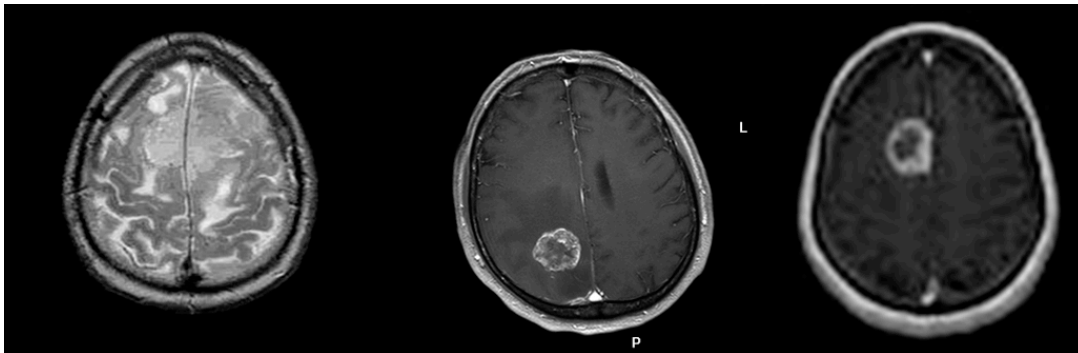
Figure 2.7 Examples of brain tumors. From left to right: meningioma, glioma, and metastatic tumor.

The specific symptoms of brain tumors are variable. The symptoms commonly seen with most types of metastatic brain tumor are those caused by increased pressure in the brain. Brain tumors are classified depending on the exact site of the tumor, the type of tissue involved, benign or malignant (cancer) tendencies of the tumor, and other factors. The cause of primary brain tumors is not well understood.

The overall incidence rate for primary brain tumors in USA has been estimated in 18.1 per 100 000 persons per year. The overall prevalence rate of individuals with a brain tumor has been estimated to be 221.8 per 100 000 in 2010. The average prevalence rate for malignant tumors (42.5 per 100.000) is lower than the prevalence for nonmalignant tumors (166.5 per 100.000) (Porter et al., 2009)

Clinical manifestations are variable and depend upon the site of the tumor. For instance: seizures, attention difficulties, headaches, and languages changes are common manifestations of brain tumors. Tumors located in the language areas are associated with aphasia-type symptomatology. However, as a general rule, the slower the growth of the tumor, the milder the symptomatology.

Although there are different types of tumors affecting the brain, gliomas (tumors originated from the glia) represent close to 50% of all brain tumors. Secondary tumors (metastatic tumors) represent a relative small percentage, close to 10% (Table 2.6).

Type of Tumor	Tumor Percentage
Gliomas	(45)
Glioblastoma	20
Astrocytoma	10
Ependymoma	6
Oligodendioglioma	5
Medulloblastoma	4
Meningioma	15
Metastatic tumors	10
Pituitary adenoma	7
Neurinoma	7
Craniopharyngioma	4
Angiomas	4
Sarcomas	4
Others	4

Table 2.6. Percentage of brain tumors

Infections

An infection appears when the body is invaded by a pathogenic micro- organism. Infectious agents include **viruses** (a small infectious agent that can replicate only inside the living cells of an organism), **bacterias** (microorganisms with a small size –some few micrometers- having a wide range of shapes, ranging from spheres –cocci- to rods –bacilli- and spirals –spirilla and spirochaetes-); **fungi** (member of a large group of eukaryotic organisms that includes yeasts, molds, and mushrooms); and **parasites** (type of non mutual relationship between organisms of different species where one organism, the parasite, benefits at the expense of the other, the host)

Infections can affect the brain tissue because they interfere with the cerebral blood flow and alter the metabolic capacity of the cells, or the characteristics of the cell membrane, changing its electric properties.

Nervous system infections are frequently secondary to infections in other parts of the body. Fever and general decrease in energy is frequently observed. Acute confusional state is frequently found in cases of brain infections: temporal-spatial disorientation, memory difficulties, naming defects, and psychomotor agitation also are found. Intracranial infections may produce widespread behavioral symptomatology. word-finding difficulties represent the most important aphasic sign in cases of brain infections.

It is interesting to refer in particular to two infections:

Herpes simplex encephalitis is a severe viral infection of the central nervous system that is usually localized to the temporal and frontal lobes. Herpes simplex encephalitis is thought to be caused by the retrograde transmission of virus from a peripheral site on the face, along a nerve axon, to the brain. The virus lies dormant in the ganglion of the trigeminal cranial nerve, but the reason for reactivation, and its pathway to gain access to the brain, remains unclear. Most individuals show a decrease in their level of consciousness and an altered mental state presenting as confusion, and changes in personality. Retrograde memory and language ability also may be impaired.

Intracerebral abscess results from the invasion of infectious organisms into the brain tissue. It is a consequence of the spread of contiguous infection from nonneural tissue, the result of hematogenous introduction from a remote site, or direct mechanical introduction as a result of penetrating trauma or a surgical procedure. A wide range of microorganisms have been recovered from intracerebral abscesses, including most types of bacteria and certain types of fungi and parasitic organisms (Figure 2.8). An intracerebral abscess can result in a focal symptomatology: when located in the brain areas supporting language, and aphasia manifestations will be evident.
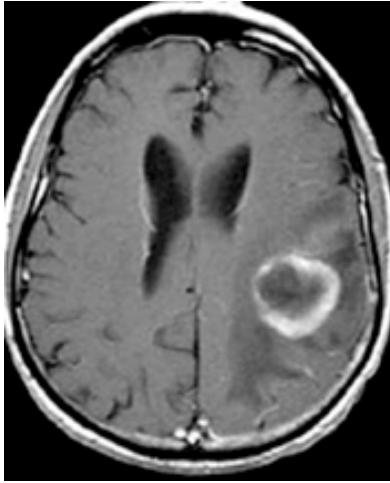
Figure 2.8. Brain abscess.

Degenerative conditions

A degenerative condition refers to a disease in which the function or structure of certain tissues or organs will progressively deteriorate over time. There is a multiplicity of degenerative conditions potentially affecting the central nervous system, such as Alzheimer's disease, Parkinson's disease, Huntington's disease, Amyotrophic Lateral Sclerosis, Progressive supranuclear palsy and multiple system atrophy. In this chapter, Alzheimer´s disease, progressive aphasia (as a special subtype of Alzheimer´s disease), and some other degenerative conditions will be examined. The specific language characteristics in different types of dementia will be examined in Chapter 8 ("Associated Disorders").

Alzheimer's disease

Alzheimer's disease is the most common cause of dementia among older people. Dementia is understood as the loss of cognitive functioning— such as thinking, remembering, and reasoning—and behavioral abilities, to such an extent that it interferes with a person's daily life and activities.

The DSM-IV-TR (APA, 2002) includes the following criteria for the diagnosis of Dementia of the Alzheimer's Type:

A. The development of multiple cognitive deficits manifested by both: 1. Memory impairment; 2. One or more of the following cognitive disturbances: (a) aphasia; (b) apraxia; (c) agnosia; (d) disturbance in executive functioning.

B. The cognitive deficits in criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.

C. The course is characterized by gradual onset and continuing cognitive decline.

D. The cognitive deficits in Criteria A1 and A2 are not due to another condition.

Dementia ranges in severity from the mildest stage, when it is just beginning to affect a person's functioning, to the most severe stage, when the person must depend completely on others for basic activities of daily living. Dementia is associated with brain atrophy and ventricular enlargement (Figure 2.9). During normal aging, brain atrophy and ventricular enlargement are also observed, but in Alzheimer's disease they are notoriously more pronounced.
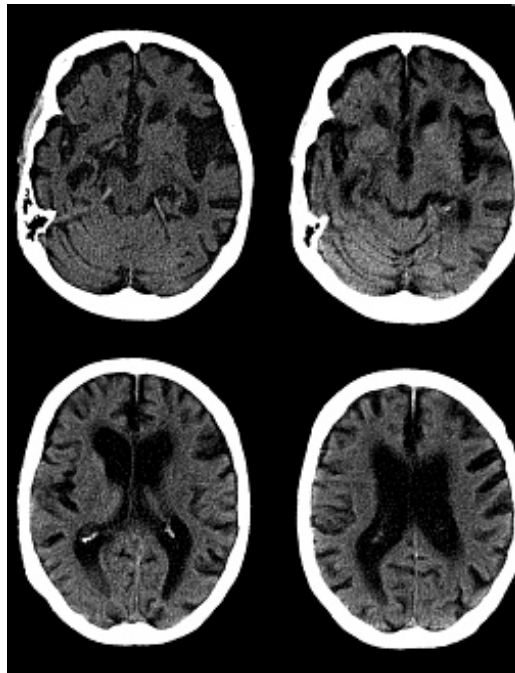
Figure 2.9. CT scan of a patient with Alzheimer's disease. Cortical atrophy and ventricle enlargement are evident.

Language disintegration follows a particular sequence: initially, word-finding difficulties and anomia are found, associated with difficulties in understanding complex language; active and passive vocabulary progressively decreases. Semantic paraphasias (semantic substitutions such as "table" instead of "chair") become more and more abundant. Later on in the disease evolution, phonological paraphasias (phonological substitutions due to phoneme additions, omissions or substitutions) also are observed. Progressively, expressive language is reduced and semi-mutism is found. However, some language abilities may remain intact even in advances stages of dementia. Language repetition and grammar are well- preserved. Mechanics of reading also may be preserved.

Progressive aphasia

Primary progressive aphasia (PPA) is a rare neurological syndrome characterized by a continuous deterioration of language. People with primary progressive aphasia may have trouble naming objects or may misuse word endings, verb tenses, conjunctions and pronouns. Symptoms of primary progressive aphasia begin gradually, frequently before the age of 65, and tend to worsen over time. When a CT scan is undertaken, usually local atrophy in the brain-language areas is observed, but later on in the evolution, not only language difficulties but also other cognitive defects are found. Eventually, the patient will present an Alzheimer´s disease, including not only language defects, but also memory impairment and other disturbances in cognition (Mesulam, 1982, 2001).

Subtypes of primary progressive aphasia have been described, in particular a non-fluent type similar to Broca´s aphasia (so-called "**progressive nonfluent aphasia**") associated with frontotemporal lobar degeneration and a fluent subtype, in occasions referred as "**semantic dementia**" (Grossman & Ash, 2004). In progressive nonfluent aphasia, agrammatism (impairment in use of grammatical and syntactic constructs of language), phonological paraphasias, apraxia of speech, and articulatory difficulties are found. In semantic dementia, word-finding difficulties, anomia, impaired comprehension and more verbal paraphasias are observed.

Not only disturbances in oral language can appear as the initial manifestation of an Alzheimer's disease (progressive aphasia). Progressive disturbances in other abilities (e.g., visual perception, writing, etc) also have been described. For instance, Ardila, Matute and Inozemtseva (2003) reported a case of a 50-year-old, right-handed female who, over approximately two years, presented with a progressive deterioration of writing abilities associated with acalculia and anomia. An MRI disclosed a left parietal temporal atrophy (Figure 2.9). Two years later, further significant cognitive decline consistent with a dementia of the Alzheimer's type was observed. Amnesia, executive dysfunction, and ideomotor apraxia were found. Writing was severely impaired, and some difficulties in reading were observed. A second MRI approximately two years later showed that brain atrophy had progressed significantly. Spontaneous writing and writing to dictation were impossible. The ability to read words was preserved, but the ability to read pseudowords was lost.
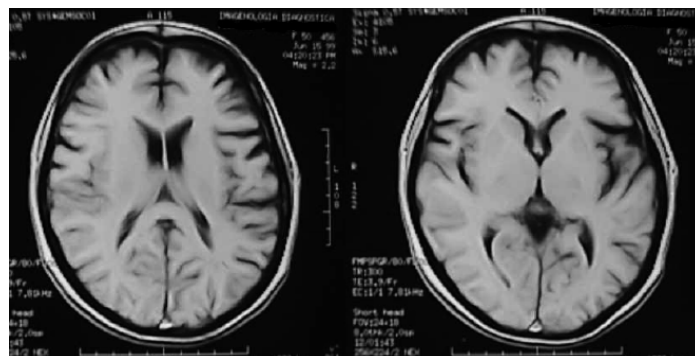
Figure 2.9. Progressive agraphia and anomia (according to Ardila et al., 2003).

Other degenerative conditions

In other degenerative conditions, such as **Parkinson's disease** and **Huntington's disease,** some language disturbances can be found. However, in these two conditions there is a subcortical dementia characterized by slowness in cognition, retrieval defects in memory, and executive functioning defects. Dysarthria is evident (hypokinetic dysarthria in Parkinson´s disease, and hyperkinetic dysarthria in Huntington´s disease) but aphasia is mild. In general, decreased verbal fluency, difficulty in the comprehension of complex commands, and word-finding difficulties (anomia) are identified.

In other degenerative conditions, speech and language impairment also may be observed. For instance, **Amyotrophic lateral sclerosis (ALS**), sometimes called **Lou Gehrig's disease,** is a rapidly progressive, invariably fatal neurological disease that attacks the nerve cells responsible for controlling voluntary muscles. This disease belongs to a group of disorders known as **motor neuron diseases**, which are characterized by the gradual degeneration and death of motor neurons. Significant motor disturbances associated with preserved cognition are observed. Dementia, however, has been occasionally reported (problem solving, attention, memory, naming defects). Speech impairments are evident corresponding to a mixed dysarthria.

Summary

Any abnormal condition affecting the brain areas involved in language processing can result in aphasia. The specific symptoms of the language impairment depend upon the particular brain area that is affected. Pathological conditions affecting the posterior frontal areas of the left hemisphere usually result in a nonfluent disorder of language, characterized by agrammatism with an accompanying apraxia of speech, whereas pathological conditions affecting the temporal and partially the parietal lobe in the left hemisphere are associated with disturbances in language understanding, word-finding difficulties, and paraphasias. Different etiologies of brain damage are recognized, but the vascular disorders and traumatic brain injury represent the two major causes of aphasia. Brain tumors, infections, and some degenerative conditions also may be associated with aphasia.

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Chapter 3

Linguistic analysis of aphasia

Introduction

Aphasia is the loss or impairment of **language** function caused by **brain damage** (Benson & Ardila, 1996)**.** In consequence, aphasia has a linguistic and a neurological dimension. In the previous chapter, the brain damage (neurological dimension) potentially associated with aphasia was reviewed. In this chapter, the specific language disturbances (linguistic dimension) observed in aphasia will be reviewed.

Language is a communication system. There are different communication systems and consequently, different types of language: sign language, animal languages, computers languages, etc. Human language is a specific example of a communication system characterized by the use of a limited amount of articulated sounds (**phonemes**), which can be combined in different ways to create meaningful units (**morphemes** and **words**) (so called "**double articulation**"; that is, speech output can be divided into meaningful elements –words-, which can be further subdivided into sound elements –phonemes-). **Tongue** is the specific verbal communication system characteristic of a human community (for instance, English, Spanish, Chinese, etc). **Speech** refers to the phonoarticulatory act that produces the acoustic signal in which phonemes and words are coded. Consequently, human language is a cognitive process, whereas speech is a neuromuscular process. Impairments in languages are referred as **aphasias**, whereas impairments in speech are known as **dysarthrias**.

In this chapter, initially some basic linguistic ideas will be introduced; later, the languages disturbances associated with brain pathology will be analyzed. Finally, a linguistic interpretation of aphasias will be presented.

Levels of analysis of language

Different levels of analysis of the language can be distinguished:

- Phonetic
- Phonemic
- Morphemic
- Morphosyntactic (grammatical)
- Semantic
- Pragmatic

Phonetic

Phonology is a branch of linguistics concerned with the analysis of speech sounds in human languages. Human languages use a relatively limited amount of sounds in communication, which are classified in the so-called International Phonetic Alphabet (Table 3.1). However, each particular tongue (e.g., English, Spanish, etc.) only uses some few (usually about 15-50) different functional sounds, known as phonemes. A phoneme, in consequence, is the smallest segmental unit of sound used to form words and meaningful language contrasts.

Table 3.1. International Phonetic Alphabet

A **phonetic variation** is a variation in a phoneme that does not change the meaning of the utterance; that means, it is not functional. For example, /r/ (alveolar tap) - /ŕ/ (alveolar trill) is a phonetic variation in English, but a phonemic contrast in Spanish. In Spanish (but not in English) /r/ and /ŕ/ are in phonemic contrast (for example, *perro* 'dog' vs *pero* 'but'). On the other hand, /ʃ/ - / t ʃ / (SH-CH) is a phonetic variation in Spanish, but a phonemic contrast in English (for example, "ship"-"chip"*).

In normal conditions, phonetic variations are due to different factors: for instance, to the specific position of the phoneme in the word (i.e. the phonemes before and after), to dialectal variations of the speaker, etc.

Phonemic

A **phoneme** is the minimal language sound capable of conveying meaning. It has been assumed that some phonemes are found in every human language (e.g. /a/), whereas other phonemes are found only in a limited amount of languages, and even in a single language. The number of phonemes in a language is variable; for instance, in English there are about 34 phonemes (Table 3.2) and in Spanish about 23 (Table 3.3) (this difference is mainly due to the increased amount of English vowels, some 12-13 vowels; while in Spanish, the number of vowels is limited, only five, but the number of diphthongs is enormous).

		bilabial	labio-dental	inter-dental	alveolar	palatal	velar	glottal
stops	voiceless	p			t		k	?
	voiced	b			d		g	
affricates	voiceless					č		
	voiced					J̌		
fricatives	voiceless		f	θ	s	š	h	
	voiced		v	ȝ	z	ž		
nasals		m			n		ŋ	
Liquids	lateral				l			
	flap				r			
	retroflex						ɹ	
semivowels		w					y	

Table 3.2. English phonological system (English consonants)

	Labial	Dental	Alveolar	Palatal	Velar
Nasal	m		n	ɲ	
Stop	p b	t d		tʃ ɟ	k g
Fricative	f	θ	s	ʃ	x
Trill			r		
Tap			ɾ		
Lateral			l	ʎ	

Table 3.3. Spanish phonological system (Spanish consonants)

Morphemic

A **morpheme** is the smallest semantically meaningful unit in a language. It is composed by one phoneme (e.g., "a" as an indefinite article) or several phonemes (e.g., "car"). It corresponds to a word (e.g., "book") or to a word element (e.g., "book" in the word "books"). Morphemes are also known as **monemes.**

Two types of morphemes can be distinguished

(1) **Free morphemes** can stand by themselves. They can be:

-**Lexical (or radical or root) morphemes** (e.g., "car" in "cars")
-**Grammatical free morphemes (connectors)** (e.g., prepositions and articles)

(2) Bound morphemes: they need to be attached to a free morpheme (e.g., the plural "s" in "cars"). They can be:

-**Derivational morphemes**: they *derive* new words (e.g., "ness" in "happiness")
-**Grammatical (inflectional) morphemes:** They encode grammatical information (e.g., the plural "s" in "cars").

The term **affix** describes where a bound morpheme is attached to a word (**prefixes:** attached at the onset of a free morpheme; **suffixes:** attached to the end; **Infixes:** affixes that occur in the middle of a word)

Example: in the following sentence there are seven words and nine different morphemes:

The boys are walking in the street

Morphosyntactic (Grammatical)

Grammar refers to the rules governing the use of language. It includes:

-**Morphology:** the study of word formation (e.g., past tense: verb+ed: e.g., want+ed)

-**Syntax:** the study of how words are combined into larger units such as phrases and sentences. (e.g., article + noun: "the house"; never noun + article: "house the")

Morphosyntaxis refers to grammatical categories or properties for which the definition criteria of morphology and syntax both apply, as in describing the characteristics of words (Crystal, 1980)

Semantic

Semantics is the study of the meaning of linguistic expressions. Each word has a particular semantic field. "Table" is a word corresponding to a semantic category (concept) from the linguistic point of view; and to a percept from the perceptual point of view. It is "a piece of furniture supported by one or more vertical legs and having a flat horizontal surface, used to lay out different articles" (usual definition), but many different objects fulfill this definition (Figure 3.1) and correspond to this semantic category.

Figure 3.1. Examples of objects included in the semantic field of the word "table"

However, some elements can be considered as "prototypes" of that semantic category, whereas others should be regarded as "peripheral" elements (Figure 3.2).

_____

Semantic category "Fruit"

| | orange 🟠 | avocado 🥑 |
| **Semantic field** | banana 🍌 | tomato 🍅 |

Prototype   *Peripheral*

_____
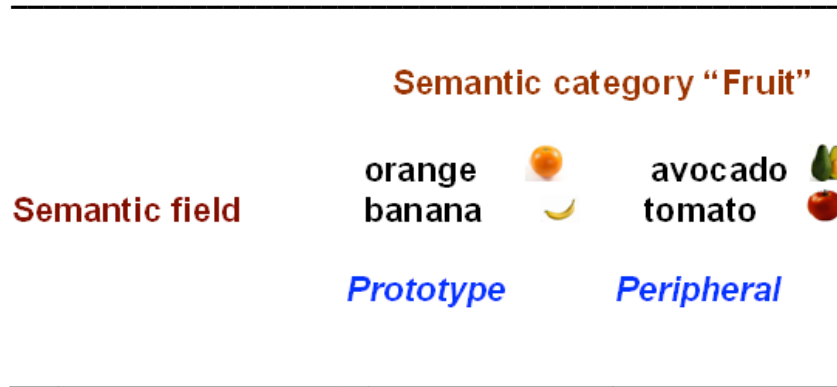
Figure 3.2. Some elements can be considered as "central" or prototypes elements in a semantic category; other may be more "peripheral".

The meanings of the words (and the concepts attached to the words) are organized hierarchically. For instance, canary is a bird, and a bird is an animal; but there are other birds, and there are other animals (Figure 3.3.)
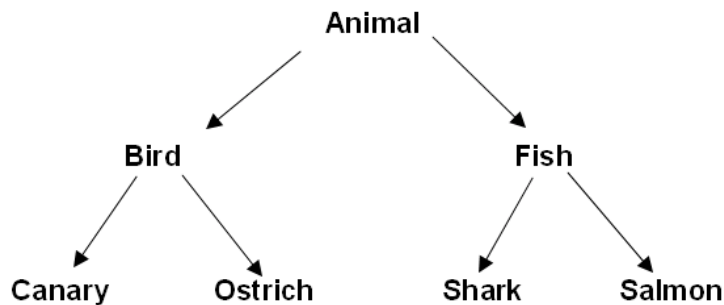
Figure 3.3. Words (and concepts) are hierarchically organized.

Finally, it is important to note that the **semantic associations** of words may correspond to different sensory systems. That is, we have a "mental" representation of the meaning of the word "house" that only corresponds to some visual associations (we know houses visually, not auditorily or tactilely); but we have both visual and auditory representations of a "phone" (we can recognize a phone from some visual and also auditory information); a "key" can be recognized using visual, tactile, or auditory information (that means, we have a visual, tactile, and auditory

representation of "keys"). An ice cream can be recognized using visual but also gustatory information (the meaning of "ice cream" is visually but also gustatorily mediated); and a flower can be recognized using visual and also olfactory information (our mental representation of flowers include visual and olfactory elements).

Pragmatic

Pragmatics is the linguistic branch concerned with the use of language in everyday social contexts. It means, how language is to be used in the real life, in real contexts.

The specific use of the language depends on the situational context and the conversational partner. As a matter of fact, the language to be selected when speaking with children is different from the language used to talk with our colleagues; when talking with children we have to use a simpler grammar, and higher frequency words, than when talking with our colleagues. The language to be used when shopping or when presenting a scientific lecture is not exactly the same; the language used to talk with our old friends from infancy and adolescence usually include words and expressions that were used during infancy and adolescence, but are probably unusual today. The way we talk to our friend and to our boss may be slightly different.

Thus, the analysis of the practical use of language corresponds to the linguistic area known as pragmatics.

Linguistic defects in aphasia

Levels of the language impaired in different aphasias

The different levels of the language (phonetic, phonemic, morphemic, morphosyntactic, semantic, and pragmatic) can be differentially affected in the different types of language disturbances. For instance, Broca's aphasia is significantly associated with grammatical defects (so-called **agrammatism** in Broca's aphasia), whereas the semantics of nouns is impaired in posterior fluent aphasias. Table 3.3 presents, in a summarized way, the levels of the language impaired in different speech/language disorders.

Level of the language	Type of speech/language impairment
Phonetic	Dysarthria, Broca's
Phonemic	Wernicke's, conduction
Morphemic	Broca
Morphosyntactic	Broca
Semantic	Extrasylvian sensory, Wernicke
Pragmatic	Extrasylvian motor (dysexecutive)

Tabla 3.3. Levels of the language impaired in different speech/language disorders.

The different aphasia subtypes are characterized by specific language disturbances. For instance, in conduction aphasia, the core language defect refers to language repetition disturbances; whereas in extrasylvian motor (dysexecutive) there is an evident fundamental defect in the pragmatic use of the language. This relationship between the type of aphasia and the core or central language impairment is presented in Table 3.4.

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Type of aphasia	Linguistic impairment
Broca's aphasia	Agrammatism
Wernicke's aphasia	Phonological discrimination Verbal memory
Conduction	Language repetition defects
Extrasylvian sensory	Semantic relations
Extrasylvian motor (dysexecutive)	Pragmantic function

───

Table 3.4. Language impairments in different types of aphasia

Language deviations

Patients with aphasia present diverse types of language deviations (Table 3.5). However, these language deviations or abnormalities vary according to the specific aphasia subtype.

─────────────────────────────

Phonetic deviations
Phonological (literal) paraphasias
Verbal paraphasias
Syntagmatic paraphasias
Circumlocutions
Neologisms
Jargon
Agrammatism
Paragrammatism

─────────────────────────────────

Table 3.5. Language deviations in aphasia (according to Ardila & Rosselli, 1993)

Phonetic deviations
They refer to misproduced phonemes that can still be recognized. They are specially observed in Broca's aphasia. Sometimes they result in kind of a "foreign accent" (phenomenon known as "**foreign accent in aphasia**").

Phonological (literal) paraphasias
They are words that are incorrect from the point of view of the phonological composition. They are also referred as **phonemic** or **literal** paraphasias. The errors can be due to:

> -**Phoneme omissions** (e.g., *pencil->pecil)*
> -**Phoneme additions** (e.g., *pencil->prencil)*
> -**Phoneme displacements** (e.g., *pencil->pelcin)*
> -**Phoneme substitutions** (e.g., *pencil->percil)*

Luria (1976) suggested that the phonological paraphasias in conduction aphasia (Luria's "afferent motor aphasia") are due to confusions in the articulation of the phonemes ("**articulemes**"), and hence, should be regarded as **articulatory paraphasias**.

Verbal paraphasias
They refer to the substitution of meaningful units in language. There are several possibilities:

> **-Formal paraphasias:** the replacing and replaced words are similar in their phonological composition but not in their meaning; they can also be interpreted as phonological paraphasias (e.g., *cat->can)*
> **-Morphemic paraphasias:** erroneous use of bound morphemes (e.g., *summerly)*
> **-Semantic paraphasias:** both words (replacing and replaced) are semantically related (e.g., *cat->dog*)
> **-Unrelated paraphasias:** sometimes aphasic patients introduce words that do not seem to have any relationship with the current linguistic context (e.g., *cat->pencil*)

Semantic paraphasias represent the most important subtype of verbal paraphasias. There are several possibilities:

> 1. Both words (replacing and replaced) correspond to the **same semantic field** (e.g., *hand->foot).*
> 2**.** They are **antonyms** (e.g., *small->big)*
> 3**.** The replacing word is a **superordinate** word with regards to the replaced one (e.g., *cat->animal).*
> 4. There is an **environmental proximity** between both words (e.g., *pencil->paper)*

Syntagmatic paraphasias
A paraphasia does not necessarily refer to a single word. Substitutions may appear with more complex linguistic units (e.g., *the aquarium of the fish -> the cage of the lion).*

Circumlocutions
When unable to find a name, the aphasic patient can replace the name by a circumlocution (e.g., *pencil->for writing).* Usually, the circumlocution refers to the function of the object; on

occasion, to the object's composition (e.g., *pencil->that long wooden object with something inside).*

Neologisms
Sometimes, the target word has been so significantly changed, that is unrecognizable (e.g., *pencil->cartin).* That is called **aphasic neologism**.

Jargon
Jargon refers to a fluent, abundant, well-articulated language output that lacks meaning for the listener. Sometimes a further distinction is established among three different types of jargon, according to the type of language deviation that predominates in the patient, making impossible to understand his/her speech: **phonological jargon**, **semantic jargon**, and **neologistic jargon**. It is supposed that the jargon is due to diverse deviation in the patients' language output. For instance, the sentence "*I use a pencil for writing*" can become:

> In **phonologic jargon**: "*I ute a telcin vor liting*"
> In **semantic jargon**: "*I throw a glass for reading*"
> In **neologistic jargon**: "*I tro a plas por leti*"

Agrammatism
Agrammatism is a disruption of the grammatical structure of the language, observed in Broca's aphasia, characterized by a reduction in the use or omission of grammatical morphemes (e.g., "*The child is playing in the yard*"-> "*child play yard*")

Paragrammatism
Paragrammatism refers to a verbal output that violates the normative rules of morphosyntactic conventions (e.g., "*The child is playing in the yard*"-> "*The, I mean one, is now on, there outside, taking something to play on that, over there*"). It is due to:

> -Overuse of grammatical words
> -Erroneous selection of grammatical words
> -Absence of defining limits of the sentences

Repetition

The ability to repeat represents one of the major elements in aphasia classification. Different aphasia groups can be separated according to the ability to repeat:

1. **Preserved repetition ability**: Extrasylvian (transcortical) aphasias
2. **Impaired repetition ability**: Perisylvian aphasias

Furthermore, it has been proposed that the ability to repeat represents a major criterion for aphasia classification. However, some authors (e.g., Ardila & Rosselli, 1992) have proposed that different mechanisms may underlie repetition deficits in aphasia: limitation of auditory-verbal short-term memory, difficulties at the level of phonological production, impairments in phoneme recognition, and semantic and syntactic comprehension; furthermore, all the aphasia

groups present at least some errors in language repetition. Errors are not only quantitatively but also qualitatively different.

Depending upon the specific task, errors can be high or low in a particular group of aphasic patients: Some patients have difficulties resulting from verbal memory limitations (anomia); other patients have difficulties at the level of phonological production (Broca's and conduction aphasia); others may have defects in grammar comprehension (Broca's aphasia) and the use of complex syntax (extrasylvian motor aphasia), etc.

Naming

Naming difficulties represent the most common defect in aphasia. Virtually all aphasic patients present impairments in naming. However, the specific characteristic of the naming defect can be significantly different in different aphasia groups.

As a matter of fact, **anomia** is a term with two different meanings in aphasiology. Anomia refers to:

(1) "*word-finding defects; naming impairment or failure*". With this broad meaning of the word, all aphasic patients may present anomia, even though the deficit can be manifested in rather different ways.

(2) Anomia has also been used to refer to the difficulties in finding words associated with circumlocutions and semantic paraphasias; that is observed in cases of temporal-occipital damage. This type of language disturbance in general corresponds to so-called ***anomic, amnesic*** or ***nominal aphasia.***

A linguistic interpretation of aphasias

Jakobson (1964, 1971; Jakobson & Halle, 1956) emphasized that there are two basic linguistic operations: **selecting** (language as **paradigm**) and **sequencing** (language as **syntagm**). Aphasia tends to involve one of two types of linguistic deficiency. A patient may lose the ability to use language in two rather different ways: the language impairment can be situated on the paradigmatic axis (**similarity disorder**) or the syntagmatic axis (**contiguity disorder**).

The similarity disorder restricts the patient's ability to select words on the paradigmatic axis. These patients (Wernicke-type aphasia) cannot find words that exist as parts of the system (vocabulary). These aphasic patients have severely limited access to this language repertoire system. Specific nouns tend to be inaccessible.

These patients cannot select among alternative names (e.g., apple, pear, banana, etc.) and may instead fill out their discourse with circumlocutions (e.g., a clock may be referred to as ''to know the time''). Words no longer have a generic (paradigmatic) meaning for these patients, and speech becomes empty. A dog can be referred to as ''fox'', ''it barks'', etc.

Aphasic individuals presenting with what Jakobson referred to as contiguity disorder (Broca-type aphasia), on the other hand, lose the ability to combine linguistic elements. Their grammar is restricted or absent, and they can produce and understand only isolated meaningful words. Words with purely grammatical functions (such as articles and prepositions) tend to be omitted. Affixes may be substituted for one another, but more likely they are simply not produced. These patients thus tend to use only very short sentences containing mostly meaningful words (nouns). In severe cases, sentences can be as short as a single word (''dog'') and in general, there is a reduction in resources available for syntactic processing (Caplan, 2006).

Luria further developed Jakobson's ideas in his paper ''On the two basic forms of aphasic disturbances'' (1972/1983). Luria emphasized that the selection disorder can be observed at different levels of language, corresponding to different aphasia subtypes: phoneme selection (aphasia acoustic agnosic), word selection (aphasia acoustic amnesic), and meaning selection (amnesic aphasia). By the same token, the contiguity disorder can be observed at different levels: sequencing words (kinetic motor aphasia—Broca's aphasia) or sequencing sentences (dynamic aphasia— transcortical motor aphasia) (Luria, 1976). It should be noted that different subtypes of Wernicke's aphasia are frequently distinguished (e.g., Ardila, 2006). Luria's acoustic agnosic, acoustic amnesic, and amnesic aphasia are indeed subtypes of the language impairment syndrome referred to as a whole as Wernicke's (sensory) aphasia.

Summary

Human language is a communication system characterized by a "double articulation". Different levels of analysis of the language can be distinguished: phonetic, phonemic, morphemic, morphosyntactic (grammatical), semantic, and pragmatic. These levels of language are impaired in a specific way in each aphasia subtype.

Patients with aphasia present diverse types of language deviations, including, phonetic deviations, phonological (literal) paraphasias, verbal paraphasias, syntagmatic paraphasias, circumlocutions, neologisms, jargon, agrammatism, and paragrammatism. Their manifestations depend upon the aphasia subtype.

The ability to repeat is a major element in aphasia classification: patients with perisylvian aphasias have repetition disturbances, whereas patients with extrasylvian (transcortical) aphasias have a normal or near normal repetition ability. It has been proposed that different mechanisms may underlie repetition deficits in aphasia.

Naming difficulties represent the most common defect in aphasia. The term "anomia" has been used in two different ways in aphasiology. Virtually all the aphasic patients present impairments in naming. However, the specific characteristic of the naming defect can be significantly different in different aphasia groups.

It has been emphasized that there are two fundamental linguistic operations, and aphasia tends to involve one of two types of linguistic deficiency. In consequence, there are two basic

language disorders in aphasia: **similarity disorder** in Wernicke's aphasia and **contiguity disorder** in Broca's aphasia. There are consequently two basic types of aphasia.

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II. CLINICAL MANIFESTATIONS

Chapter 4

Major aphasic syndromes: Wernicke's aphasia and Broca's aphasia

Introduction

Since the 19th century, it has been well established that there are two major and fundamental aphasic syndromes, named in different ways, but roughly corresponding to Wernicke-type aphasia and Broca-type aphasia (e.g., Albert et al., 1981; Alexander & Benson, 1991; Ardila, 2010, 2011, 2012; Bastian, 1898; Benson & Ardila, 1996; Freud, 1891/1973; Goldstein, 1948; Head, 1926; Hécaen, 1972; Kertesz, 1979; Lichtheim, 1885; Luria, 1976; Pick, 1931; Schuell, Jenkins, & Jimenez-Pabon, 1964; Taylor-Sarno, 1998; Wilson, 1926; see Tesak & Code, 2008, for review). This is a most basic departure point in aphasia: *Aphasia is not a single and unified clinical syndrome, but two rather different (even opposed) clinical syndromes.*

These two major aphasic syndromes have been related to the two basic linguistic operations: **selecting** (language as paradigm) and **sequencing** (language as syntagm) (Jakobson, 1971; Jakobson & Halle, 1956; Luria, 1972/1983). Jakobson (1964) proposed that aphasia tends to involve one of two types of linguistic deficiency. A patient may lose the ability to use language in two rather different ways: the language impairment can be situated on the paradigmatic axis (**similarity disorder**) (Wernicke's aphasia) or the syntagmatic axis (**contiguity disorder**) (Broca's aphasia). Table 4.1. presents some of the names that have been used to refer to these two fundamental aphasic syndromes.

Receptive	Expressive
Sensory	Motor
Posterior	Anterior
Fluent	Non-fluent
Paradigmatic disorder	Syntagmatic disorder
Decoding disorder	Coding disorder
Wernicke-type	Broca-type

Table 4.1. Names used to refer to the two fundamental aphasic syndromes. In the first one the disturbance is located at the lexical/ semantic level, whereas in the second case, the disturbance corresponds to a grammatical impairment.

In this chapter these two major (primary) aphasic syndromes will be examined. In the following chapter, other aphasia syndromes such as conduction aphasia, aphasia of the supplementary motor area, and extrasylvian (transcortical) aphasias will be analyzed. Noteworthy, a significant percentage of aphasias observed in clinical settings –probably 30%-50%- corresponds to mixed forms of aphasia, although a particular type of aphasia may predominate. For example, a patient can present a Broca's aphasia plus a milder conduction aphasia.

Wernicke's aphasia

Wernicke aphasia has been named in many different ways: **sensory aphasia** (Wernicke, 1874), **receptive aphasia** Weisenburg & McBride, 1964), **central aphasia** (Brain, 1961), **verbal agnosia** (Nielsen, 1936) and others.

Wernicke's aphasia results from pathology in Wernicke's area. Wernicke's area corresponds to the auditory association area of the left hemisphere (Figure 4.1). There is, however, some disagreement about the exact limits of Wernicke's area. The primary auditory area corresponds to Brodmann's area (BA) 41 (Heschel's gyrus, or transverse temporal gyrus), and some authors also include BA42. It could be assumed that Wernicke's area corresponds to BA22, 21, and 37; frequently BA39 is also included.
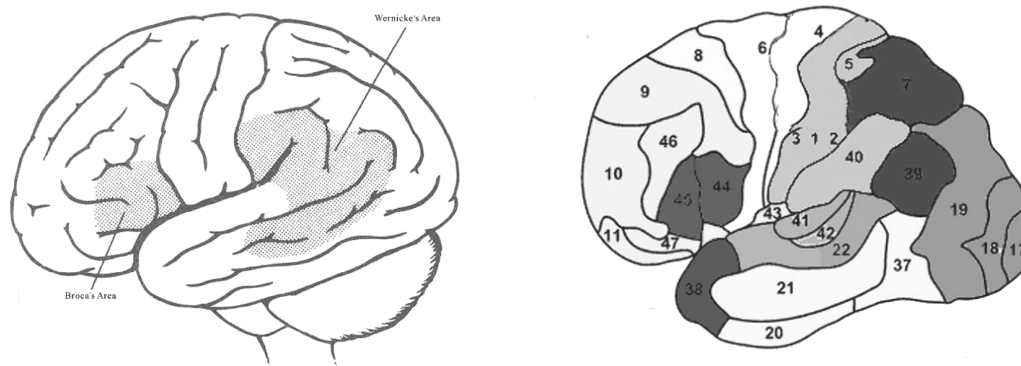
Figure 4.1. Traditionally it has been accepted that there are two major areas involved in language: frontal Broca's area (BA44 and probably BA45) and temporal Wernicke's area (BA22, 21, and 37, although BA39 is also frequently included.)

Wernicke's aphasia represents a clinical syndrome with well-defined characteristics but significant variability.

In Wernicke's aphasia the lexical repertoire (vocabulary) tends to decrease and language-understanding difficulties are evident. Wernicke's aphasia patients may not fully discriminate the

acoustic information contained in speech. Lexical (words) and semantic (meanings) associations become deficient. In Wernicke-type aphasia, evidently the language deficit is situated at the level of language sounds (phonemes) and meaningful words (nouns). Phoneme and word selection are deficient, but language syntax (contiguity: sequencing elements) is well preserved and even overused.

Speech is produced without effort. No articulatory defects (dysarthria) are observed. Fluency is normal and frequently there is excessive language output. Often extra syllables in words and extra words in sentences are found; this excessive amount of language without a clear meaning is referred as **logorrhea**. Because of the relative absence of meaningful words (so called "**empty speech**") and the excessive language output, an overuse of grammatical words – frequently incorrectly selected) is found, this phenomenon is known as **paragrammatism** (or **dyssyntaxis**). Dyssyntaaxis has been defined as "*Pathological linguistic productions in which are observed a fairly large number of sentences that transgress one or more of the normative rules of the community's morphosyntactic convention*" (Berube, 1991; page 62). Table 4.2 is a presentation of the basic language characteristics found in Wernicke's aphasia

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Conversational Language	Fluent, paraphasic
Language comprehension	Abnormal
Repetition	Abnormal
Pointing	Abnormal
Naming	Abnormal
Reading: Aloud	Abnormal
Comprehension	Abnormal
Writing	Abnormal

Table 4.2. Basic language characteristics in Wernicke's aphasia.

Paraphasias are abundant. They can be both phonological and verbal, even though phonological or verbal paraphasia can predominate in a specific patient. Frequently, neologisms are also found. When a patient presents abundant (even excessive) verbal output that is difficult to understand due to the significant amount of paraphasias and neologisms and the relative absence of meaningful words (nouns), the term **jargonaphasia** is used. In Table 4.3, an example of the expressive language found in Wernicke's aphasia is presented.

I I don't know how there is any single way, there's so many thing, you know, that I like. I like meats, I have liked beef, the Germans, you know, and what, well the French you koot the whole, I can't recall the word that I can't thay. It was the ___ where you make all the food, you make it all up today and keep it till the next day. With the French, you know, uh, what is the

name of the word, God, public serpinz they talk about, uh but I
have had that, it was ryediss, just before the storage you know,
seven weeks, I had personal friends that, that, I would cook an'
food the food and serve fer four or six mean for an evening.

Table 4.3. Speech Pattern of a Wernicke's Aphasic Responding to a Question about What He Liked to Cook (taken from Saffran, E.M. (2000). Aphasia and the Relationship of Language and Brain. Sem Neurology 20(4):409-418)

Associated neurological signs are minimal (Table 4.4); sometimes, when the lesion extends deep to involve the optic radiation, the patient can present a superior **quadrantanopia** (defective vision or blindness in one fourth of the visual field.). By the same token, if the damage extends toward the parietal lobe, some **ideomotor apraxia** (impairment in the production of learned (or skilled) movements not caused by weakness, paralysis, incoordination, or sensory loss) can be observed. At the aphasia onset, particularly in cases of extensive damage, some mild but transient motor difficulties can also be observed.

Motor system	Normal
Articulation	Normal
Cortical sensory function	Normal
Praxis	Normal
Visual field	Normal or superior quadrantanopia
Visual gnosis	Normal

Table 4.4. Associated neurological signs in Wernicke's aphasia

These patients present significant difficulties in language understanding. However, language understanding impairments are not exactly the same all the time, but present significant variations according to different contextual conditions. When short sentences are used, it is notoriously easier for the patient to understand; increasing the number of words in speech results in more severe language understanding defects. Language understanding requires an increased attention and continuous effort (similar to the attention and effort required to understand a foreign language). Usually at the beginning of the conversation the patient has very remarkable language comprehension defects, but progressively language understanding increases. The language understanding remains relatively high for some short time (may be 15 or 20 minutes) but later it begins to decrease (**fatigue phenomenon**). Furthermore, if changes in the conversational topic are introduced, language understanding immediately decreases. So, language understanding difficulties are variable according to the specific conversational conditions.

Reading ability is also impaired, but patients with Wernicke's aphasia present variability in their reading ability. Sometimes reading is relatively well preserved (at least the mechanics of reading) while other times it is significantly impaired; it has been suggested that the closer to the

primary auditory area the pathology located, the language impairment will be more similar to a **word-deafness** (that is, significant defects in auditory recognition of language; milder defects in the visual recognition of language); while if the pathology is situated closer to the occipital lobe, the language impairment will be more similar to a **word-blindness** (that is, significant defects in the visual recognition of language; milder defects in the auditory recognition of language).

As a matter of fact, written language defects are similar to the spoken language impairments. No changes in calligraphy are noted (the patient has no motor defects for speaking either); but writing contains a significant amount of verbal and literal **paragraphias** (i.e., incorrectly written words; parallelizing the spoken verbal and phonological paraphasias); neologisms can also be noted. The term **jargonagraphia** has been used to refer to abundant (even excessive) written verbal output that is difficult to understand due to the significant amount of paragraphias and neologisms and the relative absence of meaningful words (nouns).

It is assumed that there are two major defects accounting for the language understanding defects in Wernicke's aphasia: (1) Defects in phoneme discrimination; the extreme situation (i.e., complete inability to discriminate the speech phonemes) corresponds to so-called **pure word-deafness**. Usually, patients with Wernicke's aphasia have some defects in phoneme discrimination; in severe cases, the patient can suggest that he/she is unable to understand the phonological composition of speech (for instance, he/she may state that other people seemingly are speaking using a foreign language, or even, they are not really speaking but making noises); (2) defects in verbal memory: the patient cannot recall the previously learned verbal information, such as words, sentences, and in general verbal knowledge (i.e., there is a **retrograde verbal amnesia**); and the patient also has significant difficulties in memorizing new verbal information (i.e., there is an **anterograde verbal amnesia**). For instance, repetition of sentences is limited to 3-4 word long sentences. Nonetheless, for understanding conversational language, it is required that an individual is able to keep in operative (working) memory 7-8 words, and hence, to be able to repeat 7-8 word long sentences. It is consequently obvious that patient's with Wernicke's aphasia cannot correctly understand spoken language.

Consequently, at least two major subtypes of Wernicke's aphasia can be distinguished: (1) Wernicke's aphasia with predominantly defects in phoneme discrimination (**Wernicke's aphasia type I**; or **Luria's acoustic-agnosic aphasia**); and (2) Wernicke's aphasia with predominantly defects in verbal memory (**Wernicke's aphasia type II**; or **Luria's acoustic-amnesic aphasia**). The first one is associated with lesions close to the primary auditory area (first or superior temporal gyrus) while the second is associated with lesions at the level of the second or middle temporal gyrus. Often, both defects (phoneme discrimination deficits and impairments in verbal memory) appear simultaneously in the same patient. Figure 4.2 is an illustration of a typical lesion in Wernicke's aphasia.
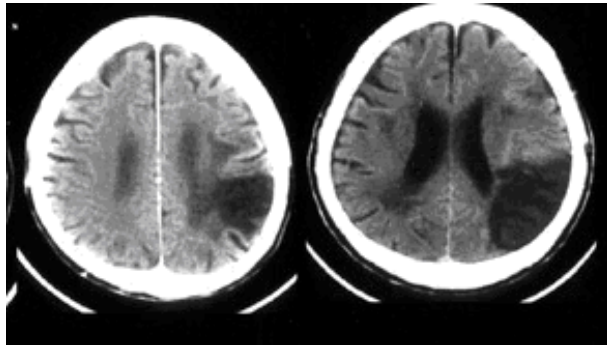
Figure 4.2. Typical lesion in Wernicke's aphasia

It has to be emphasized that Wernicke's aphasia patients can have problems not only at the level of the language sounds (acoustic-agnosic aphasia) or the memory of words (acoustic-amnesic aphasia), but also at the level of the associations between words with specific meanings (Robson, Sage, & Ralph, 2012) (so-called **amnesic** or **nominal** or **extrasylvian sensory aphasia**, associated with damage in BA37 and BA39). It is important to note that so called extrasylvian (or transcortical) sensory aphasia can be considered as another subtype of Wernicke's aphasia; indeed, many authors interpret extrasylvian (or transcortical) sensory aphasia in this way (e.g., Lecours et al., 1983) (see Chapter 5: "Other Aphasic Syndromes"). This means that in consequence not only two but three different deficits may underlie Wernicke-type aphasia: (1) **phoneme discrimination impairments** (auditory verbal agnosia; Luria's acoustic-agnosic aphasia); (2) **verbal memory impairments** (anterógrada and retrograde verbal amnesia; Luria's acoustic-amnesic aphasia; and finally (3) **lexical/semantic association deficits** (amnesic or nominal or extrasylvian sensory aphasia. Figure 4.3 is a presentation in a summarized form the model proposed by Ardila (1993) to account for the language recognition impairments observed in cases of Wernicke-type of aphasia.
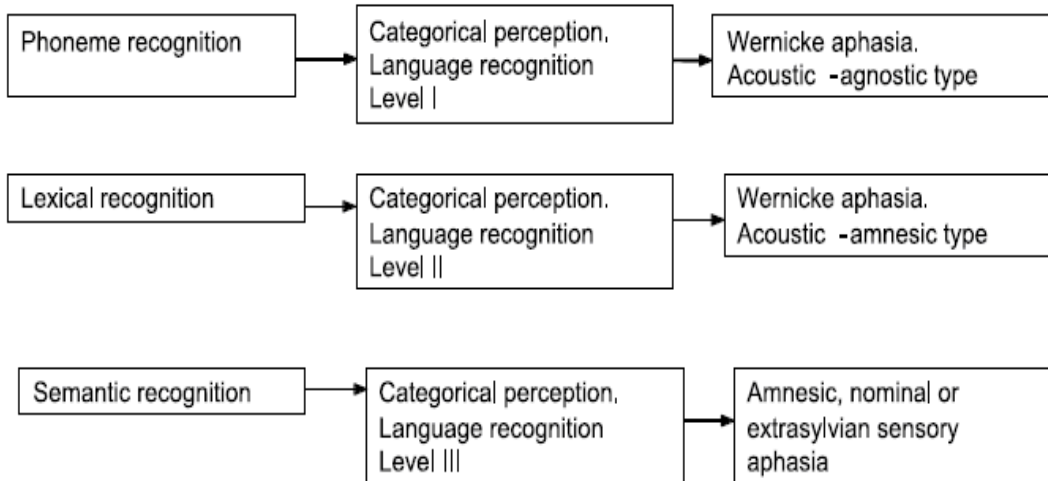
Figure 4.3. Three different deficits underlie Wernicke-type of aphasia: (1) phoneme recognition defects; (2) lexical recognition impairments; (3) lexical/semantic association deficits. Extrasylvian (transcortical) sensory aphasia can be regarded as a subtype of Wernicke's aphasia.

Nouns seem to depend on an organized pattern of brain activity. Contemporary clinical and neuroimaging studies have corroborated that different semantic categories are differentially impaired in cases of brain pathology. For instance, in anomia it has been traditionally recognized that naming body-parts, external objects, and colors depend (and are altered) on the activity of different brain areas (e.g., Hécaen & Albert, 1978). It has also been found that finer distinctions can be made with regard to naming impairments, which can be limited to a rather specific semantic category (e.g., people's names, living things, tools, geographical names, etc.) (e.g., Goodglass, Wingfield, Hyde, & Theurkauf, 1986; Harris & Kay, 1995; Lyons, Hanley, & Kay, 2002; Warrington & Shallice, 1984), and even as specific as ''medical terms'' (Crosson, Moberg, Boone, Rothi & Raymer, 1997).

Broca's aphasia

Broca's aphasia has been named in different ways, including: **aphemia** (Broca, 1863), **efferent or kinetic motor aphasia** (Luria, 1966, 1970), **expressive aphasia** (Hécaen & Albert, 1978; Pick, 1931; Weisenburg & McBride, 1935), **verbal aphasia** (Head, 1926), **syntactic aphasia** (Wepman & Jones, 1964), and **Broca's aphasia** (Nielsen, 1938; Brain 1961; Benson & Geschwind, 1971; Benson, 1979; Lecours, Lhermitte & Bryans, 1983)

Broca's area corresponds to the third frontal gyrus (F3) and is typically defined in terms of the *pars opercularis* and *pars triangularis* of the inferior frontal gyrus, represented in Brodmann's cytoarchitectonic map as areas BA44 and probably BA 45 too (Figure 4.4)
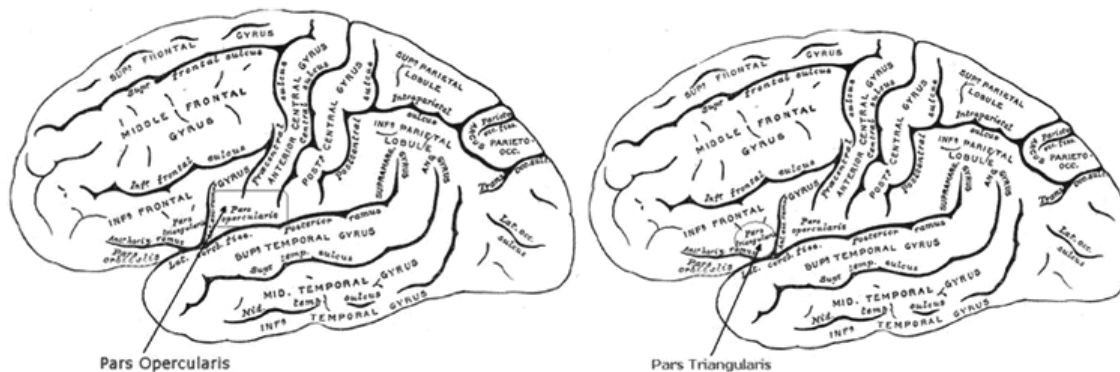
Figure 4.4. Broca's area corresponds to the pars opercularis and pars triangularis of the inferior frontal gyrus (third frontal gyrus or F3; BA 44 and probably BA 45).

Speech in Broca's aphasia is not fluent but language understanding is relatively normal. Repetition is abnormal due to the **apraxia of speech**; as a matter of fact, during repetition, the same disturbances observed in spontaneous speech are found. Pointing (e.g., "show me the ….") is relatively normal; indeed, pointing is a type of language understanding. Table 4.5 is a presentation of the basic language characteristics in Broca's aphasia,

Conversational Language	Nonfluent
Language comprehension	Relatively normal
Repetition	Abnormal
Pointing	Relatively normal
Naming	Abnormal
Reading: Aloud	Abnormal
Comprehension	Relatively normal
Writing	Abnormal

Table 4.5. Basic language characteristics in Broca's aphasia.

Motor difficulties are found in the overwhelming majority of patients with Broca's aphasia. A **right hemiparesis**, more distal (the hand) than proximal (the shoulder) is usually found. Hemiparesis varies in severity but frequently corresponds to a hemiplegia. The hemiparesis is observed in the right arm and face, but it is milder in the right leg. Because of the motor disturbance, dysarthria is almost invariable found; the dysarthria corresponds to a **spastic type of dysarthria** (damage of the upper motor neuron; see Chapter "Associated disorders"). Depending on the extension of the damage in the parietal lobe, somatosensory abnormalities can be found; such as right hemibody hypoesthesia, two-points discrimination defects, difficulties in localizing tactile stimuli in the right hemibody, etc. Because of the right hemiparesis, praxis has to be tested in the left hemibody; in a significant percentage of cases, ideomotor apraxia is found in the left hemibody. The observable apraxia on the left side is referred to as **sympathetic apraxia:** the patient presents two different motor defects: hemiparesis (at the right) and ideomotor apraxia (at the left). No visual field defects or visual recognition impairments (visual agnosia) are expected to be found. Table 4.6 is a presentation of the associated neurological signs in Broca's aphasia

Motor system	Often hemiparesis
Articulation	Abnormal
Cortical sensory function	Normal or abnormal
Praxis	Sympathetic
Visual field	Normal
Visual gnosis	Normal

Table 4.6. Associated neurological signs in Broca's aphasia

Aphasic individuals presenting Broca-type aphasia (a continuity or syntagmatic disorder according to Jakobson, 1971) lose the ability to combine linguistic elements. Their grammar is restricted or absent, and they can produce and understand only isolated meaningful words. Words with purely grammatical function (such as articles and prepositions) tend to be omitted. Affixes may be substituted one for another but more likely they are simply not produced. These patients thus tend to use only very short sentences containing mostly meaningful words (nouns). In severe cases, sentences can be as short as a single word (e.g., ''dog'') and in general, there is a reduction in resources available for syntactic processing (Caplan, 2006). This disturbance in the use of grammar is known as **agrammatism**; agrammatism is also observed in language understanding; so, these patients have difficulties understanding sentences whose meanings depend on their syntax (e.g., "*The dog was bitten by the cat"; who was bitten, the dog or the cat?).* In Table 4.7, an example of agrammatism in Broca's aphasia is presented.

> *Ah ... Monday ... ah, Dad and Paul Haney [himself] and*
> *Dad ... hospital. Two .. .ah, doctors ... and ah ... thirty*
> *minutes .. .and yes ... ah ... hospital. And, er, Wednesday*
> *... nine o'clock. And er Thursday, ten o'clock .. .doctors.*
> *Two doctors ... and ah ... teeth. Yeah, ... fine.*

Table 4.7. Example of agrammatism in Broca aphasia (Taken from www.ling.upenn.edu/ courses/Fall_2000/ling001/neurology.html)

It has to be emphasized that agrammatism (interestingly, at a certain point in aphasia history agrammatism was referred as "**telegraphic style**" of speech) is observed in different linguistic tasks (spontaneous language, language understanding, repetition, writing, and reading). Hence, it corresponds to a fundamental defect that can be observed at different language levels.

So, speech is nonfluent, poorly articulated, agrammatical, and produced with significant effort. Short utterances (singles words, nouns) are observed.

Stereotypes (restricted expression repeatedly used by the patient, as if it were the only language form available) are frequently found (for instance, the initial patient described by Broca in 1863 had a single sterotyped utterance ("tan") that he repeated when attempting to speak. Stereotypes can be short (for instance, a syllable, as in Broca's patient "tan"), or long (for instance, "/beintisinko/"); can be meaningful (e.g., "pencil") or meaningless (e.g., "sood"). Occasionally, the stereotype corresponds to a profanity (that obviously becomes particularly embarrassing not only for the patient but also for other people!). The origin of the specific stereotype is not well understood, but it has been suggested that corresponds to some language information existing exactly before the onset of the aphasia.

Patients with Broca's aphasia present a defect in making precise articulatory movements; that results in a significant amount of **phonetic deviations** (inaccurate production of phonemes), occasionally resulting in so called "**foreign accent in aphasia**". Broca's area is a premotor cortical area, and it is known that premotor damage results in so called "**kinetic apraxia**" (difficulty making precise movements in the hemibody contralateral to the brain pathology); noteworthy, Luria named Broca's aphasia as "kinetic motor aphasia" to emphasize that kinetic apraxia represented a major factor responsible for the speech defects observed in this aphasia.

In addition to phonetic deviation, patients with Broca's aphasia present a significant amount of **phonological paraphasias**. Phonological paraphasias in this type of aphasia are mostly due to phoneme omission and phoneme substitution. As a matter of fact, patients can have significant difficulties in producing certain phonemes (e.g., fricative phonemes) and complex syllables (e.g., consonant-consonant-vowel as in "tree"); fricative phonemes are replaced by stop phonemes (e.g., /s/ becomes /t/) and complex syllables become basic syllables (that is, consonant-vowel; for instance, "tree" becomes "tee"). These verbal articulatory defects in Broca's aphasia are known as **apraxia of speech**. Phonological paraphasias are a result of the apraxia of speech Indeed, it can be assumed that apraxia of speech and agrammatism represent the two distinguishing and fundamental defects responsible for the language defects in Broca's aphasia, even thought it could be argued that apraxia of speech is not exactly a language defect.

Reading aloud is particularly difficult in Broca's aphasia; as a matter of fact, the same defects observed in speaking are also found in reading aloud; that is, defective fluency, apraxia of speech, agrammatism, literal paralexias (substitutions of graphemes in reading, similar to the phonological paraphasias in speaking), etc. However, reading understanding is remarkably better than reading aloud. This reading defect on occasions has been referred as "**frontal alexia**" (see Chapter 6 "Alexia").

Writing is difficult to test because of the right hemiparesis and usually the patient has to use his/her non-preferred hand to write, representing an additional burden; writing with the left hand is usually clumsy due to the lack of practice. Interestingly, agrammatism in writing may be more severe than in spoken agrammatism, because written language requires a more precise use of the grammar; in general, spoken language is more flexible than written language (e.g., words are frequently poorly pronounced, sentences may be interrupted in the middle, etc.). Figure 4.5 is an illustration of the typical lesion in Broca's aphasia.
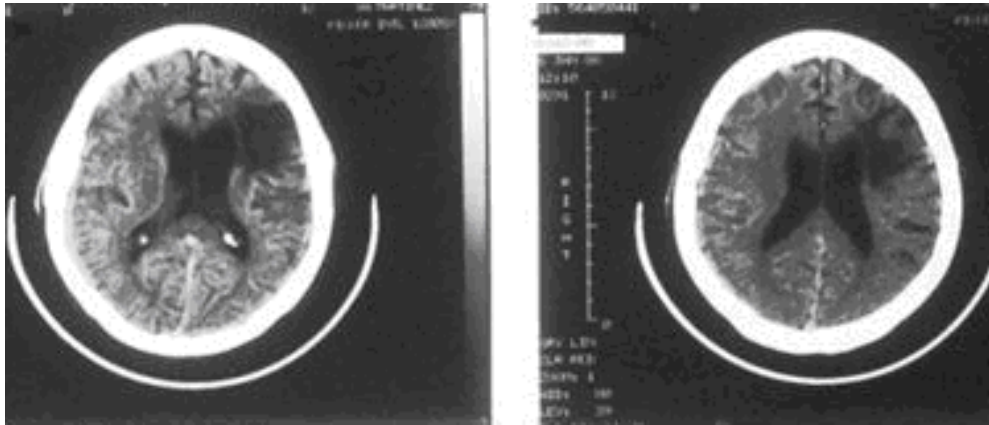
Figure 4.5. Typical lesion in Broca's aphasia

It is usually recognized that Broca's aphasia has two different distinguishing characteristics: (a) a motor component (lack of fluency, disintegration of the speech kinetic melodies, verbal-articulatory impairments, etc. that is usually referred as **apraxia of speech**); and (b) **agrammatism** (e.g., Benson & Ardila, 1996; Goodglass, 1993; Kertesz, 1985; Luria, 1976). A large part of the fronto-parieto-temporal cortex has been observed to be involved with syntactic-morphological functions (Bhatnagar et al., 2000). Apraxia of speech has been specifically associated with damage in the left precentral gyrus of the insula (Dronkers, 1996; but see Hillis et al., 2004).

If both impairments (apraxia of speech and agrammatism) are simultaneously observed, it simply means they are just two different manifestations of a single underlying defect. It has been proposed that this type of ''inability for sequencing expressive motor and phonetic elements'' could represent the single underlying factor responsible for the two components of Broca's aphasia (Ardila & Bernal, 2007). (Figure 4.6). Broca's area, most likely, is not specialized in producing language, but in certain neural activity that can support not only skilled movements required for speech, but also morphosyntax. It has been observed that indeed language networks supporting grammar and fluency are overlapped (Borovsky et al., 2007).
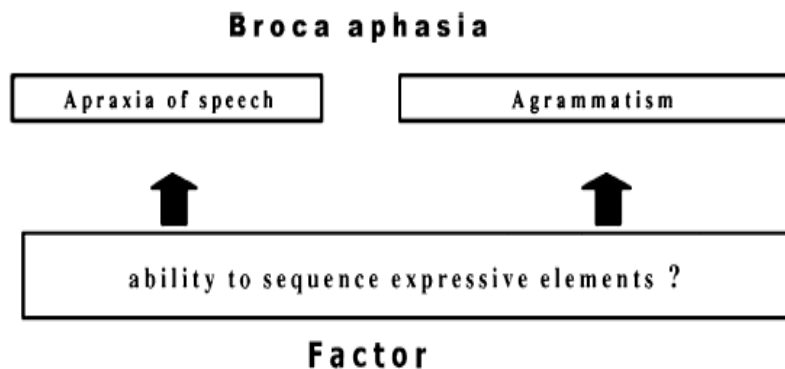
Figure 4.6. The impairment of a single factor may account for the two major clinical manifestations observed in Broca's aphasia (Ardila & Bernal, 2007).

Finally, it is important to note that with the introduction of contemporary neuroimaging techniques it was observed that lesions restricted to Broca's area are not enough to produce the complete classical syndrome of Broca's aphasia; only mild defects in articulatory agility, some "foreign accent", reduced ability to find words, and a simpler (occasionally incorrect) grammar, are observed. Hemiparesis and apraxia are minimal. This restricted form of Broca's aphasia has been named as **minor Broca's aphasia**, **aphasia of the Broca's area** (Alexander, Naeser & Palumbo, 1990) or simply **Broca's aphasia type I** (Benson & Ardila, 1996). The complete classical Broca's aphasia requires in addition to damage in Broca's area, an extension of the pathology to the lower motor cortex, anterior insula, and subjacent subcortical and periventricular white matter. This classical form of Broca aphasia could be named as **extended Broca's aphasia** or **Broca's aphasia type II**.

Summary

Since the beginning of aphasia history it has been well established that there are two major and fundamental aphasic syndromes, named in different ways, but roughly corresponding to Wernicke's aphasia and Broca's aphasia. Wernicke's aphasia results from pathology in Wernicke's area (auditory association area of the left hemisphere). In Wernicke's aphasia, the lexical repertoire tends to decrease and language-understanding difficulties are evident. Speech is produced without effort. No articulatory defects (dysarthria) are observed. Fluency is normal and frequently there is excessive language output. Paraphasias are abundant and no significant associated neurological deficits are observed. Three different deficits underlie Wernicke-type of aphasia and three different subtypes of this aphasia could be distinguished: (1) phoneme recognition defects; (2) lexical recognition impairments; (3) lexical/semantic association deficits; indeed, so called extrasylvian (transcortical) sensory aphasia can be regarded as a subtype of Wernicke's aphasia. Broca's aphasia is associated with damage in the so called Broca's, area, corresponding to the third frontal gyrus. Speech in Broca's aphasia is not fluent but language understanding is relatively normal. Repetition is abnormal due to the apraxia of speech. Pointing is relatively normal. Motor difficulties (including dysarthria) are found in the overwhelming majority of patients with Broca's aphasia. Speech is nonfluent and poorly articulated whereas language output is agrammatical. It is usually recognized that Broca's aphasia has two different distinguishing characteristics: (a) apraxia of speech, and (b) agrammatism; both could be the result of a single underlying defect (sequencing expressive elements). Indeed, the restricted damage to Broca's area is not enough to produce the complete clinical picture of Broca aphasia and two variants of Broca's aphasia can be distinguished: aphasia of the Broca's area (first subtype) and extended ("classical") Broca's aphasia (second subtype).

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Chapter 5

Other aphasia syndromes:
Conduction aphasia, extrasylvian (transcortical) aphasias, supplementary motor area aphasia, subcortical aphasia, global aphasia

Introduction

In addition to the two major aphasic syndromes (Broca's aphasia and Wernicke's aphasia), different aphasia classifications generally include a diversity of additional language disturbances, such as conduction aphasia, transcortical (extrasylvian) aphasia, anomic aphasia, etc. Indeed, some aphasic syndromes can eventually be considered as variants of the Broca's and Wernicke's aphasias. For instance, amnesic or anomic or nominal aphasia (usually due to damage in the vicinity of BA37) can be interpreted as a subtype of Wernicke's aphasia in which the semantic associations of the words are significantly impaired. By the same token, extrasylvian (transcortical) sensory aphasia can be regarded as a subtype of Wernicke's aphasia, and indeed, that is the interpretation proposed by different authors (e.g., Lecours et al., 1983).

In this chapter, these additional aphasia syndromes will be reviewed.

Conduction aphasia

Conduction aphasia has been named as **motor or kinesthetic afferent aphasia** (Luria, 1966, 1980), **central aphasia** (Goldstein, 1948), **efferent conduction aphasia** (Kertesz, 1985), or simply **conduction aphasia** (Benson & Ardila, 1994; Benson, 1979; Hécaen & Albert, 1978; Lecours, Lhermitte & Bryans, 1983; Wernicke, 1874).

Conduction aphasia was initially described by Wernicke in 1874, and interpreted as a disconnection between the superior temporal gyrus (Wernicke's area) and the inferior frontal gyrus (Broca's area). Wernicke's interpretation was supported by Geschwind during the 1960s (the so-called **Wernicke-Geschwind model of language**), who put it in terms of modern anatomic nomenclature, attributing to the **arcuate fasciculus** the main role in the speech repetition disturbances. According to Geschwind (1965), disconnection syndromes were higher function deficits that resulted from white matter lesions or lesions of the association cortices; conduction aphasia was usually presented as the prototypal example of a disconnection

syndrome. This is up to now its most frequent interpretation (e.g., Damasio & Damasio 1980): conduction aphasia is usually due to a lesion affecting the arcuate fasciculus (Yamada et al., 2007) and sporadically an indirect pathway passing through the inferior parietal cortex (Catani, Jones, & Ffytche, 2005) (Figure 5.1).
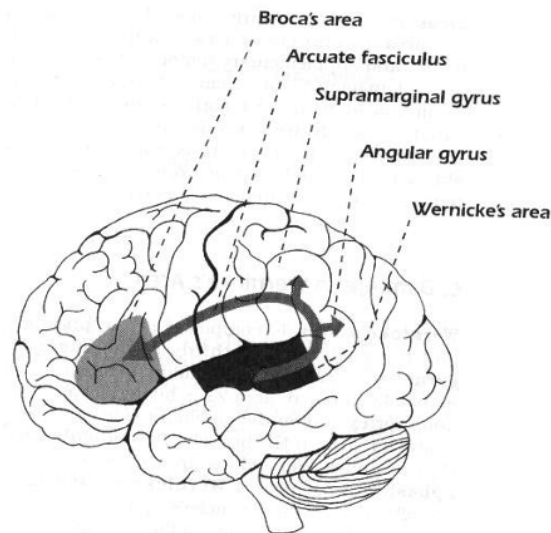
Figure 5.1. Explanation of conduction aphasia as a disconnection between Wernicke's and Broca's area.

Alternatively, conduction aphasia has also been interpreted as a **segmentary ideomotor apraxia** (e.g., Ardila & Rosselli., 1990; Brown, 1975; Luria 1976, 1980). According to this second interpretation, conduction aphasia could be regarded as a **verbal apraxia**, an ideomotor apraxia impairing the movements required for speaking, or simply as a **kinesthetic apraxia of speech**. Luria (1976) suggested that paraphasias in conduction aphasia (Luria's kinesthetic motor or afferent motor aphasia) are articulatory-based (articulatory literal paraphasias). These errors are due mainly to phoneme substitutions and phoneme deletions; they result basically in switches in phoneme manner and place of articulation (Ardila, 1992). Similarities between errors in ideomotor apraxia and conduction aphasia language deficits have been suggested.

According to Benson et al. (1973), conduction aphasia has three fundamental and five secondary characteristics; so-called secondary characteristics are frequently but not necessarily found in conduction aphasia. The three basic characteristics are: (1) fluent conversational language; (2) comprehension almost normal; and (3) significant impairments in repetition. Secondary characteristics include: (1) impairments in naming; (2) reading impairments; (3) variable writing difficulties (apraxic agraphia); (4) ideomotor apraxia; and (5) additional neurological impairments. Bartha and Benke (2003) report that conduction aphasia patients present as relatively homogenic in their aphasic manifestations: severe impairment of repetition and fluent expressive language functions with frequent phonemic paraphasias, repetitive self-corrections, word-finding difficulties, and paraphrasing. Repetitive self-corrections frequently result in so-called *conduit d'approche* (behavior of approximation). Language comprehension

(auditory and reading) is only mildly impaired. Table 5.1 is a summary of the basic language characteristics in conduction aphasia.

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Conversational Language	Fluent, paraphasic
Language comprehension	Good to normal
Repetition	Abnormal
Pointing	Good to normal
Naming	Abnormal
Reading: Aloud	Abnormal
Comprehension	Good to normal
Writing	Abnormal

―――――――――――――――――――――――――――――――

Table 5.1. Basic language characteristics in conduction aphasia

Some neurological abnormalities can be found in conduction aphasia (Table 5.2); mild hemiparesis is frequent at the onset of aphasia, but tend to disappear, unless the damage extends to the frontal lobe. Articulation is usually normal, but frequently somatosensory defects (such as hypoesthesia, difficulties for localizing tactile stimuli, etc.) are found. Ideomotor apraxia is generally found, and even some authors have proposed that conduction aphasia could be interpreted as a segmentary ideomotor apraxia (e.g., Luria, 1976). Visual field defects and visual agnosia are not expected to be found.

―――――――――――――――――――――――――――――――

Motor system	Mild hemiparesis
Articulation	Normal
Cortical sensory function	Usually somatosensory defects
Praxis	Ideomotor apraxia
Visual field	Normal
Visual gnosis	Normal

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Table 5.2. Associated neurological signs in conduction aphasia

When attempting to repeat (but also on occasion in spontaneous language) the patient with conduction aphasia presents successive approaches to the target word (*conduit d'approche*); every time he/she produces the word, the patient recognizes it has been incorrectly produced (because language understanding is preserved), and attempts to correct it. A significant amount of phonological paraphasias are observed and from time to time, verbal paraphasias are also found. Sometimes it is impossible to produce the word during repetition, but not in spontaneous language. Figure 5.2 is an illustration of a typical lesion in conduction aphasia
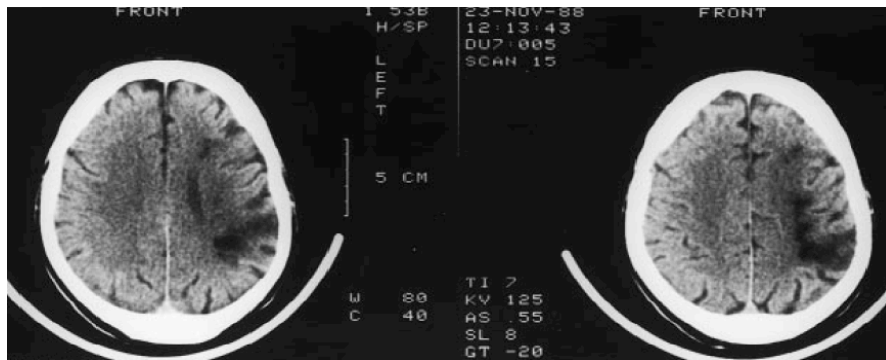
Figure 5.2 Typical lesion in conduction aphasia

Reading aloud is defective (similar to naming or repetition) whereas reading comprehension is nearly normal. Writing defects (**afferent motor agraphia**, according to Luria, 1977) are variable in severity; usually literal paragraphias (parallelizing the phonological paraphasias) are found. In cases of extended damage in the left parietal lobe, an apraxia for writing (apraxic agraphia) can be found (see Chapter 7: "Agraphia")

The possibility of several mechanisms, each of which is capable of giving rise to deficient repetition, led to the postulation of two different forms of conduction aphasia named as **efferent conduction aphasia** and **afferent conduction aphasia** (Kertesz, 1985); or **reproduction** and **repetition** (Shallice & Warrington, 1977); or **supra-** and **infrasylvian** (Axer et al., 2001); or simply **parietal** and **temporal** (Bartha & Benke, 2003). The efferent-reproduction type involves the phonemic organization and representation of words and is correlated with parietal and insular damage, whereas the afferent-repetition conduction aphasia involves short-term memory defects and affects the repetition of large strings of material. This second subtype of conduction aphasia has been described more frequently with lesions of the temporal lobe and indeed correspond the the acoustic-amnesic subtype of Wernicke's aphasia.

Of note, language repetition impairments are not restricted to conduction aphasia and can be observed in different aphasia syndromes. Ardila and Rosselli (1992) analyzed 38 aphasic patients divided into six groups (transcortical motor, Broca's, conduction, Wernicke's, anomic, and global aphasia) in the three repetition subtests of the Boston Diagnostic Aphasia Examination (Goodglass & Kaplan, 1983). Repetition errors were generally associated with perisylvian aphasias (Broca's, conduction, and Wernicke's). However, in all aphasic groups some repetition errors were observed. These errors were not only quantitatively but also qualitatively different. It was concluded that, depending on the specific repetition task, errors may be evident or unnoticed in a particular aphasic group. The authors proposed that different mechanisms may underlie repetition deficits in aphasia: limitation of auditory-verbal short-term memory, difficulties at the level of phonological production, impairments in phoneme recognition, and semantic and syntactic comprehension (Table 5.3).

Boston Diagnostic Aphasia Examination	Words	High-Probability	Low-Probability
Transcortical motor	98.0	95.0	67.5
Broca	46.0	50.0	45.0
Conduction	63.0	53.7	21.2
Wernicke's	74.0	45.0	22.5
Anomic	100.0	71.2	52.5
Global	27.0	0.0	0.0

Table 5.3. Percentage of correct repetition for each patient group on the three repetition tasks from the Boston Diagnostic Aphasia Examination (Adapted from Ardila and Rosselli, 1992)

The arcuate fasciculus is a brain association tract composed of arched fibers that is assumed to connect the posterior temporal language understanding area (Wernicke's area) and the anterior frontal language production area (Broca's area). The arcuate fasciculus is the main part of a larger tract located lateral to the corticospinal tract, known as the superior longitudinal fasciculus. The really crucial question becomes: Is it invariably the arcuate fasciculus affected in cases of conduction aphasia? (Ardila, 2010).

Bernal and Ardila (2009) observed that transferring of speech information from the temporal to the frontal lobe uses not only one but two different streams (the arcuate fasciculus and an indirect pathway passing through the inferior parietal cortex); and furthermore, conduction aphasia can be found in cases of cortical damage without subcortical extension (Quigg, Geldmacher & Elias,2006). Tractography demonstrates that the arcuate fasciculus connects the posterior temporal lobe with BA6 (premotor area) and BA4 (primary motor area), not with BA44 (Broca's area). Together, these observations strongly suggest that the arcuate fasciculus is not required for repetition, but it could have a subsidiary role in it. Bernal and Ardila (2009) further proposed a new language network model emphasizing that the arcuate fasciculus connects posterior brain areas with Broca's area via a relay station in the premotor/motor areas (BA6 and BA4). Thus, the connection with Broca's area would not be direct, but indirect (Figure 5.3).
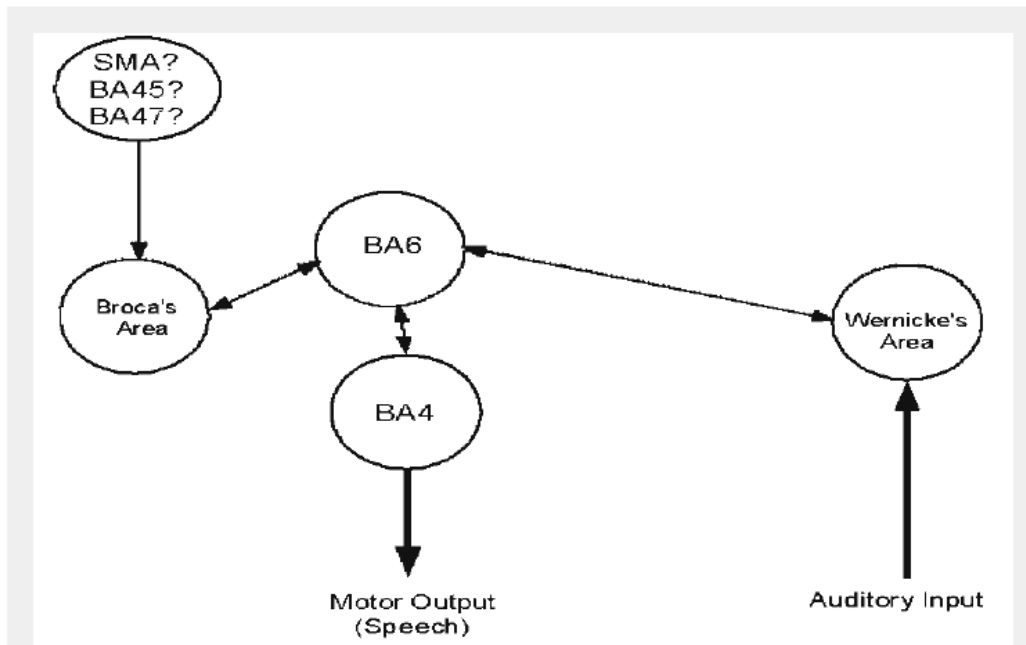
Figure 5.3. The arcuate fasciculus connects posterior brain areas with Broca's area via a relay station in the premotor/motor areas (BA6 and BA4) (taken from Bernal & Ardila, 2009).

Extrasylvian (transcortical) sensory aphasia

Extrasylvian (transcortical) sensory aphasia (TSA) has been a polemic syndrome; frequently it is considered as a subtype of Wernicke's aphasia. Seemingly, the polemic is related to the way TSA is defined and the elements included in its definition. Some authors have even simply denied the existence of such a syndrome. Two integrative revisions of TSA are available (Berthier, 1999; Boatman et al., 2000).

In general, it is considered that extrasylvian (transcortical) sensory aphasia includes the following elements: (1) Good repetition (the patient repeats words and sentences presented by the examiner, regardless if they are incorrect and even in a foreign language); (2) Fluent conversational language; (3) Significant amount of verbal paraphasias and neologisms; and (4) Empty speech. TSA presents similar deficits as in Wernicke's aphasia, but repetition ability is spared and phoneme discrimination impairments are not found. Some authors also include a semantic jargon in the definition of TSA (Goodglass, 1993; Kertesz, 1982; Lecours et al., 1981). But jargon is not a required symptom for the diagnosis of TSA. By the same token, other language impairments can also be found, such as poor naming, and preserved oral reading with impaired reading comprehension, but their presence is not essential to establish the diagnosis of TSA (Berthier, 1999). Table 5.4 is a presentation of the basic language characteristics in extrasylvian (transcortical) sensory aphasia*.*

Conversational Language	Fluent, paraphasic echolalic
Language comprehension	Defective
Repetition	Good to excellent
Pointing	Defective
Naming	Defective
Reading: Aloud	May be preserved
Comprehension	Defective
Writing	Defective

Table 5.4. Basic language characteristics in extrasylvian (transcortical) sensory aphasia

The associated neurological signs in extrasylvian (transcortical) sensory aphasia are presented in Table 5.5. No motor (including articulatory) defects are observed; but because of its location in the brain, cortical sensory function can be defective and ideomotora apraxia can be present, depending upon the extension of the pathology to the parietal lobe. Similarly, the extension of the damage to the occipital lobe may result in visual agnosia and visual field defects.

Motor system	Normal
Articulation	Normal
Cortical sensory function	Often defective
Praxis	May be defective
Visual field	Normal to defective
Visual gnosis	May be defective

Table 5.5. Associated neurological signs in extrasylvian (transcortical) sensory aphasia

Because repetition is spared, phonological processing is assumed to be preserved, at least partially, while lexical-semantic information included in the word meaning is impaired (Boatman et al., 2000). Usually, it is accepted that TSA is associated with relatively extensive posterior lesions including the temporo-parieto-occipital junction of the left hemisphere but sparing the areas around the primary auditory cortex (Berthier, 1999). Damasio (1991) observed that TSA is associated with lesions involving the temporal-occipital area (BA37), the angular gyrus (BA39) (Figure 6.3), or the white matter underlying these regions, but sparing the primary auditory cortex (BA41 and 42), and BA22. Damasio suggested that the core area for TSA is the temporal-occipital area (BA37) with variable extension to the occipital lobe and the angular gyrus (Figure 5.4).
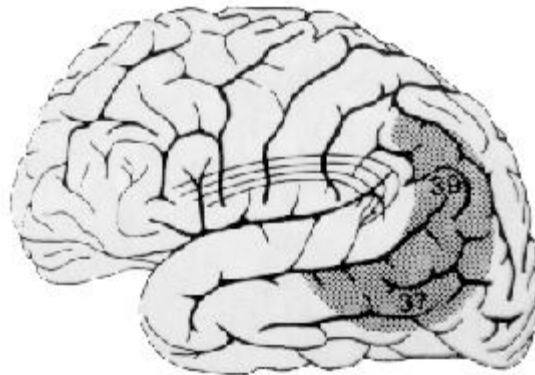
Figure 5.4.   Area of the extrasylvian (transcortical) sensory aphasia. Two different subtypes can be distinguished: Lesions in BA37 result in amnesic (or anomic or nominal) aphasia (or first subtype of transcortical sensory aphasia) whereas lesions in BA39 result in semantic aphasia or semantic anomia (or second subtype of transcortical sensory aphasia)

Recent reports support the assumption that TSA is usually found associated with extensive lesions of the left hemisphere (Figure 5.5) (e.g., Warabi et al., 2006), generally involving large portions of the temporal-parietal-occipital areas. According to Alexander, Hiltbrunner, and Fischer (1989), the critical lesion for transcortical sensory aphasia in these patients involved pathways in the posterior periventricular white matter adjacent to the posterior temporal isthmus, pathways that are most likely converging on the inferolateral temporo-occipital cortex. However, frequently the variability in the lesions responsible for TSA account for the variability observed in its clinical manifestations, suggesting that TSA does not necessarily represent a single aphasic syndrome. When the lesions are restricted to BA 37 or BA 39, specific and well-described language impairments are observed (Benson & Ardila, 1996; Luria, 1976). With more extended lesions, additional clinical manifestations, such as jargon, can be found. These additional clinical manifestations are only observed in the acute stage of the brain pathology, and progressively disappear (Kertesz, 1979). Dronkers and Larsen (2001) state that ''*transcortical sensory aphasia always resolves into mild anomic aphasia*'' (p. 29).
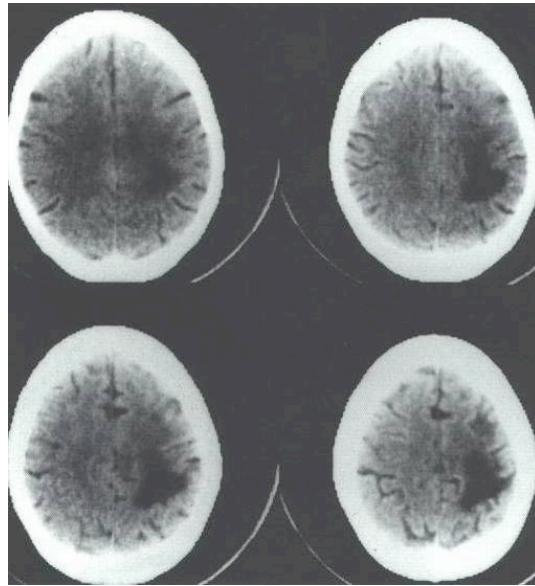
Figure 5.5. Typical lesion of extrasylvian (transcortical) sensory aphasia (according to Berthier, 1999)

Benson and Ardila (1996), considering this variability in TSA, distinguished two subtypes: the first one similar to Luria's **amnesic** aphasia (BA37) (also known as **anomic** or **nominal** aphasia), and the second one corresponding Luria's **semantic** aphasia (BA39). This distinction is coincidental with the neuroanatomical correlates of TSA found by Damasio (1991).

In the first one (left temporal-occipital –BA37- syndrome), fluent spontaneous language is observed with poor comprehension and good repetition. Semantic paraphasias and neologisms are abundant. As a matter of fact, the damage in this area results in the highest amount of semantic paraphasias. Comprehension at the word level is defective and there are significant defects in naming; but the presentation of phonological cueing is effective. Because of the location of the pathology (temporo-occipital), minor or moderate visual agnosic defects are found; indeed, the patient presents a significant defect in visualizing for him/herself the meaning of the words (i.e., how a "book", or a "dog" or whatever noun looks like). Thus, it is a language defect at the level of the semantics of the words.

The second subtype (angular and parietal-occipital –BA 39- syndrome; **transcortical sensory aphasia second subtype**; Kertesz, 1983); partially corresponds to **semantic aphasia** (Head, 1926; Luria, 1966, 1980; Ardila et al., 1989), and **semantic anomia** (Benson, 1988). Some verbal amnesia is usually found. There is fluent language, with few paraphasias; comprehension relatively good, and repetition is normal. But the patient presents significant word-finding difficulties; it is usually frequently associated with the so-called Gerstmann's (angular gyrus) syndrome (right-left disorientation, finger agnosia, acalculia and agraphia).

Head (1926) defined semantic aphasia as an inability to simultaneously recognize the elements included in a sentence. Luria (1973, 1976) emphasizes that language deficiencies are observed in: (1) sentences with a complex system of successive subordinate clauses (e.g., "the person that I think that you mentioned that would come…") ; (2) reversible constructions, particularly of the temporal and spatial type (e.g., "I read the newspaper before taking the breakfast"); (3) constructions with double negative (non existing in English, but existing in Spanish, Russian and other languages); (4) comparative sentences (e.g., "Peter is taller than John but shorter than Bob"); and (5) passive constructions (e.g., "The earth is illuminated by the sun"). He 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.

Finally, it is most important to emphasize that extrasylvian (transcortical) sensory aphasia can be interpreted as a subtype of Wernicke's aphasia; indeed, many authors interpret extrasylvian (transcortical) sensory aphasia in such a way (e.g., Lecours et al., 1983) (see Chapter 4).

Extrasylvian (transcortical) motor aphasia ("dysexecutive aphasia")

Extrasylvian (transcortical) motor aphasia has been named as dynamic aphasia (Luria, 1966, 1980), loss of verbal initiative (Kleist, 1934) or just transcortical motor aphasia (Goldstein, 1948; Hécaen & Albert, 1978; Benson & Geschwind, 1971; Benson, 1979). It is associated with left convexital prefrontal damage. Figure 5.6 is an illustration of the typical lesion in this type of aphasia
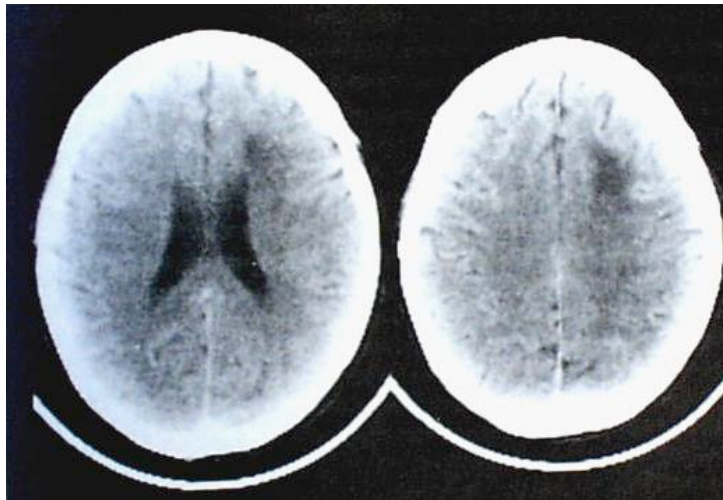
Figure 5.6. Typical lesion of transcortical motor aphasia (according to Berthier, 1999)

Extrasylvian (transcortical) motor aphasia is characterized by non fluent language, good comprehension, and good repetition. Prosody, articulation, and grammar are preserved. The

patient presents long latencies in language when beginning to speak or when answering questions. Open questions are slow and incomplete, and the patient tends to repeat the words included in the question. Expressive language is limited with some tendency to echolalia and perseveration; occasionally verbal paraphasias are observed. This type of aphasia could be interpreted as a language disturbance at the pragmatic level (use of the language according to the specific social context). Table 5.6 is a presentation of the basic language characteristics in extrasylvian (transcortical) motor aphasia

Conversational Language	Sparse, echolalic
Language comprehension	Relatively normal
Repetition	Good to normal
Pointing	Normal
Naming	Mildly abnormal
Reading: Aloud	Defective
Comprehension	Good to normal
Writing	Defective

Table 5.6. Basic language characteristics in extrasylvian (transcortical) motor aphasia

Depending upon extension of the damage, some motor weakness may exist, but quite frequently, strength is normal and no articulation defects (dysarthria) are found. However, the patient usually presents primitive (pathological) reflexes (reflexes normal in newborns, that disappear with the maturation of the brain; they can reappear en cases of frontal lobe damage) such as palmar grasp reflex, palm mental reflex, snout reflex, and plantar reflex (Babinski sign). No somatosensory abnormalities, visual field defects, apraxia, or agnosia are observed. Table 5.7 is a summary of the associated neurological signs in extrasylvian (transcortical) motor aphasia.

Motor system	Hemiparesis may exist pathological reflexes
Articulation	Normal
Cortical sensory function	Normal
Praxis	Normal
Visual field	Normal
Visual gnosis	Normal

Table 5.7. Associated neurological signs in extrasylvian (transcortical) motor aphasia

Extrasylvian (transcortical) motor aphasia could be interpreted as an executive function defect specifically affecting language use. The ability to actively and appropriately generate language appears impaired while the phonology, lexicon, semantics, and grammar are preserved. Simply speaking, the question is: Should the ability to correctly generate language be regarded as a linguistic ability (i.e., cognitive ability)? Or rather, should it be considered as executive function ability (i.e., metacognitive ability)?

It does not seem difficult to argue that the ability to correctly organize language sequences can be interpreted as an executive function and as a metacognitive ability rather than a purely linguistic ability. Some rationales to support this interpretation are: (1) It could be argued that in extrasylvian (transcortical) motor aphasia there is a defect in verbal initiative rather than in language knowledge (Kleist, 1934). (2) Some authors (Luria 1976, 1980) have emphasized that this type of aphasia shares the general characteristics of prefrontal (i.e., dysexecutive) syndrome but specifically with regard to verbal processes. This means, it is the prefrontal (dysexecutive) syndrome affecting the verbal processes; (3) Furthermore, the impairment in extrasylvian (transcortical) motor aphasia does not affect language understanding, and fundamental linguistic processes are preserved (Berthier, 1999). And finally, (4) it could be argued that the prefrontal cortex does not participate in basic cognition, but rather in metacognition (e.g., Ardila & Surloff, 2013). Consequently, extrasylvian (transcortical) motor aphasia does not necessarily have to be interpreted as a primary aphasic syndrome, but rather as a language disturbance due to a more general intellectual impairment (dysexecutive syndrome). In this regard, it is a secondary—not primary—aphasia syndrome. Extrasylvian (transcortical) motor aphasia could indeed be referred to as ''**dysexecutive aphasia''**.

Some authors have previously interpreted extrasylvian motor aphasia in a similar way (e.g., Luria 1976, 1980). Alexander (2006) suggested that transcortical motor aphasia could be more accurately defined as an executive function disorder rather than aphasia. 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 and/or lateral frontal to medial frontal. It is noteworthy that successful functional communication is significantly associated with executive function in aphasia (Fridriksson. Nettles, Davis, Morrow, & Montgomery, 2006).

Mixed extrasylvian (transcortical) aphasia

Mixed extrasylvian (transcortical) aphasia is an extremely unusual aphasic syndrome and just some few cases have been reported in the aphasia literature (e.g., Carota, Annoni, Marangolo, 2007; Rapcsak, Krupp, Rubens & Reim, 1990). It is also referred as "**isolation syndrome**", because supposedly the language area becomes isolated from the rest of the brain. In this aphasia, Broca's and Wernicke's areas are intact but their surrounding areas are impaired. It is thought that damage to these association areas leaves Broca's and Wernicke's areas completely isolated from the rest of the language system, thus precluding the production of spontaneous speech and the comprehension of spoken and written language. The most common cause of mixed transcortical aphasia is a watershed zone (areas of the brain along the "border zones" between major arteries receiving dual blood supply) (Cauquil-Michon, Flamand-

Roze & Denier, 2011) stroke of the language association areas as a result of severe internal carotid stenosis. Figure 5.7 is an illustration of a case of mixed extrasylvian (transcortical) aphasia
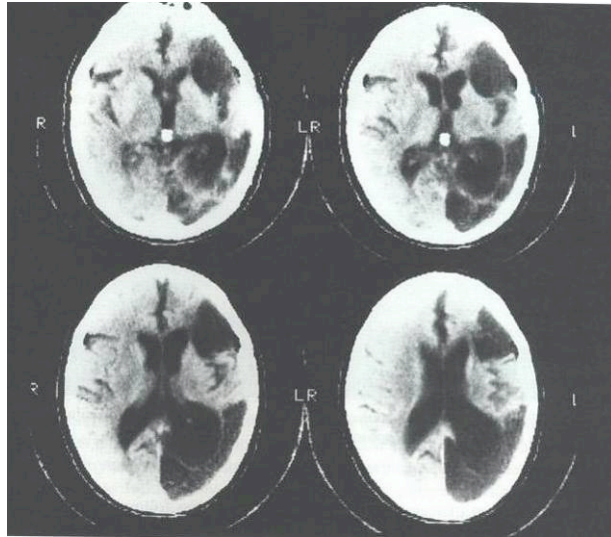
Figure 5.7. Typical lesion in mixed extrasylvian (transcortical) aphasia (according to Berthier, 1999)

In this unusual syndrome, spontaneous language is absent and speech production is virtually limited to repetition; frequently echolalia is observed, but articulation is good and automatic language is relatively preserved. Table 5.8 is a summary of the basic language characteristics in mixed extrasylvian (transcortical) aphasia; indeed, the only language ability that is maintained is language repetition. Indeed, its only difference with global aphasia is the relatively preserved language repetition ability.

Conversational Language	Nonfluent, echolalia
Language comprehension	Defective
Repetition	Relatively good
Pointing	Defective
Naming	Defective
Reading: Aloud	Defective
Comprehension	Defective
Writing	Defective

Table 5.8. Basic language characteristics in mixed extrasylvian (transcortical) aphasia

The associated neurological signs correspond to the extended location of the pathology, that includes so pre-rolandic as post-rolandic areas (Table 5.9): paresis, pathological reflexes, apraxia, frequently visual field defects and visual agnosia.

Motor system	Paresis, pathological reflexes
Articulation	Normal
Cortical sensory function	Often disordered
Praxis	May be defective
Visual field	Normal to defective
Visual gnosis	May be defective

Table 5.9. Associated neurological signs in mixed extrasylvian (transcortical) aphasia

Supplementary motor area (SMA) aphasia

In 1940, Brickner reported that electro-cortical stimulation of SMA (mesial aspect of BA6; Figure 5.8.) resulted in continuous perseveration. Penfield and Welch (1951) observed arrest of speech associated with stimulation of this cortical region. However, language disturbances associated with SMA pathology were reported relatively late in the aphasia literature. Clinical characteristics of this type of aphasia were described by Rubens (1975, 1976). Jonas (1981) later referred to the participation of the SMA in speech emission.
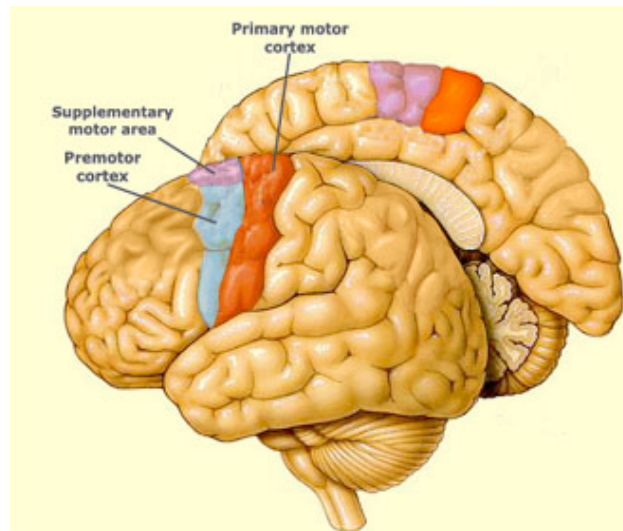
Figure 5.8. The SMA (in purple) corresponds to the mesial extension of the BA6 (premotor cortex).

The occlusion of the left anterior cerebral artery is the most frequent etiology, but it has also been reported in cases of tumors and traumatic head injury (e.g., Ardila & Lopez, 1984). Speech is characterized by (1) an initial mutism lasting about 2–10 days; (2) later, a virtually total inability to initiate speech, (3) nearly normal speech repetition, (4) a normal language understanding, and (5) absence of echolalia. A right leg paresis and right leg sensory loss are observed; a mild right shoulder paresis and Babinski sign are also found. Language recovery is outstanding and it is usually observed during the following few weeks or months. Table 5.10 is a summary of the basic language characteristics: spontaneous language is limited, but language understanding and language repetition are normal; there is a significant difficulty in initiating and maintaining speech, regardless that the patient makes significant effort to speak; reading aloud is defective but reading understanding is nearly normal; writing is slow and painstaking. Noteworthy, this type of aphasia has sometimes been interpreted as an extrasylvian (transcortical) motor aphasia.

Conversational Language	Sparse, effortful
Language comprehension	Normal
Repetition	Good to normal
Pointing	Normal
Naming	Mildly abnormal
Reading: Aloud	Defective
Comprehension	Good to normal
Writing	Slow with paragraphias

Table 5.10. Basic language characteristics in SMA aphasia

Legs have a motor representation in the mesial aspect of the frontal lobe motor areas. Consequently, hemiparesis right leg represents the most important neurological abnormality; frequently, an extension of the pathology toward the parietal lobe is found, and hence, some right leg sensory loss is observed. Table 5.11 is a summary of the associated neurological signs in SMA aphasia

Motor system	Hemiparesis right leg
Articulation	Mild defects
Cortical sensory function	Right leg sensory loss
Praxis	Normal
Visual field	Normal
Visual gnosis	Normal

Table 5.11. Associated neurological signs in SMA aphasia

The SMA is a mainly mesial premotor area involved in the ability to sequence multiple movements performed in a particular order (Tanji & Shima, 1994). SMA participates in initiating, maintaining, coordinating, and planning complex sequences of movements; it receives information from the posterior parietal and frontal association areas, and projects to the primary motor cortex (Kandel, Schwartz & Jessell, 1995). SMA damage is also associated with slow reaction time (Alexander et al., 2007). It has been observed that activation of the SMA precedes voluntary movement (Erdler et al., 2000); a crucial role in the motor expression of speech processing has also been assumed (Fried et al., 1991). Nonetheless, the SMA is located some distance—and indeed far away— from the classic language area postulated by Dejerine (1914) and assumed in most anatomical models of aphasia (Figure 5.9).
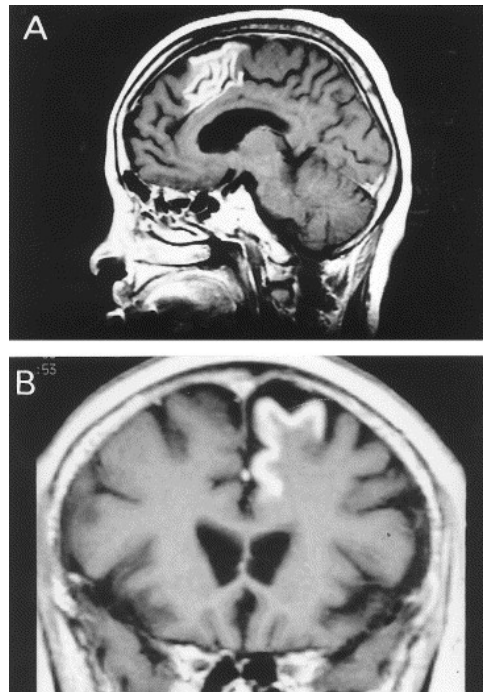
Figure 5.9. Typical lesion in aphasia of the supplementary motor area

It has been suggested that SMA has a close connectional relationship with the prefrontal cortex and plays a critical role in the update of verbal representations (Tanaka, Honda, & Sadato, 2005). Neuroimaging studies in humans have demonstrated that SMA is active when performing various cognitive tasks, such as spatial working memory (Jonides et al., 1993), verbal working memory (Paulesu, Frith, & Frackowiak, 1993), arithmetic tasks (Dehaene et al., 1996; Hanakawa et al., 2002), spatial mental imagery (Mellet et al., 1996), and spatial attention (Simon et al., 2002).

Evidently, the SMA is a complex motor cortical area, not primarily a language related brain area. Its role in language seemingly refers to the motor ability to initiate and maintain voluntary speech production.

Subcortical aphasia

Since Wernicke (1874), it has been assumed that aphasia can represent the consequence of damage to neural networks including both cortical and subcortical structures. Marie (1906) stated that the subcortical damage involving the basal ganglia (an area further known as "Marie's quadrilateral space"; Figure 5.10.) would result in dysarthria, not really in aphasia.
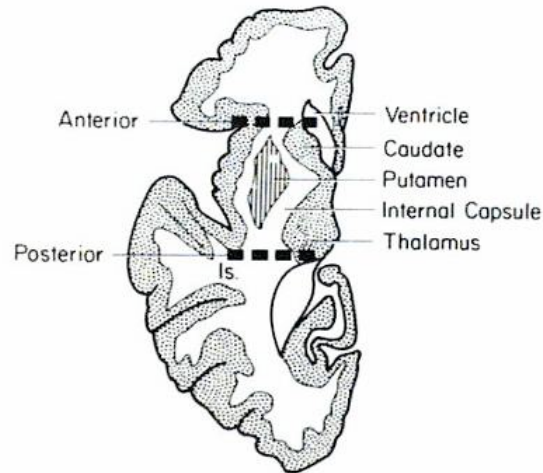
Figure 5.10. "Marie's quadrilateral space"

When Dejerine (1914) described the brain's "language area" no specific mention to subcortical structures was made. The idea of "subcortical aphasia" was somehow forgotten during the following decades. Only with the introduction of the CT scan it was observed that aphasia was frequently associated with subcortical pathology, and the discussion and interpretation of subcortical aphasia re-emerged. Contemporary neuroimaging techniques have permitted far better understanding of subcortical pathology involved in aphasia. Nonetheless, whether true aphasia results from isolated subcortical brain damage, or whether it is due to a cortical extension or cortical deactivation, remains unanswered (e.g., Craver & Small, 1997). Subcortical pathology frequently includes altered speech (dysarthria), often beginning with total mutism followed by hypophonic, slow, sparse output, and poorly differentiated, amelodic speech. In addition to dysarthria, sometimes language impairments are also found.

Two neuroanatomical areas are most frequently discussed in subcortical aphasias: the **striatocapsular** region and the **thalamus**. Patients with striatocapsular damage show significant articulation impairments. Their language output appears truncated, but it is not agrammatic. Speech mechanisms are generally impaired resulting in impairments in articulation and prosody. Comprehension is intact for casual conversation but breaks down when complex syntax is presented. Word-finding problems may be noted. Alexander and colleagues (1987) have proposed six subtypes of verbal output impairment. These subtypes are dependent on the specific neuroanatomical locus of striatocapsular damage, demonstrating that considerable

variation in speech and language impairment can follow this type of pathology. Figure 5.11 is an illustration of a typical lesion in subcortical aphasia.

Figure 5.11. Typical lesion in subcortical aphasia. An extensive subcortical hemorrhage is observed in the left hemisphere.

Frequently, extension that involves the cortex is present in these cases. Extensive subcortical damage is required to produce a pure striatocapsular aphasia, but prognosis is worse when the posterior limb of the internal capsule is involved (Liang et al., 2001). Mega and Alexander (1994) evaluated 14 cases of striatocapsular aphasia. The clinical profiles of the patients were quite similar, varying in severity in rough proportion to lesion size and varying in quality in proportion to anterior paraventricular extent. Large lesions were associated with impaired ''executive'' and ''generative'' language functions. Similar aphasia profiles in patients with deep frontal and paraventricular white matter lesions suggest that damage to a frontal-caudate functional system underlies a ''core'' aphasia profile in these patients. Nadeau and Crosson (1997), after a critical review of the literature, suggested that linguistic impairments associated with striatocapsular pathology are predominantly related to sustained cortical hypoperfusion and infarction not visible on structural imaging studies.

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 (Kalefa, Hodorog

& Stefanache, 2008). Frequently, aphasia is observed in cases of left pulvinar nucleus pathology; interesting to note, the pulvinar nucleus projects to an extensive cortical area, frequently related with extrasylvian sensory aphasia (Figure 6.12.)
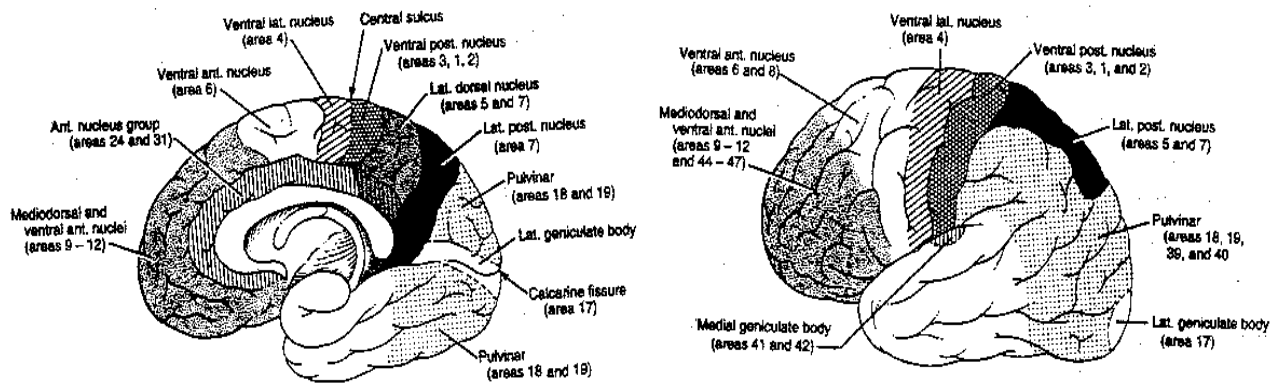
Figure 5.12. The thalamic pulvinar nucleus projects to BA18, 19, 37, 39 and 40.

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). It has been further proposed that the left thalamus seems to bring online the cortical network involved in language processing (Metz-Lutz et al., 2000).

In brief, aphasia is sometimes associated with subcortical lesions, particularly left striatocapsular and thalamic pathology. To account for subcortical aphasia it has been proposed that aphasia may result from a cortical extension or cortical deactivation. The idea of a cortical deactivation seems to prevail (e.g., Choi et al., 2007; Hillis et al., 2004). The question of subcortical aphasia suggests the existence of cortical-subcortical circuits in language, as observed in other forms of cognition (e.g., Lichter & Cummings, 2001). The analysis of subcortical aphasia can significantly advance the understanding of the language representation in the brain, but indeed it does not affect the issue of aphasia classification.

Global aphasia

Global aphasia is an extended aphasia observed in cases of damage involving the complete perisylvian area of the left hemisphere (frontal, temporal and parietal areas). Its most frequent etiology is the occlusion of the major trunk of the middle cerebral artery of the left hemisphere, but it can also be the result of multiple lesions ((Pai et al., 2011). Patients with global aphasia present both, expressive and receptive defects, and as a matter of fact, it could be interpreted as a mixed Broca's, Wernicke's and conduction aphasia. Global aphasia is initially the most common type of aphasia in stroke patients (Vidović, 2011); it is observed in about one third of

the stroke patients in the acute stage, but tends to improve during the following months (Pedersen et al., 2004).

Typically, initially the patient presents an absence of speech or an expressive speech limited to a stereotype. Right hemiparesis is significant. Language understanding is seriously impaired and may be limited to just some few short commands (e.g., stand-up). Repetition is impossible, and reading and writing are severely limited. Figure 5.13 is a presentation of a typical lesion is global aphasia.

Figure 5.13. Typical lesion in global aphasia

Although some improvement is observed during the following months and years, language recovery is usually very modest. Usually, the patient learns some few utterances (e.g., "thank you"; "it is OK", etc) that are used in a correct way. Language understanding usually progresses, and the patient generally becomes able to understand some high frequency words, and stereotyped expressions (e.g., "good bye"). In a significant percentage of cases, alternative and augmentative communications systems are required to compensate for the severe language impairment.

Summary

In addition to the two major aphasic syndromes (Broca's aphasia and Wernicke's aphasia) different aphasia classifications generally include a diversity of additional language disturbances, including: (1) conduction aphasia (fluent conversational language associated with nearly normal comprehension and significant impairments in repetition); (2) extrasylvian

(transcortical) sensory aphasia (characterized by good repetition, fluent conversational language, significant amount of verbal paraphasias and neologisms, and empty speech); (3) Extrasylvian (transcortical) motor aphasia (characterized by a non fluent language, good comprehension, and good repetition); (4) supplementary motor area aphasia (characterized by an initial mutism lasting about 2–10 days; later, a virtually total inability to initiate speech; nearly normal speech repetition; a normal language understanding, and absence of echolalia; significant and rapid recovery is observed); (5) mixed extrasylvian (transcortical) aphasia (spontaneous language is absent and speech production is virtually limited to repetition; frequently echolalia is observed, but articulation is good and automatic language is relatively preserved); (6) subcortical aphasia (sometimes observed in cases of striatocapsular and thalamic pathology); and (7) global aphasia (extended aphasia observed in cases of damage involving the complete perisylvian area of the left hemisphere characterized by significant expressive and receptive language defects).

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Chapter 6

Alexia

Introduction

Alexia (or acquired dyslexia) refers to an **acquired disorder in reading caused by brain pathology** (Benson & Ardila 1996). Two case reports published by Dejerine in 1891 and 1892 represent important milestones in the study of alexia (Dejerine 1891; 1892). In the 1891 paper, he described a patient who suffered a cerebrovascular accident that produced some degree of right-sided visual field defect and mild difficulty in naming and in understanding spoken language together with a complete loss of the ability to read. The patient could write nothing but his signature. Spoken language improved, but the alexia and agraphia remained basically unchanged until his death. Postmortem examination showed an old infarct in the left parietal lobe involving three quarters of the angular gyrus and extending deep to the lateral ventricle (Dejerine 1891) (Figure 6.1).
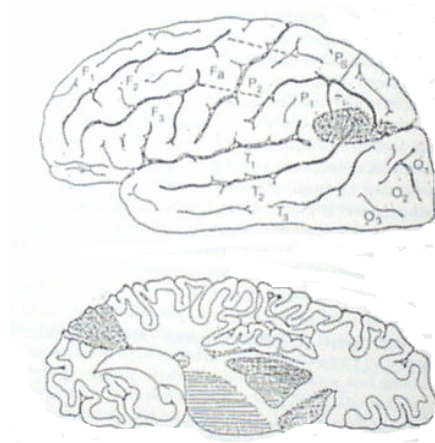
Figure 6.1. Alexia with agraphia associated with an angular gyrus cerebro-vascular accident (Dejerine, 1891)

One year later, Dejerine reported a second patient who noted an inability to read, but no other language disturbances. The only neurologic finding was a right hemianopia. Unlike the former case, this patient, although unable to read except for a few individual letters, could write adequately. Four years later, a second vascular accident led him to death. Postmortem examination revealed 2 different infarcts: 1 infarct was a large softening that involved the left angular gyrus and was obviously of recent origin, and the other infarct was an old gliotic

infarct that involved the medial and inferior aspects of the left occipital lobe and the splenium of the corpus callosum. The old infarct was the source of the alexia without agraphia (Dejerine 1892) (Figure 6.2).
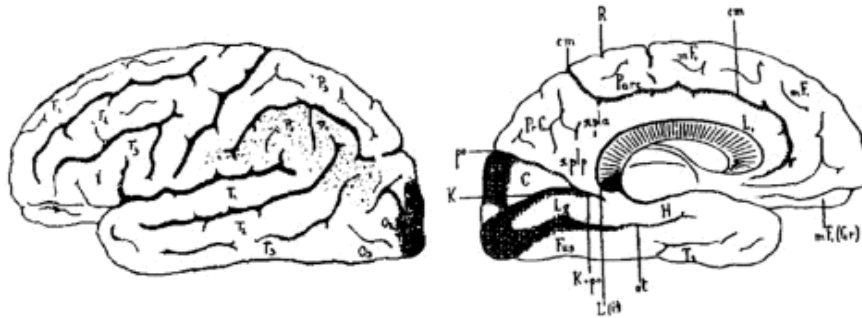
Figure 6.2. Alexia without agraphia associated with an occipital vascular accident (Dejerine, 1892)

Alexia without agraphia (also known as **occipital alexia** or **pure alexia**) and **alexia with agraphia** (**parietal-temporal alexia** or **central alexia**) were extensively investigated and corroborated during the years following Dejerine's discoveries. A third, clinically distinct alexia syndrome, **frontal alexia**, which is associated with pathology in the frontal language areas, also has been proposed (Benson 1977). Reading difficulties associated with right hemisphere pathology having a visuospatial basis, on the other hand, have been noted for quite a long time. However, only a few studies, have investigated these visuospatial reading defects using large samples of patients with right hemisphere pathology (Hécean & Marcie, 1974; Ardila & Rosselli 1994).

These four types of alexias (without agraphia, with agraphia, frontal, and spatial) represent the **neurologic, classic, or neuroanatomically-based classification of alexias.** Significant variability, however, has been observed in the pattern of disturbances, particularly in parietal-temporal alexia (with agraphia). During the 1970s and 1980s, a new approach to the analysis of alexia was developed (Marshall and Newcombe 1973; Caramazza et al 1985). This new approach to alexias is usually known as the **psycholinguistic or cognitive perspective of alexias**. Interest shifted from the anatomical correlates of acquired reading disturbances to the functional mechanisms underlying alexias. It should be noted that, in the psycholinguistic or cognitive interpretation of alexias, the name "**acquired dyslexia**" is preferred, rather than "alexia"; this latter usage is more commonly used in England, where these approaches were initiated.

The linguistic and cognitive approaches to alexia required the development of **models for normal reading**. Several partially coincidental cognitive models of normal reading have been proposed (Coltheart, 1978; Caramazza et al., 1985; Friedman, 1988) (Figure 6.3). In general, most of these models propose that after initial letter identification, reading proceeds along two linguistically different routes: (1) the direct route, wherein the written word is associated with a visual word in lexical memory; and (2) the indirect route, wherein the written word is

transformed into a spoken word following a graphophonemic set of rules, and the meaning of the word is attained through its phonological mediation. If one or the other of these reading systems is altered, different error patterns can be observed. In some cases, both systems can be disrupted simultaneously.
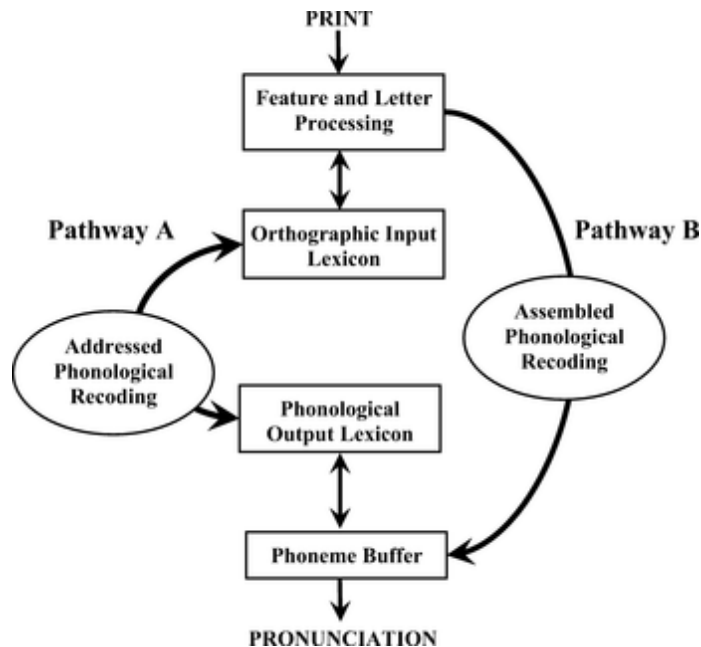
Figure 6.3. Example of dual-route model of reading (Coltheart et al. , 2001).

Classical alexia subtypes

The classic alexic syndromes include alexia without agraphia, alexia with agraphia, frontal alexia and spatial (or visuospatial) alexia.

Alexia without agraphia

The syndrome has been given many different names including **alexia without agraphia**, **pure alexia**, **pure word blindness**, **agnosic alexia, occipital alexia, posterior alexia, verbal alexia,** and more recently**, letter-by-letter reading**. The core clinical features include a serious disturbance in reading contrasted with a preservation of writing competency. Patients with occipital alexia find themselves unable to read what they have just written. Reading letters (literal reading) is relatively preserved, and reading words (verbal reading) is seriously impaired.

Sometimes, the patient fragments the letter when reading and reads only the initial letter segment (eg, "K" is read as "l"). Letter-by-letter reading aloud eventually can result in word

recognition. Patients with this type of reading disorder appear to use an inefficient eye movement strategy in reading, fixating to the left of the usual normal viewing location of words; consequently, less of the word is processed, with the refixation rate increasing and reading becomes slower (McDonald et al., 2006). It is notable that not only is the recognition of letters and words clearly impaired but also the recognition of fragmented pictures, suggesting an inefficient build-up of sensory representations (Starrfelt et al., 2010).

The process of reading individual letters aloud to recognize the word is slow and open to error, particularly on long words; reading time is proportional to the number of letters in a word, but this effect differs according to the degree of associated hemianopia (Sheldon et al 2012). **Morphological paralexias** (the misreading of the final morphemes) is a common characteristic of occipital alexia (eg, "closing" is read as "closed"). Patients with occipital alexia can recognize words spelled out loud to them, and they can recognize letters outlined on the palm of the hand. They can also match letters written with different writing forms.

Damage usually includes the left medial and inferior occipital region, particularly the fusiform and lingual gyri and the posterior segment of the geniculocalcarine pathway (Figure 6.4). Left occipital damage may result in alexia for two reasons, which may coexist depending on the distribution of the lesion. A lesion of the left lateroventral prestriate cortex or its afferents impairs word recognition ("pure" alexia). If the left primary visual cortex or its afferents are destroyed, resulting in a complete right homonymous hemianopia, rightward saccades during text reading are disrupted ("hemianoptic" alexia) (Leff et al., 2000). Impairments in oculomotor behavior during reading have been documented in this group of patients; they present a disproportionate increase in the number and duration of fixations per word and in the regressive saccades per word, suggesting that pure alexia could be the result of a general reduction of visual speed and span (Starrfelt et al 2009). It has been suggested that brain lesions in patients with pure alexia and functional imaging data support that the abstract letter identities (visual word form) are subtended by a restricted patch of left-hemispheric fusiform cortex, which is activated during reading (Kleinschmidt & Cohen 2006). Cortical stimulation of the left posterior fusiform and inferior temporal gyri results in pure alexia (Mani et al 2008). Noteworthy, associative visual agnosia is frequently observed in pure alexia, but prosopagnosia is rarely found.
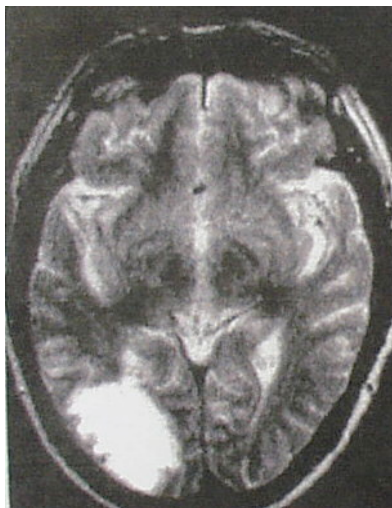
Figure 6.4. Typical lesion in pure alexia (alexia without agraphia).

Alexia with agraphia

Other names used to refer to this reading disorder are **central alexia, parietal-temporal alexia, literal alexia,** and **letter-blindness**. The characterizing features of this alexia are the impairments of reading and writing: alexia and agraphia. The ability to read aloud and to comprehend written language is disturbed. The alexia is a literal alexia (inability to read letters) resulting in a **total alexia**. Patients will fail to recognize a word when it is spelled aloud. The writing disturbance is usually equal in severity to the alexia. Their ability to copy written and printed words is far superior to their ability to write them spontaneously or from dictation. They also have difficulty in transposing cursive to printed forms and vice versa (Benson 1985). Some residual reading abilities (such as some preserved ability to recognize shape and canonical orientation of letters) have been reported, but these residual abilities probably are supported by the right hemisphere (Volpato et al 2012). This type of alexia has been informally referred to as "acquired illiteracy." Reading of other symbolic systems, such as musical notation, is also likely to be impaired. Reading numbers is usually at least partially impaired but occasionally can be spared.

Parietotemporal alexia can result from cerebrovascular disease involving the angular branch of the left middle cerebral artery (Figure 6.5). Trauma, abscess, tumor, or any pathology involving the posterior parietal area and the temporal-parietal region can be associated with alexia. Similarly, damage involving the Brodmann area 19 and white matter in the left inferior parietal lobe can result in alexia with agraphia.
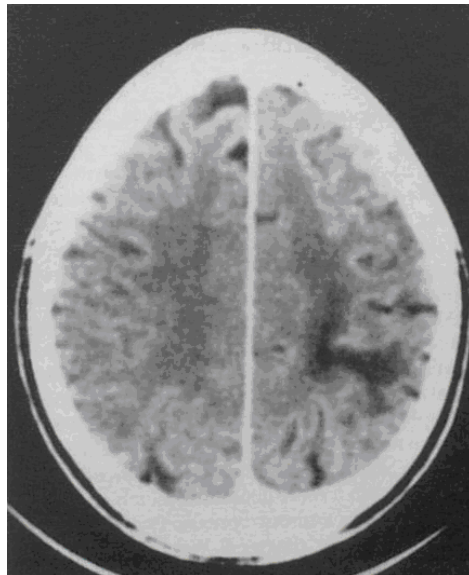
Figure 6.5. Typical lesion in alexia with agraphia.

Frontal alexia

For many years, clinicians have noted that patients with Broca aphasia had either lost the ability to read or found the task difficult. Most patients with Broca aphasia do understand some written material, but this is usually limited to individual words. The words that can be recognized are almost exclusively content words (nouns and verbs). Reading aloud as well as spoken language, is agrammatic. The difficulty that patients with frontal alexia have comprehending written material closely resembles the auditory comprehension disturbance demonstrated in patients with Broca aphasia (Benson 1977). Even though patients are able to read, they insist they cannot read and avoid reading. Patients with frontal alexia will read some meaningful words, but fail when asked to read the individual letters of a word. Although they can recognize some words spelled aloud, they have difficulty in comprehending most words. As a general rule, reading comprehension is superior to reading aloud. When reading aloud, the same speech and language problems observed in spontaneous language are noted. Any abnormal condition affecting the posterior area of the left frontal lobe can result in a frontal alexia (Figure 6.6).
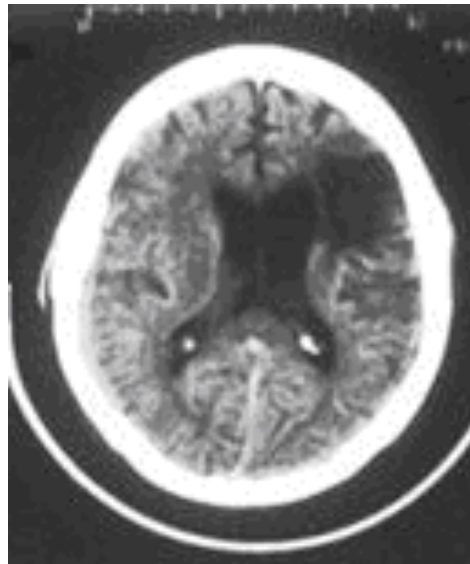
Figure 6.6. Typical lesion in frontal alexia.

Spatial alexia

Right hemisphere pathology can be associated with significant spatial disturbances. Spatial disturbances will be observed in different tasks, including reading, but specific representation of neglect for words may be independent of representational neglect for objects (Arduino et al

2012). Spatial (or visuospatial) alexia is characterized by deficits in recognizing the visuospatial arrangement of words and texts, usually associated with hemi-spatial neglect. Because of its association with hemi-neglect, frequently the name "**neglect alexia**" (or "**neglect dyslexia**") also has been used to refer to this reading disturbance. Often, patients with reading impairments of a spatial type have considerable difficulty in comprehending written material (Table 6.1).

Before landing on the island, Crusoe's father wants him to be a good, middle-class guy. Crusoe, who wants nothing more than to travel around in a ship, is definitely not into this idea. He struggles against the authority of both his father and God and decides to thumb his nose at both by going adventuring on the sea instead.

Table 7.1. Example of reading in spatial alexia. What is presented in yellow was omitted (neglected) by the patient. Noteworthy, that the extent of neglected hemi-space in each line is variable.

According to Hécean and Marcie (1974), spatial alexia is characterized by: (1) inability to fix gaze on the word or text and to move from one line to another, and (2) neglect of the left side of the text. They report that spatial alexia was observed in 23.4% of a series of 146 right-hemisphere damaged patients. In a series of 138 consecutive patients with right hemisphere stroke, Lee and colleagues (2009) found hemi-neglect in 58% of the cases and spatial alexia in 22.5%. Hemi-neglect severity and visual field defects significantly predicted reading difficulties. Spatial alexia typically has been reported in relation with spatial agraphia, spatial acalculia, and other spatial deficits. Ardila and Rosselli (1994) studied 21 patients with right hemisphere damage and found that reading errors included literal errors (substitutions, additions, and omissions of letters), substitutions of syllables and pseudo-words for meaningful words, left hemispatial neglect, confabulation, splitting of words, verbal errors (substitutions, additions, and omission of words), grouping of letters belonging to two different words, misuse of punctuation marks, and errors in following lines of text on a page. They proposed that spatial alexia is characterized by:

  • Some difficulties in the recognition of spatial orientation in letters.
  • Left hemispatial neglect.
  • Inability to follow lines of text when reading and sequentially explore the spatial

distribution of the written material.

      • Grouping and fragmentation of words, most likely as a consequence of the inability to correctly interpret the relative value of spaces between letters.

Right parietal, parietal-occipital, and parietal-temporal pathology usually results in significant spatial disturbances, including spatial alexia (Figure 6.7). Lee et al. (2009) observed that in patients with hemi-neglect, brain lesions were located in the superior and middle temporal gyri, inferior parietal lobule, and posterior insular cortex of the right hemisphere; when reading disturbances were found, additional lesions in the lingual and fusiform gyri were also observed.

Figure 6.7. Typical lesion in spatial alexia.

Psycholinguistic models of alexias (dyslexias)

Psycholinguistic models of alexias usually introduce a major distinction between **central** and **peripheral alexias**. In central alexias, the patient can perceive a word correctly but has difficulties recognizing it with either semantic or phonological processing. Three different types of central alexias are distinguished: (1) **phonological,** (2) **surface**, and (3) **deep**. Each features a specific pattern of reading errors (paralexias). In the peripheral alexias, the reading impairment appears to have more connection to a perceptual disturbance. The patient has difficulty attaining satisfactory visual word processing. Usually, three different types of peripheral alexias are recognized: (1) **letter-by-letter reading**, (2) **neglect alexia**, and (3) **attentional alexia**.

Phonological alexia

Phonological alexia is characterized by the inability to read legitimate pseudo-words, despite relatively well-preserved ability to read real words. This dissociation implies that the phonological (indirect) reading route is impaired, and reading must rely on the lexical (direct) route. Word frequency (probability of appearance) plays a crucial role; high frequency words are likely to be read, whereas pseudo-words (zero frequency) are usually impossible to read. Real words are stored in lexical memory, whereas pseudo-words are not present in the lexicon. Patients with phonological alexia cannot use the spelling-to-sound correspondence (graphophonemic) rules in written language. When reading, **visual paralexias** are frequently observed; thus, the patient will read real words as other words that are visually similar to the target word. The target word and the paralexic error have many letters in common (eg, "mild" is read as "slid"). Hamilton and Coslett (2007) reported a patient with phonological alexia who was impaired in writing affixed words (i.e., words including a root and an affix, such as "flowed"; "flow" corresponds to the root and "ed" to the affix) but did not demonstrate that defect when reading affixed words.

Phonological alexia has been reported in cases of diverse brain pathology. In general, however, the middle cerebral artery territory is involved, most frequently the superior temporal lobe and angular and supramarginal gyri of the left hemisphere. Functional neuroimaging studies have suggested that the left frontal operculum is more active when normal participants read pronounceable pseudo-words as compared to most word types. Damage in this area results in defects at reading pseudo-words associated with a relatively intact word reading ability, a pattern observed in phonological alexia (Fiez et al 2006).

Surface alexia

The indirect route (graphophonemic) reading system is available to patients with surface alexia, whereas the lexical (direct) route is impaired. Consequently, surface alexia represents an acquired disorder characterized by the superior reading of regular words and legitimate pseudo-words in comparison to irregular words. Legitimate pseudo-words can be easily read because they rely on the indirect (phonological) route. The overuse of the preserved phonological route will result in "regularization errors". According to Friedman (1988), surface alexia is characterized by:

• Regularization errors that are always observed (irregular words are phonologically read), but with variable frequency.
• Frequency effects, grammatical category effect, and length effect are reported in only a few cases.
• Surface alexia is associated with lexical (surface) agraphia.
• Fluent aphasia is found in most cases.
• Almost all patients present a left temporal or temporoparietal lesion.

Interestingly, significant activation in the left anterior middle temporal gyrus is associated with healthy individuals reading irregular words (which is impaired in surface alexia) (Wilson et al 2012).

Deep alexia

If both the lexical (direct) and phonological (indirect) routes are impaired, only limited residual reading ability will remain. Some distinguishing characteristics have been proposed for deep alexia:

   • **Semantic paralexias** are always observed (e.g., "lawyer" is read as "attorney"). Varieties of semantic paralexias have been proposed.
   • Success in reading a word is affected by the grammatical category and imageability (concrete nouns are read better than abstract nouns).
   • Pseudo-words cannot be read.
   • Visual and derivational (i.e., morphological) paralexias are always observed.
   • Deep alexia is always associated with aphasia and agraphia.

It has been proposed that deep alexia represents reading that relies extensively on right-hemisphere orthographic and semantic processing (Coltheart 2000). Colangelo and Buchanan (2005) studied a patient with deep dyslexia who was able to read aloud a series of ambiguous (e.g., bank) and unambiguous (e.g., food) words as well as perform a lexical decision task using these same items. When required to explicitly access the items (i.e., naming), the patient showed relative impairment for ambiguous compared to unambiguous words. The authors proposed that errors in production were due to a failure to inhibit spuriously activated candidate representations. Warrington and Crutch (2007) reported a subject who presented a better ability to read concrete than abstract words; furthermore, reading concrete words corresponding to living items was more accurate than reading concrete words corresponding non-living items. The authors interpreted this pattern of performance as evidence for a degree of autonomy for the semantic processing of written words.

Anatomical lesions causing deep alexia are commonly extensive left-hemisphere insults, including the Broca area.

Attentional alexia

Shallice and Warrington (1977) reported two patients who were able to read single words but unable to read multiword displays or to name the constituent letters of the word. They presented deep left parietal tumors, and both presented right homonymous hemianopia. Their impairment was not specific for letters, but included all the stimuli in which more than one item of the same category was simultaneously present in the visual field (numbers and even pictorial material). The underlying problem in attentional alexia is attributed to a deficit in selective attention, which is not specific to orthographic (i.e., written) material.

Regardless the apparent differences between both interpretations of alexias, as a matter of fact, the classifications of reading disturbances presented in the neurological ("classical") and psycholinguistic approaches are not contradictory; indeed, they can be easily equated, as observed in Table 6.2.

Neurological Classification	Psycholinguistic Classification
	Central alexias (dyslexias)
Parietal-temporal alexia	Surface alexia Phonological alexia
Frontal alexia	Deep alexia
	Peripheral alexias (dyslexias)
Occipital alexia	Letter-by-letter reading
Spatial alexia	Neglect alexia

Table 6.2. Correspondence between the neurological ("classical") classification of alexias, and the psycholinguistic classification.

Other alexias

Aphasic alexia

Aphasic patients present characteristic reading difficulties that can be related directly to their basic language defect. In conduction aphasia, for example, reading comprehension is better than reading aloud, just as auditory comprehension is superior to repetition of spoken language. When reading aloud, **literal paralexias** are observed, parallel to the phonological paraphasias in spoken language. Patients with extrasylvian motor aphasia may show "frontal deficits" when reading; thus, they can misread a phrase due to perseveration. They usually read pseudo-words as real words (the pseudo-word is mispronounced to sound like a visually similar real word) (Ardila et al., 1989). Reading defects in Broca aphasia are usually significant, particularly for reading grammatical words and reading aloud. Alexia in Broca aphasia corresponds to so-called frontal alexia. Patients with Wernicke aphasia may produce substitutions, omissions, additions, and even neologistic reading. Comprehension of written language is often severely impaired. Extrasylvian sensory aphasias are associated with some reading difficulties, even though severity of alexia can vary. Anomic aphasia patients have defects in interpreting the meaning of written words. When damage extends posteriorly,

some degree of occipital alexia may be present.

Hemialexia

Following surgical section of the posterior corpus callosum, some patients have significant difficulties in reading material visualized to the left visual field, but normal reading for the material presented to the right visual field can be observed. This condition has been termed as hemialexia. Hemialexia can occur with any pathology (e.g., tumors) that destroys the splenium of the corpus callosum.

Alexia in phonological and logographic writing systems

It has been proposed that characteristics of alexia correlate with the idiosyncrasies of writing systems (Coltheart 1982). The lexical organization and processing strategies that are characteristic of skilled reading in different orthographies are affected by different developmental constraints in different writing systems (Ziegler & Goswami 2005). In bilingual speakers, alexia can be restricted to only one language (Kremin et al., 2000). Alexias, however, have been studied mostly in Indo-European language writing systems, and cross-linguistic analyses are scarce. Psycholinguistic models of alexias have been developed especially in English and French, two languages with rather irregular writing systems. In English, with a significant amount of irregular words (words that cannot be read using grapheme-to-phoneme correspondence rules and can only be recognized as a whole), the existence of two different reading strategies or reading routes (indirect and direct) is understandable. Developmental dyslexia has been found to be more frequent in irregular writing systems, such as English or French, than in regular orthographic systems, such as Italian (Paulesu et al 2001). The applicability of the double route reading models to regular (phonologic) writing systems has been challenged (Lukatela & Turvey, 1998; Karanth, 2003).

Reports about alexia in logographic writing systems (e.g., Chinese) are extremely scarce. With the exception of some studies on the Japanese Kana and Kanji reading systems, comparative research on alexias and agraphias in non-Indo-European languages has been extremely limited (Yamada et al., 1990; Sakurai, 2004). Pure alexia, selectively impairing Kana (but not Kanji) reading, has been reported in cases of left posterior occipital lobe damage (Sakurai et al. 2001) similar to the anatomy of pure alexia in other phonographic systems. Conversely, alexia with agraphia in Korean Hanja (logographic), but preserved Hangul (phonographic) reading and writing have been reported after a left posterior inferior temporal lobe infarction (Kwon et al., 2002). Sakurai and colleagues (2006) distinguished two different types of pure alexia: pure alexia for Kanji (and Kana; fusiform type: pure alexia for words) characterized by impairments of both whole-word reading, as represented in Kanji reading, and letter identification; and different from pure alexia for Kana (posterior occipital type: pure alexia for letters) in which letter identification is primarily impaired. Thus, individuals using two different writing systems (e.g., ideograms and phonograms as found in Japanese and Korean) may present a dissociated alexia. Yamawaki et al. (2005) observed in a specific form of alexia that oral reading of Kanji words significantly correlates with naming pictures corresponding to the words, suggesting that naming the objects and reading the logographic Kanji words share common underlying mechanism.

These studies, as a whole, indicate that reading strategies and alexia characteristics are

under the influence of the idiosyncrasies of the individual reading systems (Karanth 2003).

Some special forms of alexia

Alexia has been reported in blind people for Braille reading following bilateral (Hamilton et al 2000) or right-sided occipital damage (Perrier et al 1988). Therefore, in blind people, reading Braille depends at least in part on occipital lobe activity. The right occipital area, in particular, seems to play a major role in reading Braille; however, paralexias for Braille reading have also been observed in cases of right parietal pathology. Braille alexia might be interpreted as a tactile agnosia (Larner 2007).

Kinesthetic alexia (inability to read following the letter with the fingers) with preserved visual reading has been associated with left parietal damage (Ihori et al 2002). Noteworthy, kinesthetic reading has been used as a successful procedure in the rehabilitation of alexia without agraphia.

Summary

Brain pathology frequently is associated with disturbance in the reading ability (alexia). Since the 19th century, two major types of alexias have been recognized (alexia with and without a preserved ability to write). In the mid-20th century, two additional types of alexias were proposed (alexia due to spatial disturbances and alexia associated with frontal pathology). During the 1970s and 1980s, a new approach to the analysis of alexia was developed. This new approach (psycholinguistic or cognitive perspective) shifted the focus from the anatomical correlates of acquired reading disturbances to the functional mechanisms underlying alexias. A major distinction between central (phonological, surface, and deep alexia) and peripheral (letter-by-letter reading, neglect alexia, and attentional alexia) is introduced. Significant parallelism between both approaches (classical or neurological; and psycholinguistic or cognitive) can be established. Some special types of alexias such as alexia for Braille reading have also been reported. Contemporary neuroimaging studies have significantly contributed to a better understanding of brain organization of reading processes and reading disturbances.

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Chapter 7

Agraphia

Introduction

Agraphia (or acquired dysgraphia) refers to an **acquired disorder in writing caused by brain pathology** (Benson & Ardila 1996). The ability to write can be impaired as a result of linguistic defects (aphasic or linguistic agraphias) or non-linguistic defects (e.g., motor or spatial) (non aphasic or linguistic agraphias).

Writing requires diverse abilities such as: knowledge of the language codes (phonemes, words), an ability to convert phonemes into graphemes, an understanding of the orthographic system, a motor skill to perform some specific fine movements, and an appropriate use of the space to distribute, joint, separate letters and words. It is understandable that diverse types of agraphia can be found associated with brain pathology.

Historical Development

In 1867 Ogle coined the term **agraphia** to refer to the acquired disturbances in writing due to abnormal brain conditions. Exner (1881) proposed a "**writing center**" (base of the second frontal gyrus, in front of the primary motor area of the hand; currently known as Exner's area; see Figure 8.5). Dejerine (1891) described the "alexia without agraphia" syndrome. Gerstmann (1940) proposed that agraphia can appear with acalculia, right-left disorientation, and finger agnosia in a single syndrome (**Gerstmann's** or **angular gyrus syndrome**)

Various attempts at the classification of agraphias are found. Goldstein (1948) distinguished two main types of agraphia: apractoamnesic and aphasoamnesic. Luria (1976, 1980) referred to five different groups, three of them associated with aphasic disorders (sensory agraphia, motor afferent agraphia, and motor kinetic motor) and two resulting in visuospatial defects. Hecaen and Albert (1978) distinguished four varieties of agraphia: pure, apraxic, aphasic and spacial. Recently linguistic classifications been proposed (e.g., Roeltgen, 1985) including phonological, lexical, and deep agraphias. Table 7.1 presents a classification of agraphias.

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NEUROLOGICAL CLASSIFICATION

Aphasic Agraphias
      Agraphia in Broca Aphasia
      Agraphia in Wernicke Aphasia
      Agraphia in Conduction Aphasia
      Other Aphasic Agraphias

Non-Aphasic Agraphias
      Motor Agraphias
          Paretic Agraphia
          Hypokinetic Agraphia
          Hyperkinetic Agraphia
          Other Motor Agraphias
      Pure Agraphia
      Apraxic Agraphia
      Spatial Agraphia
Other Writing Disturbances
      Hemiagraphia
      Frontal Writing Disturbances
      Confusional States
      Psychogenic Agraphia

PSYCHOLINGUISTIC CLASSIFICATION OF AGRAPHIAS

      Central agraphias
      Phonological agraphia
      Lexical (surface) agraphia
      Deep agraphia
      Peripheral agraphias
      Spatial (afferent) agraphia
      Apractic agraphia

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Table 7.1 Agraphia classification (according to Benson & Ardila, 1996)

This chapter initially discusses the classical forms of agraphia (aphasic and non aphasic agraphias) and then analyses the psycholinguistic classifications of writing impairment. The classification model proposed by Benson and Ardila in 1996 will be used.

Aphasic Agraphias

Patients with aphasia present fundamental linguistic defects manifested both in their expressive oral language, and in their writing. The agraphia is then a consequence of this fundamental language defect and is parallel to the linguistic difficulties in oral language.

Agraphia in Broca's Aphasia

Patients with Broca's aphasia present a writing disturbance clearly correlated with the fundamental linguistic defect (Table 7.2). Writing is slow, clumsy, painstaking, short and agrammatic. Literal paragraphias due to anticipations (*take ->kake*), perseverations (*take -> tate*), and letter omissions, particularly in syllabic clusters (*glass->gass*) are observed. Calligraphy is poor. Figure 7.1 presents and example of agraphia in Broca's aphasia.

Broca's aphasia	Agraphia in Broca's aphasia
SPOKEN OUTPUT	**WRITTEN OUTPUT**
Sparse output	Sparse output
Effortful	Effortful
Poor articulation	Clumsy calligraphy
Short sentences	Abbreviated output
Dysprosody	----
Agrammatism	Agrammatism
Phonological paraphasias	Literal paragraphias

Table 7.2 Comparison between oral and written production in Broca's aphasia (according to Benson & Ardila, 1996)

Right hemiparesis is usually observed, and the patient has to use his/her left hand in writing. This change implies an additional problem. Writing difficulties, in consequence, are not only the result of the linguistic defects (aphasic agraphia) but also of the clumsiness in writing with the non-preferred hand.

It is common to find that the spelling is inadequate: there are omissions, particularly of grammatical elements, and writing in general is scarce and agrammatic. Interestingly, agrammatism may be more evident in written language than in spoken language. If the patient writes with his right hand (using a special device) writing can improve, suggesting that in his/her writing with the left hand not only are there elements of a linguistic agraphia (as a result of the aphasia), but also of a hemiagraphia due to interhemispheric disconnection. In practice, the lesions usually extend beyond the Broca's area and include connections between the cortex

and the basal ganglia and the primary motor area of the hand. Consequently, agraphia in Broca's aphasia could be interpreted not only as an aphasic agraphia, but also as a motor agraphia (not aphasic) and even a hemiagrafia due to interhemispheric disconnection.
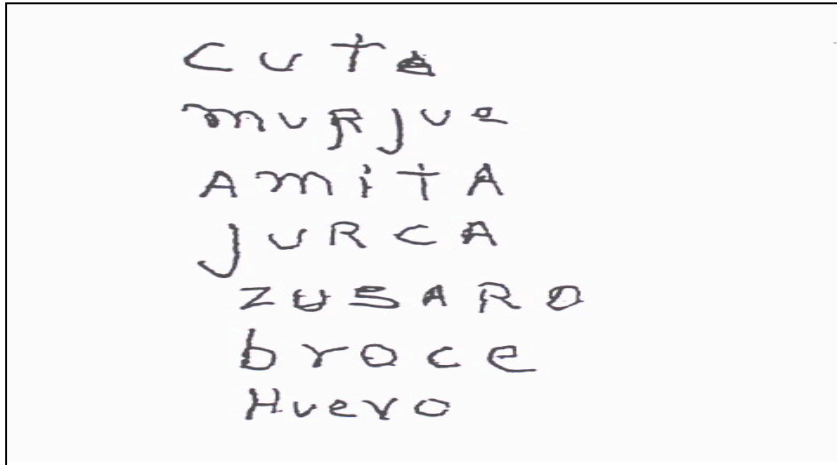
Figure 7.1. Example of agraphia en Broca's aphasia.

Agraphia in Wernicke's Aphasia

Patients with Wernicke's aphasia present an impairment in writing ability characterized by a fluent writing, well-formed letters, combined in an inappropriate way. Table 8.3 presents a comparison between oral and written production in Wernicke's aphasia. Literal paragraphias (additions, substitutions and omissions of letters), verbal (word substitutions) paragraphias and neologistic (non understandable pseudo-words) are also found. Written language deficit parallelizes the oral language defect. Grammatical elements are observed; frequently, these grammatical elements are overused. Sentences may lack clear limits; nouns may be under-represented. Writing, even though fluid, may not be understandable (jargonagraphia). Figure 8.2 illustrates the writing defects in Wernicke's aphasia.

Writing in word-deafness syndrome is theoretically preserved, except of course, writing to dictation.

In summary, the defect in writing associated with Wernicke's aphasia, is parallel to the oral defect. Since Wernicke's aphasia is a relatively heterogeneous syndrome, it is also natural to expect some heterogeneity in agraphia. Table 7.3 presents a comparison between oral and written production in Wernicke's aphasia.

Wernicke's aphasia   **Agraphia in Wernicke's aphasia**

SPOKEN OUTPUT   **WRITTEN OUTPUT**

Normal articulation   Normal calligraphy
Fluent   Fluent
Normal phrase length   Normal sentence length
Normal prosody   ----
Paragrammatism   Paragrammatism
Paraphasias   Paragraphias

Table 7.3 Comparison between oral and written production in Wernicke's aphasia (according to Benson & Ardila, 1996)
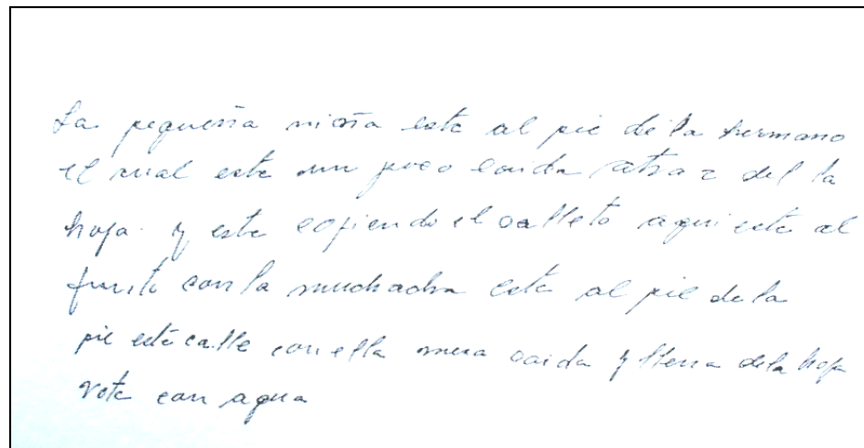
Figure 7.2. Example of agraphia in Wernicke's aphasia.

Agraphia in Conduction Aphasia

Agraphia in conduction aphasia is variable, depending upon the extent of the damage. Luria (1977, 1980) referred to this agraphia as an "afferent motor agraphia" (that is, the agraphia of the afferent motor aphasia, that is the name used by Luria to refer to conduction aphasia). Spontaneous writing is much better than writing by dictation (as spontaneous language is better than language repetition). Literal paragraphias (substitutions, omissions and additions of letters) are abundant, particularly in complex and unusual words. The patient recognizes that the word is incorrectly written, but when attempting to correct it, new errors are observed. Writing contains a significant number of self-corrections (*conduit d'approche*). The patient may state that he/she knows the word (and even repeats it for him/herself), but cannot remember how to write it. Writing is slow and difficult. Some ideomotor apraxia may exist, and consequently letters are poorly formed, even though in general they are recognizable. Sometimes an evident apraxic agraphia is found. Figure 7.3 illustrates the writing defects in conduction aphasia.
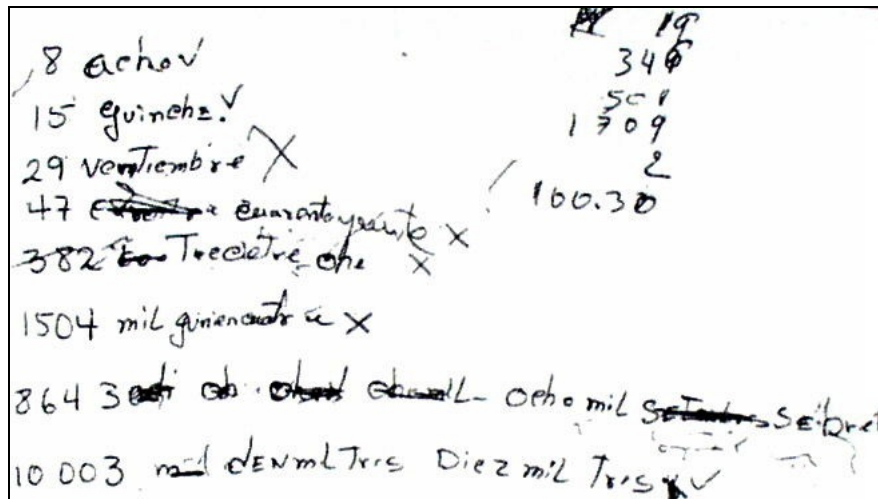
Figure 7.3 Example of writing in conduction aphasia.

The agraphia associated with conduction aphasia is variable. Sometimes the defect is mild and it is found only in writing by dictation. Other times the defect can be so severe that the patient is totally unable to write.

Other Aphasic Agraphias

Patients with global aphasia present severe defects in writing. Production is difficult to understand and is sometimes limited to certain features or poorly formed letters. In mixed

extrasylvian aphasia, there is a severe writing defect, but some ability to copy is preserved (such as the ability to repeat). Writing by dictation is impossible.

In extrasylvian sensory aphasia the patient presents an important word-finding difficulty that is also observed in writing; verbal paragraphias are consequently abundant. The writing defect is variable depending upon the extension of the pathology to the parietal lobe. In extrasylvian motor aphasia there is an import difficulty to write that may correspond to a "dysexecutive agraphia"; reading, however, is much better (as language understanding is much better in general than language production).

In alexia with agraphia there is a significant decrease in writing ability; the patient frequently cannot recognize and cannot write letters, and a literal agraphia is consequently observed. Associate oral language disturbances are variables, but they can include some Wernicke's aphasia, extrasylvian sensory aphasia, and quite often the Gerstmann's syndrome (right-left disorientation, finger agnosia, acalculia and agraphia). Occasionally, it can be associated with some apraxic agraphia

Non-Aphasic Agraphias

In addition to language skills, writing also depends on complex spatial and motor skills. The motor defects alter writing, and spatial defects cause disruption in the spatial organization of writing.

Motor Agraphias

Motor alterations in writing may appear as a result of injury to the central nervous system that involves the basal ganglia, the cerebellum and the corticospinal tract; or as a result of injuries affecting the peripheral nerves and the mechanical aspects of hand movements (Benson & Cummings, 1985).

Paretic agraphia

The alterations of peripheral nerves, either by neuropathy or by nerve entrapment can alter writing. In particular, the commitments of the radial and ulnar medial nerves affect the ability to write. The lower motor neuron dysfunction may also affect the upper extremity muscles needed for writing. In injuries affecting upper motor neurons, spastic rigidity occurs. The patient with a paretic hand tends to write block letters with poorly formed, unusually large characters.

Hypokinetic agraphia

Extrapyramidal dysfunction may be manifested in hypokinesia, as occurs in Parkinsonism, or a hyperkinetic disorder as seen in chorea. Two different types of micrographia (Figure 7.4) have been pointed out in Parkinson's disease; in one, letters are always small, while in the other, there is a progressive decrease in their size.
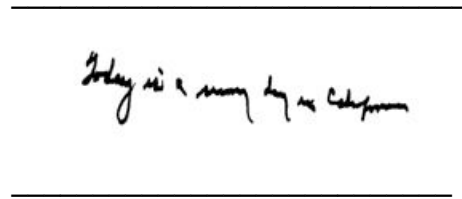
Figure 7.4. Example of micrographia in Parkinson's disease

Hyperkinetic agraphia

Hyperkinetic movements of the upper limbs alter the ability to write. This may be due to tremors, tics, dystonia, and chorea. Of the three types of tremors (Parkinsonian, postural, and cerebellar), the last two particularly affect the ability to write. Postural tremors may appear in a variety of clinical conditions and are exacerbated during stress. Choreiform movements can produce a total inability to write. Neuroleptic-induced tardive dyskinesia often includes choreiform movements of the hand and fingers, but does not usually disabling writing.

A highly controversial pathology in literature is the so-called "writer's cramp". It consists of an inability to write resulting from dystonia. It progressively appears prematurely during the act of writing, and often occurs in people who spend long periods of time writing. Although no specific neuropathological changes have been identified, it seems to be the result of a dysfunction of the neurotransmitters that change the activity of the basal ganglia (Benson & Cummings, 1985)

Pure agraphia

Exner (1881) proposed a "center for writing," located at the base of the second frontal gyrus, in front of the primary motor area of the hand; so-called "Exner's area" (Figure 7.5). Since then, the existence of some "pure agraphia" associated with Exner's area pathology has been polemic. Some authors name the apraxic agraphia as pure agraphia. Dubois, Hécaen, & Marcie (1969) reported six cases of pure agraphia, four of them associated with a frontal lesion. Some other reports have been published supporting the existence of a significant writing defect in cases of damage to the left second frontal gyrus (Exner's area). Writing is a complex activity; consequently, it is very sensitive to brain pathology. It can be conjectured that patients with minor frontal pathologies around the Broca's area can present minor language defects only observed in the written language.
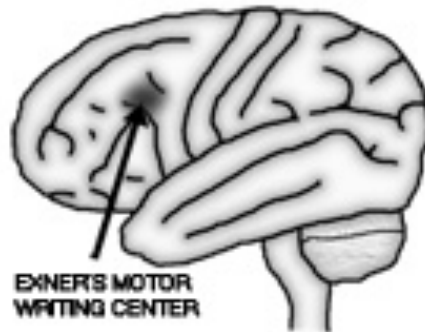
Figure 7.5 Location of the Exner's area corresponding to the second frontal gyrus of the left hemisphere.

Apraxic agraphia

For a long time now, it has been mentioned that there is a significant association between agraphia and apraxia. Kleist (1923) distinguished several types of apraxic agraphia (apraxia for holding the pencil, apraxia for writing texts, and apraxia for writing letters) and Goldstein (1948) referred to apractoamnesic agraphia. Hécaen and Albert (1978) define apraxic agraphia as the inability to form graphemes; letter distortions and inversions are observed. The patient may retain the ability to spell words and form words with letters written on cards. Agraphia is evident in all modes: spontaneous writing, copying, and dictation. Eventually, the patient is able to write short sentences, but there are obvious paragraphic errors. Sometimes apraxic agraphia has been called "pure agraphia". Hécaen and Albert (1978) distinguished between two forms of apraxic agraphia: in one, the patient has no associated aphasia and alexia, but ideomotor apraxia is evident in his left hand and apraxic agraphia in his right hand. Very often, the apraxic agraphia is associated with other signs of the parietal syndrome; alexia and certain difficulties in language comprehension are frequent. However, agraphia does not depend on aphasia, but represents an inability to program the movements required to form letters and words. Crary and Heilman (1988) emphasize that apraxic agraphia can appear without other manifestations of ideomotor apraxia (pure agraphia). Therefore, apraxic agraphia is often equated with pure agraphia (Auerbach & Alexander, 1981).

Spatial Agraphia

Spatial or visuospatial agraphia has been considered as a non-aphasic writing disorder, resulting from visuospatial defects that impair orientation and correct sequencing in writing. It has been defined as a disturbance in graphic expression due to impairment of visuospatial perception resulting from a lesion in the non-language-dominant hemisphere (Hécaen & Albert, 1978).

According to Hécaen and Albert (1978), spatial agraphia has the following characteristics:

(1) Some graphemes are produced frequently with one, two, or even more extra strokes.

(2) The lines of writing are not horizontal but slanted at variable angles of inclination to the top or bottom of the page.

(3) Writing occupies only the right-hand part of the paper.

(4) Blanks are inserted between graphemes that make up the word, disorganizing the word and destroying the unity.

It is usually associated with spatial alexia, spatial acalculia, left hemi-spatial neglect, constructional apraxia, and general spatial difficulties (Table 7.4). Some degree of spatial agraphia is observed in about 75% of the patients with right retro-Rolandic lesions, and about 50% of patients with right pre-Rolandic lesions.

The improper use of space while writing appears as one of the salient features (e.g., *the man walks down the street -> theman wal ksdownth ester et*). There is also a lack of respect for the space used and the patient can write in a manner overlapping what s/he had previously written. The iteration of features in letters (especially m and n), and letters in words (especially in graphemes using duplicate letters, such as "ss" and "tt") are observed more frequently in patients with pre-Rolandic lesions (e.g., *written -> writttten*) (Hécaen & Marcie, 1974). However, features and letter omissions are also found. There is also an inability to maintain a horizontal line in the script and the patients' writing follows an oblique or unsteady direction (Table 7.4). The use of left margins too large, and its progressive increase, leads to the so-called "cascade phenomenon" in writing (Ardila & Rosselli, 1993). There is also a tendency to change the type of writing from premorbid writing: right-injured patients tend to prefer writing using print. This change in calligraphy could be associated with some general disautomalization of the act of writing, which is also manifested in other forms of automatic writing (such as is the patient's signature).

Neglect represents one of the factors responsible for the difficulties in writing found in patients with right retro-Rolandic lesions. The iterations of features and letters represent the most important defect in the case of pre-Rolandic lesions (Ardila & Rosselli, 1993). Neglect is manifested in the presence of the aforementioned "cascade phenomena", and even in the "mental representation" of words. One patient, for example, when dictated words and sentences, tended to write only the right part of the words (e.g., *Peter is walking -> er ing*).

	Visual defects	Spatial neglect	Constr. apraxia	Spatial agraphia	Spatial alexia	Spatial acalculia
Left hemiparesis	0.00	0.07	0.14	0.28	0.07	0.00
Visual field defects		0.14	0.20	−0.11	0.28	0.33
Spatial hemi-neglect			0.27	0.36	0.55	0.39
Constructional apraxia				0.49	0.49	0.22
Spatial agraphia					0.60	0.37
Spatial alexia						0.58
Spatial acalculia						

Table 7.4. Correlations between different neurological and neuropsychological disorders observed in cases of right hemisphere pathology; a significant association between spatial agraphia and constructional apraxia, hemi-spatial neglect and spatial alexia is observed (according to Ardila & Rosselli, 1993).

The iterations could be explained as a consequence of some motor disautomatization and the tendency to persevere, not inhibit, a previous movement. Patients with right hemispheric lesions also tend to have iterations in expressive language (Ardila, 1984), somehow similar to an acquired stuttering. It has been proposed that the phenomenon of iteration during writing, in patients with right hemispheric lesions, can match different forms of perseveration in speech (Marcie et al., 1965).
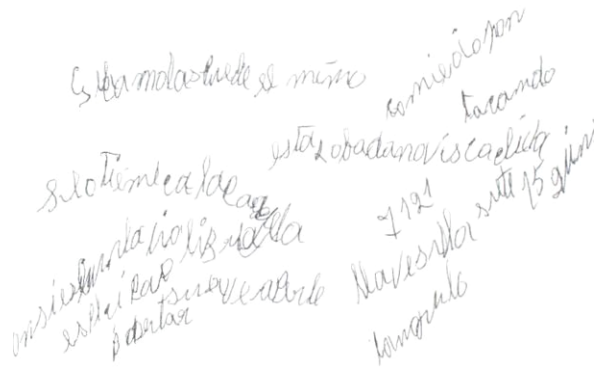
Figure 7.6. Example of spatial agraphia.

To sum up, spatial agraphia is characterized by: (1) feature and letter omissions and/or additions; (2) inability to correctly use the spaces to join and separate words; (3) difficulty in maintaining a horizontal written line; (4) increased left margins and persistence in continuing the left margins (so-called "cascade phenomenon"); (5) disregard of spaces and spatial disorganization of the written material; (6) disautomatization and changes in handwriting style; and (7) constructional apraxia for writing.

According to Ardila and Rosselli (1993) writing defects associated with right hemisphere damage are a consequence of:

(1) Left hemi-spatial neglect, manifested as an inconsistent increases in left margins

(2) Constructional difficulties in writing manifested in disautomatization and changes in handwriting style, as well as grouping of elements in writing.

(3) General spatial defects: inability to use correctly the spaces between words, difficulties to maintain a horizontal writing, and general spatial disorganization

(4) Some motor disautomatization and tendency to perseverate.

Dystypia

Writing has significantly changed during the last decades. Writing is not anymore equivalent to handwriting, but in a significant extent, writing means to use a computer word processor. Progressively, contemporary people use more a more computers for writing; and handwriting, as a matter of fact, is becoming relatively unusual.

Writing using a computer keyboard obviously is not the same cognitive, motor, and spatial task as using a pencil and a paper. Although the conceptual knowledge of written language can be

the same, the motor activity and the spatial abilities that are used are rather different (Ardila, 2013).

Regardless the enormous amount of agraphia patients reported in the neurology and neuropsychology literature, very few cases of typewriting disturbances have been documented. For instance, Boyle and Canter (1987) described a skilled professional typist who after a left cerebrovascular accident, sustained a disturbance of typing disproportionate to her handwriting disturbance. Typing errors were predominantly of the sequencing type, with spatial errors much less frequent. Depriving the subject of visual feedback during handwriting greatly increased her error rate. The authors suggested that impaired ability to utilize somesthetic information—probably caused by the subject's parietal lobe lesion—may have been the basis of the typing disorder.

Otsuki and colleagues (2002) reported on a 60-year-old right-handed Japanese man who showed an isolated persistent typing impairment without aphasia, agraphia, apraxia, or any other neuropsychological deficit. They proposed the term "dystypia" for this peculiar neuropsychological manifestation. The symptom was caused by an infarction in the left frontal lobe involving the foot of the second frontal convolution and the frontal operculum. The patient's typing impairment was not attributable to a disturbance of the linguistic process, since he had no aphasia or agraphia; nor was it attributable to an impairment of the motor execution process, since he had no apraxia. Thus, it was deduced that his typing impairment was based on a disturbance of the intermediate process where the linguistic phonological information is converted into the corresponding performance. The authors hypothesized that the foot of the left second frontal convolution and the operculum may play an important role in the manifestation of "dystypia."

No question, toward the future the analysis of agraphia should include no only the study of handwriting disturbances, but also the writing disorders observed when using the new writing technologies (computers, cellular phones, etc.).

Other Writing Disorders

Hemiagrafia

When the corpus callosum is cut, the patient can normally write with their right hand, but fails completely when trying to write with their left hand. It has been proposed that the left hemisphere controls the activities needed to write, and writing with the left hand in right-handed subjects is achieved using commissural fibers of the corpus callosum (Geschwind, 1965). This type of impairment in writing has been called hemiagrafia or disconnection agraphia (Lebrun, 1987) (Figure 7.7).
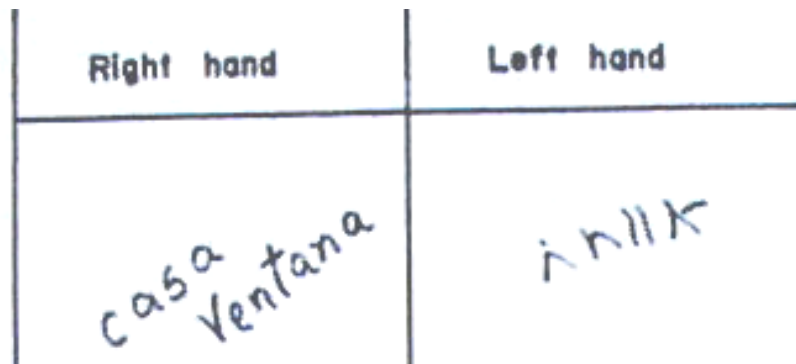
Figure 7.7. Writing with the right and left hand in a patient with a section of the corpus callosum.

Frontal ("dysexecutive") agraphia

It should be emphasized that patients with prefrontal lesions may present an obvious difficulty in writing. These patients, however, have a normal ability to read.

This is not, however, a primary defect in writing. These patients frequently have a defect in spoken language (extrasylvian motor aphasia) characterized by a decrease in spontaneous speech, with good repetition and understanding. This deficit is manifested also in the literacy system. Agraphia originating in the frontal lobe might be called "dysexecutive agraphia".

For this group of patients, reading (recognition) is notably higher than writing (production). As in oral language, written production is scarce, and often the patient does not finish what they try to write. Copying may be higher than spontaneous writing. Perseveration is manifested in writing as in any other motor acts. Perseveration may include words, letters or features. Perseveration is not only seen in focal frontal damage, but also in dementias, particularly in Pick's disease (frontotemporal dementia), a disease associated with significant pathology of the frontal lobes.

Confusional states

Chedru and Geschwind (1972) found that patients in confusional states resulting from various etiologies, could speak, understand, repeat, name and read, but they could not express their ideas in writing. Writing was slow, clumsy, and with vague meaning. The authors emphasize the susceptibility of the written output to any disturbance in the brain function.

Psychogenic Agraphia

Hysterical paralysis frequently involves the left hemibody, and therefore it is not usual to report hysterical agraphia. The hysterical agraphia can be due to a conversion reaction or a hand tremor (Benson & Cummings, 1985), but most likely many of the reported cases of hysterical

agraphia in classical literature were suffering from idiopathic focal dystonia (writer's cramp). In hysterical paralysis, reflexes are normal and there are only moderate changes in muscle tone. Sometimes the sensitivity is also affected.

Ardila (1989) reported an unusual case in a patient with a hysterical personality, who had learned the bizarre ability to write backwards (from the last to the first letter) and could sign with both hands in any direction. The patient also had a history of difficulties in learning to read and mixed handedness.

Psycholinguistic Models of Agraphias

In recent years, there has been great interest in the development of psycholinguistic models of agraphias (Roeltgen, 1993). Different levels of language processing have been correlated (phonological, lexical, semantic) with particular defects in written language.

In these models it is usual to distinguish two main groups of agraphias (disgraphias): central and peripheral (Ellis, 1988), similar to the distinction between central and peripheral alexias (Shallice & Warrington, 1980). Table 7.5 presents a comparison between classical agraphic syndromes (neurological or anatomical classification of agraphias) and the agraphic syndromes according the psycholinguistic approach.

Central agraphias (dysgraphias)

Central agraphias affect one or more of the processes involved in the spelling of familiar and non-familiar words and pseudowords. Central agraphia affects spelling in all ways: handwriting, typing, oral spelling, etc. Three sub-types are distinguished: phonological, lexical (surface) and deep.

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NEUROLOGICAL CLASSIFICATION	PSYCHOLINGUISTIC CLASSIFICATION
Aphasic agraphias	**Central agraphias**
Classified according to the basic language disorder associated with agraphia: agraphia in Broca aphasia (non-fluent agraphia), agraphia in Wernicke aphasia (fluent agraphia), etc.	Lexical (surface) agraphia Phonological agraphia Deep agraphia
Non-aphasic agraphias	**Peripheral agraphias**
Motor agraphia Spatial agraphia Apraxic agraphia	Spatial (afferent) agraphia Apraxic agraphia

_____

Tabla 7.5. Correspondence between the neurological and psycholinguistic classification of agraphias.

Phonological agraphia

In this type of disturbance, the patient maintains the ability to write familiar words, both regular and irregular, but is unable to spell nonwords. There is a good performance in legitimate words, observed even in low-frequency words that contain unusual spelling patterns. In contrast with the ability to write regular words, there is a striking inability to write legitimate pseudowords under dictation (Bub & Kertesz, 1982; Baxter & Warrington, 1985). Spelling errors observed in this group of patients are not usually phonologic, but may present a high degree of visual similarity with the target word (Roeltgen, 1983; Shallice, 1981).

Roeltgen (1985) relates the phonological agraphia with lesions in the supramarginal gyrus and adjacent insular region. Alexander et al. (1992), based on their own cases and cases previously reported in the literature, concluded that phonological agraphia might appear as a result of injuries in an extensive perisylvian region, which is involved in phonological processing.

Lexical (surface) agraphia

In this this type of agraphia, a dysfunction of the lexical system is assumed. The disorder is characterized by difficulties in spelling irregular and ambiguous words with a preserved ability to spell regular words. The patient's ability to write decreases as the orthographic ambiguity of the target word increases. As in surface alexia, the word frequency plays a decisive role. In may correspond to dysorthography in Spanish.

It has been proposed that at least in some languages, such as English, there are two possible systems for the spelling of words: lexical and phonological (Beauvois & Dérouesné, 1981; Hartfield & Patterson, 1983; Roeltgen, 1985; Roeltgen et al., 1983;

Shallice, 1981). The lexical system is necessary for the spelling of irregular words (for example, "knight"), and ambiguous words (words with sounds that can be represented by different letters or combinations of letters), requiring the use of the visual image of the word (Roeltgen, 1985). However, the lexical system can also be used to spell orthographically regular words, which could also be written using the phonographemic system. This disorder has been termed lexical (or surface) agraphia.

The lexical agraphia patient cannot spell irregular words, but is able to spell words and legitimate pseudowords. The ability to write decreases when spelling ambiguity is increased. As in the surface alexia, the frequency of the word plays a decisive role; suggesting that the orthographic vulnerability of the units depends on the subject's previous reading history (Bub, Cancelliere & Kertesz, 1985).

Generally, these patients tend to present a "regularization" in writing: the words are written in a way that seems phonologically correct, although their spelling is incorrect. These errors would result in overuse of the phonographemic system, associated with a decrease in the ability to use the visual form of words. In languages with high heterophonic homography (e.g., French), it is expected that this defect is important.

It has been proposed that this particular defect in writing appears with lesions in the angular gyrus and parietal-occipital lobe damage (Roeltgen, 1993). Yet Rapcsak, Arthur, and Rubens (1988) reported a case of lexical agraphia with a focal lesion in the left precentral gyrus. Interestingly, in Spanish, spelling defects are associated not only with Wernicke's aphasia, but also with right hemisphere pathology (Ardila, Rosselli & Ostrosky, 1996). It has been proposed that the use of Spanish spelling is significantly associated with the ability to visualize the written form of words.

Deep agraphia

Deep agraphia refers to a writing disorder characterized by: (a) the inability to spell nonwords and function words; (b) better spelling of high imageability nouns than low imageability nouns; (c) semantic paragraphias. It is also associated with phonological agraphia, and consequently these patients present lesions at the level of the supramarginal gyrus and the insula, but their lesions are notoriously more extended.

Peripheral agraphias (dysgraphias)

The peripheral agraphias affect one production mode of writing. Generally, good oral spelling associated with writing difficulties is reported (Baxner & Warrington, 1986; Papagno, 1992).

Spatial (afferent) agraphia

The spatial agraphia usually associated with right hemispheric lesions, has been relatively well analyzed in the literature (see above) (Ardila & Rosselli, 1993; Ellis, Young & Flude, 1987; Hecaen, Angelergues & Douziens, 1963;). Lebrun (1976) proposed to call this form of agraphia, "afferent agraphia".

Apraxic agraphia

Writing is correct from the point of view of the spelling, but the letters can be seriously distorted (Baxter & Warrington, 1986; Papagno, 1992; Roeltgen & Heilman, 1983). Generally the copy is almost normal. This type of agraphia was described above.

Summary

Brain pathology is frequently associated with disturbance in writing ability (agraphia). A major distinction is usually established between aphasic (or linguistic) and non-aphasic (or non-linguistic) agraphias. Aphasic agraphias are the manifestation of a fundamental linguistic defect in writing and parallelize the aphasic (spoken language) disturbance. Non-aphasic agraphias include motor, apraxic, and spatial agraphia; sometimes a kind of "pure" agraphia is also recognized. During the 1970s and 1980s, a new approach to the analysis of agraphia was developed. In this approach, a major distinction was established between central agraphias affecting spelling in all the ways: handwriting, typing, oral spelling, etc. (phonological, lexical –surface- and deep agraphia); and peripheral agraphias affecting only one spelling modality (spatial and apractic agraphia).

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III. SPECIAL PROBLEMS IN APHASIA

Chapter 8

Associated disorders

Introduction

Patients with aphasia may present a series of associated disorders, such a hemiparesis, sensory defects, apraxia, agnosia, and acalculia. Depending upon the lesion location and extension, these disorders can be mild, moderate, or severe; or simply absent.

The following groups of disorders will be reviewed: (1) Disorders of awareness; (2) Motor disorders; (3) Sensory disorders; and finally, (4) Disorders of cognitive function.

Disorders of awareness

Patients with aphasia, particularly in cases of some etiologies, such as traumatic aphasia, can present awareness disturbances. These awareness disturbances include:

Confusional states

A confusional state refers to the inability to maintain a coherent line of thought. It is associated with aphasia etiology (most frequently in traumatic aphasia) and aphasia type (it is most frequent in extrasylvian or transcortical motor aphasia involving the frontal lobe system). At the onset of aphasia, confusion is frequently found; it improves in most patients, but a residual degree of attention abnormality is often present.

Inattention

Although contralateral inattention (**hemi-inattention syndrome**) is most frequently found in cases of right hemisphere pathology, some right unilateral inattention may also be found in cases of left hemisphere damage associated with aphasia, particularly if the frontal eye field (Brodmann area 8) is involved. In these cases, the patient has difficulties in visually exploring the contralateral visual field, and some right visual field neglect can be found.

Motor neglect

Motor neglect refers to the underutilization of one side of the body, contralateral to the brain pathology, without defects in strength, reflexes or sensibility. As with inattention, it is notoriously more frequent in cases of right hemisphere pathology (left motor neglect), but some motor neglect can be found in cases of left frontal damage.

Motor disorders

Motor disorders in aphasia include hemiparesis, dysarthria, extraocular motor palsies, pseudobulbar palsy, and apraxia.

Hemiparesis

Broca's aphasia is usually associated with a motor defect in the right hemibody. Severity is variable, depending upon the extension of the lesion. Sometimes it is minimal, sometimes is very severe, even representing a hemiplegia.

The hemiparesis specially affects the hand and the face, and is milder in the leg. Furthermore, the hemiparesis is more distal (hand) than proximal (shoulder). Because it is a lesion at the level of the upper motor neuron, it is a spastic hemiparesis characterized by an increase in the muscle tone.

The hemiparesis may affect the articulatory organs (lips, tongue, etc), and in such a case it is manifested as a dysarthria, usually a **spastic dysarthria**.

In conduction aphasia, it is frequent to find some initial hemiparesis, but it tends to disappear or at least decrease in severity. In cases of aphasia of the supplementary motor area, a right leg hemiparesis is found, associated with gait difficulties.

Patients with Wernicke's aphasia usually do not present motor defects, but occasionally a mild motor defect is observed at the onset of the aphasia.

Patients with extrasylvian (transcortical) motor aphasia (dysexecutive aphasia) usually do not present hemiparesis, even though they may present deficits in controlling eye movements and defects in visual scanning.

Dysarthria

Dysarthria is a neurologic motor speech impairment that is characterized by slow, weak, imprecise, and/or uncoordinated movements of the speech musculature. It represents an impaired execution of movements of speech production. There are five different types of motor impairments, and in consequence, five different types of dysarthria can be distinguished (Table 8.1).

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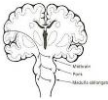
Types of motor impairments	Types f dysarthria
Pyramidal system	
Upper motor neuron: spasticity	Spastic dysarthria
Lower motor neuron: flaccidity	Flaccid dysarthria
Basal Ganglia	
Hypokinesia	Hypokinetic dysarthria
Hyperkinesia	Hyperkinetic dysarthria
Cerebellum	
Ataxia	Ataxic dysarthria

_____

Table 8.1. Types of motor disturbances and associated speech impairments (dysarthria)

The dysarthria observed in aphasia, specifically in Broca's aphasia, corresponds to a spastic dysarthria (upper motor neuron damage), and is characterized by imprecise consonants, monopitch, reduced stress, harsh voice quality, monoloudness, low pitch, and a slow speech rate.

Extraocular motor palsies

Brain damage involving the frontal eye field (Brodmann´s area 8) (Figure 8.1) will result in impairment of eye movements, particularly by verbal command. It is not unusual to find extraocular motor disturbances in extrasylvian (transcortical) motor aphasia (dysexecutive aphasia).
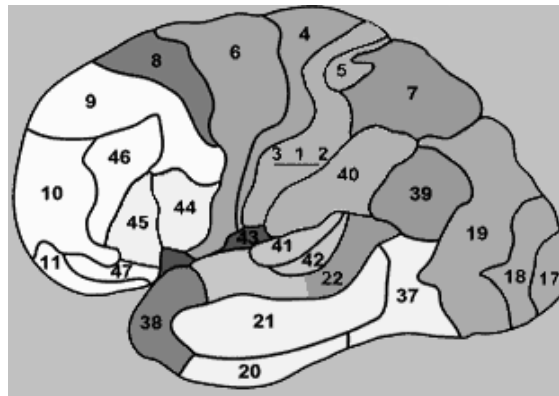
Figure 8.1. The frontal eye field corresponds to BA8. Primary somatosensory area corresponds to the postcentral gyrus (BA3, 1 and 2)

Apraxia

Apraxia is a disorder in motor activity in which the person is unable to perform tasks or movements when asked, even though the patient correctly understands the command and makes attempts to perform the requested movement, the muscles needed to perform the task work properly, and the task may be a well-learned task.

Apraxia is frequently associated with aphasia. It is assumed that about 40% of the aphasia patients present an ideomotor apraxia. Furthermore, some types of aphasia, in particular conduction aphasia and partially Broca´s aphasia, have been interpreted as segmentary apraxias.

The type of aphasia most frequently associated with ideomotor apraxia is conduction aphasia, and even this aphasia has sometimes been regarded as a segmentary ideomotor apraxia involving the articulatory movements (**verbal apraxia**). In Wernicke's aphasia, ideomotor apraxia suggests an extension of the damage to the parietal lobe.

Callosal lesions produce apraxia of the left hand, because the right hemisphere is incapable of organizing the plan of movement independently. In cases of left-

hemisphere pathology, a bilateral apraxia may be observed. When the left-hemisphere lesion also destroys the primary motor zone (usually seen in Broca's aphasia), the right arm is paralyzed and the apraxia is masked. The observable apraxia on the left side is referred to as **sympathetic apraxia**: the patient presents two different motor defects: hemiparesis (at the right) and ideomotor apraxia (at the left)**.** This is seen in many individuals with right hemiplegia and Broca's aphasia.

Apraxia of speech

Apraxia of speech represents a disorder due to an impairment in planning and programming the sequences of movements required for speech production. It is considered –with agrammatism– one of the elements underlying Broca aphasia.

Apraxia of speech is characterized by abnormalities in phoneme production (phonetic deviations), omissions, and substitutions of speech sounds. Speech rate is decreased. Speech is not fluent and produced with effort. Errors are inconsistent. Automatic language is notoriously better produced than language repetition. It can be associated with oral (buccofacial) apraxia (ideomotor apraxia resulting in difficulty carrying out movements of the face and the mouth on demand) and/or dysarthria.

Verbal apraxia

This term has been used in different ways, and hence, has become somehow confusing. In general two types of verbal apraxia have been distinguished: pre-Rolandic and retro-Rolandic. The first one (frontal kinetic apraxia) would correspond to the "apraxia of speech" (one of the two distinguishing elements of Broca's aphasia).

Post-Rolandic type of verbal apraxia is associated with conduction aphasia. As mentioned before, it has even been suggested that verbal apraxia and conduction aphasia are the same disorder. That is, according to some authors, conduction aphasia could be regarded as a verbal apraxia.

Sensory disorders

Two major types of sensory disorders are frequently associated with aphasia: somatosensory defects and visual field defects.

Somatosensory defects

The somatosensory system is the part of the sensory system concerned with the perception of touch, pressure, pain, temperature, position, movement, and vibration, which arise from the muscles, joints, and skin. Somatosensory information is projected contralaterally to the primary somatosensory cortex (Brodmann's areas 3, 1 and 2), which is located in the postcentral gyrus (Figure 8.1). Somatosensory defects are more evident in conduction aphasia, but they can also be found in a lesser degree in Broca and Wernicke aphasia, depending upon the extension of the lesion.

In conduction aphasia, it is not unusual to find hypoesthesia, difficulties in two-point discrimination and tactile extinction.

Visual field defects

Right homonymous hemianopsia (Figure 8.2) is almost invariable in alexia without agraphia. In those cases, the pathology involves the primary visual area (Brodmann's area 17). However, in aphasia, visual field defects are not common. In Wernicke aphasia, occasionally, a right superior quadranopsia is found. By the same token, in conduction aphasia, an inferior quadranopsia can be found if damage extends deep into the brain.
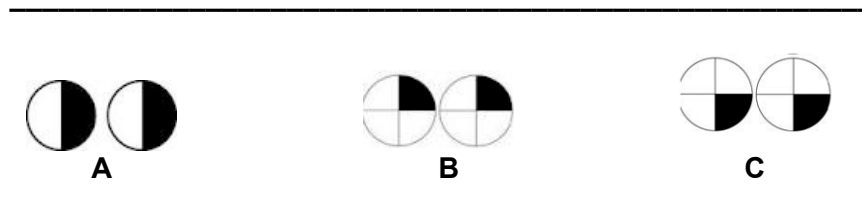
Figure 8.2. Different types of visual field defects potentially associated with aphasia. A) Right homonymous hemianopsia, found in cases of occipital damage, and almost invariably associated with pure alexia. B) Right superior quadranopsia occasionally observed in Wernicke's aphasia when the lesion extends deep into the brain partially damaging the optic radiation. C) Right inferior quadranopsia potentially found in conduction aphasia due to the extension of the pathology to the optic radiation.

Disorders of cognitive function

As a matter of fact, aphasia is a defect in a particular type of cognition (language). But additionally, patients with aphasia frequently present other disorders in cognition. The additional cognitive syndromes frequently found in aphasia include amnesia, agnosia, acalculia, Gerstmann's syndrome, and dementia.

Amnesia

Due to their linguistic limitations, in general, patients with aphasia present difficulties with memorizing verbal information. Performance in different memory tests, such as memorizing words and sentences, is decreased with aphasia. However, depending upon the specific type of aphasia, the memory defect can be more evident at a specific language level (e.g., memory for words, memory for sentences, etc).

A particular sub-type of Wernicke aphasia (Luria's acoustic-amnesic aphasia) has been interpreted as a specific verbal amnesia (both anterograde and retrograde amnesia). In those cases the patient cannot recall the previously learned verbal information, such as words, sentences, and in general verbal knowledge (i.e., there is a verbal retrograde amnesia); and the patient also has significant difficulties with memorizing new verbal information (i.e., there is a verbal anterograde amnesia); for instance, repetition of sentences is limited to maybe 3-4 word long sentences. As mentioned in Chapter 5 ("Major Aphasic Syndromes"), in order to understand conversational language, it is necessary to be able to keep 7-8 words in working (operative) memory, and hence, to be able to repeat 7-8 word long sentences.

If damage extends to other areas (e.g., prefrontal areas, hippocampus, etc.) involved in the brain memory system, a broader amnesia can be found (Figure 8.3).
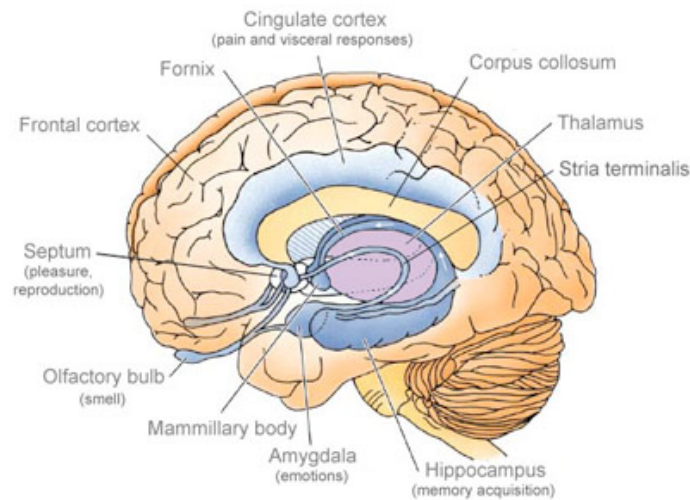
Figure 8.3. Brain memory system includes the hippocampus, the amygdala, the fornix, the mammillary bodies, the dorsomedial nucleus of the thalamus, and the prefrontal cortex.

Agnosia

Agnosia refers to a perceptual disturbance, regardless of a preserved sensation. It is due to pathology in the sensory association areas. In particular, reference to visual agnosia, auditory agnosia, and somatic agnosia will be made in this section.

Visual agnosia

Visual agnosia implies the inability to recognize (perceive) and/or understand visual stimuli. It is observed in cases of damage to the visual association areas (extrastriate cortical areas corresponding to Brodmann areas 18 and 19). Visual agnosia can be associated with aphasia, particularly in case of damage to the posterior portion of the temporal lobe (Brodmann area 37). In this specific condition, the patient presents significant naming difficulties and an important amount of semantic paraphasias; a mild or moderate agnosic disorder is frequent. Anomia in these patients is partially due to some difficulties in revisualization (representation for oneself) of the objects.

Visual agnosia can also be associated with alexia. Alexia without agraphia (also called agnosic alexia) is always associated with visual agnosia. Alexia without agraphia can even be interpreted as a particular type of visual agnosia. It can be also associated with central achromatopsia (inability to see the colors due to a cortical damage; also known as cortical achromatopsia) or achromatognosia (inability to correctly relate the color with the objects; for instance, the grass is green and the sky is blue).

Auditory agnosia

There is a specific subtype of Wernicke's aphasia that has been interpreted as a verbal auditory agnosia (Luria's acoustic-agnosic aphasia). **Word deafness** refers to a syndrome characterized by severe difficulties in understanding spoken language, with sparing written language understanding, and language production. Patients with this disorder can hear, but they cannot discriminate the language sounds (phonemes). When a clear and overt dissociation between understanding of oral and written language is observed, frequently the naming "pure word deafness" is used. Because of

the preserved ability to understand written language, it can be interpreted as an auditory processing defect (verbal auditory agnosia). Word deafness has been regarded either as a subtype of Wernicke's aphasia, or as an independent aphasic disorder, or just as one of the underlying disturbances in Wernicke's aphasia. Some phoneme discrimination defects are usually found in cases of Wernicke's aphasia and left temporal lobe damage, but cases of pure word deafness are extremely unusual. Reported cases have found a left or bilateral superior temporal lobe pathology, including Heschl's gyrus or the auditory projection to this region (Figure 8.4). In Luria's acoustic-agnosic aphasia there are some difficulties in the recognition of language sounds, associated with other aphasic phenomena, such as paraphasias.

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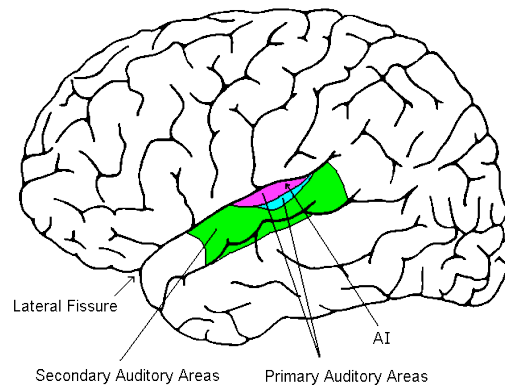
Figure 8.4. Location of the primary auditory area (Heschl's gyrus).

Somatic agnosia

Aphasia can impair the ability to know the body using the language. **Autotopagnosia** is a brain syndrome characterized by the inability to name or locate parts of one's own (or, in some cases, another person's) body. It can be interpreted as a restricted anomia for body-parts, and it is associated with left parietal pathology. It has been proposed that this disorder can be attributed to deficits in "mental images," visual body schema, semantic representations or visual structural descriptions of the human body and its parts.

Finger agnosia and even **right-left disorientation** can also be interpreted as specific types of anomia (anomia for fingers and anomia for the body lateral dimensions). That is, when knowledge of the body is mediated by language, difficulties can be observed in aphasia.

Acalculia

Acalculia has been defined as an acquired disturbance in computational ability. Calculation ability under normal circumstances requires not only the comprehension of numerical concepts, but also that of conceptual abilities, language, and other cognitive skills. Several classifications have been proposed for acalculias. The most traditional classification however, distinguishes between a primary acalculia and secondary acalculias (Table 8.2).

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      PRIMARY   Anarithmetia

      SECONDARY   Aphasic acalculia
                           Alexic acalculia
                           Agraphic acalculia
                           Frontal acalculia
                           Spatial acalculia

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Table 8.2. Classification of acalculias (according to Ardila & Rosselli, 2002).

Primary acalculia

Anarithmetia corresponds to primary acalculia. It represents a basic defect in computational ability. Patients with anarithmetia present a loss of numerical concepts, inability to understand quantities, defects in using syntactic rules in calculation (e.g., "to borrow"), and deficits in understanding numerical signs. However, they may be able to count aloud and to perform some other rote numerical learning (e.g., the multiplication tables). They may conserve some numerical knowledge, but fail in comparing numbers (magnitude estimation) and performing arithmetic operations. In primary acalculia, the calculation defect must be found in both oral and written operations. That is, anarithmetia is a fundamental calculation defect, and is not restricted to a specific type of output (oral or written). Anarithmetia could be interpreted as an acquired defect in understanding how the numerical system works. It is not easy to find cases of pure primary acalculia (anarithmetia) without associated aphasic defects. Usually, it has been assumed that left posterior parietal damage is associated with primary acalculia. Conduction aphasia is the type of aphasia most frequently associated with anarithmetia.

Aphasic acalculia

Calculation difficulties are generally found in aphasic patients, correlated with their linguistic defects. As a result, patients with Wernicke's aphasia exhibit their verbal memory defects in the performance of numerical calculations. Patients with Broca's aphasia have difficulties handling the syntax when applied to calculations. In conduction aphasia, repetition defects may affect successive operations and counting backwards, which, like repetition, require subvocal rehearsal. This means that, ultimately, the calculation defects could very well have originated and been correlated with general linguistic difficulties in aphasic patients.

Alexic and agraphic acalculia

Calculation defects can be correlated with reading difficulties. This is an alexic acalculia or alexia for numbers. The particular manifestation of alexic acalculia is obviously different in alexia with agraphia, alexia without agraphia, frontal alexia, and spatial alexia. Calculation errors can also appear as an inability to write quantities. Specific difficulties will be correlated with the specific type of agraphia.

Frontal acalculia

Patients with prefrontal injuries frequently develop calculation difficulties that are not easily detected. Patients with damage in the prefrontal areas of the brain may display serious difficulties in mental operations, successive operations (particularly backward operations; e.g., 100 – 7), and solving multistep numerical problems. Written arithmetic operations are notoriously easier than mental operations. Difficulties in calculation tasks in these patients correspond to different types: (1) attention difficulties, (2) perseveration, and (3) deficiency of complex mathematical concepts. Attention deficits are reflected in the patient's difficulty in maintaining concentration on the problem. Attention difficulties result in defects in maintaining the conditions of the tasks and impulsiveness in answers. Perseveration is observed in the tendency to continue presenting the very same response to different conditions. It can be found in extrasylvian (transcortical) motor (dysexecutive aphasia).

Gerstmann´s syndrome

Angular gyrus (Gerstmann's) syndrome is classically described as finger agnosia, right-left disorientation, agraphia and acalculia in association to lesions in the left angular gyrus (Figure 8.5). Aphasia is not typically described as part of this syndrome. Ever since its description, the existence of a Gerstmann's syndrome has not been free of debate and questioning in literature. In part, this debate emerges because this syndrome usually unfolds as either an `incomplete' tetrad or in association to other cognitive deficits, particularly, aphasia, alexia, and perceptual disorders. Because of the location of the pathology (left angular gyrus) the possible association with aphasia is very high. Furthermore, it has been pointed out that some specific type of aphasia (so called "semantic aphasia," corresponding to a subtype of Wernicke's aphasia) has frequently been overlooked (Ardila, Concha & Rosselli, 2000).

Agraphia would correspond to an apraxic agraphia (non aphasic), not necessarily associated with alexia. It has been proposed to replace agraphia with semantic aphasia; or simply, to consider semantic aphasia as the fifth characteristic of the Gerstmann´s syndrome (Ardila, López & Solano, 1989). Gerstmann´s syndrome (or angular gyrus syndrome) would include acalculia, finger agnosia (or a more extended form of autotopagnosia), right-left disorientation and semantic aphasia. Sometimes agraphia without alexia is observed, but agraphia would be the consequence of the extension of the lesion toward the inferior parietal lobe.
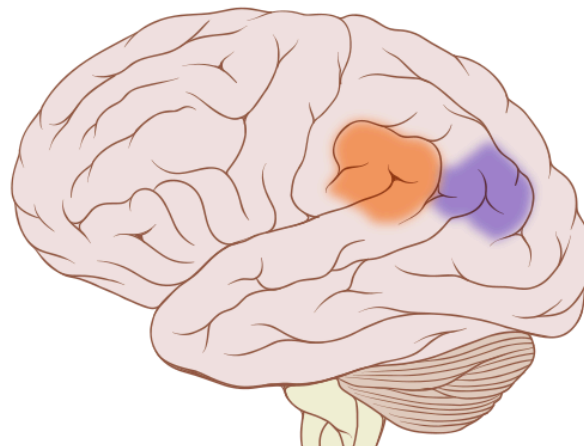
Figure 8.5. Location of the angular gyrus (Brodmann area 39; in purple) and supramarginal gyrus (Brodmann area 40; in orange).

Dementia

Dementia is a progressive decline in cognition due to damage or disease in the brain (degenerative condition, multiple brain infarcts, etc.) beyond what might be expected from normal aging. Particularly affected areas may be: memory, attention, language and executive functions.

Alzheimer disease

The language impairment found in **Alzheimer's disease** has been equated with an anomia (initially), with an extrasylvian sensory aphasia (later), and with a mutism or semi-mutism in the terminal stages (Table 8.3).

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Sequence of language disintegration in Alzheimer's disease

-word-finding difficulties
-semantic paraphasias
-phonological paraphasias
-difficulties in understanding complex sentences
-use of a simple and concrete language
-semi-mutism or mutism

Best preserved language abilities in Alzheimer's disease

-language repetition
-grammar
-mechanics of reading

_____

Table 8.3. Sequence of disintegration of the language and the best preserved language abilities in Alzheimer's disease

One of the earliest stages in dementia language disorders is anomia, usually secondary to a defect in the process, rather than a semantic perceptual abnormality, although there can be both linguistic and perceptual errors. Anomia progresses in parallel with dementia deterioration.

Conversational language comprehension is affected only in advanced stages of dementia. In early stages, difficulty comprehending semantically complex orders, such as those used in the Token test is evident. Poor performance on this test does not seem to be associated with defects in the volume of semantic memory, but with defects in verbal comprehension.

The ability to repeat numbers, words, and phrases of high frequency is generally preserved. Repeating phrases with complex lexical content is, however, altered. Aphasia-like features that develop in Alzheimer's disease have been equated to transcortical sensory aphasia characterized by fluent paraphasic language with defects in understanding, while retaining the ability to repeat.

Alzheimer's disease also affects written language. Most patients conserve the ability to read aloud, except for those very severe deterioration cases. Understanding sentences

and texts decreases as dementia progresses. Defects in writing appear in advanced stages of dementia. This is particularly true in spontaneous writing and dictation.

The generation of words in phonological and semantic categories is usually another linguistic function impaired by Alzheimer's disease. Some authors have noted a major difficulty producing words within semantic and phonological categories among these patients. There are different levels of difficulty when producing words within different semantic categories; the generation of animal names being the easiest task, followed by vegetables and fruits.

Vascular dementia

Vascular dementia is the second most common form of dementia. Vascular disease produces either focal or diffuse effects on the brain and causes cognitive decline. Focal cerebrovascular pathology occur secondary to thrombotic or embolic vascular occlusions. The three most common mechanisms of vascular dementia are: multiple cortical infarcts, a strategic single infarct, and small vessel disease. In multi-infarct dementia, the combined effects of different infarcts produce cognitive decline. Aphasia depends on the location of the infarcts in the brain, and indeed it can be found associated with other cognitive disturbances.

The onset is usually sudden, with staggered cognitive impairment and fluctuating course; there is a slight improvement after each vascular infarct. Antecedents such as a history of hypertension or vascular lesions should be sought out. On neurological and neuropsychological examinations, focal signs can be seen (e.g., hemiparesis, aphasia, etc.).

From a clinical standpoint vascular dementia may be confused with Alzheimer's disease; indeed, it is common to find a combination of the two conditions (degenerative and vascular) in approximately 15% of patients with Alzheimer's disease. The key diagnostic differences are based on the patient's medical history and clinical features of dementia. In Alzheimer's disease, both the onset and deterioration are progressive in nature, while vascular dementia may have a more sudden onset with staggered fluctuating deterioration. Antecedents like vascular disease or hypertension point more towards vascular dementia. Likewise, the presence of focal neurological or neuropsychological signs is also characteristic of the vascular disease.

Language disturbances can also be found in other dementia conditions, such as: subcortical dementia (e.g., Parkinson´s disease), normal pressure hydrocephalus, AIDS dementia complex, etc. Some of these conditions will be briefly reviewed.

Parkinson's disease.

The disease that bears this name is characterized by tremor, rigidity, and bradykinesia. Tremor is primarily resting and undertakes hands, ankles and/or head. The increased muscle tone rigidity occurs predominantly in the flexors. The stiffness gives the patient the characteristic posture of leaning forward with slight flexion of the knees, neck, and shoulders. Motor problems manifest themselves in difficulty initiating movement and a slowdown in the execution thereof. Control of fine motor movements is severely altered, resulting in compromised coordination and absence of mimicry. The lack of facial expression (hypomimia), bradykinesia, and rigidity gives the appearance of weakness and lethargy. Speech loses its intonation and prosody, similar to that of dysarthria (hypokinetic dysarthria). In advanced stages of the disease, patients with Parkinson's disease may develop mental changes, that added to their motor difficulties,

constitute subcortical type dementia (Albert, 1978) with defects of attention, executive functions, and language (e.g., difficulty finding words). Although most of these patients demonstrate severe depressive frames, not all show evident cognitive deterioration. It is estimated that between 19 and 40% of patients with Parkinson's disease develop dementia (Aarsland, Andersen, Larsen, & Kragh-Sorensen Lolk, 2003).

Lewy body dementia

Disease of Lewy bodies usually starts between the ages of 60 and 70 years and is characterized by Parkinsonian symptoms and neuropsychological defects of the fronto-subcortical type, mainly with alterations in attention (McKeith et al., 1996). Histologically, the so-called Lewy bodies are observed in cases of Parkinson's disease, and even in normal individuals without dementia (Wakisaka et al., 2003). The similarity between dementia of Parkinson's disease and dementia with Lewy bodies has led to the suggestion that these two disorders have a common etiology.

Huntington´s disease

Huntington's disease is another degenerative disease of the central nervous system transmitted in an autosomal dominant manner, and characterized by the presence of choreiform movements and cognitive or behavioral changes of a psychotic type. The disease is the result of an abnormal gene on chromosome 4, with a 100% expressivity; it is expected that half of the offspring of an affected individual will develop Huntington's, with equal incidences for both sexes. The age of onset of the disease is variable, but the highest incidence is in the fourth or fifth decade of life.

At the beginning of the disease are mild involuntary shakes of the hands, fingers, shoulders or muscles of the face, that can be hidden by the patient by making them part of a chain of voluntary movements. These involuntary movements become more abrupt, rapid and repetitive, compromising a larger group of muscles. Some patients develop associated cognitive and behavioral changes. Decreased attention span reduced memory, depressive behaviors, apathetic and sometimes paranoia, reduced vocabulary, word-finding failures, and reduced understanding of complex language have been described.

Normal pressure hydrocephalus

The term hydrocephalus refers to the increase in size of the ventricles as a result of either an obstruction in the flow of cerebrospinal fluid, or as compensation for a cortical atrophy. In normal pressure hydrocephalus, an obstruction can be secondary from trauma, infection, or tumor. With the obstruction, the pressure increases on the lateral ventricles, causing them to widen; as the ventricles become enlarged to adapt to the new condition, the pressure of the cerebrospinal fluid returns to normal.

The clinical picture is characterized by gait disturbance, incontinence, and cognitive impairment. The first symptom (apraxic gait) is characterized by small steps, and is described as magnetic gait because the individual cannot lift their feet off the floor, as if they were attached to it. However, the movement can be achieved after examiner's verbal command of "lift your right foot, now left...", etc. Later, sphincter incontinence, which may initially manifest as urinary urgency, is observed. Cognitive and behavioral disturbances are the last symptom to appear. The patient has marked slow mentation and bradykinesia, with alterations in attention, and orientation difficulties, associated with a reduction in expressive language. Language difficulties are due to the effect of

the ventricle enlargement in the frontal lobes; and consequently, the language defect is similar to extrasylvian (transcortical) motor aphasia.

<u>AIDS dementia complex</u>

AIDS dementia complex is characterized by sluggishness in cognition, poor concentration, apathy, and forgetfulness. A patient's defects in language are characterized by verbal learning failures and word finding difficulties. As the disease progresses, there are increases in bradykinesia, ataxia, hypertonia, and motor weakness appear. These cognitive and motor disorders are accompanied by behavioral changes such as apathy, social isolation, and decreased spontaneity, constituting a frank picture of dementia that has been called HIV encephalopathy or acquired dementia complex (ADC).

Sidtis and Price (1990) proposed a severity scale for HIV-associated dementia complex extending from zero (normal) to stage 4 (terminal), passing through stages 0.5 (subclinical), 1 (mild), 2 (moderate) and 3 (severe). In the subclinical stage, neurological soft signs may be seen as abnormal reflexes or motor slowing, without loss of strength or gait defects; cognitive defects are not obvious and the patient can maintain daily activities and work. During stage 1, or mild, neurological and neuropsychological (memory and motor deficits) signs are evident, but the subject can still keep active in the workforce. The inability to normally work marks the beginning of the moderate stage. During the most severe stage different cognitive functions are involved: there are significant memory defects, reduction in expressive language and understanding, marked bradykinesia and abstraction defects. Motor problems are increased to the point that it becomes difficult to walk. Stage 4, the terminal state, would correspond to a near-vegetative state, accompanied by mutism, incontinence, and few responses to the environment.

<u>Other types of Dementia</u>

There are a number of pathological conditions that can lead to diffuse brain involvement and therefore to dementia, associated with language disorders, such as hematoma, secondary to minor trauma, neoplasms, and metabolic and toxic conditions

Surgical intervention in chronic subdural hematoma can completely reverse the dementia. In boxers, dementia derived from the repeated microtraumas these patients receive during fights (dementia pugilistica) has been described.

Neoplasms, particularly in the frontal lobes, can produce a global deterioration of cognitive function. Depending on the characteristics of the tumor, resection may reverse or at least temporally stop dementia. Neoplasms in language areas result in aphasia, whose manifestations are correlated with the growth rate of the tumor: in fast growing tumors there are noticeable defects in language, while slow growing tumors will demonstrate minor defects.

Chronic alcohol abuse can cause cognitive impairment, different from Korsakoff syndrome induced by thiamine deficiency. About half of chronic alcoholics have some degree of neuropsychological impairment. This decline is most evident in subjects over 65 years. Perseveration, attention deficits, bradykinesia, disorientation, memory defects and abstraction are the main features of this dementia (Ropper & Samuels, 2009). The deterioration is slowly progressive. Alcoholic dementia is at least partially reversible if the subject remains abstinent; improvement in neuropsychological functioning is seen, but there is no complete reversal of the deficit (Cummings &

Benson, 1992). The language in these patients is described as a concrete language associated with a reduction of active vocabulary.

Psychiatric disorders are sometimes associated with dementia. Such is the case of patients with chronic schizophrenia, whose intellectual capacity, memory abilities, and visual perceptual behavioral changes fit the definition of dementia. In depressed patients it is manifested in the form of a pseudodementia, because patients appear cognitively impaired as a result of the intense depression. Schizophrenic patients may develop defects in language that will eventually resemble in Wernicke's aphasia.

Summary

Aphasia is frequently associated with diverse brain disorders, such as: hemiparesis, sensory defects, apraxia, agnosia, and acalculia. The severity of these associated disorders is variable.

Disorders of awareness are frequently found with a lesion extending to the frontal lobes; confusional states, hemi-inattention and motor neglect can be found. Right hemiparesis and spastic dysarthria are usually associated with Broca's aphasia. Apraxia can also be found in cases of frontal (kinetic apraxia) and parietal (ideomotor apraxia) damage. Sensory disorders, including somatosensory disturbances and visual field defect can be found in retro-Rolandic lesions; somatosensory disturbances are associated with conduction aphasia; visual field defects are sporadically found in cases of aphasia due to lesions involving the visual radiation. Disorders in cognitive function include amnesia, agnosia, acalculia, Gerstmann's syndrome, and dementia.

Recommended readings

Papathanasiou, I., Coppens, P. & Potagas, C. (2012). Aphasia and Related Neurogenic Communication Disorders. Jones & Bartlett Learning.

Ropper, A.H. & Samuels, M.A. (2009). Adams & Victor's Principles of Neurology. New York: The McGraw-Hill Companies, Inc., 9th Ed.

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Chapter 9

Aphasia in special populations

Introduction

Aphasia most often has been analyzed in monolingual, right-handed, literate adults, speakers of some few Indo-European languages (mainly English, French, German, Russian, Italian, and Spanish). Aphasia, however, can appear in special populations, presenting some specific manifestations. In this chapter, a description of aphasia in bilinguals, children, left-handers, illiterates, deaf-signers, and speakers of non-Indo-European languages will be presented.

Aphasia in bilinguals

Worldwide, some 6,800 different languages are spoken (http://www.ethnologue.com), and over half of the world's population is bilingual or multilingual. This means that over half of the cases of aphasia are bilingual aphasias. Bilingualism, however, is quite variable across different countries. Table 9.1 presents the number of languages spoken in some selected countries.

Country	Number of languages spoken
Algeria	18
Argentina	25
Brazil	181
Canada	86
China	292
Colombia	80
Congo	62
Cuba	2
Egypt	12
Germany	27
Haiti	2
Iceland	2
Israel	33
Mexico	291
Papua New Guinea	830
Russia	100
Spain	14
United Kingdom	12
United States	176

Table 9.1 Number of languages spoken in some selected countries (taken from http://www.ethnologue.com/).

Types of bilingualism

It is difficult to establish a clear criterion for bilingualism. According to Grosjean (1994), a bilingual is a person who uses two or more languages or dialects in his/her everyday life. A bilingual individual is not necessarily a balanced ambilingual (an individual with native competency in two languages), but a bilingual of a specific type who, along with other bilinguals of many other different types, can be classified along a continuum. Some bilinguals possess very high levels of proficiency in both oral and spoken language. Others display varying degrees of proficiency in understanding and/or speaking skills, or reading/writing skills, depending on the immediate area of experience in which they are required to use their two languages.

Bilingualism is, in consequence, a very heterogeneous phenomenon and it is difficult to even find two identical bilinguals. Bilingualism varies according to different variables, such age of acquisition of the second language, language proficiency, frequency of use of the two languages, similarity between both languages, etc. We shall briefly review the first two variables (age of acquisition and language proficiency), which are usually considered the most important ones.

<u>Age of acquisition</u>

Bilinguals can be distinguished according to the time of acquisition of the second language (e.g., Bialystok & Hakuta, 1999; Birdsong, 1992; Genesee & Nicoladis, 1995; DeKeyser, 2000; Flege, 1999). Some distinctions have been proposed:

Simultaneous bilingualism (sometimes also named as **authentic bilingualism**). Learning two languages as "first languages" (two native languages). Infants who are exposed to two languages from birth will become simultaneous bilinguals. If exposure to the second language occurs after age 3-5 years, the term **sequential bilingual** is used.

Early bilingualism. The second language (L2) is acquired before completing the acquisition of the first one (L1).

Late bilingualism. L2 is acquired after completing the acquisition of L1. L2 is learned mediated by L1. Sometimes the term **consecutive** or **successive bilingualism** is used to refer to this learning of one language after already knowing another.

<u>Language proficiency</u>

A frequently used distinction in bilingualism refers to the mastery of both languages (Weinreich, 1953**)**. Three situations can be distinguished:

Coordinate bilingualism: the linguistic elements (words, phrases) are all related to their own unique concepts. A coordinate bilingual acquires the two languages in different contexts (e.g., home and school), so the words of the two languages belong to separate and independent systems. This means that an English-Spanish bilingual speaker of this type possesses different associations for 'table' and for '*mesa'* ("table" in Spanish). There are, in consequence, two lexical and two semantic systems.

Compound bilingualism: speakers of this type attach their linguistic elements (words, phrases) to the same concepts. For them, 'table' and '*mesa*' are two words for the

same concept. The person acquires one notion with two verbal expressions. There are, in consequence, two lexical systems, but only one semantic system.

Subordinate bilingualism: the linguistic elements of one of the speaker's languages are only available through elements of the speaker's other language. This type is typical of, but not restricted to, beginning L2-learners. "*Mesa*" means "table" and table has certain semantics. There is one semantic system, and lexicon in the second language is accessed using the first language lexicon.

It is important to note that a bilingual can simultaneously be classified in more than one category. When learning a second language, mastery progressively increases. Figure 9.1. illustrates these three types of bilingualism.

Figure 9.1. Coordinate bilingualism (two lexicons, two meanings), compound bilingualism (two lexicons, one meaning), and subordinate bilingualism (meaning in the second language is achieved though the first language).

Patterns of aphasia

Different clinical observations have demonstrated that bilingual aphasics do not necessarily manifest the same language disorders with the same degree of severity in both languages (Albert & Obler, 1978). Aphasia can be **parallel** (both languages are impaired in a similar way) or **dissociated** (there is a different aphasia profile for each one of the languages). Fabbro (2001) observed, in a sample of 20 bilingual aphasics, parallel aphasia in 65% of the subjects; in the rest (35%) aphasia was dissociated: 20% showed a greater impairment of L2, while 15% of the patients showed a greater impairment of L1. These percentages can be considered relatively typical.

It is assumed that parallel aphasia is usually found in early bilinguals, whereas dissociated aphasia is characteristic of late bilinguals. As a matter of fact, language representation of both languages can be regarded as coincidental in early bilinguals, whereas language representation of L1 and L2 is not completely coincidental in late bilinguals. On one hand, L2 seems to be acquired through the same neural structures responsible for L1 acquisition; however, neural differences may be observed, in terms of more extended activity of the neural system mediating L2 processing (Abutalebi, 2008). Indeed many studies have reported that later acquired languages may involve broader activation locations than the first acquired language; largely overlapping, but sometimes distinct cortical areas are involved in the comprehension and production of first and second languages (Obler et al., 2007).

For example, Kim et al. (1997) used an fMRI study, in which participants used widely different L1 and L2 pairs of languages (English-French, Korean-English, Spanish-

English). While silently describing to themselves what they did during the morning, afternoon or evening of the previous day, it was found that within the frontal-lobe language area (Broca's area), second languages acquired in adulthood (late bilinguals) are spatially separated from native languages. However, when acquired during the early language acquisition stage of development (early bilinguals), L1 and L2 tend to be represented in common frontal cortical areas. In both late and early bilingual subjects, the temporal-lobe language-sensitive regions (Wernicke's area) also showed effectively little or no separation of activity based on the age of language acquisition.

In cases of dissociated aphasia, usually the most impaired language is L2, but sometimes, it can be L1. For instance, Ardila (2008) reported the case of a 63 year-old right-handed female native Spanish speaker, who had been living in the U.S.A. for 38 years. She never studied English in a formal way, but after years of having been exposed to it, she had learned some English. Suddenly, she presented an extensive left temporal intracerebral hemorrhage. A significant language understanding defect was found, associated with severe impairments in verbal memory (Wernicke's aphasia), difficulties in language repetition, severe anomia with phonological and semantic paraphasias and neologisms, alexia, and aphasic agraphia. The naming defect was more severe in Spanish than in English; furthermore, there was also a clear tendency to answer in English, to switch to English, and mixing English and Spanish. The patient presented a dissociated aphasia with a better conservation of L2 (English) than L1 (Spanish).

Occasionally, it has been reported that bilinguals can present a different pattern of aphasia in L1 and L2. For example, Silverberg and Harold (1979) reported two cases of dissociated aphasia; following a left parietotemporal lesion, moderate nonfluent aphasia was found in the native language of the first patient, in contrast to less severe, fluent aphasia in the patient's L2. Conversely, mild anomia was found in L1 of a second patient, while global aphasia was found in L2. His lesion was located in the left posterior frontal area.

Patterns of recovery

Two opposite points of view were proposed during the XIX century to explain the language recovery in bilingual aphasics.

Ribot's law or **Ribot's rule** (1883) states that the language best recovered by polyglot aphasics is the mother language.

Pitres' law or **Pitres' rule** (1895). He described seven cases of bilingual aphasics presenting differential recovery of the two languages. Pitres suggested that patients tended to better recover the language that was *most familiar* to them prior to the aphasia onset, regardless it was not the mother tongue.

Paradis (1977) refers to six different patterns of aphasia recovery in bilinguals.

1. **Differential.** Each language is impaired separately and recovered at the same or different rate

2. **Parallel.** Different languages are similarly impaired and restored at the same rate.

3. **Antagonistic**. Recovery of one language progresses, while the other regresses.

4. **Successive**. One language does not show any recovery until another has been restored.

5. **Selective**. One language is not recovered at all.

6. **Mixed.** Both languages are used in some combinations

However, most patients present the first (differential) or second (parallel) recovery pattern. The other patterns are indeed unusual. Fabbro (1999) reports a parallel recovery in about 40% of the cases; a better recovery of L1 in 32% of the patients, and a better recovery of L2 in about 28% of the cases.

What about therapy?

A major question in bilingual aphasia is selecting the language for therapy. In other words, in what language to provide therapy. Or should therapy be provided in both languages, given that indeed the patient has two languages?

There are some obvious answers to this question: (1) in what specific language does the patient prefer therapy to be provided? The patient will most likely prefer his most emotionally-linked language, the language he/she feels as his/her dominant language; usually, but not necessarily, it will be L1; (2) in what specific language can aphasia therapy be provided, given the existing conditions? For instance, it is unlikely to find a speech-language pathologist able to provide speech therapy in Tibetan to a Tibetan/English bilingual patient in U.S.; (3) which one is the most functional language for the patient? For instance, for a Finnish/German aphasic, living for decades in Germany, with a German-speaking family, it is more functional to speak German than Finnish; and hence, it may be preferred to provide language therapy in German rather than in Finnish.

However, regardless of the language in which therapy is provided, at least some generalization to the untreated language can be anticipated. Unfortunately, it is not well understood what specific variables contribute to the generalization of therapy to the untreated language. For instance, it has been observed that in naming treatment, generalization is observed for **cognates** (a cognate can be defined as "a word in one language which is similar in form and meaning to a word in another language because both languages are related"; Richards & Schmidt, 2002, p. 829; e.g., English *flower*, Spanish *flor*) but not for other types of words (Kohnert, 2004).

Aphasia in Children

Childhood aphasia refers to language impairment due to an acquired brain pathology, occurring during the period of language development. The crucial question is: at what age is language development completed? It can be assumed that basic language acquisition (which means, phonology, basic vocabulary, and basic grammar) is completed around the age of four to six years (Hoff, 2008). It is further supposed that a second language can be acquired with proficiency similar to a native speaker up to the age of about 10-12 years (Bialystok & Hanuka, 1999)

The major question with aphasia in children through history has been: how similar or different is childhood aphasia from the aphasia observed in adults? Traditionally it had been assumed that:

- Usually childhood aphasia is a **nonfluent (expressive) type of aphasia**; that means, after the pathological condition (most frequently a head injury), expressive language decreases (or even disappears), whereas language understanding is just minimally impaired.
- There is an increased frequency of **crossed aphasias** (right hemisphere damage).
- **Language recovery is significantly better** in children than in adults (so-called **Kennard principle**: equivalent brain damage to a child and an adult would lead to less problems and better recovery in a child than in the adult); a virtually complete language recovery in children suffering aphasia has even been suggested in some cases.

This traditional point of view has been challenged during the last decades (e.g., Lauterbach et al., 2010; Narbona & Crespo-Eguílaz, 2008). Some authors, however, continue supporting it.

For instance, Martins (1997) pointed out that the syndrome of childhood aphasia is more similar to adult aphasia than had previously been assumed. The prognosis is less favorable than previously supposed; language sequelae and academic difficulties are observed. Aphasic manifestations in adults are similar to those observed in children with similar brain lesions; furthermore, the same brain areas participate in language recovery. Paquier and Van Dongen (1996) observed that case studies show a great variety of aphasic symptomatologies in childhood aphasia, including auditory comprehension disorders, paraphasias, neologisms, logorrhea, jargon, impaired repetition abilities, and a host of linguistic deficits in reading and writing. Not only the typology of the aphasias, but also the recently established clinic-radiological correlations, appear to resemble those found in adults. Also, recovery from childhood aphasia shows to be less complete than previously thought.

These findings bear consequences for theories on cerebral organization of language in childhood. It appears that already in infancy, the two cerebral hemispheres are unequal substrates for language representation. Therefore, the prognosis and final outcome of childhood aphasia are not uniformly favorable. Marien et al. (2001) analyzed crossed aphasia in children reported in history. Since 1975, five cases of crossed aphasia were reported (out of 180 cases), equivalent to 2.7%. However, three cases are ambiguous and only two are evident. Consequently, the frequency of crossed aphasia in children would be 2/180=1.1%, exactly the same percentage observed in adults

According to recent literature it can be concluded:

- Crossed aphasia does not seem more frequent in children than in adults.
- Plasticity and recovery may be more limited than they was considered some time ago.
- Receptive language seems to be more bilaterally represented. Because of that, expressive impairments are more evident.
- Probably, old reports of childhood aphasia may contain errors about the lesion localizations.

A special type of childhood aphasia is the **Landau-Kleffner syndrome.**

Landau-Kleffner syndrome

Landau-Kleffner syndrome (LKS) (also known as **infantile acquired aphasia, acquired epileptic aphasia** or **aphasia with convulsive disorder**)**,** first described by William Landau and Frank Kleffner in 1957, is a rare, childhood neurological disorder characterized by the sudden or gradual development of aphasia (the inability to understand or express language), and an abnormal electroencephalogram (EEG) (Pearl, Carrazana & Holmes, 2001). Typically, the child presents a normal language, but then loses their language ability for no apparent reason. The cause of LKS is unknown, and all of the children with LKS appear to be perfectly normal until their first seizure or the start of language problems.

Approximately 80 percent of the children with LKS have one or more epileptic seizures that usually occur at night. Behavioral disorders such as hyperactivity, aggressiveness, and depression can also accompany this disorder.

LKS occurs most frequently in normally developing children who are between 3 and 7 years of age. For no apparent reason, these children begin having trouble understanding what is said to them. The auditory agnosia may occur slowly or very quickly. Parents often think that the child is developing a hearing problem or has become suddenly deaf. Hearing tests, however, show normal hearing. Children may also appear to be autistic or developmentally delayed. The inability to understand language eventually affects the child's spoken language, which may progress to a complete loss of the ability to speak (mutism). Children who have learned to read and write before the onset of auditory agnosia can often continue communicating through written language. The loss of language may be preceded by an epileptic seizure that usually occurs at night. The seizures usually stop by the time the child becomes a teenager. All LKS children have abnormal electrical brain activity on both the right and left sides of their brains (Loddenkemper, Wyllie & Hirsch, 2012),

Complete language recovery has been reported; however, language problems usually continue into adulthood. If recovery takes place, it can occur within days or years. Generally, the earlier the disorder begins, the poorer the language recovery (Duran et al., 2009). Most children outgrow the seizures, and electrical brain activity on the EEG usually returns to normal by age 15.

Aphasia in left-handers

It is well known that there is a significant association between language lateralization and handedness. However, pinpointing the exact association has been elusive.

Using the **Wada test** (or sodium amytal test: essentially, sodium amobarbital is introduced into one of the internal carotid arteries. It is injected into one hemisphere at a time; a transient aphasia will be observed when injected to the "linguistic" hemisphere) Rasmussen and Milner (1977) observed in a sample of 262 subjects (right-handed=140; left-handed=122) that in 96% of right-handers, language was lateralized to the left hemisphere, and in only 4% was language lateralized to the right hemisphere. In left-handers the situation was quite different: in 70% of left-handers, the language was lateralized to the left hemisphere, in 15% to the right hemisphere, and in the remaining 15% a bilateral representation was found.

Knecht et al (2000) measured lateralization directly by **functional transcranial Doppler sonography** (neuroimaging tool measuring cerebral perfusion changes due to neural activation) in 326 healthy individuals using a word-generation task. The incidence of right-hemisphere language dominance was found to increase linearly with

the degree of left-handedness, from 4% in strong right-handers (handedness = 100) to 15% in ambidextrous individuals and 27% in strong left-handers (handedness = –100).

Since the XIX century, cases of aphasia in right-handers associated with right-hemisphere damage have been observed. Byron Bramwell (1899) coined the term **crossed aphasia** to indicate a language pathology following a cerebral lesion ipsilateral to the preferred hand. Currently, this term ("crossed aphasia") is mostly used to refer to aphasia in a right-hander after a right hemisphere pathology.

Aphasia in left-handers is, in over 50% of the cases, associated with left hemisphere damage. But it has been suggested that aphasia is also quite frequently associated with right-hemisphere lesions (Basso & Rusconi, 1998); some authors have reported that up to 50% of left-handers with right hemisphere lesions present aphasia, although currently the accepted percentage is notoriously lower.

The aphasia profile in general, is similar between right and left-handers; although it has been suggested that left-handed aphasics are less frequently impaired in comprehension and writing, although they do have reading disorders more frequently than right-handed aphasics (Hecaen & Sauguet, 1971). Comparing the aphasia due to right and left hemisphere pathology in left-handed individuals, just minor differences are found. By the same token, comparing aphasia recovery in right and left-handed individuals, only small and non-significant differences are found (Basso & Rusconi, 1998), regardless of the fact that, in the past, it was accepted that aphasia recovery was significanlty better in left-handers.

Aphasia in illiterates

Two opposing points of view have emerged in the literature regarding the influence of education on brain organization of language in general, and aphasia in particular. Cameron, Currier, and Haerer (1971) reported a lower frequency of aphasias associated with injuries of the left hemisphere among right–handed illiterate patients than among educated ones. The authors concluded that language is more bilaterally represented in the illiterate group. In contrast, Damasio and colleagues (1976) claimed that there is no qualitative or quantitative difference between the aphasias of educated and illiterate patients. The aphasias of schooled literates did not differ from that of illiterates in the prevalence rate, distribution of clinical types, or semiological structure.

Matute's research (1988) supports the Damasio and colleagues (1976) conclusions. She compared three groups of right–handed Mexican adults: brain–damaged illiterates, brain–damaged literates, and normal illiterates. An aphasia test was given to all three groups in the context of a more extensive neuropsychological assessment. No differences between the two brain damaged groups were evident with regard to language measures related to repetition, oral expression, and comprehension. However, when analyzing incidence of aphasia due to unilateral brain lesions, all left hemisphere–damaged illiterates presented aphasia, although the lesion locus for some of them were out of the perisylvian area, whereas none presented aphasia after right hemisphere damage. Thus, the data obtained in this study suggest a less intrahemispheric specialization for language on the left hemisphere in illiterates.

The hypothesis that illiterates might have a different cerebral organization for language and, therefore, deviate from literates in their clinical profile, the severity of their aphasia, and their prognosis, was evident as early as 1867 with Scoresby–Jackson's observation of an illiterate patient. This author presented the case of an illiterate patient with a severe motor aphasia. After the postmortem exam, the author found that

although the lesion was very extended in the left hemisphere, the frontal lobe was attainted only in the posterior part of the third frontal circumvolution (Broca's area). Since it was only the posterior part of the third frontal circumvolution that was attainted, Scoresby–Jackson suggested that a bigger part of this circumvolution would participate in language only with a grater language acquisition (given by reading acquisition). Thus, in illiterate people, it would be only the posterior extreme of the third frontal circumvolution that will be active in language expression, whereas in a literate person, all the circumvolution will be employed.

Critchley (1956) was probably the first author to suggest schooling could influence that hemispheric functional asymmetry. He suggested that low-educated persons tended to have less left hemispheric lateralization of language. This suggestion was supported by Wechsler's (1976) interpretation of the crossed aphasia in his illiterate patient (Castro-Caldas et al., 1987). Fonseca and Castro-Caldas (2002) compared the recovery process of literate and illiterate aphasics. They studied 24 illiterates and compared those with 42 schooled literates matched for age, gender, and type of aphasia. Generally, all scores obtained in subtests of the Aphasia battery were lower in illiterates than they were in literate controls. Patients were tested in the first month of their disease and 6 months later. The global scores of aphasia improved similarly in both groups; however, the correlation between the test scores suggested that the process of recovery was different for each group.

Lecours and colleagues (1987a, 1987b, 1988) studied the relationship between brain damage and schooling with regards to aphasic impairments of language. On the basis of their findings, the authors concluded that: (a) there was a greater right–hemisphere language involvement in illiterates than in the well–educated patients; and (b) left–stroke school–educated patients seemed to be "sicker," as it were, than their illiterate counterparts, that is: (i) The classical symptoms of aphasia (suppression stereotype, jargon aphasia) were more apparent among left stroke schooled literates than among left stroke illiterates; and (ii) auditory comprehension was more frequently impaired among the left stroke literate patients. Lecours and colleagues (1987b) also studied the influence of education on unilateral neglect syndrome. They analyzed a large sample of right–handed unilingual brain–damaged individuals: illiterates (left stroke and right stroke) and schooled literates (left stroke and right stroke). Evidence of a unilateral neglect syndrome was found in both left- and right-brain-damaged schooled literates and illiterates. Their results provided no indication that spatial attention was globally stronger depending on the side of the lesion or the educational level of the patients. Rosselli, Vergara, and Ardila (1985), however, reported a higher frequency of right hemispatial neglect in low–educated patients.

In summary, studies of brain–damaged illiterates, when compared with brain–damaged literates, have indicated that: (a) literacy does not change the dominance of the left hemisphere for language; illiterates as well as literates present aphasia most often after left brain damage, and not after right brain damage; and (b) the right hemisphere appears to have a disproportionate involvement in language in illiterates when compared with literate individuals: left-damaged literates present a larger number of errors in aphasia tests than left-damaged illiterates (Lecours et al., 1988; Matute, 1988), and right–damaged illiterates more frequently present poorer performance in aphasia tests than right-damaged literates (Lecours et al., 1987a, 1987b).

Aphasia in deaf-signers

Aphasia for sign language is of very special interest for understanding the brain organization of language. In general, it has been observed that sign language, as well as phonological language, is represented in the left hemisphere for most people. Thus, in deaf-signers, aphasia is associated with left hemisphere lesions, whereas lesions in the right hemisphere result in visuospatial disturbances (Poizner, Klima & Bellugi, 1981; Vaid, Bellugi & Poizner, 1989), as it is also observed in hearing individuals.

After reviewing the available literature, Corina (1998) concluded that there is ample evidence for left hemisphere mediation of sign language in deaf individuals. Furthermore, aphasia for sign language, observed in cases of left hemisphere damage, is not attributable to more general problems in motor or symbolic processing. Impairments in using sign language are separable from apraxic defects. Functional neuroimaging studies, however, show a more extended bilateral brain activity in deaf signers than in normally hearing individuals during speaking, suggesting the participation of more spatially and visually-based abilities in signing (e.g., Hickok, Bellugi & Klima, 1996). Saito, Otsuki, and Ueno (2007) reported a deaf signer who showed substantial sign language aphasia with severe impairment in word production due to a left occipital lesion, suggesting that sign language uses a visuospatial modality through visual information.

Falchook et al. (2012) reported a 55-year-old right-handed congenitally deaf woman with a 2-year history of progressive memory loss and a deterioration of her ability to communicate using Sign Language. Episodic memory impairments as well as defects in the production and comprehension of fingerspelling and grammatically complex sentences were found. She also presented signs of anomia as well as an ideomotor apraxia and visual-spatial dysfunction. The authors suggest that, in many respects, the cognitive disorders in this patient mirror those of patients with dementia who had normally learned to speak.

Aphasia across different languages

Some attempts to compare aphasia across different languages are currently available. For instance, Menn and Obler (1990) published a collection of papers about agrammatism in aphasia across different languages. Paradis (2001) compared the aphasia manifestation in some selected languages; as anticipated, similarities were found, but so were differences. Bates and Wulfeck worked for years comparing aphasia across languages, especially with regard to grammatical impairments (e.g., Bates, Wulfeck & MacWhinney, 1991). However, the number of languages included in these cross-linguistic comparisons were limited; furthermore, they were mostly western Indo-European languages.

Aphasia has been poorly studied in non-Indo-European languages. For instance, there is not a single published case of aphasia in an Amerindian language, regardless that Amerindian languages represent close to 50% of the languages spoken worldwide.

There has been special interest in understanding brain organization of language through the analysis of **tonal languages** (**tone** is the use of pitch in language to distinguish lexical or grammatical meaning; Figure 9.2). This interest is directly related with the research program developed by J. Gandour (e.g., Gandour et al., 1993, 1998, 2000) particularly in Thai (official language of Thailand) but also in other tonal languages. Many African, East Asia, and Amerindian languages are tonal languages. Chinese (Mandarin) represents the largest currently spoken tonal language (about 845 million of speakers). It has been observed that left-damaged non-fluent aphasic

speakers of Chinese experience a tonal production deficit (Packard, 1986). It has been suggested that in tonal languages, lexical specification of tone contour information results in left hemisphere lateralization of that information, thus making the tonal phonemes vulnerable to left hemisphere damage. The hemispheric processing of Mandarin tones reveals that, for native speakers, it is lateralized in the left hemisphere, suggesting that tones are processed as linguistic units, just like the segmental properties (Hsieh, Gandour, Wong, & Hutchins, 2001).
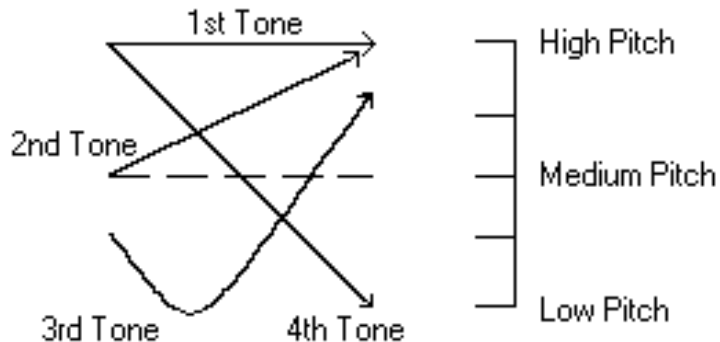

Figure 9.2. Chinese language has four different tones. These four tones are used to communicate different meanings. Many words have the same sounds, but different tones, and different meaning.

Van Lancker (1988) suggested a **scale of hemispheric specialization**, ranging from "most linguistically structured pitch contrasts" (such as Chinese tones and Norwegian word accents) related to left hemisphere specialization, to "least linguistically structured pitch contrasts" (such as emotional prosody and personal voice quality) depending on right hemisphere specialization. Gandour (1998) concludes that "language representation in the brains of tone language speakers is essentially the same as that in non-tone language speakers" (p. 135). Tones obviously are linguistically structured contrasts.

Summary

In this chapter the aphasia manifestations of some "special" populations (bilinguals, children, left-handers, illiterates, deaf-signers, and speakers of non-Indo-European languages) was reviewed. From this review it can be concluded that:

1. Bilingualism is quite heterogeneous, and it has been established that bilingual aphasics do not necessarily manifest the same language disorders with the same degree of severity in both languages. Different variables affect the clinical manifestations and the pattern of recovery of aphasia; the two most seemingly important are age of acquisition proficiency in the second language.

2. Traditionally, it has been assumed that childhood aphasia: (a) is usually an expressive type of aphasia; (b) has an increased frequency of crossed aphasias (right hemisphere damage); and (c) results in significantly better (even complete) language

recovery in children. During the last decades, these traditional points of views have been challenged, and currently it is accepted that aphasia in children is more similar to adult aphasia than previously was supposed.

3. Comparing aphasia in right and left-handers, it can be concluded that aphasia profile in general, is similar between right and left-handers.

4. Studies of brain–damaged illiterates have demonstrated that: (a) literacy does not change the dominance of the left hemisphere for language, illiterates as well as literates present aphasia most often after left brain damage; and (b) the right hemisphere appears to have a disproportionate involvement in language in illiterates when compared with literate individuals.

5. In deaf-signers, aphasia is associated with left hemisphere lesions, whereas lesions in the right hemisphere result in visuospatial disturbances, as is also observed in hearing individuals.

6. Although aphasia has been poorly studied in non-Indo-European languages, it has been found that the hemispheric processing of tones in tonal languages is lateralized to the left hemisphere.

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IV ASSESSMENT AND REHABILITATION

Chapter 10

Assessment of aphasia

Introduction

Usually aphasia patients are individuals who suffered a stroke or traumatic head injury; sometimes they are patients with brain tumors or dementia. Most often, language evaluation is carried out either at the institutional level (usually a hospital), or at the professional's office. When the patient is assessed at the hospital, frequently a shorter (bedside) evaluation is required. An extensive language evaluation may take one hour or even more, but of course, it depends upon the specific language impairment (for example, the evaluation of a global aphasia is usually very short) and the testing procedure that is selected (for instance, a short language test battery; a diversity of different language tests, etc.).

The patient's evaluation has different goals. Five fundamental purposes can be distinguished in aphasia assessment:

1. **To determine if patient's language is normal or abnormal, usually --but not necessarily-- after pathological brain conditions.** This is indeed the basic aim of any clinical exam, to determine whether or not there is an abnormal condition.

2. **To analyze the symptoms** (i.e., what is reported by the patient, for instance, "*I forget words")* **and signs** (i.e., what is found during the exam, for instance, paraphasias) **in order to identify fundamental syndromes.** "Aphasia" is a clinical syndrome; "dementia" is also a syndrome; "dysarthria" can be regarded as a clinical syndrome, or as a sign of a more extended "motor syndrome". "Wernicke's aphasia" is a subtype of aphasia; "subcortical aphasia" refers to the topography (location) of the pathological process responsible for the aphasia (topographical diagnosis). "Traumatic aphasia" points to the etiology (cause) of the aphasia (etiological diagnosis).

3. **To propose rehabilitation procedures.** Rehabilitation strategies in aphasia depend not only on the specific aphasia subtype (e.g., Broca's aphasia, Wernicke's aphasia, etc.), but also on other conditions, such as the etiology of aphasia, the patient's age, etc.

4. **To establish a differential diagnosis between apparently similar conditions** (e.g., aphasia and dysarthria). However, a patient can simultaneously present different conditions, and as a matter of fact, Broca's aphasia is usually associated with dysarthria. Aphasic patients can   present a diversity of associated clinical conditions (for example, amnesia, apraxia, executive function disturbances, etc.) depending upon the specific location and extension of the pathological process.

5. **To propose pathologies potentially responsible for the language impairment.** Depending upon the specific aphasia characteristics (for instance, language fluency, progression of the impairment, etc.), the underlying pathological process can be different. As an example, a sudden severe impairment in language understanding may suggest a stroke at the level of the left temporal lobe.

Language domains

In aphasia assessment, usually six different language domains are included: expressive language, language understanding, repetition, naming, reading, and writing.

Expressive language

An initial step in language evaluation is to obtain a patient's language sample. For this purpose, different strategies can be used; such as asking the patient to describe a picture (for instance, plate #1 from the Boston Diagnostic Aphasia Examination; Goodglass et al., 2000; Figure 10.1), or asking the patient to tell the history of his/her condition.

Different speech/language characteristics are observed:

- fluency
- articulation
- prosody and volume
- phonology
- word-selection (lexicon)
- grammar
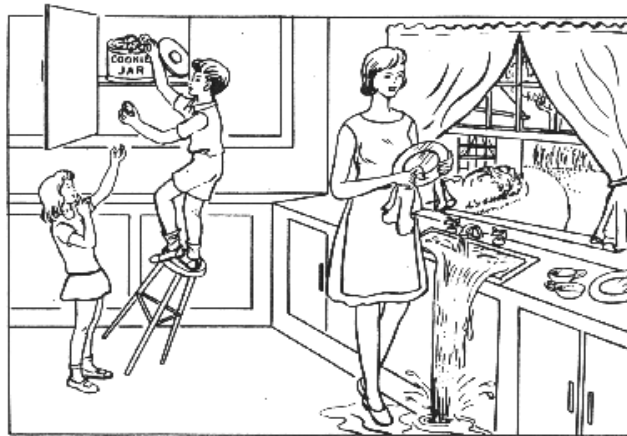
Figure 1. Plate #1 ("The cookie theft") from the Boston Diagnostic Aphasia Examination (Goodglass et al., 2000).

Fluency
Fluency in aphasia includes several characteristics related to the flow of speech, such as number of words per minute (wpm), smoothness (effort), etc. Table 10.1 presents the major characteristic of non-fluent and fluent speech in aphasia. Motor aphasias (Broca's aphasia) are considered as non-fluent aphasias, whereas sensory aphasias (Wernicke's aphasia) are regarded as fluent aphasias.

Non-fluent speech	Fluent speech
sparce (less 50 wpm)	normal (100-200 wpm)
increased effort	normal
dysarthric	normal
short sentences	normal (5-8 wps)
dysprosodic	normal
excess substantive words	lack of substantive words
few paraphasias	many paraphasias

Table 10.1. Characteristics of non-fluent and fluent speech in aphasia.

Articulation

"**Verbal agility**" refers to the ability to correctly produce all the native language phonemes at a normal speed. "**Oral agility**" refers to the agility to make movements with the articulatory organs (tongue, lips, etc.) not related to language (for instance, to move the tongue in different directions, to whistle, etc.).

Articulation disorders are usually observed in Broca's aphasia; they can also be found in conduction aphasia at the aphasia onset, or when the pathological process responsible for the aphasia extends toward the primary motor area. The type of dysarthria most frequently associated with aphasia is **spastic dysarthria**, because the pathological process is located at the level of the upper motor neuron of the pyramidal system.

Prosody and volume

Prosody is normal in posterior (fluent) aphasias. Prosody is abnormal in motor aphasias, due to the articulatory effort. By the same token, phonation is normal in fluent aphasias, but can be abnormal in motor aphasias, particularly when there is a subcortical extension. Frequently, no phonation is found at the aphasia onset.

Phonology

Frequently, phonological abnormalities (**phonological paraphasias**) are observed in aphasias. Phonological paraphasias are found in different aphasia subtypes, but they are particularly abundant in Wernicke's aphasia and conduction aphasia. They can also be found in Broca's aphasia, but frequently involve phoneme omissions, particularly in complex syllables.

Word-selection (lexicon)

Word-finding difficulties are frequent in aphasia, particularly in some subtypes of Wernicke's aphasia. Word-finding difficulties are quite often associated with **circumlocutions** and **verbal paraphasias.**

Grammar

Two major types of grammatical abnormalities are found in aphasia: (1) **agrammatism**, observed in Broca's aphasia, characterized by a reduction in the use of grammatical elements in language; and (2) **paragrammatism**, observed in Wernicke's aphasia, characterized by an over-use of grammatical elements, frequently wrongly-selected.

Language understanding

Different strategies are used to test language understanding. First, there is a level of language understanding required to follow a normal conversation. So, the first question in language understanding is: Can the patient maintain a simple conversation? This level is usually referred as "**understanding of conversational language**."

Language understanding is formally tested by: (1) asking the patient **to point** at something (usually, objects, body parts, colors, and actions); and (2) presenting **verbal commands** with increasing complexity. When asking the patient to point ("show me where …."), different categories should be used because they can be differentially impaired in aphasia; for instance, a particular patient can have severe difficulties to point at body parts, and just mild difficulties in pointing at external objects, and no difficulty at all in pointing at colors or actions. The second strategy (presenting verbal commands with increasing complexity) is used in several language understanding tests, such as the Token Test (De Renzi & Faglioni, 1978).

Repetition

Frequently, a distinction is established between **aphasias with impaired repetition ability** (perisylvian aphasias) and **aphasias with a preserved ability to repeat** (extrasylvian or transcortical aphasias).

When testing language repetition, it is important to include different types of items: at least, short-long verbal information and meaningful-meaningless utterances.

There is a test that is particularly informative in aphasia: **repetition of progressively longer sentences**. This type of test is included in some test batteries, such as the Multilingual Aphasia Examination (Benton et al., 1994).

Naming

Naming is an extremely important section in aphasia testing. When testing naming, different categories should be included: **external objects, body parts, colors** and **actions** (naming actions is not always is tested). The opposite of naming ("What is the name of this …?") is pointing ("Show me where the … is"). Pointing is indeed language understanding ability (a word is presented and the patient has to find the meaning), whereas naming is a word-retrieval skill (the meaning is presented and the patient has to find the word). As mentioned above, the ability to name/point different categories can be dissociated in aphasia.

Reading

Traditionally two different reading abilities are tested: (1) the **mechanics of reading**, that is, the ability to convert visual signs into spoken language. The mechanics of reading is tested at different levels: reading letters, syllables, words, sentences, and texts. (2) **Reading comprehension**. Reading comprehension can be tested using written commands; asking the patient to read, and then asking questions about what the patient read; and finally, matching a word or sentence with a visual representation, as included in the 'Reading comprehension of words and sentences' subtest of the Multilingual Aphasia Examination (Benton et al., 1994).

With the introduction of the psycholinguistic approaches to alexias (e.g., Marshall & Newcombe, 1973; Caramazza et al., 1985), testing for reading somehow changed.

Psycholinguistic approaches to alexias use a single test, Mechanics of reading, including only single words to read aloud; but words are controlled according to frequency (high, low, pseudowords), regularity (regular, irregular), imageneability (high, low), and grammatical category (content words, grammatical words).

Writing

Testing for writing is the opposite of testing for reading. Traditionally, writing has been tested using three strategies: **spontaneous writing, writing by dictation,** and **copying**. Different levels of complexity can be used: Letters, syllables, words, sentences, and texts.

Psycholinguistic approaches use a procedure similar to reading testing: writing single words that are controlled according to frequency (high, low, pseudowords), regularity (regular, irregular), imageneability (high, low), and grammatical category (content words, grammatical words).

Aphasia test batteries

Historically, the first aphasia test battery was proposed by Henry Head in 1926. He insisted on the need to have similar language testing procedures across different clinicians, in order to be able to compare aphasia characteristics. During the following years, however, most of the researchers in aphasia continued using informal testing procedures.

Diverse aphasia test batteries have been developed during the last decades. Some of them have become particularly influential in the area. The following aphasia test batteries will be reviewed:

- Boston Diagnostic Aphasia Examination
- Multilingual Aphasia Examination
- Minnesota Test for Differential Diagnosis of Aphasia
- Western Aphasia Battery
- Bilingual Aphasia Test

Boston Diagnostic Aphasia Examination

The Boston Diagnostic Aphasia Examination Battery (BDAE) (Goodglass & Kaplan, 1972, 1983, 2000) is one of the most widely used aphasia test batteries worldwide (Figure 10.2). It is a comprehensive, multifactorial battery designed to evaluate a broad range of language impairments. It has been translated and adapted to different languages, including Spanish (Figure 10.3); two Spanish translations and adaptations were published by Editorial Médica Panamericana in 1979 and 1996.

The BDAE is designed to go into the components of language dysfunctions (symptoms) that have been shown to underlie the various aphasic syndromes.

Figure 10.2. Boston Diagnostic Aphasia Examination, third edition.

Figure 10.3. Spanish version of the Boston Diagnostic Aphasia Examination (*Evaluación de la afasia y de trastornos relacionados*).

This test battery evaluates various perceptual modalities (e.g., auditory, visual), processing functions (e.g., comprehension, analysis), and response modalities (e.g., writing, articulation).

Severity scale. Eight different characteristics are scored on a 7-point scale, to pinpoint the aphasia severity: Melodic line, Phrase length, Articulatory agility, Grammatical form, Paraphasias in running speech, Repetition, Word finding, and Auditory comprehension. Figure 10.4 presents the typical profile for Broca's aphasia and Figure 10.5 presents the typical profile for Wernicke's aphasia; in each figure, two cases are included.

Figure 10.4. Deficit pattern for Broca's aphasia (from Goodglass, & Kaplan. The Assessment of Aphasia and Related Disorders. 2nd ed. Philadelphia: Lea & Febiger; 1983).
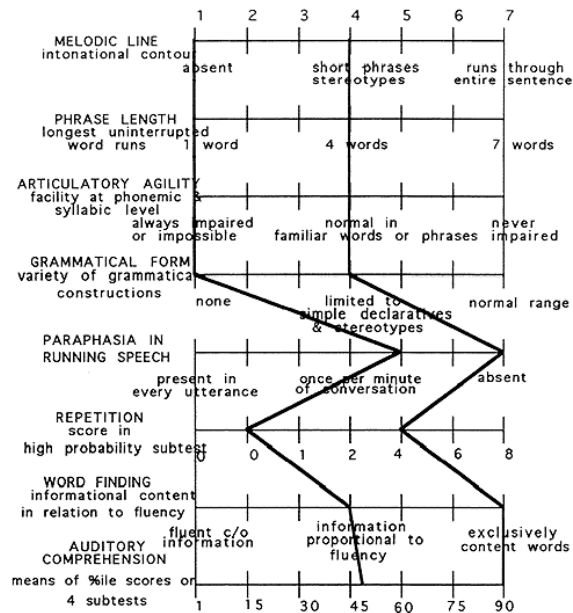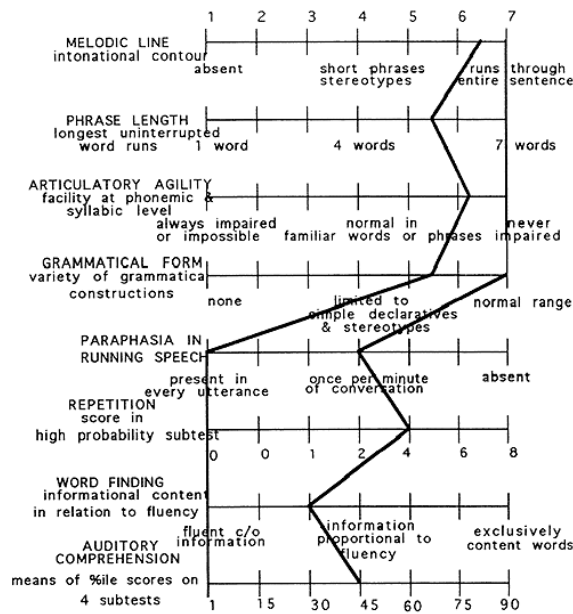

Figure 10.5. Deficit pattern for Wernicke's aphasia (from Goodglass, & Kaplan. The Assessment of Aphasia and Related Disorders. 2nd ed. Philadelphia: Lea & Febiger; 1983).

The Boston Diagnostic Aphasia Examination includes the following areas and tests:

Auditory comprehension
Word discrimination
Body part identification
Commands
Complex material

Oral expression
Automatic speech: Automatized sentences, Singing & Rhythm
Repetition: Words, High probability, Low Probability
Naming: Responsive naming, Confrontation naming, Body part naming, Animal naming

Reading comprehension:
Symbol discrimination
Word recognition
Oral spelling
Word – picture matching
Sentences – paragraphs

Writing
Mechanics
Serial Writing
Primer-level dictation
Written confrontation naming
Spelling to dictation
Sentences to dictation
Narrative writing

Administration time is variable, depending on the specific patient, but may be about 45-60 minutes.

Multilingual Aphasia Examination

This is a relatively short and easy to administer test battery (Benton, Hamsher & Sivan, 1978, 1994) (Figure 10.6); there is a Spanish version (Rey & Benton, 1991).

It is interesting to take into consideration that scores are adjusted according to the patient's age and education.

Figure 10.6. Multilingual Aphasia Examination.

The Multilingual Aphasia Examination includes the following subtests:

1. Visual naming
2. Sentence repetition
3. Controlled word association
4. Oral spelling
5. Written spelling
6. Block spelling
7. Token test
8. Aural comprehension of words and sentences
9. Reading comprehension of words and sentences
10. Rating of articulation
11. Rating of praxic features of writing

Administration time is variable, depending on the specific patient, but may be about 30-40 minutes.

Minnesota Test for Differential Diagnosis of Aphasia

The Minnesota Test for Differential Diagnosis of Aphasia (Schuell, 1953, 1973) (Figure 10.7) was one of the first systematic test batteries to be developed. It is a large and comprehensive battery; administration can take several hours. Results are summarized in a diagnostic scale divided into functional categories. The results are expected to be especially useful in planning therapeutic procedures.

It contains 59 subtests grouped into five different areas:

- Auditory disturbances (9 subtests)
- Visual and Reading disturbances (9 subtests)
- Speech and language disturbances (15 subtests)
- Visuomotor and Writing disturbances (10 subtests)
- Numeral Relations and Arithmetical Processes (4 subtests)

Figure 10.7. The Minnesota Test for Differential Diagnosis of Aphasia

Western Aphasia Battery

The Western Aphasia Battery (Kertesz, 1982, 2006) (Figure 10.8) represents a further development of the Boston Diagnostic Aphasia Examination. As a matter of fact, some of the items have been taken from the Boston Diagnostic Aphasia examination. It

includes 4 oral language subtests that allow drawing five scores. These scores are converted in a 10-point scale, in order to create a performance profile.

Figure 10.8. Western Aphasia Battery.

The examiner first engages patient in conversation, and then scores the informational content and fluency of spontaneous speech according to the scale provided in test booklet.

Three different quotients are calculated: Cortical Quotient, Aphasia Quotient and Performance Quotient. The Aphasia Quotient and Performance Quotient are combined to obtain the Cortical Quotient.

Test items used to calculate Aphasia Quotient include responding to questions; identifying objects, body parts, pictures, letters, and numbers; following directions; imitating words; and naming objects. A normal score corresponds to 100. Discrepancies from this total score are informative about the aphasia severity. Reading, writing, arithmetic, praxis, and constructional ability tests, as well as a test similar to the Raven's Progressive Matrixes, are included in the Performance Quotient.

There is a Spanish adaptation of the Western Aphasia Battery carried out by Pascual Leone in Spain (Kertesz, Pascual-Leone & Pascual Leone, 1990), and it has been used in different Latin American countries, for example in México.

Bilingual Aphasia Test

Paradis has been working for many years, developing a battery to assess bilingual aphasia subjects (Paradis, 1987). This battery is known as the *Bilingual Aphasia Test* (BAT). The BAT was designed to assess each of the languages of a bilingual or multilingual individual with aphasia in an equivalent way. The various versions of the BAT are thus not mere translations of each other, but culturally and linguistically equivalent tests. The battery has to be administered in both languages, and further, the ability to translate is analyzed.

This test is available in dozens of different pairs of languages, including Spanish-English (Paradis & Ardila, 1989) (Table 10.2).

Amharic | Arabic (Jordanian, Maghrebian) | Armenian (Eastern, Western)| Azari | Basque | Berber | Bosnian | Bulgarian | Carinthian | Catalan | Chinese (Mandarin, Cantonese) | Croatian | Czech | Danish | Dholuo | Dutch | English | Farsi | Finnish | French | Friulian | Galician | German | Greek | Hebrew | Hindi | Hungarian | Icelandic | Inuktitut | Italian | Japanese | Kannada | Korean | Kurdish | Latvian | Lithuanian | Luganda | Malagasy | Norwegian | Oriya | Polish | Portuguese (European, Brazilian) | Romanian | Russian | Sardinian | Serbian | Slovenian | Somali | Spanish (Castilian, American) | Swahili | Swedish | Tagalog | Tamil | Tulu | Turkish | Ukrainian | Urdu | Vietnamese | Yiddish |

Table 10.2. Languages in which the BAT is available.

The BAT includes three sections:

PART A
History of bilingualism (items 1-50)

PART B
Language background (items 1-17)
Spontaneous language (items 18-22)
Pointing (items 23-32)
Simple commands (items 32-37)
Semi-complex commands (items 38-42)
Complex commands (items 43-47)
Verbal-auditory discrimination (items 48-65)
Syntactic comprehension (items 66-152)
Semantic categories (items 153-157)
Synonyms (items 158-162)
Antonyms (items 163-167)
Antonyms II (items 168-172)
Grammaticality judgment (items 173-182)
Semantic acceptability (items 183-192)
Repetition and judgment (items 193-252)
Sentence repetition (items 253-259)
Series (items 260-262)
Verbal fluency (items 263-268)
Naming (items 268-288)
Sentence construction (items 289-313)
Semantic opposites (items 314-323)
Derivational morphology (items 324-343)
Description (items 344-346)
Mental Arithmetic (items 347-361)
Text listening comprehension (items 362-366)
Reading words aloud (items 367-376)
Reading sentences aloud (items 377-386)
Text reading comprehension (items 387-392)
Copying (items 393-397)
Word dictation (items 398-402)
Sentence dictation (items 403-407)
Word reading comprehension (items 408-417)

Sentence reading comprehension (items 418-427)
Spontaneous writing
Post-test analysis

PART C
Four tasks in each direction
Recognition of translation equivalents
Production of translation equivalents
Translation of sentences
Grammaticality judgments

Currently, the Bilingual Aphasia Test is available online:
http://www.mcgill.ca/linguistics/research/bat/

Assessment of specific linguistic abilities

In addition to the test battery assessing language in a comprehensive way, there is a variety of tests directed at evaluating specific linguistic abilities. The following four tests will be reviewed:

- Boston Naming Test
- Token Test
- Verbal Fluency tests
- Cross-Linguistic Naming Test

Boston Naming Test

The Boston Naming Test (BNT) (Kaplan, Goodglass & Weintraub, 1978) is a well-known naming assessment instrument, used to evaluate confrontation naming. The BNT contains 60 line drawings graded in difficulty from "bed" (easy, high frequency) to "abacus" (difficult, low frequency) (Figure 10.9).
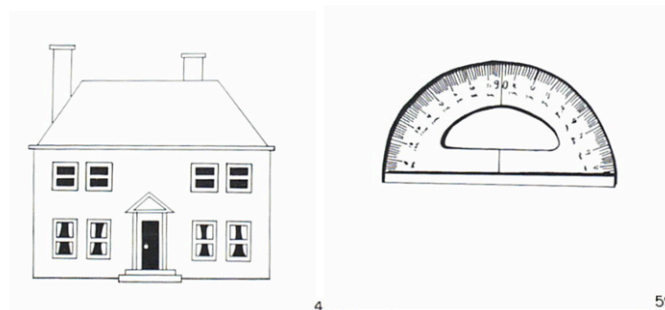
Figure 10.9. One of the easiest and one of the hardest items in the BNT.

The patient is instructed to tell the examiner the name of each picture; 20 seconds to respond for each trial are allowed. If the patient fails to give the correct response, the examiner may give a semantic cue (a description of the function); if the patient cannot answer, a phonemic cue (the initial sound of the target word) is provided. Scores are significantly correlated with the subject's age and educational level.

There are versions of the BNT adapted to other languages, including Spanish. There are also shorter versions of the BNT.

Token Test

The Token Test is a short test very sensitive to language understanding defects. There are two different versions: an extended one (De Renzi & Vignolo, 1962) and an abbreviated one (De Renzi & Faglioni, 1978). Norms for the short version include a correction score according to the subject's education. There are several adaptations; for instance, the Multilingual Aphasia Examination (Benton & Hamsher, 1976) and the Neuropsi (Ostrosky, Ardila & Rosselli, 1999) both include an adaptation of the Token Test.

The test includes 20 tokens (Figure 10.10):

- Two shapes (circles, squares)
- Two sizes (small, big)
- Five colors (red, green, yellow, blue, white)
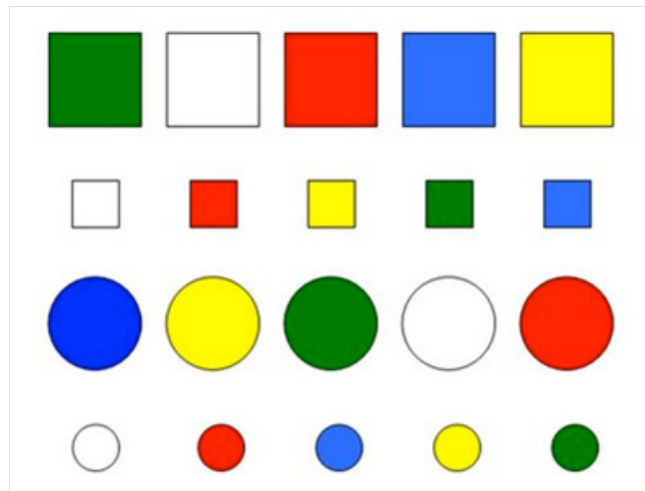
Figure 1010. Tokens used in the Token Test.

Different commands with an increasing level of difficulty are presented. The test is divided into sections according to the length of the command, syntactic complexity, and working memory demand; the commands become more difficult within each section and across sections. These are some examples of the different levels of difficulty:

1. Touch a circle
8. Touch a yellow square
12. Touch the small white circle
16. Touch the red circle and the green square
20. Touch the large white circle and the small green square
24. Put the red circle on the red square

Verbal Fluency tests

Verbal fluency tests are short and easy to administer. They assess the ability to find words according to a semantic or phonemic characteristic. Two different conditions are used:

(1) **Category or semantic verbal fluency** (to say as many words as possible corresponding to a specific semantic category –such as animals, fruits, or vegetables– in one minute).

(2) **Phonemic (letter) fluency** (to say words beginning with a particular letter; usually F, A, and S; one minute is usually provided).

Typically, the amount of correct words produced in one minute is counted. A normal person can produce about 12 words beginning with a specific letter, and about 16 words corresponding to a semantic category in one minute (Table 10.3 and Table 10.4)

Educational level	Age				
	56-60	**61-65**	**66-70**	**71-75**	**>75**
0-5 years	12.8 (9.4)	12.2 (8.4)	11.2 (7.2)	11.1 (7.2)	9.4 (7.0)
6-12 years	16.0 (12.2)	15.9 (12.1)	13.3 (9.9)	12.7 (9.3)	11.0 (7.2)
> 12 years	16.1 (13.3)	16.1 (12.9)	15.8 (12.5)	13.7 (12.4)	11.7 (9.1)

Table 10.3. Semantic and phonemic (in parentheses) verbal fluency in a sample of 346 normal subjects (Ardila & Rosselli, 1989).

	English monolinguals (n=45)	Spanish monolinguals (n=18)	Bilinguals	
			English	**Spanish (n=19)**
F	12.9(5.4)	11.7(4.1)	12.5(5.0)	11.3(4.3)
A	10.7(5.1)	11.8(4.6)	10.7(5.4)	12.3(4.6)
S	13.8(5.4)	11.4(3.8)	12.4(3.9)	11.6(5.4)
Animals	16.8(5.2)	16.7(3.8)	14.2(4.1)	14.5(3.8)

Table 10.4. Some norms for the Verbal Fluency tests in monolingual and bilingual participants (60-65 years; 12-14 years of education) (Rosselli et al., 2002).

Cross-linguistic naming test

Proceeding from the basic **universal vocabulary** proposed by Swadesh (1952, 1967), a naming test potentially usable in any language, was developed (Ardila, 2007). Six different semantic categories were included:

- **body-parts** (10 words),
- **natural phenomena** (non-touchable) (5 words),
- **external objects** (potentially known through sight and touch) (5 words);
- **animals** (5 words),
- **colors** (5 words)
- **actions** (10 words).

This test has two major advantages: on one hand, **it is readily available in hundreds of different languages**; and on the other, **it is not a "fixed" test**, but it includes photographs that can be replaced. Theoretically, norms are not required, and it represents a low-ceiling test. Word frequency can be used as a criterion for the level of difficulty. Table 10.5 presents the words corresponding to body-parts included in the test, in six different languages, and Figure 10.11 illustrates some pictures that can be used in the Cross-Linguistic Naming Test.

English	Spanish	Turkish	Basque	Russian	Sikuani
ear	oreja	kulak	belarri	uxa	muxu
eye	ojo	göz	begi	glaz	itaxu
nose	nariz	burun	sudor	nos	pumu
mouth	boca	ağız	aho	rot	koibo
tooth	diente	diş	hortz	zub	wono
tongue	lengua	dil	mihia	jazyk	ebanu
knee	rodilla	diz	belaun	koliena	matabaka
belly	estomago	karın	sabel	zhivot	koto
neck	cuello	ense	lepo	ceja	wosi
foot	pie	ayak	oin	noga	taxu

Table 10.5. Body-parts included in the Cross-Linguistic Naming Test in different languages (Sikuani is an Amerindian language from the Amazonian jungle).
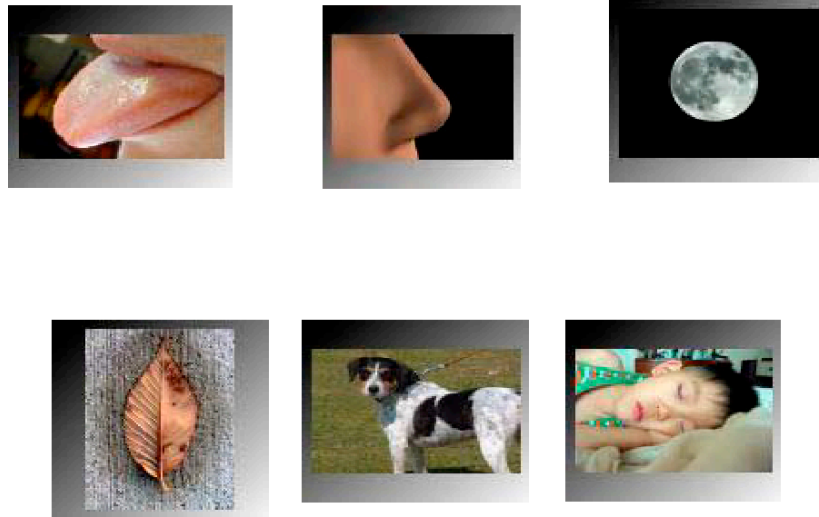
Figure 10.11. Some pictures that can be used in the Cross-Linguistic Naming Test.

Summary

The major goal of language evaluation is to determine if patient's language is normal or abnormal. If abnormal, it is necessary to pinpoint what specific syndrome the patient presents. Six different aspects of language have to be assessed: expressive language, language understanding, repetition, naming, reading, and writing. There are several extensive test batteries that include all these aspects, such as the Boston Diagnostic Aphasia Examination, the Multilingual Aphasia Examination, the Minnesota Test for Differential Diagnosis of Aphasia, the Western Aphasia Battery and the Bilingual Aphasia Test. There is also a diversity of tests directed at evaluating specific linguistic abilities, including the Boston Naming Test, the Token Test, the Verbal Fluency tests, and the Cross-Linguistic Naming Test.

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Chapter 11

Recovery and prognosis in aphasia

Introduction

The recovery of language after a pathological brain condition is a question that has interested health professionals since Hippocrates (~400 years BC). Aphasia is a relatively frequent condition and the amount of references throughout recent human history to the "loss of speech" associated with brain disorders and head injuries, is not surprising. During the XVI century, some specific reports of spontaneous recovery after brain pathologies are found; different authors express interest in the potential improvement of communication ability after an abnormal brain condition.

World wars during the XX century were social conflicts resulting in an enormous amount of wounded people, including people with traumatic aphasias. Death tolls in both, WWI and WWII, are counted in dozens of millions of people, and it is easy to assume that the number of wounded individuals also corresponds to dozens of millions. Many of them were people with traumatic aphasias.

During and after WWI (1914-1918), in some hospitals, particularly in Germany, aphasia rehabilitation sections were created and a significant interest in aphasia recovery and treatment was observed. Goldstein (1917), Head (1926), and Nielsen (1936, 1938) are just some of the many clinicians that approached the question of aphasia rehabilitation during this time.

However, it was only during WWII (1939-1945) that aphasia rehabilitation became a central clinical issue. During the time of the war, Luria was working in a rehabilitation hospital in the Urals (Russia); he had the specific goal of developing rehabilitation procedures for war-wounded soldiers. After the WWII, Luria published his classical book "Restoration of functions after brain injury" (1948/1963), which became the most important book ever published about cognitive rehabilitation. During the following years, this book was translated to many different languages and became a milestone, not only for language rehabilitation, but also for cognitive rehabilitation in general.

Interest in aphasia recovery and rehabilitation has continued growing to the present day. Currently, new technological instruments, such as contemporary neuroimaging techniques (for instance, functional magnetic resonance, fMRI; and positron emission tomography, PET) have become available; these new technologies have allowed for a more precise understanding of the neurological processes accounting for aphasia recovery and rehabilitation.

Stages of language recovery

After a pathological brain condition, some recovery is expected. This recovery, observed without the application of any language intervention techniques, is known as "spontaneous recovery" (Figure 11.1) (Demeurisse et al., 1980; Lendrem & Lincoln, 1985). Of course, spontaneous recovery is only observed after a stroke, a traumatic

brain injury, or another "static" condition; when aphasia is due to a progressive condition, such as a brain tumor or a degenerative disease, no spontaneous recovery is observed.
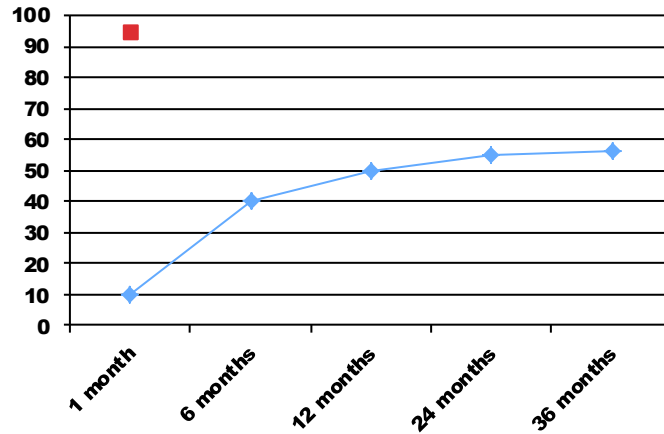

Figure 11.1. Typical curve of spontaneous recovery.

Two stages in spontaneous recovery are usually distinguished (Kertesz, 1988; Lomas & Kertesz, 1978):

Stage 1 (early recovery)

It refers to the rapid recovery observed during the initial weeks and month after the aphasia onset. Indeed, most of the spontaneous recovery is observed during the initial 3 months following the pathological brain condition. It has been assumed that some neurophysiological processes (such edema decrease, disappearance of hemorrhages, etc.) are responsible for this initial rapid language improvement. An association between early recovery of spoken word comprehension and reperfusion (restoration of blood flow) of Wernicke's area has been demonstrated in stroke aphasia patients (Hillis & Heidler, 2010).

Nonetheless, spontaneous recovery becomes progressively slower.

Stage 2 (late recovery)

Language continues improving during the following months, but recovery is progressively slower and slower. It is usually accepted that after about 2-3 years, no further spontaneous recovery is observed. **Relearning** and **reorganization of the language in the brain** are considered the two basic mechanisms accounting for this late language recovery. In general, relearning (i.e., re-training) and language reorganization (i.e., the use of compensatory strategies to overcome the specific communication deficits) have been considered the two means for language rehabilitation in aphasia.

Factors affecting recovery

Various factors affect language recovery in aphasia. Some of them can be regarded as major factors, whereas others represent secondary factors.

Lesion site

Lesion site is associated with aphasia type; this means that left temporal lobe damage will result in a Wernicke's-type aphasia, and a left posterior frontal pathology will be associated with a Broca's-type aphasia. In some, recovery from language defects is very limited (e.g., agrammatism associated with Broca's aphasia), whereas the recovery from other language defects can be significantly better (phoneme discrimination in Wernicke's aphasia).

Lesion size

The association between language recovery and lesion size is obvious: the smaller the lesion, the milder the language defect and the larger the remaining intact brain tissue that can be used to re-learn and compensate the language deficit. Conversely, the larger the lesion, the greater the language defect and the smaller the remaining intact brain that can be used to re-learn and compensate the language deficit.

Age

Frequently, it has been assumed that children recover more rapidly than adults suffering from the same type of brain lesion; this assumption is known as the "**Kennard principle**" (Kennard, 1936). According to the 'Kennard Principle', "there is a negative linear relation between age at brain injury and functional outcome. Other things being equal, the younger the lesioned organism, the better the outcome" (Maureen, 2010; page 1043). This principle has been recently challenged (e.g., Lauterbach et al., 2010; Narbona & Crespo-Eguílaz, 2008). Some authors, however, continue supporting it.

Etiology

Aphasia recovery tends to be better in the case of traumatic head injury than in cerebrovascular accidents. In degenerative conditions, no recovery is anticipated. Aphasia associated with tumors is variable and recovery depends on the specific type of tumor. Recovery after tumor removal may be excellent in a usually benign tumor, such as meningioma. No recovery is expected in a rapidly growing malign tumor, such as glioblastoma. Recovery, however, is associated not only with the type of tumor, but also with other factors such as size, location, patient's age, etc.

Aphasia profile

Language recovery depends on the specific type of aphasia. Language recovery is worst in cases of global aphasia. In general, the larger the brain damage, the more severe the aphasia and the lesser the expected recovery. Some types of aphasia are considered to be very limited in their recovery; for instance, recovery in Broca's aphasia is frequently very modest. Furthermore, aphasia profile may change over time; for instance, a Wernicke's aphasia can become an anomia.

Temporal factors

Temporal factors refer to the speed of the pathological process (i.e., etiology) accounting for the aphasia. The general rule is: when the pathological process has a sudden installation (e.g., traumatic head injuries and strokes), the initial symptomatology is severe, but recovery is good; conversely, when the pathological

process has a progressive installation (e.g., slow growing tumors), the initial symptomatology is mild, but recovery is limited.

Time from onset

It is known that in aphasia, language therapy should begin as soon as possible. It is generally accepted that the sooner it begins, the better the recovery will be. During some time it was (wrongly) assumed that 2-3 years after the aphasia onset, the observed language defects were permanent and aphasia therapy was no longer effective.

Handedness

Frequently, it is assumed that left-handers as a group have a more bilateral representation of language; because of this more bilateral representation, language recovery in cases of aphasia is more rapid and more complete. Basso (1992) presents an extensive review of prognostic factor in aphasia recovery; she concludes that handedness and gender play just a minor role in recovery from aphasia

Gender

The influence of gender on aphasia recovery has been controversial. Assuming that females have a more bilateral representation of language, it has been suggested that they present a better aphasia recovery. For instance, Pizzamiglio et al. (1985) analyzed 91 adult aphasics of both sexes before and after a 3-month period of language therapy. Although no initial difference was found in severity of language disorders between sexes, females within the global aphasic group showed significantly greater improvement in three tests of language comprehension. It was suggested then that more bilateral representation of language functions in the female brain may account for this greater improvement. Some other authors have reported minimal differences between genders (e.g., Basso, 1992), where other clinicians have not found any difference at all (e.g. Pedersen et al., 1995).

Treatment

The effect of aphasia treatment represents a major factor affecting recovery. This has been firmly established for a long time (Basso et al., 1979) and has been corroborated in many different studies using diverse methods (e.g., Basso, 2003; Helm-Estabrooks & Albert, 2004).

Motivation and personality

It has been suggested that motivation and personality play a crucial role in aphasia recovery. For example, people used to reading will be especially motivated to recover their reading ability in the case of alexia. By the same token, certain personality factors (e.g., tenacity) can be crucial in any type of therapy; therapy is usually not only time consuming, but also effort-demanding.

Associated disorders

Aphasia is frequently associated with a diversity of disorders, such as hemiparesis, apraxia, acalculia, agnosia, amnesia, etc. (See **Chapter 8: Associated Disorders**). Of course, a patient with hemiparesis (or other disorders) will have more limitations, and hence the recovery can be slower and the therapy harder to administer.

Effects of therapy

It has been well established that aphasia therapy results in a higher performance on diverse language tests at every moment of the aphasia evolution (Figure 11.2). In a pioneer study Basso et al (1979) selected 281 aphasics (162 reeducated and 119 controls); they were subjected to a second examination no less than six months after the first. Presence or absence of rehabilitation between first and subsequent examination was studied. It was found that rehabilitation had a significant positive effect on improvement in all language skills. This study was particularly important because of the large sample of participants; taking into consideration the size of the sample, potential confounding variables capable of affecting the results were randomly distributed. This positive effect of language therapy has been extensively corroborated using different methods (e.g., Basso, 2003; Helm-Estabrooks & Albert, 2004).
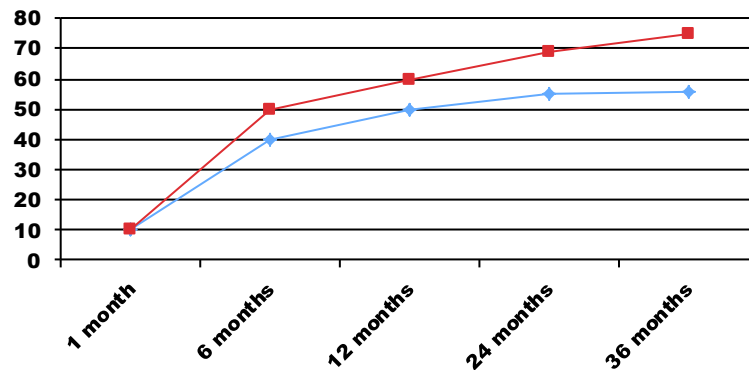

Figure 11.2. Hypothetical curve of aphasia evolution. Comparison of the aphasia recovery with (red) and without (blue) therapy.

Brain damage symptoms

Goldstein (1948) defines two types of symptoms observed after a brain pathological condition:

1. Direct (or **negative**, according to Jackson, 1864). They represent a direct consequence of the brain damage; for example, word-finding difficulties due to pathology in the posterior left temporal lobe.

2. Indirect or **positive**. Behavioral or cognitive changes intended to compensate the deficits. They are affected by the previous personality and current environmental conditions. For instance, people with language understanding difficulties frequently attempt to pay an increased attention to some secondary information such as the gestures, the face expressions, and the lip movements.

Why recovery?

It is presumed that recovery is due to two major mechanisms: relearning (re-training) and compensatory techniques (reorganization of the functional system) (Levin & Grafman, 2000; Luria, 1980). Practice plays a decisive role in the re-learning process.

Re-learning (re-training)

Regardless of the brain damage, language can be re-learned to some extent. It is likely that homologous areas of the contralateral (right) hemisphere participate in this relearning process (Raboyeau et al., 2008). It has been observed that the practice in of ability (language or other) results in an increase in the size of the cortical brain area involved in that particular ability (Levin & Grafman, 2000).

Compensatory techniques (reorganization of the functional system)

This means that an alternative way to process the information is used to perform the task. The possibility of using alternative strategies is a consequence of the brain's plasticity. Plasticity is defined as the brain's capacity to be shaped by experience, its capacity to learn and remember, and the ability to reorganize and recover after injury (Gleissner et al., 2005; Kolb et al., 2003). For instance, the aphasic patient can use speech prosody in an extended way to communicate (prosody is potentially preserved in cases of aphasia; it is more related to the right hemisphere activity; Ross & Monnot, 2008; and prosody production and understanding are impaired in cases of right hemisphere pathology).

Rehabilitation Goals

A rehabilitation program for aphasia, as a matter of fact, has different goals. They can be summarized in the following five points:

To keep the patient verbally active

This is the basic rule in any type of rehabilitation: keep the patient active. Frequently, because of the communication difficulties, there is a certain tendency to verbally isolate the aphasic patient. The family frequently attempts to communicate using only gestures. If the aphasic patient is not intensively exposed to language, and is not required to practice in a continuous way, recovery will be limited. Speech/language therapy plays a crucial role in this regard.

To re-learn language

To a significant extent, therapy is directed at re-learning language. Regardless of age, and the abnormal brain condition, it is still possible to at least learn some language. This re-learning process has to follow a specific sequence: from the simpler to the more complex. There is a gradual sequence to re-learn the vocabulary or the grammar. The simpler part has to be re-learned first.

To provide strategies to improve language

Linguistic abilities can improve if certain strategies are used (i.e., there is a reorganization of the functional system). These strategies depend on the particular type of aphasia and the specific conditions of the patient. For instance, so called

*Melodic Intonation Therapy (*Albert et al., 1973; Norton et al., 2009*)* is very useful in cases of Broca's aphasia, but is not particularly effective in Wernicke's aphasia.

To teach the family to improve communication

Language is used to communicate in different social contexts, but family represents the major and most significant communication context, particularly for an individual with some limitations. One major function of the speech/language therapist is to explain to the family how to maximize the effectiveness of communication with the patient. Some simple and easy to use strategies can be considered, such as:

  • To avoid interference, especially, but not only, verbal interference
  • To keep the conversational topic
  • To use redundant information
  • To speak slow but not too slow
  • To use prosody and other paralinguistic information
  • To be aware that the patient's communication ability fluctuates
  • To be aware that the use of language is specially effort- and attention- demanding for the aphasic patient

To provide psychological support

Because of the communication difficulties, aphasic patients usually have a feeling of isolation and solitude. Communication with somebody capable of understanding them (the therapist) becomes particularly important and reinforcing. Usually the patient has significant internal conflicts (e.g., family, finances, etc.) due to his/her condition. By the same token, the patient usually has important questions about the future (What is going to happen? What language recovery to expect? etc.). Additionally, it is not unusual (particularly in some aphasias) to be depressed. All of these difficulties and conflicts can be reflected in the rehabilitation context. Encouraging the patient, providing realistic goals, etc., represents a very important psychological support for the aphasic patient.

Summary

The recovery of language after a pathological brain condition is a question that has interested health professional for a long time. During WWI, but very especially during WWII, the question of aphasia recovery and rehabilitation became most crucial. After a pathological brain condition, some recovery is expected. This recovery observed without the application of any language intervention technique is known as "spontaneous recovery". Two stages in recovery are usually distinguished (early and late recovery). There is a series of factors that have proven to affect recovery, including lesion site, size, age, etiology, aphasia profile, temporal factors, time from onset, handedness, gender, treatment, motivation and personality, and associated disorders. Two types of symptoms have been described after a brain pathological condition: direct and indirect. It is assumed that recovery is due to two major mechanisms: re-learning (re-training), and compensatory techniques (reorganization of the functional system). A rehabilitation program for aphasia has different goals, including: to keep the patient verbally active, to re-learn language, to provide strategies to improve language, to teach the family to improve communication, and to provide psychological support to the patient.

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Chapter 12

Aphasia rehabilitation

Introduction

Patients with aphasia may present some spontaneous language improvement (so-called "spontaneous recovery"), but systematic therapeutic programs can significantly contribute to a more rapid and complete language recovery.

During the last decades, it has been observed that an increased number of aphasia patients have had the opportunity to participate in rehabilitation programs; this situation has resulted in a better quality of life for a significant number of aphasic individuals. Unfortunately, however, there are still many aphasic patients that, due to a range of conditions, cannot attend language therapeutic programs; language recovery for them is more limited, and the quality of life is somehow lower.

By the same token, research in aphasia rehabilitation has significantly grown during the last decades. New research has contributed not only to having a better understanding of the rehabilitation processes, but has also proposed new avenues to treat speech and language disorders associated with brain pathology. New technological advances, such as contemporary neuroimaging techniques, have significantly increased our insight of the neurological processes underlying language recovery in aphasia. It can be anticipated that this interest in aphasia rehabilitation will continue growing toward the future.

In this chapter some major general guidelines in aphasia rehabilitation will be analyzed. An attempt to integrate contemporary approaches to aphasia rehabilitation (Albert et al., 2012; Basso, 2003; Helm-Estabrooks et al., 2012) will be made. Later, some examples of specific rehabilitation techniques will be presented.

Obviously, the starting point for any rehabilitation program is a good language evaluation in order to pinpoint the specific language components impaired in the patient. Rehabilitation programs should be tailored to the specific linguistic needs of each patient. However, it can be considered that language includes two major dimensions: lexical and grammatical (Ardila, 2011, 2012). The first one is mostly impaired in the sensory aphasias, whereas a disturbance of the second one is characteristic of Broca's aphasia. Basso (2003) specifically analyzes the rehabilitation of each one of these two language levels. Her rehabilitation strategies for each one of these components are presented below.

Rehabilitation of lexical and sentence disorders

Different specific components of the language lexicon can be distinguished (auditory analysis, word identification, semantics associations, etc.). However, many components and/or processes are generally impaired in the same patient.

Auditory analysis system

According to Basso (2003) **the same tasks used to evaluate the integrity of a component can also be used for its treatment**. So, if the patient has difficulties in phoneme discrimination, pairs of phonemes can be presented in order to say whether they are the same or different. Initially both phonemes can be different in several features, such as,

/p/ - /r/

And progressively the task moves to phonemes that are different in only one feature, such as,

/b/ -/p/

Abstract letter identification

The use of computers seems appropriate in cases of damage to the abstract letter identification system. The patient is shown pairs of letters in different fonts and has to say whether they represent the same letter or not. Initially, differences can be obvious (e.g., R and r) and progressively they become more and more similar (e.g., K and k). The task can become progressively more complex, as pairs of letters and words are introduced.

Input lexicon (comprehension)

It is interesting to note that comprehension disorders have been shown to be the first to recover spontaneously in a significant number of aphasic patients. Comprehension disorders are mostly associated with Wernicke's-type fluent aphasias.

A classical approach for the treatment of comprehension disorders has been word-picture matching; this is when a word is orally presented (e.g., pencil) and the patient is required to point to the picture representing a pencil. However, this strategy does not distinguish between input lexicon (decoding and integrating the sequence of phonemes included in the word pencil); and semantic system disorders (associating the word "pencil" with the representation –meaning– of the word).

The task of choice for the evaluation of the input lexicons is the so-called "lexical decision task"; that is, determining whether or not a string of letters (or phonemes) corresponds to a real word or not. Written stimuli can be used for the orthographic system and spoken for the phonological system. For example:

CAR: Is it a real word or not?
PAR: Is it a real word or not?

Basso (2003) suggests that an exercise that the patient can do alone is to look up in a small dictionary containing only frequently used words, and focus on those that s/he is unsure about. The patient is encouraged to look the orthographic form and to read the definition.

The semantic system

Damage to the semantic system will prevent the correct performance of any task requiring the comprehension or production of words. The semantic system is mostly impaired in Wernicke's aphasia and so-called transcortical (extrasylvian) sensory aphasia, which indeed can be regarded as a subtype of Wernicke's aphasia.

Tsvetkova (Hanninen, 1985) observed that patients with lesions in the temporal-occipital area of the left hemisphere have significant difficulties in drawing the meaning of words. For instance, if these patients are required to draw a "squirrel" and a "rabbit", they may know that "both are animals; and animals have a head, ears, tail, body and legs"; but these patients are unable to draw the specific features distinguishing both animals (i.e., squirrel has big tail and small ears; whereas rabbit has small tail and big ears) (Figure 12.1).
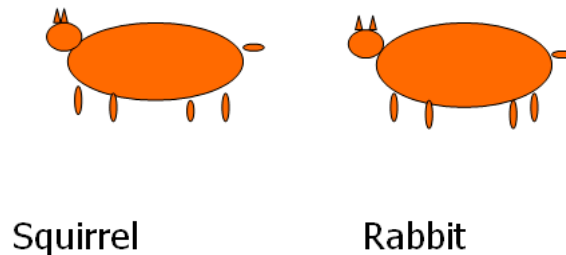

Squirrel   Rabbit

Figure 12.1. Typical drawings of a squirrel and a rabbit in patients with left temporal-occipital lesions. The patient is unable to use the distinguishing features, characteristics of a squirrel and a rabbit.

The complete knowledge that we have about a word, including visual aspects, function, sensory attributes, associated gestures, category, and so forth, correspond to the "semantic system" (concept) of that particular word. However, the sensory representations are different depending on the specific knowledge that we have about that particular word; indeed, they are modality-specific semantic systems (e.g., visual, auditory, etc.). For instance, these are the associations of the following words:

House*: only has a visual representation
Phone: has a visual and also an auditory representation
Key: has visual, tactile, and auditory representations
Ice cream: has a visual and a gustatory representation
Flower: has a visual and an olfactory representation

Classification tasks are frequently used to treat the semantic deficits in aphasia and restore the semantic representations of the words. Initially, the patient can be requested to make classification of objects represented in cards (for instance, animals, furniture, and fruits) without using language. A name is then given to each category; and emphasis is made on the features distinguishing each category. Further categories are introduced, for instance, pets and wild animals; later, different representations of the same animal (e.g., a cat) can be used to emphasize the common features. This type of classification task can also be developed using written words, instead of the direct visual representation. The purpose is to restore the semantic field of the words (Figure 12.2)
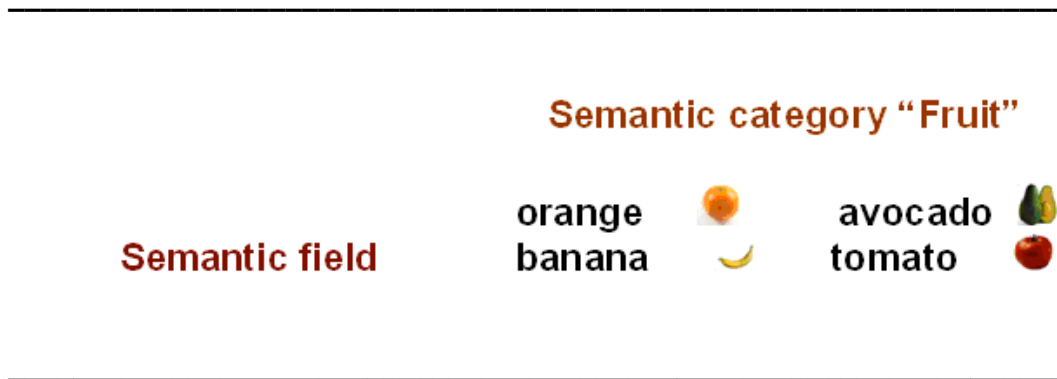
_____

_____

Figure 12.2. Semantic field of the word "fruit". Pictures and words can be used.

Output lexicon

Difficulties in using vocabulary words can be due to defects in storage (i.e., knowledge) or access (i.e., retrieval of the word). According to Basso (2003), several criteria can be used to distinguish storage and access disorders.

Storage disorders have the following characteristics: (1) responses are consistent; (2) there is a "frequency effect" (that means that high frequency words are easier than low frequency words); (3) it is easier to make decisions about superordinate than subordinate information; (4) a "priming effect" is not observed; (5) there is no effect from the rate of presentation (Warrington & Shallice, 1979).

Access disorders on the other hand, present the following characteristics: (1) there is inconsistency in the responses; (2) the frequency effect is weak; (3) superordinate and subordinate information are equally damaged; (4) there is a positive "priming effect"; (5) performance is better at lower rates of presentation.

Priming is an increase in the speed or accuracy of a decision that occurs as a consequence of a prior exposure to some of the information in the decision context, without any intention or task-related motivation. Since priming occurs in tasks where memory for previous information is not required, and may even have detrimental effects, it is assumed to be an involuntary and perhaps unconscious phenomenon.

One of the original demonstrations of priming occurred in a **lexical decision task,** in which a series of decisions is made about whether or not letter strings correspond to real words. Priming is shown to occur in cases where two successive letter strings were semantically related words. For example, the decision that 'doctor' is a real word is faster when the preceding letter string is 'nurse,' compared to 'north' or the non-word 'nuber'.

In most cases patients rehabilitated for anomia are required to produce the target words, but the strategies used are different. Most frequently **cuing,** both **phonemic** (the initial sounds) and **semantic** (describing the meaning) has been used. However, **orthographic** cuing (the initial letters included in the target word) has also proven to be effective in facilitating naming. Other strategies can also be useful; for instance, including the target word in a "high probability sentence" (e.g., "I write with a…").

Conversion rules

Some so-called conversion rules have been analyzed in aphasia. These conversion rules can be impaired in aphasia, and it is required to re-learn these rules. For instance:

- *Input to output phoneme conversion impairment (prevents repeating nonwords)*
- *Grapheme to phoneme conversions impairment (prevents reading nonwords)*
- *Phoneme to grapheme conversions impairment (prevents writing nonwords from dictation)*

Multiple disorders

As a matter of fact, in clinical practice, it is unusual to find patients with damage to a single component. Most patients have difficulties at different levels. Strategies used in aphasia therapy depend on the specific components that are impaired.

Sentence level

Difficulties in producing sentences represent the most salient diagnostic sign of agrammatism associated with Broca's aphasia. Indeed, agrammatism is difficult to treat and represents a frequently long-term sequel of motor and global aphasia. When treating agrammatic patients, it is advisable to use words in grammatical contexts, not isolated (e.g., instead of referring to "pencil" to refer to "the pencil", "there is a pencil", "the pencil writes", etc.).

Some studies about agrammatism treatment are available (e.g., Ballard & Thompson, 1999; Bastiaanse et al.,2006; Helm-Estabrooks & Ramsberger, 1986; Marini et al., 2007). For instance, Thompson and Shapiro (2005) reported that the so-called Treatment of Underlying Forms (TUF), a linguistic approach to treatment of sentence deficits in patients with agrammatic aphasia, is effective for treating sentence comprehension and production in patients who present language deficit patterns like those seen in Broca's aphasia. Patients receiving this treatment show strong generalization effects to untrained language material.

Wisenburn et al. (2010) performed a meta-analysis of the therapy efficiency for agrammatism in aphasia. Twenty-one therapy studies for agrammatism were included in this analysis. Results showed impressive gains for most therapy approaches, with an overall mean effect size of 1.32. That means that regardless of the difficulties in treating agrammatism, different therapeutic strategies have proven to be useful in recovering grammar and sentence production.

Global aphasia

In global aphasia all the language levels and modalities are impaired. Expressive language is usually reduced to some few stereotyped words or sounds. Comprehension is generally limited to some high frequency words and verbal formulas (e.g., "thank you"). Although language recovery in global aphasia is very limited, these patients can still develop some communication abilities. They are able to discriminate between the native language and a foreign language; they can recognize the pragmatic

intent of communication; recognize emotional content; follow commands that involve the whole body movements, comprehend gestures; and learn to manipulate symbols in syntactically acceptable manners to construct phrases (Alexander & Loverso, 1992).

Therapy aims, to improve a patient's ability to communicate as best as possible; their remaining abilities can be used to restore language abilities as much as possible, to compensate for language problems, and to learn other methods of communicating (Basso, 2003).

A variety of treatments are useful to assist in the improvement of language and communicative functions in the global aphasic individual (e.g., Albert et al., 2012; Alexander & Loverso, 1992; Helm-Estabrooks et al., 1982; Helm-Estabrooks et al., 2012). For instance, Helm-Estabrooks et al. (1982) selected eight globally aphasic patients who had not responded to traditional treatment and administered Visual Action Therapy (VAT), a non-vocal approach directed at training the patient to produce symbolic gestures for visually absent stimuli. Statistical analyses of pre and post VAT scores earned on the *Porch Index of Communicative Ability* (PICA) showed highly significant improvement on those subtests which measure pantomimic and auditory comprehension skills.

Treatment may be offered in individual or group settings, although individual treatment appears to be more beneficial to the patient. Some of the therapies available include the use of augmentative and alternative communication (AAC), drawing, gestures, exercises, and family involvement in home treatment.

Patients presenting severe language limitation and with little or no functional speech frequently rely on AAC devices to augment or replace natural speech. Speech generating devises are electronic AAC systems used to supplement or replace natural speech (Figure 12.3). They include communication aids and graphic symbol software programs that produce synthetic speech upon activation, such as the DynaVox. Studies involving AAC intervention with patients presenting global aphasia have demonstrated that these individuals are able to access, identify, recognize, manipulate, and combine graphic symbols in order to produce simple phrases and basic communication.

Figure 12. 3. Example of a speech generating devise.

Examples of some rehabilitation techniques

Stimulus facilitation technique

Wepman (1951, 1955) and Schuell et al. (1964) proposed a series of therapy principles. Schuell emphasized the importance of adequate stimulation, but stressed controlling the rate, the complexity, and even the loudness of language presentation. One language modality can be used to stimulate another. The use of topics that are of interest to the patient is beneficial.

Wepman (1951) emphasized three approaches: (1) *stimulation*: an organized presentation of stimuli sufficient to produce a reaction; (2) *facilitation*: repeated practice to increase the patient's efficiency in language tasks as they are accomplished; (3) *motivation:* to encourage the patient to continue the therapy process. These three approaches represent the bulk of traditional aphasia therapy.

Deblocking

Weigl (1968) emphasized the use of intact (or less damaged) language channels to compensate and actually improve the operation of malfunctioning channels. For instance, presenting the patient with the printed word simultaneously with the spoken word, when the patient understand better through the visual channel (reading) than through the auditory one.

When a word (and also a phoneme, syllable, phrase) is produced, the probability of it being produced again in a new context increases, even using different conditions; for example, if the patient gets to read the word PENCIL, the probability of correctly naming a pencil increases. Therefore, what is most important in aphasia rehabilitation is getting the patient to produce as many phonemes, syllables, words, etc. as possible, because language has to be "updated". According to Weigl, language in aphasia is blocked. A major function of therapy is deblocking it.

Functional system reorganization

Luria (1963, 1980) proposed that reorganization of the damaged functional system is required after a pathological brain condition. Better preserved levels of language can be used as a base point from which to achieve the communication goal. For instance, emotional and prosodic language is better preserved in aphasia; hence, emotional and prosodic language can be useful to recover communication ability. This is the basic idea in developing some rehabilitation techniques, such as the Melodic intonation therapy (Albert et al., 1973; Norton et al., 2009),

Luria insisted that each disorder calls for a unique rehabilitation program based on the analysis of the underlying deficit. Hence, the initial step in rehabilitation is to pinpoint the basic disturbances accounting for the language impairment. The rearrangement of basic language processes may be required to achieve the communication goal.

Melodic Intonation Therapy

Melodic intonation therapy (MIT) (Albert et al., 1973; Norton et al., 2009) in aphasia is a well-defined and effective technique for rehabilitation of non-fluent aphasia. MIT is characterized by slow and precise tempos that facilitate articulation and reduce paraphasic errors by drawing attention to the three elements that comprise MIT. These elements are melodic line, rhythm, and points of stress.

Starting from a series of carefully intoned sentences and phrases, the aphasic subject is guided through a sequence of steps which increase the lengths of the units, diminish the dependency on the clinician, and diminish reliance on intonation. At the end of the

program, the patient is capable of using spoken prosody for uttering the sentences embedded in the program structure

Four progressive levels of difficulty can be used:

Level I has no linguistic component and simply requires that the clinician hum the melody of the target phrase and aid the patient in accurately hand-tapping the rhythm of each utterance.

Level II involves moving from asking the patient to hand-tap the utterance, to asking him/her to actually repeat it.

Level III involves reducing the participation of the clinician. Strategies such as enforced delay of responses are used in order to force an element of retrieval and later requiring that the patient give appropriate responses to intoned questions.

Level IV primarily involves facilitating the return of the patient's normal speech prosody. Strategies used in this final stage include latency between stimulus and response, training longer and more complex sentences, and the use of speech-song. The melodic line remains the same, as in the levels prior to this one; however, pitch becomes variable and more alike to that of normal speech.

Augmentative and Alternative Communication

AAC consists of various methods used to facilitate communication skills as without speech per se. These can easily be described as gestures, signs, facial expressions, and writing used in everyday life. Advantages and drawbacks of some AAC systems as a therapy approach for aphasic patients should be considered.

For example, pictures and boards are used so that the patient can point to a specific need without having to use speech at all. Picture boards are limited to the needs that are shown on the board, so it is somewhat limited in its flexibility. This form of communication approach is still beneficial in a patient with severe language limitations. Drawing is another useful alternative communication tool.

Gesture systems, keyboards, word/picture communication books/boards, speech-generating devices, and communicating partner techniques are used in addition to or instead of speech (Figure 12.4). Specialized gestures; sign language; Morse code; communication aids such as charts, bracelets, and language boards, which might also consist of pictures, drawings, letters, words, sentences, special symbols, or a combination of these are also used.

Figure 12.4. Some Augmentative and Alternative Communication devices.

AAC is particularly useful in cases of global aphasia, when limited linguistic resources are available.

Summary

Research in aphasia rehabilitation has significantly grown during the last decades; today, a variety of techniques and procedures are available to the clinician to treat aphasia patients. The starting point for any rehabilitation program is a good language assessment, in order to pinpoint the specific language components impaired in the patient.

Different specific components of the lexicon can be distinguished (auditory analysis, word identification, semantics associations, etc.). However, many components and/or processes are generally paired in the same patient. Depending on the specific level that is impaired, the rehabilitation strategy to be used will vary. Agrammatism and sentence level disorders are difficult to treat; however, research has demonstrated that regardless of the difficulties in treating agrammatism, different therapeutic strategies have proven to be useful in recovering grammar and sentence production. In global aphasia, therapy aims to improve an individual's ability to communicate by helping the individual to utilize remaining abilities, to restore language abilities as much as possible, to compensate for language problems, and to learn other methods of communicating.

An assortment of language rehabilitation techniques and strategies have been proposed, such the Stimulus facilitation technique, the deblocking, the reorganization of the functional system, the so-called Melodic Intonation Therapy, and the use of Augmentative and Alternative Communication .

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