

Aphasia

Introduction

The term “aphasia” refers to an acquired disorder of language function caused by brain damage. It was introduced in 1864 by Trousseau to replace the term “aphemia”, used in the classical 1861 description of the syndrome by Broca. However, the term “aphemia” continues to be used, referring usually to severely impaired, non-fluent speech output, considered by some as an independent phenomenon, by others as a form of aphasia. Some authors prefer the term “dysphasia” to “aphasia”, to stress that the syndrome is usually associated with a dysfunction rather than a complete loss of language abilities. Aphasia is often accompanied by disorders of reading and writing. A disorder of reading is referred to as “alexia” or “dyslexia” (some authors use the first term for an acquired, the second for a developmental disorder). A disorder of writing can be described as “agraphia” or “dysgraphia”.

Aphasia can be distinguished on the one hand from peripheral disorders of speech and articulation, such as dysarthria, speech ataxia and speech apraxia, on the other hand from other central deficits affecting memory, attention or executive function. The distinction is, however, not always straightforward. Firstly, different deficits can co-occur, e.g. aphasia and dysarthria. Secondly, given the close interaction between language and other cognitive functions, the same clinical picture, such as semantic dementia, can be interpreted either as a form of aphasia or as a semantic memory disorder with linguistic ramifications. Another problematic distinction is that between aphasia and language abnormalities seen in psychiatric patients, such as “schizophasia”. It is still a matter of considerable debate to what extent abstract thinking can be independent of language and hence, not affected by aphasia.

History

Aphasia is one of the earliest documented neurological disorders and has played a central role in advancing our knowledge of brain function. A description of a language disorder following an injury to the temple by the Egyptian surgeon Imhotep (2800 BC) is possibly the earliest recorded case of a localised brain lesion causing a neurological deficit. Descriptions of a sudden loss of language can also be found in the Bible as well as in the medical treatises of ancient Greece and Rome. The observation of dissociations between different aspects of lan-

guage (e.g. loss of speech with preserved singing, selective noun and verb impairment) in the 18th century prepared the ground for the idea of the dissociability of mental faculties, which, in the next century, led to the first elaborated models of brain function based on clinico-pathological correlation studies. In the 20th century the study of aphasia influenced the development of many scientific theories, from the modularity of mind to connectionist modelling and it continues to play an important role in the current debates on innateness, functional specialisation, neuronal plasticity.

Etiology

By far the most common cause of aphasia is stroke and the majority of studies of aphasia have been conducted in stroke patients.^{1,2} Other common causes include space occupying lesions and head injury. Aphasia also occurs in neurodegenerative diseases, such as Alzheimer’s disease (AD), frontal (fv) and semantic dementia (SD) variants of Frontotemporal Dementia (FTD), Parkinson’s disease (PD) and Huntington’s disease (HD). It is the defining characteristic of Primary Progressive Aphasia (PPA) and can be a prominent and even presenting feature in diseases traditionally considered to affect mainly the motor system, such as Corticobasal Degeneration (CBD), Progressive Supranuclear Palsy (PSP) and Motor Neurone Disease (MND).³

Classification

Based on the observation of different forms of aphasia Wernicke and Lichtheim constructed what was to become the most influential model in cognitive neurology. It assumed the existence of two language centres, a sensory and a motor one, connected by a direct and an indirect pathway, the latter connecting both centres with a putative conceptual centre. The model predicted different forms of aphasia on the basis of lesion location (Table 1). Broca’s and Wernicke’s aphasia were due to direct damage of the motor and sensory centre respectively. A disruption of the direct pathway between the motor and the sensory centre lead to conduction aphasia, while a disconnection between either of them and the conceptual centre produced transcortical-motor and transcortical-sensory aphasia. Extensive damage to different parts of the system resulted in global aphasia, affecting all aspects of language.



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Table 1: The classical forms of aphasia

Diagnosis:	Main characteristics:	Associated deficits:	Localisation (all left hemisphere):
Broca’s aphasia	slow, non-fluent, laboured speech, telegraphic speech, agrammatism*	right-sided hemiparesis, dysarthria	medial cerebral artery territory (anterior branch)
Wernicke’s aphasia	fluent speech, paragrammatism** paraphasias, comprehension deficit	right-sided hemianopia, dyslexia	medial cerebral artery territory (posterior branch)
Conduction aphasia	fluent speech with phonemic paraphasias good comprehension, repetition deficit	right-sided hemisensory deficits	arcuate fasciculus (inferior parietal lobe) supramarginal gyrus
Transcortical-motor aphasia	reduced, dysarthric speech output preserved comprehension & repetition	mutism	frontal lobes (supplementary motor area) deep subcortical
Transcortical-sensory aphasia	fluent, semantic paraphasias, impaired comprehension, preserved repetition	variable	typically temporo-parietal junction, but also anterior perisylvian and deep subcortical areas

* agrammatism – loss of grammatical morphemes (e.g. the third person marker “s” in words like “he runs”) and function words (e.g. “at”, “also”, “because”).

** paragrammatism - blending of different sentence structures.

Table 2: Aphasia examination

What to test?	How to test?	What to look for?
Spontaneous speech	asking the patient questions (e.g. the history of the disease etc.), asking to describe a picture	decreased fluency and speed of speech, dysprosody, slurred articulation semantic and phonemic paraphasias, self-correction, conduit d'approche*
Naming	naming of real objects, pictures, drawings naming to description (e.g. What do you call an African animal with a very long neck?)	word finding difficulty, semantic and phonemic naming paraphasias, substitutions, use of superordinate category ("animal" for "giraffe"), neologisms, circumlocutions, perseverations
Repetition	asking the patient to repeat words and sentences	paraphasias, incomplete sentences, breaking up in the middle
Comprehension	asking the patient to answer questions, perform actions or point to objects, people or pictures	failure to follow commands, incorrect answers, performing incorrect actions or performing correct actions in a wrong order
Sentence completion	asking the patient to complete a phrase (e.g. he posted a letter without a...)	facilitates speech production in patients with dynamic aphasia, particularly in sentences with only one plausible answer (... stamp)
Definition	asking the patient to define a concept (e.g. what is an elephant? a violin?)	particularly sensitive to deficits in semantic knowledge, e.g. in patients with semantic dementia with otherwise fluent and grammatically correct speech
Phonological processing	asking the patient to judge whether two words start with the same sound, if they rhyme etc.	a useful way to distinguish central phonological deficits (inability to process phonological information) from pure speech output deficits (e.g. dysarthria)

* repeated attempts to articulate the word, getting closer and closer to the target

It is a remarkable achievement of Wernicke and Lichtheim that the terms they introduced are still widely used today^{4,1} and that the basic assumption of their model, that of a direct and indirect route between the two language areas, has been recently confirmed using the modern technique of tractography.⁵ However, despite the use of the same terms different authors interpret the basic syndromes in very different ways. The agrammatism of Broca's aphasia, a phenomenon which has attracted particular attention of researchers, has been interpreted among others as a selective loss of a particular syntactic module, a breakdown of automatic speech processing, an impairment of rule-based grammar (as opposed to memory-based lexicon) or a compensation strategy. Each interpretation focuses on a particular aspect of the phenomenon but none succeeds in capturing the full range of the observed symptoms. To minimise the theoretical bias many aphasiologists prefer, therefore, to divide aphasia into two broad, descriptive categories: "non-fluent" vs. "fluent".

Since the traditional classification of aphasia is based mainly on the study of chronic stroke patients⁶ it is less appropriate for describing acute aphasia, aphasia recovery or aphasias of non-vascular origin. Anomic aphasia is a relatively pure word-finding difficulty, seen in recovery from stroke as well as in neurodegeneration. Dynamic aphasia, originally described by Luria in patients with traumatic brain injury, is also observed in patients with tumours and neurodegenerative processes affecting the frontal lobes. Patients with this form of aphasia have difficulty producing spontaneous speech but are much better on constrained tasks such as naming or sentence completion.

The most extreme form of fluent aphasia is jargon aphasia, in which the speech production is characterised by frequent paraphasias, neolo-

gisms, perseverations, non-sense syllables and different words run together into one. This form of aphasia has been observed in patients with subcortical lesions. Despite its dramatic appearance it can be associated with a good recovery.

Localisation

Although generally speaking anterior lesions tend to produce a non-fluent, posterior lesions a fluent aphasia, Broca's and Wernicke's aphasia cannot always be identified with lesions to Broca's (posterior inferior frontal gyrus) and Wernicke's (superior temporal gyrus) area respectively.⁷ Broca's aphasia has been observed in patients with intact Broca's area while lesions in Broca's area do not automatically lead to Broca's aphasia. The same applies to Wernicke's aphasia and area. Many other brain regions have been implicated in different forms of aphasia: inferior parietal cortex and supra-marginal gyrus (BA 40) have been implicated in conduction aphasia, angular gyrus (BA 39) in written language, supplementary motor cortex (BA 6) in transcortical motor aphasia, the region anterior to Broca's area (BA 45) in dynamic aphasia, anterior insula in non-fluent progressive aphasia.⁸ The role of the right hemisphere, cerebellum and basal ganglia in the pathogenesis of aphasia are topics of intense debate.⁹

Aphasia – a language specific phenomenon?

The majority of studies of aphasia has been conducted in a comparatively small number of related languages such as German, French and English. It is questionable, however, that the observations made in a particular language can be generalised to others.¹⁰ The use of free stems (words without grammatical endings), for example, considered to be a hallmark of agrammatism in English, does not occur to the same

extent in highly inflected languages such as Greek or Polish. Language-related differences are of particular importance in multilingual patients. The same aphasic process can affect differently the different languages spoken by the same person. The native tongue is often better preserved than languages learned later in life, but the opposite pattern has also been reported.¹¹ A new and fascinating area of cross-linguistic research is aphasia in sign language, where clinical pictures analogous to Broca's and Wernicke's aphasia have been described.¹²

Assessment

A brief conversation with a patient can provide a lot of valuable information about the language function: speech rate (fluent/non-fluent distinction), pronunciation (articulation, phonology), choice of words (semantics), use of past and present tense (syntax), self-correction of aphasic errors (awareness of the deficit). Table 2 lists simple tests that can easily be performed at the patient's bedside. Table 3 gives examples of some standardised tests for a more comprehensive aphasia assessment. The individual test batteries differ greatly in their length, composition and focus and the choice of an appropriate test depends on the relevant questions. Diagnostic tests (BDAE, WAB, CAT) provide a measure of the severity of aphasia and classify the patients into the main syndromes. They can be useful in the initial assessment and help to choose the right treatment strategy. The tests of communicative abilities (PICA, ANELT), in contrast, focus on practical communication skills, which reflect not only the underlying linguistic deficit but also the compensation strategies as well as an interaction with motor, cognitive and behavioural symptoms, such as depression, apraxia or anosognosia. They are particularly useful in assessing the results of aphasia treatment.

Table 3: Examples of standardised aphasia tests (BDAE, WAB, CAT), a comprehensive test battery (PALPA), tests of communicative abilities (PICA, ANELT), aphasia screening test (TT), and tests of specific language functions such as comprehension (TROG) and semantic processing (PPT, KDT).

Name of the test & its authors:	Structure:	Scope:
Boston Diagnostic Aphasia Examination (BDAE) H. Goodglass & E. Kaplan	34 subtests examining different components of language as well as aspects of perception and problem solving	well known and comprehensive but also long (180 mins) provides a profile of impairment and severity rating
Western Aphasia Battery (WAB) A. Kertesz	contains 10 subtests examining different aspects of language, such as naming, repetition, comprehension	shorter than BDAE (60 mins), determines severity and allows taxonomic categorisation into the main syndromes
Comprehensive Aphasia Test (CAT) K. Swinburn, G. Porter, D. Howard	34 subtests, including a cognitive screening, a language battery (21 subtests) and a disability questionnaire	a new test designed to be completed in 90-120 mins can also be used in the early stages of aphasia
Psycholinguistic Assessment of Language Processing (PALPA) Kay et al	a selection of different individual aphasia tests e.g. phonological processing, reading, writing etc.	individual subtests can be selected and used separately depending on the specific questions in each case
Porch Index of Communicative Ability (PICA), B.E. Porch	18 verbal, gestural and graphic subtests, using 10 common items (e.g. pen, key); 16 point scoring system	a sensitive measure of the ability to communicate and of changes in performance, useful in monitoring treatment
Amsterdam-Nijmegen Everyday Language Test (ANELT), Blomert et al	10 situations in which the patient has to demonstrate practical communication skills, 5 point rating scale	ecologically valid but the rating can be difficult frequently used to evaluate aphasia treatment
Token Test (TT) E. De Renzi & L. Vignolo	a set of geometric tokens of different size and colour patient asked to perform actions (e.g. touch the squares)	used for aphasia screening, quantitative but not qualitative influenced by motor symptoms e.g. apraxia, bradykinesia
Test of the Reception of Grammar (TROG) D. Bishop	20 blocks examining syntactic structures of growing complexity, from single words to embedded phrases	qualitative assessment of comprehension (15-20 mins) applied in language acquisition as well as in aphasia
Pyramids & Palmtrees Test (PPT) D. Howard & K. Patterson	52 pages with triplets of pictures depicting objects and the same number of pages with corresponding words	examines separately picture and word association, assesses verbal and non-verbal semantic knowledge
Kissing and Dancing Test (KDT) T. Bak & J. Hodges	an extension of the PPT, containing the same number of pictures/words in the same format, but depicting actions	assesses specifically the knowledge of actions/verbs, which can dissociate from that of objects/nouns

One of the most important yet often neglected parts of language examination is the assessment of comprehension. The extent of comprehension deficit is easily underestimated, particularly if the patient has a chance to rely on non-verbal cues, the context or the mimic and gestures of the examiner. Many commands widely used to test comprehension, such as “close your eyes” require only understanding of a single word: there is not much more that the patient can do with his/her eyes than to close them. Commands and questions used to test comprehension should be, therefore, more complex and less obvious, e.g. “point to the window after touching the bed” or “how many people in this room are not standing?” There is also the opposite danger: to underestimate the patient’s communicative ability. Patients with MND-associated aphasia might still be able to write full sentences at a time when they are virtually mute. Patients with dynamic aphasia or bradykinesia might have little spontaneous speech and be extremely slow in answering open questions. Giving the right tasks, however, such as sentence completion or object naming, one might exhibit a remarkably intact language function.

Treatment and Recovery

The growing recognition of neural plasticity and the functional reorganisation of the brain after injury has had profound impact on our understanding of aphasia. Aphasic syndromes believed to result from specific damage to well-defined parts of the language system became reinterpreted as compensation strategies. These

theoretical developments have also increased the interest in aphasia therapy. Some treatments, like constraint-induced aphasia therapy, build explicitly on the experiences of physiotherapy in the rehabilitation of stroke patients. Others make use of the facilitation of language production through association with singing or chanting (Melodic Intonation Therapy MIT) or focus on practical strategies to cope with everyday requirements (Promoting aphasic’s communicative effectiveness PACE). In many cases the treatment combines elements from different techniques to adjust to the needs of an individual patient. Despite practical difficulties in evaluating aphasia therapy, a number of meta-analyses could establish their utility in aphasia treatment.¹³ Transcranial magnetic stimulation (TMS) has been proposed as a complementary treatment, based on the assumption that the overactivation of the right hemisphere constitutes a maladaptive strategy interfering with recovery.¹⁴ Growing experience with the pharmacotherapy of aphasia suggests that the efficacy of each drug (Piracetam, Dopamine, Bromocriptine, Dexamfetamine, Donepezil) might depend on the stage of aphasia (acute vs. chronic) as well as on its type (fluent vs. non-fluent).²

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