

Panel: Unusual Aphasias: Some Criteria for
Evaluating Case Studies in Aphasiology

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Broca's brain is interesting. Tan Tan, nee Leborgne's, brain as described by Broca is and remains (1984) an important influence on aphasia and on aphasiology's notions about the neural substrates of language comprehension and use. Similarly the myriad case studies published in the modern aphasiology literature differ in their worth. All, however, are worth something.

Case studies are descriptions. As such, they lack many of the traditional experimental controls. Their preparation is reasonably simple, assuming the clinical researcher is a good observer and a careful recorder, but their publication is increasingly difficult. It may even be that Broca could not publish LeBorgne's description today. Nonetheless, even with the threat of rejection (and the rejection of Broca's report would have been a travesty) the writer's job is easier than the reader's. Writers get excited by certain patients and believe publication of their cases can be important. Readers, on the other hand, must decide how much to be excited by, and believe in, published cases.

This short discussion's purpose, coming as it does after three case studies, is to propose some criteria for evaluating these and other case studies. The criteria are limited to those which might guide evaluation of reports whose purpose it is to describe unusual aphasias. In addition, the criteria are being described, not prescribed.

A Definition of Aphasia. A definition of aphasia strengthens a case study whether or not the topic is "usual aphasias," but is especially critical when notions about aphasic symptoms or neural substrates are being addressed. Without a definition, it is impossible to know what the writer means by "aphasic patient," and assumptions about what might be meant can be dangerous. Benson (1979) defines aphasia as "loss or impairment of language caused by brain damage" (p. 1). His definition hardly prepares one for his book, Aphasia, Alexia, and Agraphia (1979), a title which suggests that a language deficit present in writing and reading is different from a language deficit present in verbal expression and comprehension. The appeal for a definition of aphasia as part of case studies is pretty bland stuff. More interesting is the creation or selection of the specific definition to be featured.

McNeil (1984) suggests that a definition of aphasia must address five issues: unimodality versus multimodality, performance versus competence, verbal versus nonverbal, cortical damage versus subcortical damage and unidimensional versus multidimensional. His discussion of these issues should be read in the original, but one can get the flavor of what he is requesting by reading his own definition.

Aphasia is a multimodality physiological inefficiency with greater than loss of verbal symbolic manipulations (e.g., association, storage, retrieval, and rule implementation). In isolated form it is caused by focal damage to cortical and/or subcortical structures of the hemisphere(s) dominant for such symbolic manipulations. It is affected by and affects other physiological, information processing, and cognitive processes to the degree that they support, interact with, or are supported by the verbal symbolic deficits (p. 129).

His definition addresses the issues as does Darley's (1982) widely disseminated definition of aphasia as:

Impairment, as a result of brain damage, of the capacity for interpretation and formulation of language symbols; multimodality loss or reduction in efficiency of the ability to decode and encode conventional meaningful linguistic elements (morphemes and larger syntactic units); disproportionate to impairment of other intellectual functions; not attributable to dementia, confusion, sensory loss, or motor dysfunction; and manifested in reduced availability of vocabulary, reduced efficiency in application of syntactic rules, reduced auditory retention span, and impaired efficiency in input and output channel selection (p. 42).

McNeil presciently insists that modern definitions reflect the writer's attitude about the relationship of language comprehension and use to cognitive processes (or to other cognitive processes, if one believes that language comprehension and use are themselves cognitive processes). Traditionally, aphasiologists of one persuasion (Darley, 1982) have been content to distinguish aphasia from the language of dementia. Those of another persuasion (Appell, Kertesz, and Fisman, 1982) use the traditional aphasia types as labels for the speech and language deficits of dementing illnesses. Probably both of these persuasions simplify the issues.

Nowhere is the need for a definition that includes a statement about the relationship of cognitive processes and language comprehension and use more crucial than in discussions of unusual aphasias such as those we have heard today. Patients with crossed aphasia also have deficits in attention, arousal, and visual-spatial processing. Patients with subcortical aphasia also have cognitive deficits, some of which persist (Fromm, Holland, Swindell, and Reinmuth, 1985). The issue in slowly progressive aphasia is perhaps the most trenchant of all.

Probably because of the complexity, it has become fashionable to rely on aphasia's smell for a differential diagnosis and perhaps even as the basis for identifying an "unusual aphasia." If it does not smell like aphasia, it is not aphasia. Aphasiology's science is threatened by a reliance upon odor. We have not proven that paraphasias smell sweet to everyone or even that everyone can detect their odor. Indeed the flap over whether or when aphasia exists in dementia suggests that some of us wrinkle our noses at quite different things.

Case studies in aphasiology which begin with a definition -- or at least reference to a definition -- are more likely to be informative and less likely to be merely interesting. The appeal is not for a common definition but for the common use of some definition, perhaps even a statistical one.

Lesion Information. In aphasiology's early days, lesion data were rare and the validity and reliability of the data that existed were nearly impossible to judge. A tradition of providing either no or only very general lesion data has continued, at least in some reports, into the present. A few would say that even the elegant lesion data now available from magnetic resonance imaging (MRI) and positron emission tomography (PET) are of little relevance to most studies in aphasia. Recent data (Metter and Hanson, 1985) suggest that quite the opposite may be true. However, the argument is not that to use sophisticated methods is the best science and to know the locus is to know the patient. The argument is that a reader's confidence can be enhanced by data on etiology, locus, extent, and number of lesions. Consider

these examples. A diagnosis of thalamic aphasia is more convincing if the etiology is embolus rather than hemorrhage (Mohr, 1983). The conclusion that a patient has only one lesion in the distribution of the parietal branch of the left middle cerebral artery is more convincing if the data are provided by MRI rather than CT and if the neurodiagnostician's reliability has been confirmed statistically. This is not to say that CT is bad, that MRI is good, and that radiologists are unreliable. It is to say that important cases deserve the most sophisticated study possible and that clinical examinations may differ substantially from research examinations. Finally, the conclusion that a patient has a subcortical aphasia is more credible if angiography (see Weinrich, this volume) and PET rather than CT or even MRI alone are provided.

This criterion of complete lesion information is not meant to be exclusionary. Informative cases can as easily come from the worst-funded as from the best-funded centers. Its rather more democratic purposes are 1) to remind clinical researchers that interesting cases are more likely to preserve the traditional and the usual than to proclaim the new and the unusual and 2) to remind writers to give readers more than just their word.

Time Post Onset. Time post onset's importance in interpreting aphasia treatment data is common knowledge. It seems, however, that cases reported for other than treatment reasons are studied whenever they come into the reporter's view or whenever the reporter is ready to launch a project. Time post onset is always potentially important, and it is a variable best not left to chance.

Aphasia usually changes over time. For example, the majority of patients begin with global aphasia. Many evolve into or through other types -- Broca's, apraxia of speech, and so on. Even when the type of aphasia does not change, performance in one or more of the modalities may. Globally aphasic people begin comprehending more, for example, despite their near total speechlessness. Rubens (1976) describes some reasons for these changes. It is not only the aphasia that changes. Aphasic people change as well. Chronic aphasic people do not communicate the way they do merely because their lesions offer no other choices. In other words, they do not merely have their aphasia -- they adjust, develop strategies, and sometimes even give up.

As a result both of changes in the aphasia and in the aphasic person, case studies at different times post onset make different contributions to aphasiology's data base. We are not yet smart enough to know what questions to ask of what patients. Writers strengthen their reports, however, if they add a section outlining their attitudes about duration's influence on their reason for presenting a case.

Risk Factors at Onset. Case studies are enhanced by systematic reporting of each patient's risk factors -- e.g., smoking, drinking, obesity, diabetes, high blood pressure. Data suggest that risk factors may be associated with differences among "normal" speakers on brain image testing. Similarly, aphasic patients may present or evolve differently depending on the number of their risk factors. Few otherwise healthy people suddenly have a stroke and aphasia. It may be that these are the patients who do especially well with (and even without) treatment. Perhaps certain conditions such as recovery from a total left middle cerebral artery lesion stroke are rare because people with no risk factors are rare.

It may be that each patient's medical past is as important as the medical present. Therefore, a review of risk factors as part of patient

description seems a good idea. Some unusual aphasias may actually be quite usual if one knows each patient's risk profile. It may eventually evolve that patients, especially stroke patients, can be assigned a clinically relevant number reflecting their risk status. If clinical aphasiologists are careful to attend to their patient's risk factors, they may contribute to such an index.

Additional Medical History. Aphasiologists are often less conscientious than their medical colleagues about reporting the medical histories (including the previously discussed risk factors) of patients they describe in the literature. Aspects of the medical history, especially if that history has been collected by a competent practitioner with publication in mind, enhances a case report. Obviously the medical history of many aphasic people could easily overwhelm the speech, language, cognitive, or other data which are frequently a case study's major focus. Certain kinds of medical data seem crucial, however.

The presence, severity, duration, and response to medication of infections, metabolic disturbances, and seizures, are among potential influences on language comprehension and use. Seizures and the medications to treat them are of special interest. Type, locus, and duration of seizures, type of medication, adequacy of seizure control, and the relationship of seizure activity to the timing of speech and language testing may all be especially critical (Lesser, Lüders, Wyllie, Dinner, and Morris, 1986).

Until aphasiologists know more about all the influences on language comprehension and use, it may be advisable to report more rather than less medical data. Or at very least, it may be wise to have the data available upon request or in an Appendix. Aphasia is a bit like fever. It is measurable. It is also only part of a patient's disease, and often not the most important part, even for the aphasiologist.

Complete, Standardized Speech, Language, and Cognitive Testing. Modest clinical research aims can nearly always be supported by case reports. A liberal interpretation of what modest means, however, has permitted a hopeless ruck of cases that are described with incomplete, informal testing. The reader's burden when reading such cases is to decide if the author is trustworthy. All aphasiologists know other aphasiologists whose work is to be trusted. A label -- severe, Broca's aphasia, for example, or a label and brief summary of informal observations -- may be enough from such a colleague, at least for some purposes. However, we do not all trust the same professionals, even trustworthy people make mistakes, and some of this generation's findings may outlast their authors and even their author's reputations. Standardized test batteries, supplemented by completely described informal measures, are best. Like Broca's LaBorgne, some modern patients may be remembered or resurrected by future practitioners. It behooves us to provide more than a bare-bones description.

Finale. A complication of using single cases to influence definitions and notions about language's neural substrates is that as Ojemann and Whitaker (1978) say, brains can be as unique as faces. Another complication is that centers from which cases emerge have different sensitivities and capabilities. Nonetheless, aphasiologists can control the care with which each case is studied and presented. They can provide the reader with a definition of aphasia and with the criteria that behavior must meet to be called aphasic. They can identify, describe, and sometimes even quantify all potential influences upon each patient's language comprehension and use. They can provide complete, formal speech and language data. Cases prepared

in this way allow the reader to decide if it is the aphasia or the aphasic person that is unusual -- whether the person's nose is really above his eyes or if he is merely doing a handstand.

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