Acquired Aphasia in Children
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Acquired childhood aphasia is rare but has important conceptual implications for developmental neuropsychology. The last 15 years have seen major changes in their clinical description, which have led to the awareness that the syndromes in acquired childhood aphasia are more similar to the syndromes in adult aphasia than previously thought. This article briefly discusses the definition and differential diagnosis of acquired childhood aphasia from the point of view of the child neurologist and adds new perspectives afforded by neurolinguistic examinations. It reviews the main causes and syndromes of acquired childhood aphasia. Prognosis is less favorable than usually supposed, in terms of both language sequellae and academic failure. Finally, suggestions regarding the basis for aphasic children's nonverbal deficiencies are presented.

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Acquired Childhood aphasia (ACA) refers to the language deficits that may follow a brain lesion sustained after the age of acquisition of first sentences. Although it is rare, compared with the prevalence of developmental language disorders and of aphasias in the adults, ACA has major implications for pediatric neuropsychology. Since their first descriptions, the language consequences of brain lesions acquired in childhood were placed in the overall context of the current views on the functional maturation of the brain. They turned out to provide the very foundations of our current understanding of brain-behavior relationships in children. We have moved away from Lenneberg’s 1967 theories, which proposed that lateralization of language to the left hemisphere was progressive, and from the idea of a uniform symptoms irrespective of the localization of the lesion. The clinical diversity of ACAs and of their lesional counterparts makes it possible to reason by analogy, drawing from the many varieties of acquired aphasia in adults; it points to greater similarity than previously imagined in language organization in the child, compared with the adult brain.

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from aphasia, with which it is still often confused, because mutism often precedes the emergence of the aphasia.

**Differential Diagnosis**

There are several items to be considered in the differential diagnosis of ACA:

- **Acquired dysarthria** is where the sounds of speech are distorted by a subcortical or peripheral lesion, for instance in cortical dysarthria, bulbar palsy, or peripheral facial palsy. Phonic errors are less variable than in aphasia, and there are other neurological signs of orofacial involvement.

- **Acquired stuttering** occurs in some aphasic children, especially those who have word-finding difficulties, often with autocorrections in which the repetition of syllables must be differentiated from syllabic iterations caused by troubles with the speech effectors. The scores for naming to confrontation is impaired in the first condition.

- **Congenital aphasia owing to prenatal or perinatal lesions.** Although unilateral brain damage may be responsible for abnormal development of babbling and early language (see also Nass’s article elsewhere in this issue), for example, subtle metalinguistic problems later on, the lesion has to be bilateral and encroach on subcortical areas for the language disorder to be severe and persistent. In such severe cases, recovery is very slow and limited.

- **Developmental dysphasia:** in this often familial group of conditions, language is usually deviant from the start, although autism and autistic-like conditions may present as language deterioration following a “free interval” of normal acquisition of language milestones (see Tuchman’s article elsewhere in this issue).

- **Mutism:** “true mutism” usually results from large frontal or midbrain lesions or from diffuse axonal shearing. Unlike “aphasic mutism,” which has similarities with hypopontaneity of speech, for example, lack of spontaneous speech production, true mutism differs from it because it is not possible to alleviate it by stimulation.

**Neurolinguistic Testing**

Before the end of the 1970s, the usual practice was to test aphasic children with tests devised for adults. Many of the adult tests have since been standardized for children of different ages, and some new tests, more specifically aimed at the assessment of child language, have been developed. Comprehensive batteries enable one to evaluate the different aspects of language (such as fluency, naming, auditory comprehension, repetition, development of syntax, and articulatory agility). These batteries enable the categorization of each case of ACA on the basis of an overall language profile identifying the specific language abilities affected, as well as those spared. A detailed analysis of this type can contribute to the intervention plan, which usually aims at enhancing deficient abilities and reinforcing stronger ones.

Provided scores are expressed in standard deviations from the mean, availability of standardized tests also provides the opportunity to make comparisons among the profiles of children of different ages. This is useful in view of the usually small number of cases of children with ACA in any particular series.

In addition to generating an overall language profile, one also needs to classify the specific “positive symptoms” of aphasia. These include the following:

- **paraphasias:** substitutions of words (verbal paraphasias) or of phonemes (phonemic paraphasias)
- **perseverations:** providing the same answer in answer to different questions or stimuli
- **echolalia:** compulsive immediate or delayed repetition of heard words or sentences
- **neologisms:** such marked distortion of words by well-articulated phonemes as to render target words unintelligible
- **jargon:** production of unintelligible speech because of a large number of neologisms

It may also be useful to measure the length of the pauses that precedes a target word and the duration of individual words. In addition, experiments may be required to determine the source of some of the aphasic symptoms.

**ETIOLOGY**

Epileptic aphasia is not considered here because it is discussed in Tuchman’s article elsewhere in this issue. The main causes of ACA have changed drastically since the 1940s, when the leading cause was vascular, whereas at present the major cause is craniocerebral trauma. According to Woods and Teuber, before the advent of antibiotic therapy
and proper rehydration, the vascular predominance resulted from the high frequency of encephalopathic complications of systemic infections and electrolyte disturbances, which were often responsible for cerebrovascular thrombosis. Modern treatments have greatly reduced this cause.

**Vascular Lesions**

In contrast to the causes in older series, bacterial infections are no longer a major vascular cause of ACA. Currently, most ACAs arise from the complications of systemic disorders (especially hematological and cardiac conditions). The paucity of their symptoms and good prognosis are often stressed. One might speculate that it was the predominance of vascular causes in the "classic series," with their better outlook, that contributed to the standard doctrine on the favorable evolution of ACA (see below). In fact, prognosis varies considerably from series to series. In the Cranberg cohort, symptoms were severe and prognosis rather unfavorable even though the outlook was more optimistic than for adults with similar lesions and localizations.

**Trauma**

Trauma is now the chief cause of ACA, and mutism is particularly frequent with this cause. Sometimes it is preceded by coma and accompanied by overall movement reduction (akinetism). As in adults, symptoms are often mild and prognosis is favorable. What plays a bigger role than age and origin is the extent and severity of the lesion. Although mild anomic aphasias are frequent, cases of severe sensory (receptive) aphasia may also occur.

**Tumors**

Cortical hemispheric tumors are less frequent than posterior fossa tumors in young children. They too may cause a mild anemia, but specific subtypes of aphasia have been described, for example, a child with a transcortical sensory aphasia.

**Infections**

Although antibiotic therapy has eliminated some infectious causes of ACA, modern antiviral treatment has spawned new origins. For instance, herpes simplex encephalitis, which was often lethal, is now treatable but may leave sequellae in its wake. The specific tropism of herpes virus for the temporal lobes may have contributed to the inflation in the number of posterior types of ACA in the current literature.

**APHASIC SYMPTOMS AND SYNDROMES IN ACA**

It was widely believed before the late 1970s that ACA was mostly characterized by "negative symptoms," that is, mutism, dysarthria, reduction in phrase length, and telegraphic speech. Other rarer symptoms were thought to occur chiefly in children approaching puberty. Since the "unexpected" description by Woods and Teuber of a case of jargonaphasia in a 5-year-old boy who did not have Landau-Kleffner's syndrome (acquired epileptic aphasia), suggestion that the "anterior-like picture" of ACA might be artifactual and attributable to hemiparesis as criterion for determining the unilaterality of the lesion has largely been substantiated. Different symptoms thought never to occur in young children ("positive symptoms," such as logorrhea, paraphasias, perseverations, neologisms) have, in fact, been reported in every recent unselected case series. Moreover, these positive symptoms correlate with overall clinical aphasic profiles and lesion localizations similar to those of adults. Indeed, equivalent ACA syndromes to most of the aphasic syndromes of adults have been encountered.

**Sensory (Receptive) Aphasias**

The classical auditory comprehension disorder is characterized in children by fluent, well-articulated speech, often containing paraphasias either in spontaneous speech or in naming tasks. Neologisms may occur and, if frequent, gave rise to "jargonaphasia." The rhythm of speech may be accelerated, and real logorrhea with anosognosia may occur (Wernicke’s aphasia). However, in contrast to adults where the different components of fluency usually evolve in parallel, in children under 8 years old, one can observe an inverse relationship between the severity of the jargon and that of pressure of speech. The lesion is usually located in posterior brain regions, with the extent of the lesion in Wernicke’s area proportional to the severity and duration of symptoms.

**Anomic Aphasia**

Anemia refers to difficulty coming up, within an appropriate time-frame, with the appropriate name for a target, either in response to visual presentation
or verbal description. Anomia is present in almost every form of aphasia, but it owes its name to its occurrence in isolation or as the most prominent aphasic sign. Van Hout et al.\textsuperscript{17} described cases of modality-specific (tactile) or category-specific (color) anomias in children. In some cases, as in adults, this form of aphasia occurs during the recovery stage of a sensory aphasia,\textsuperscript{20} and some authors\textsuperscript{16} consider that it may represent the most persistent sign of a fluent aphasia. Anatomic correlations usually point to a parietotemporal localization, although there are descriptions of cases with frontal or parieto-occipital involvement.

**Conduction Aphasia**

In this often fluent aphasia, the “lack of words” (difficulty retrieving words) may be so pronounced that pauses and self-corrections give it the appearance of nonfluency evocative of stuttering. The main feature is dramatic impairment of repetition, with multiple phonemic substitutions at the word level and preservation of verbal comprehension. Martins and Ferro\textsuperscript{19} and Van Hout et al.\textsuperscript{17} described cases of conduction aphasia with parietal lesions. Tanabe et al.\textsuperscript{22} were able to demonstrate discrete pathology localized to the arcuate fasciculus by MRI, thus supporting the current explanation for this syndrome as a disconnection between the receptive (Wernicke’s) and the expressive (Broca’s) language areas.

**Transcortical Aphasias**

In almost direct contrast to the symptoms of conduction aphasia, this group of aphasias is characterized by striking preservation of the repetition of words or sentences, which often take on a compulsive, echolalic quality. Isolation of the language cortices from other cortical areas provides an explanation for the transcortical aphasias, with preservation of the arcuate fasciculus, which connects the anterior and posterior language areas, allowing repetition to occur. This group of aphasias can be divided into two large subgroups: the sensory transcortical aphasias and the motor transcortical aphasias. In the *sensory transcortical aphasias*, comprehension is severely disturbed and ongoing speech is fluent, often of a jargon type. Martins and Ferro\textsuperscript{11} and Van Hout et al.\textsuperscript{17} have described cases that fit this diagnosis (in the latter series there was even a 5-year-old boy shown to have a large area of hypoperfusion in the left hemisphere). In Cranberg et al.’s series,\textsuperscript{15} an adolescent boy with this syndrome presented, as do some adults, with a left subcortical lesion extending to the internal capsule. In the *motor sensory aphasias*, expression is severely reduced, to near muteness, and speech initiation is extremely slow. Cases of vascular origin have been reported by Cranberg et al.\textsuperscript{15}

**Motor (Expressive) Aphasias**

It is difficult to distinguish, in the classic literature, what have been called “motor symptoms of ACA” from true, adultlike motor aphasias. Authors writing before the 1980s have offered different interpretations for their global reduction in all speech parameters. These views reflected different points of view, some authors regarding these aphasias as “motor,” others as the result of a global “hypospontaneity” of speech. According to this latter view, mutism was considered a symptom intermediate between motor aphasia and speech hypospontaneity, with a paucity of verbal productions and failure to initiate spontaneous speech.

In the literature, articulatory disorders have been regarded either as regression to more primitive stages of speech articulation or as being equivalent to the syndrome of phonetic disintegration in adults.\textsuperscript{23} Reduction in mean length of utterances in the absence of other specific syntactic deviance has been regarded either as a return to the infantile “holophrastic stage” (single words to express a sentence) or as a form of Brocalike agrammatism.

As in the case of fluent aphasias, the recent use of neurolinguistic batteries has brought out the existence of true motor aphasias with Brocalike characteristics. Language is nonfluent, slowly and laboriously emitted. There are conspicuous syntactic errors, for example, verbs erroneously used in the infinitive form. There are also many pauses in ongoing speech, and phonemes are distorted (cortical dysarthria) owing to a reduction in and abnormalities of the articulatory movements. Many children of various ages with Brocalike clinical pictures have now been described.\textsuperscript{24,15}

**Atypical Symptoms**

Although nearly all the subtypes of aphasias that occur in adults have now been described in ACA with essentially similar lesional substrates, children may have atypical symptoms whose exploration might throw light on the brain mechanisms that
underlie language development. Their frequency might provide an explanation for some of the discrepancies between the classic symptom descriptions in ACA and the "new" ones. Some of these often overlooked symptoms consist, for example, in a dissociation between the different fluency parameters. In adults, these parameters evolve in parallel, whereas in children, at least in those under 8 years old,\(^5\) fluency may be reduced during the more acute phase of jargon aphasia, and there may be an inverse relationship between mean length of utterances and an increase in the proportion of neologisms.\(^5,17,25\) Moreover, hypospontaneity may be a permanent symptom apparent long after all other language parameters have returned to normal.

**LANGUAGE TESTS IN ACA**

Since the 1970s, comprehensive neurolinguistic batteries have been developed to probe the various aspects of language\(^{26,27}\) including the following:

- **Fluency** or the ease of speech emission as an effortless articulation also with a normal amount in the overall production rhythm. Its measures are usually calculated on a sample of free or spontaneous language (ie, for picture description or answers to simple open questions). Either the number of words or that of syllables per minute are computed or the "mlu" (mean length of utterances). It may also be useful to measure the number of pauses either before content words (their increase usually means anomia) or within the words themselves (for instance, in cases of articulation slowness).

- **Naming** is usually measured on a visual confrontation task for objects or pictures of various frequencies or after oral description. It is mandatory to disclose anomia to confront the active vocabulary to its passive counterpart (one cannot evoke an unknown word) for instance in multiple choice pointing tasks.

- **Auditory comprehension** is evaluated by pointing to pictures depicting different situations for sentences of various lengths or syntactic complexity. Realization of orders is also used particularly in younger children and "yes or no" questions may be used.

- **Repetition tests** usually consist in reproduction of a material of different length and complexity, either semantic or phonologic. The digit span is evaluated and words and nonwords of increasing syllable lengths are applied. Sentence repetition is of limited diagnostic value in the acute stages of aphasia being nearly always abnormal as a rule.

- **Articulation** may be scored for repetition or spontaneous production of various phonemes and phonemic clusters.

All too often, however, the batteries still consist of the mere standardization by age of measures devised for aphasic adults. Psycholinguistic tests that are more specifically designed to evaluate young children's language tend to be insufficiently "clean," that is, dissociable language dysfunctions may account for a same erroneous answer, in contrast to what is the case for most tests designed for adult aphasics. Nonetheless, these "adult aphasic tests" do bring out sufficient dissociations for diagnosing aphasic subtypes, which is the first step toward gaining an understanding of the way in which language has been affected. These specialized batteries tend to be insufficiently sensitive to detect subtle alterations in children who are recovering from an aphasia or to evaluate its sequellae. Among the drawbacks of standardized "adult tests" adapted to children, one may cite the widely used Token Test.\(^{28}\) This test, which is supposed to measure syntactic comprehension out of context, suffers from uncontrolled variations in its load on memory and attention factors. Indeed, the Token Test may not be adequately discriminatory in view of the fact that scores are lowered in sensory aphasias as well as other aphasic subtypes. Moreover, recovered aphasic children may still earn abnormal scores in the long term.\(^{29}\)

**PROGNOSIS AND RECOVERY**

According to the authors of the classic series, language recovery in ACA is usually "rapid and complete." This statement has been widely attributed to Lenneberg.\(^2\) However, going back to the source, one notes that what Lenneberg said was that the younger the child the longer the recovery process might take, a view that is quite the opposite of the one generally attributed to him.\(^3\) Yet in papers that appeared before the early 1980s, one finds many statements about the relatively benign nature of aphasia in younger children, particularly in traumatic cases. When they reviewed the prognosis of ACA in series dating before 1978, Satz and Bullard-Bates\(^{30}\) reported in 1981 that, in fact, 25% to 50% of children still had aphasic symptoms 1 year after onset. In the series by Van Hout et al,\(^{17}\)
residual "positive symptoms" were still detectable in the severe group more than 2 years after onset, and Woods and Carey,29 applying finely tuned linguistic (metalinguistic) tests to formerly aphasic children, were able to discriminate them on the basis of persisting deficits from brain-lesioned children who had not sustained aphasia.

Moreover, even if language returns to normal, nonlinguistic abilities are usually affected, and as a rule, the children encounter academic problems whose occurrence does not seem to depend on age or the cause of the lesion.31 Two interpretations have been proposed to try to explain those "secondary symptoms."3 One invokes the "crowding effect," according to which the transfer of language to right-brain areas not normally devoted to language takes the "place" of some nonverbal functions, which decline as a result. The other invokes the "price of recovery": homolateral or heterolateral areas that take over the deficient language functions are usually immature zones whose own function has not yet fully developed. Their involvement in the elaborate functions of language jeopardizes what would have been their own functional development, with the result that nonverbal later-developing functions are sacrificed in favor of the verbal ones.

There is a third possibility, which has rarely been invoked. Perhaps the insult responsible for the left brain lesion that caused the aphasia also damaged areas involved in nonverbal functions. Unfortunately, these nonverbal functions have been investigated less well than the language disorders. These other lesions, in more immature cortex, would produce less spectacular acute deficits than those responsible for the aphasia, and might give rise to dyslexia, dysorthographia, or dyscalculia, deficits that would go undetected until the child goes to school. Thus, the supposed "price to pay" for language recovery might well depend on undetected concomitant direct brain effects of the lesion.

CONCLUSIONS

The last decade has witnessed major conceptual changes in our understanding of ACA. In fact, every earlier statement regarding its symptoms and their evolution has been successfully challenged. In particular, all aphasic subtypes described in adults, thought never to occur in young children, are now known to occur. Their existence points to early specification of the neural basis of language and helps establish the foundations for the study of brain-behavior relationships. However, many unexplained areas remain, for instance, regarding atypical symptoms, cognitive explanatory mechanisms, and the role of the lesion itself or of brain reorganization in the "price of recovery."

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