Editorial

Acquired Aphasia in Children

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There are few disorders in child neurology, even in medicine, about which thinking has changed more drastically over the last quarter century than acquired aphasia in children. The thinking at the time is exemplified by Lenneberg's 1967 seminal book *Biological Foundations of Language*,[1] which contrasts childhood acquired aphasia with aphasia in adults. Lenneberg summarized the literature and, based on his review of 17 adequate reports from the literature and eight personal cases, divided children into four groups. He stated that when a focal brain lesion is sustained between ages 20 and 36 months, children recapitulate all the stages of language learning, from vocalization and babbling to single words, primitive two word phrases, etc until perfect speech is achieved, and he concluded that recovery is due to reacquisition rather than symptom reduction. He reported that between ages 3 and 4 years, aphasic symptoms last no more than a few weeks and that aphasia is invariably expressive. He stated that between 4 and 10 years of age, children evince the classic aphasic syndromes of adults, except that fluent "logorrheic" receptive aphasia does not occur; complete recovery is universal, although recovery may extend over several years, in contrast to adults, in whom recovery is minimal after 5 months. After puberty, residual symptoms, including word finding difficulty and verbal and phonemic paraphasic errors may persist, and after the midteens, all the characteristics of adult aphasia prevail.

The advent of modern neuroimaging and more rigorous neuropsychologic and psycholinguistic study of childhood acquired aphasia, together with the publication of a number of individual cases, several case series, and two books[2,3] have changed our views substantially since Lenneberg's book. I will focus here on the following issues: (1) cerebral maturation and lateralization; (2) site of lesion and age of the child; (3) cerebral plasticity and recovery from aphasia; (4) subtypes of aphasia; and (5) acquired epileptic aphasia.

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CEREBRAL MATURATION AND LATERALIZATION

The question of whether cerebral dominance for language develops progressively or is innate was a burning issue in Lenneberg's time. The then-prevalent view was that the two hemispheres are equipotential at birth and that hemispheric lateralization for language develops progressively during childhood. Two arguments in support of the hypothesis were, first, that virtually all children with a congenital or infantile hemiplegia are verbal and, it was said, acquire language at the expected age regardless of the side of the lesion, and second, that hemispherectomy, whether performed on the right or the left, does not result in aphasia, provided the original lesion was sustained in infancy.[4]

The idea of progressive maturation of cerebral dominance for language has become largely discredited on anatomic, physiologic, and behavioral evidence that was unavailable to Lenneberg. Witelson and Pallie[5] found that the asymmetry in the planum temporale described by Geschwind and Levitsky[6] in the majority of adult brains is already discernible in the neonate and, according to Wada et al,[7] in the fetus at 20 weeks of gestation. There are already demonstrable interhemispheric differences in the amplitude of auditory event-related potentials in neonates, with speech eliciting larger-amplitude responses on the left, and music doing so on the right.[8] Bertoncini et al[9] were able to show in neonates a right ear (left hemisphere) advantage for dichotically presented speech sounds and a left ear (right hemisphere) advantage for music.

SITE OF LESION AND AGE OF THE CHILD

It was also believed at the time that aphasia after right-sided lesions was much more common in children than in adults: Basser[4] reported that 47% of right hemispheric lesions sustained after age 2 years resulted in aphasia—certainly less than the 85% he reported after left-sided lesions but many more than would be the case in adults, virtually all of whom are now known to be left-brained for language if right-handed, and at least two thirds of whom are substantially left-brained for language even if left-handed. Woods and Teuber,[10] who studied 65 children aged 2 to 14 years with strictly documented nonprogressive unilateral brain lesions, noted that 25 (74%) of the 34 with left-sided lesions were aphasic, compared to four (13%) of the 31, two of them left-handed, with right-sided lesions. They attributed the earlier reports of a

higher prevalence of aphasia after right-sided lesions in children than adults to the inclusion of cases with unrecognized bilateral damage due to traumatic and infectious causes rather than to incomplete hemispheric specialization in early life.

Bates and colleagues[11] carried out systematic longitudinal studies of language development in infants with unilateral hemispheric lesions documented by imaging before age 6 months. All the infants were moderately to severely delayed in babbling, preverbal communication (eg, direction of gaze and gestures), vocabulary (single words), and syntax (sentences). From ages 12 to 18 months, delay affected both comprehension and production of speech, irrespective of the site, size, or side of the lesion, whereas between 18 and 36 months of age, expression was more severely delayed in children with left-sided lesions than right-sided lesions, especially in those posterior (rather than anterior!) left-sided lesions. In contrast, site, side, or size of lesions made no difference in the severity of comprehension deficits. A provocative finding was that the style of language acquisition differed depending on whether the lesion was on the right or the left: it was more holistic/fluent in children with right hemisphere damage, more analytic/less fluent in those with left lesions. These findings indicate that the brain regions that mediate language acquisition in early life are distinct from those involved in the overlearned processing of language in adults. It will be interesting to find out whether any of these very early lesions have long-term consequences for the acquisition of written language despite probably fully adequate oral skills.

CEREBRAL PLASTICITY AND RECOVERY FROM APHASIA

The idea in the 1960s was that recovery from aphasia in children was rapid and complete and was due to takeover by the right hemisphere because language had not yet become strongly lateralized to the left. Again, understanding has undergone substantial change. In a recent review of the literature, Satz[12] indicates that interhemispheric reorganization with switching of dominance for language to the right depends in part on age at lesioning: language switching to the right occurred in 78% of cases when the lesion was sustained before age 2 years and in only 28% when sustained after that age. In a later study Strauss et al[13] suggest that interhemispheric switching may also depend on sex, with girls limited to the first year, due to earlier brain maturation than in boys, in whom switching may occur up to puberty. Milner[14] had shown much earlier the importance of the size and location of the lesion in patients being considered for surgery for epilepsy: interhemispheric switching occurs mainly in large lesions of the major perisylvian language areas of the left hemisphere, whereas intrahemispheric reorganization is likely when damage is less extensive. When a large postnatal lesion on the left leads to the development of language on the right, the individual may show deficits not so much in language as in visuospatial skills. These have been attributed to "crowding" of activities that would normally take place in right hemisphere circuits now engaged in language operations, indicating that there is a limit to cerebral plasticity.[13] Aram and Eisele[15] reviewed some of the cellular and systems changes that underlie recovery from focal lesions in the immature brain.

Views on the completeness of recovery from childhood aphasia have also changed. Although many reviews took it for granted that, at least in early childhood, recovery is complete, in 1965 Alajouanine and Lhermitte[16] had already reported that recovery of speech is not tantamount to full recovery. Twenty-four of the 32 children in their series had recovered normal or near normal speech. However, not one of the 32 was able to attend a normal class in school, because of disorders of written language and difficulty acquiring new knowledge due to intellectual impairment not directly attributable to their language deficit. Hécaen[17] stated that reading disorders disappear rapidly and completely but confirmed the frequency of writing disorders, which he found in 63% of his cases. These observations show that there is a price to pay, at least in some cases, for the recovery of language.

SUBTYPES OF CHILDHOOD APHASIA

Another early tenet about childhood aphasia was that its symptomatology is stereotyped and predominantly expressive and dysfluent, often after a transient period of mutism and failure to respond that make it difficult to evaluate comprehension. Most early investigators relied on bedside assessments of speech, often during relatively short follow-up periods. Systematic longitudinal investigation of language, using both standardized tests and detailed linguistic analysis of recorded conversational speech, disclose an entirely different picture (reviewed by van Hout[18]). Essentially all of the aphasic syndromes observed in adults are encountered in children, albeit with different base frequencies. Brain imaging shows that correlations between type of aphasia and localization of lesions parallel those in adults. For example, the paper by Hynd et al[19] in this issue details anomic aphasia in a 10-year-old girl with a previous intracerebral hematoma of the left temporoparietal region. Even logorrhea, stated not to occur in earlier studies, is described in several case reports and was observed transiently by the author after bilateral posterior traumatic injury in a 4-year-old child with a fluent aphasia.[20]

Part of the reason for differences in the prevalence of aphasic syndromes in children and adults may have to do with the differential maturation of language circuits disclosed by the early lesion studies of Bates and colleagues[11] alluded to earlier. However, it may also have to do with the relative rarity in children, compared to adults, of strictly focal circumscribed lesions. Many more adults than children harbor the static consequences of cerebrovascular accidents; these provide the most favorable opportunity for detailed aphasiologic study and anatomic

correlation. In children, infections, trauma, and supratentorial tumors are evolving conditions unlikely to interfere selectively with one or another language subroutine. Also, in very young children language processing probably engages more widespread brain circuitry, judging from the data from the Bates group.[11]

ACQUIRED EPILEPTIC APHASIA

There is one form of acquired aphasia specific to childhood, acquired epileptic aphasia, to which Landau and Kleffner[21] drew attention in 1957. Loss of language is associated with either clinical seizures (generalized, partial, partial complex, or absence) or with an electroencephalogram showing unilateral or bilateral paroxysmal activity that is often accentuated in slow-wave sleep. This aphasia, which is rarely associated with discernable pathology on brain imaging though it may be associated with decreased temporal blood flow, differs from lesional epilepsy in that it tends to be much more long lasting. Reception is usually severely to profoundly affected, resulting in word deafness or verbal auditory agnosia when language comprehension is impaired at the stage of phonologic decoding. Because it occurs at the language-learning age, expression is also affected, often even more severely than reception. There are exceptional children with predominantly expressive deficits in whom dysfunction involves more anterior sylvian areas.[22] Contrary to lesional aphasia in children, prognosis for recovery from acquired epileptic aphasia is generally worse in children under age 5 years than in older children. This syndrome overlaps with rolandic epilepsy, with status epilepticus in slow-wave sleep, and with autistic regression and disintegrative disorder associated with unilateral or bilateral centrotemporal spike/spike-wave discharges.[23] The prognosis for seizure control and disappearance of the epileptic electroencephalographic discharges is excellent, but prognosis for recovery of language is much less predictable.[24] Because language impairment may persist for months and even years, and because it is modality-specific in that visual processing is usually unaffected, it is critical to provide these children with language through the visual channel using gestures, communication boards, signs, computers, or reading. Reversal of the aphasia in response to anticonvulsant drugs or to steroids or corticotropin is unpredictable and effective in only a minority of children.

CONCLUSION

A major knowledge gap about childhood aphasia is lack of information about cerebral reorganization in the immature lesioned brain. There is a need for many more systematic case studies of aphasic children in whom modern morphometric analysis of lesions is coupled with detailed longitudinal analysis of their language over the entire course of recovery. Application of event-related potentials and functional magnetic resonance imaging during the performance of specified verbal and nonverbal tasks is a promising approach for clarifying some of the unanswered questions about the interface between development and functional plasticity. The occasional children with acquired aphasias caused by well-defined focal lesions, especially if they result in one of the less common aphasic syndromes like the anomic aphasia described by Hynd and collaborators,[19] richly deserve full study because they provide a welcome and privileged window for learning about the range of strategies responsible for functional recovery.

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Vignette

John's Eulogy

How do you celebrate a life like John's? How do you celebrate a tragic life and death? How do you find any redeeming value to the life of a retarded boy who suffered from severe and bizarre emotional problems? How do you celebrate a life of dead expectations?

When John was born, we were ecstatic to have two bouncing twin babies. We had all the arrogant expectations of the best and the brightest. Then we learned that John was a victim of tuberous sclerosis. At first, Kathy and I hoped that John would have a mild case. We hoped that he would run and play. We hoped that he could continue to live at home with us. But over the years, gradually it became apparent that he had a full-blown case. His was one of the worst case scenarios. As John grew, so did the tumors and his dysfunction. He got progressively worse.

After 5 years of sleepless nights and 3 more years of bizarre, violent behavior, we learned that we simply could not handle John at home. And so our deepest expectation, that John could live at home, was dead. We committed John at age 8 years to a life of 5 years of institutionalization, first at a state psychiatric hospital, then at a residential school, and finally at a private hospital. Taking your 8-year-old child to the locked ward of a psychiatric facility, notwithstanding his sobbing pleas to the contrary, is like death, too.

So how do you celebrate a life like John's? They say that expectations die hard. John's life was full of expectations that died hard, but John taught us many lessons. They have certainly been expensive lessons, and we would never have voluntarily paid this dear a price, but the value of these lessons and John's life, to us, is profound.

We learned to live life after the death of our expectations. We learned to love our child, the child that we sometimes had thought would be better off dead. We learned to love our child, although he was perceived by some to have no value. We found that John had great value. We learned to accept him on his terms. We learned not to discount his life because he was handicapped.

We learned to respect John's right to live. John loved his life after he went to a group home in Richmond. He loved his freedom. He loved the freedom of living in an unlocked building. He loved to go out in public—to the malls, the movies, the Braves games, and the batting cage. He loved to go up to strangers and introduce himself. He loved to come home every Sunday for dinner. When I brought him home, he loved to burst through the door and call his mother's, sister's, and brother's names. He loved eating dinner with us and having his father give him a shave afterwards. And for the first time in 5 years, we saw John smile and laugh and sing again.

And so despite the Christmases that John spent in padded cells injuring himself and our thoughts that he would be better off dead, we learned to respect his right to live and gave him all the love and support humanly possible. We learned to respect the differences in people. We learned to respect people of limited functioning. We learned that there is life after the death of a "normal" life. We learned that, when a tragedy occurs, it redefines your life, and that your life before that tragedy is no longer possible. We learned that we had to build a new life with different expectations. We learned to develop positive responses to overcome tragedy. We learned to build a new life based on love, faith, and hope. We learned to live life a week at a time. We learned to find redeeming value in life, to appreciate the daily beauty and joys of life. And we do. We learned to be as healthy and happy as possible and to have fun and to limit our grief. And so, in the end, we got nothing that we wanted in our child, John, and everything that we could have ever needed. John gave us love that gives life a new meaning, that gives us an agenda, that gives us courage and strength, and that never leaves us alone.

John had his good days and his bad days. When his behavior was bad, he was very violent and abusive to his staff. When he had good days and his behavior was good, he always said he was having a "thumbs up" day. On the last day of his life, he sat in his bed with his shaved head, an intravenous tube in each arm, and his hands tied to the bed by restraints. When asked how he felt, John told his doctors he was having a "thumbs up" day. He told his mother, "Mom, do you know what I like?" She said, "No, John," and he said, "You!"

Today our hearts are broken, but we have learned to live with a broken heart and to enjoy life anyway. We have been inspired by John's love and positive, persevering spirit. And we will live our lives with as many "thumbs up" days as possible.

John and Katherine Conrad
Richmond, Virginia