Landau-Kleffner syndrome treated non-surgically

(continued from page 1)

Corticosteroid therapy for LKS is not new; three previous reports (two single-case studies, and one with three corticosteroid-treated children) indicated that the treatment might have potential. "It is somewhat surprising," Lerman et al. say, "that steroids have not been given a more extensive trial after the encouraging results obtained" by these studies. The researchers plan to rectify this situation, saying that a multicenter trial is being planned for Europe and Israel.

LKS usually strikes previously normal children between the ages of three and seven. Typically, children with LKS first lose their ability to understand others, and then lose the ability to speak. (The disorder is also known as "acquired epileptic aphasia;" aphasia is a term meaning loss of, or defect in, the ability to communicate.) Symptoms may appear gradually or suddenly, and frequently wax and wane.

A classic symptom of LKS is the development of abnormal epileptiform EEGs, often detectable only on extended sleep EEG recordings. Two-thirds of children with LKS develop seizures; anticonvulsant drugs usually control the seizures but have little or no effect on other symptoms.

Children with LKS may exhibit many autistic behaviors, including aggression, withdrawal, failure to make eye contact, unusual eating habits, insistence on sameness and rituals, poor sleep, hyperactivity, attention deficit, insensitivity to pain, and an appearance of being deaf to some sounds. Like autistic children, children with LKS may become completely mute or use echolalic or "telegraphic" speech consisting mostly of nouns and a few verbs, and their voices may be nasal or monotonic.

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"Effect of early corticosteroid therapy for Landau-Kleffner syndrome," Pinchas Lerman, Tally Lerman-Sagie, and Sara Kivity; *Developmental Medicine and Child Neurology*, 33, 1991, pp. 257-266. Address: Pinchas Lerman, EEG Laboratory, Beilinson Medical Centre, 49 100 Petah Tikva, Israel.

Fragile X gene

(continued from page 1)

Fragile X defect have no symptoms — and symptomless male carriers can pass the gene on to their daughters who also have no symptoms, but then have children with the syndrome. A mutation that prevents genes from "turning on" properly may cause these unusual patterns, according to Mandel et al.

Women have two X chromosomes, so women's bodies compensate for their double dose of X genes by inactivating one X chromosome in each cell. An inactivated X chromosome normally is turned on again before it is passed on to a child.

Suspecting that this process might be disrupted in Fragile X, Mandel et al. analyzed the area of the X chromosome where the defective gene was later found. They found that the Fragile X sites of 21 affected men showed abnormal patterns of methylation, a chemical process that shuts genes off. No abnormal methylation was seen in controls or symptomless male Fragile X transmitters. Nonaffected female carriers had normal results, while retarded female carriers had abnormal methylation patterns.

Mandel et al. say this supports the theory of Charles Laird of the University of Washington that Fragile X could be linked to imprinting, a process in which genes inherited from the mother behave differently from those inherited from the father. Perhaps, Mandel et al. say, Fragile X is caused by "a mutation that affects reactivation of an inactive X chromosome during differentiation of female germ cells."

Laird believes the size changes in the DNA at the Fragile X site may be caused by abnormal methylation, while Mandel says it is not clear which occurs first — the methylation changes or the size changes.

Sutherland, however, says that the change in size alone is adequate to explain all of Fragile X genetics. Questioning the imprinting theory, he notes that cells from a single affected individual may have Fragile X sites of varying sizes, indicating that the size changes can occur whenever the cell copies its DNA—not just during passage through the mother.

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A list of the six references used in preparing this article is available from ARRI; send self-addressed, stamped envelope and ask for Fragile X references.

The PDD issue: What's in a name?

Calling the creation of the term "pervasive developmental disorders" (PDDs) to categorize autism and autistic-like disorders "a serious mistake," a prestigious group of researchers from several countries recently called for the term to be dropped.

The term "pervasive developmental disorders" was first used in the 1980 edition of the *Diagnostic and Statistical Manual of Mental Disorders*, Edition III (DSM-III) — the first issue of this manual to include autism as an official diagnostic category. (DSM is the standard reference guide to psychiatric diagnoses in the U.S.) Seven years later, the American Psychiatric Association, which publishes the manual, altered the categories of PDDs somewhat in a revised edition known as DSM-III-R. This newest version of the DSM lists two categories of PDD:

—***Autistic disorder***, described as "a severe form of pervasive developmental disorder, with onset in infancy or childhood." The diagnosis requires that the individual exhibit at least eight of 16 criteria in three categories (social impairment, communication impairment, and restricted repertoire of activities), meeting a minimum number in each category.

—***Pervasive Developmental Disorder Not Otherwise Specified (PDDNOS)***, used when the quality of social and communication skills is impaired, but criteria for autistic disorder (or schizophrenia or schizoid-type disorders) are not met.

The 16 researchers from Europe and the U.S. say that "we are already aware of people with autistic-like conditions who have not been given their lawful 'autism rights' because they were diagnosed with PDD." They argue that the term PDD is little used ("Did you ever think about Rain Man as PDD?"), poorly conceived, and—in their opinion—inaccurate. "Autism and autistic-like conditions are not generally 'pervasive,'" they say. "Rather, they constitute disorders in which *specific* social/cognitive capacities have become dysfunctional."

"The conceptual unsoundness of PDD becomes striking," they say, "when one considers the fact that the most pervasive of all developmental disorders, viz. severely-profound mental retardation, is not included among the pervasive developmental disorders in the current classification systems."

The researchers suggest terms such as "autism and autisticlike conditions," "autism spectrum disorders," or "autism continuum" as replacements for PDD.

In a separate article, Peter Tanguay makes his own suggestion for a candidate to replace PDD. "Is it time to consider a new diagnostic category: social communication spectrum disorder, of which autism would be the earliest and most severe manifestation?" he asks. "Such a concept would admit to a continuum of social communications deficits and might permit a more specific description of symptoms than does PDDNOS."

Tanguay notes that many autistic individuals have a good command of grammar and vocabulary; "What they appear to lack specifically," he says, "is the ability to encode and decode meaning through changes in the pitch, loudness and rhythm of speech, or to communicate subtle affectual signals by gestures and facial expressions." They also have no understanding, he says, of the social rules of conversation or the mental states of others.

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"Autism is not necessarily a pervasive developmental disorder" (letter), G. Baird, S. Baron-Cohen, M. Bohman, M. Coleman, U. Frith, Christopher Gillberg, Carina Gillberg, P. Howlin, G. Mesibov, T. Peeters, E. Ritvo, S. Steffenburg, D. Taylor, L. Waterhouse, L. Wing, M. Zapella ; *Developmental Medicine*, 33(4), April 1991, pp. 363-364.

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"Editor's note: infantile autism and social communication spectrum disorder," Peter Tanguay, *Journal of the American Academy of Child and Adolescent Psychiatry*, 29:54, 1990. Address: Peter Tanguay, UCLA NPI, 760 Westwood Plaza, Los Angeles, CA 90024.