LETTERS TO THE EDITOR

LKS diagnosis and treatment

Editor's Note: the following letter is longer and more technical than those usually published in the ARRI. We have made an exception in this case because the diagnostic information presented by Dr. Stefanatos may be of great interest to many readers.

To the Editor:

Your recent article on Landau-Kleffner syndrome (ARRI 5/1) provided a lucid account of LKS and contributed an invaluable service in underscoring the point that this syndrome, like a number of other distinguishable clinical entities, may be confused with autism. However, the article also raises several points regarding the diagnosis and treatment of LKS that could be misleading if not appreciated in a broader context.

LKS actually represents quite a diverse group of children who have in common an inexplicable retardation of speech development occurring in the context of electrographic disturbances and after a period of seemingly normal development. [Given this diversity], it should not be surprising that there are also potentially different outcomes of treatment based on the relationship between the seizure disorder and the language disturbance, and caution is therefore warranted in evaluating treatments for LKS.

The most common characteristic of LKS is an initial period of hypoacusis [diminished auditory sensitivity] resembling transient deafness which subsides in time to language deficits including impaired comprehension and expressive disorder (ranging from distorted speech to near speechlessness). However, we have seen a couple of LKS children whose language impairment appears predominantly to involve expressive language with rather better comprehension. They do not appear deaf, nor do they have obvious problems in understanding simple conversation, although assessment will reveal that a comprehension difficulty does exist. Such children are clearly less likely to appear autistic when compared to children with severe receptive language impairment.

When a receptive language impairment is primary, it is often characterized by a predominant difficulty in decoding speech sounds ("verbal auditory agnosia"). This results in a severe auditory *language* comprehension problem, [while there is] some preservation of the ability to discriminate *environmental* sounds.

The dense receptive impairment may be key to confusing such children with autistic disorders, but it is noteworthy that this condition can arise developmentally and be unassociated with a seizure disorder or evidence of electrographic disturbances; in this case it is regarded as a form of receptive developmental dysphasia (RDD), and the features which distinguish it from autism have been well outlined by Bartak and colleagues.

The nature of the receptive language impairment in these conditions has intrigued us for a number of years. In pursuing this phenomenon, we have concluded that indeed there are similarities to the underlying dysfunction resulting in verbal auditory agnosia in RDD and LKS. It appears to be based in problems in processing specific features of speech sounds critical for language comprehension. We have developed and studied a specialized evoked potential measure which assesses the function of the mechanisms which process these sounds. We believe this method may be of considerable use in diagnosing these children, along with more traditional assessment using brain stem evoked responses.

While it is important to assess brain stem evoked responses, in most instances these are normal in both RDD and LKS and indeed in many autistic children. This indicates that auditory information is processed up to the level of medial geniculate nucleus, and thus is useful for ruling out deafness or brain stem dysfunction. By contrast, our new procedure tells us whether or not specific types of [sound] essential for speech processing are being processed at the cortical level and, consequently, provides us critical information that other procedures have not. We have found anomalies of these responses in LKS children whose epileptiform disorders have receded with time, but who continue to have auditory comprehension problems, suggesting that the pathophysiology underlying LKS extends beyond the electrographic disturbance.

"Our new procedure [for diagnosing LKS] . . . provides us critical information that other procedures do not."

This observation points out a second feature which I hope to highlight, that significantly different factors may result in a picture of LKS. Indeed, Deonna et al. consider that there may be at least three separate conditions. The first is associated with rapid onset and recovery from the aphasia with a fairly direct link between the epileptic activity and the language impairment. In the second there is a rather poor recovery of language after repeated seizures, possibly indicating more permanent pathophysiology. The third subgroup is associated with variable recovery; here seizures are infrequent or may not occur at all, and for the most part, improvement in seizures is not associated with consequent improvement of the aphasia.

It is critical to appreciate the implications of the differences between these subgroups. [Some] researchers believe the epileptiform disorder in LKS, particularly in the latter two subgroups, [is not a cause of the language disturbance, but rather that both are caused by an underlying neural dysfunction.] Under such circumstances, it seems unlikely that a surgical procedure addressing the epileptic activity would have any significant impact on improvement of the language disorder. A correlation between the epileptic activity and the language disturbance would need to be clearly delineated by other means before reasonable consideration of surgical intervention is warranted.

It is unclear at this point what circumstances merit surgical intervention. Surgery, even in cases where it is directed to treatment of intractable epilepsy, is regarded as a last resort when other modes of medical intervention have failed. Such caveats may especially apply to the use of surgery to treat LKS, since there is a tendency toward spontaneous recovery and/or improvement with age. [Editor's Note: see ARRI 5/2 for an article on non-surgical treatment of LKS.]

Taken together, these points will hopefully enlighten readers to the pitfalls as well as the promise of surgical intervention for LKS. It is essential to be cautious, to appreciate the necessary indications for such an undertaking, and to have sufficient facts to justify the procedure. In other words, one should not be drawn into any unreasonable or unwarranted expectations, especially when other modalities, such as drugs, also seem to effect changes in these children.

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Editor's Note: The new brain-wave procedure for clarifying the diagnosis of LKS referred to by Dr. Stefanatos is currently available only at his laboratory. For information, contact him at the address above.

"Community, My Foot!"

To the Editor:

Your article, "Community, My Foot!," (5/3) is of special interest to us because we have been Intervenors in a recent lawsuit which resulted in a court order to move some of our children, relatives and wards into community settings. We have acted to defend our institution and find that we must continue to do so as the state Department of Health seems determined to reduce substantially the role of the institutions in the care of developmentally disabled citizens.

May we have your permission to circulate copies of your article to our state legislators and others in the state administration? I would also like to send a copy to the national organization, Voice of the Retarded, which consists of parent groups and other associations working for the defense and preservation of institutional facilities where they are threatened. VOR is located at 2800 Central Road, Rolling Meadows, Illinois 60008, phone (708) 253-6020.

Your article on "The Non-Urban Alternative" is also appropriate because the state wants to close down our facility at Fort Stanton, a rural setting where the state has had gratifying success with disabled clients who are ambulatory and high-functioning.

Charles E. Woodhouse, Chairman
Los Lunas Hospital and Training School
Parents and Guardians Association

Editor's Note: Permission granted—enthusiastically! Everyone has the right to live in a safe and secure environment.

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