

Clinical Forum

Landau-Kleffner Syndrome: Acquired Epileptic Aphasia in Children

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Abstract

Acquired epileptic aphasia, or Landau-Kleffner syndrome (LKS), once thought to be a rare syndrome, may occur more frequently in the pediatric population than once thought. This syndrome is typically characterized by an abrupt or gradual loss of language ability and inattentiveness to sound, sometimes called auditory agnosia, with onset during the first 5 years of life. This interruption in communication ability is generally closely preceded, accompanied, or followed by the onset of seizure activity and/or abnormal electroencephalographic (EEG) findings. This report describes two cases of LKS evaluated at the same hearing and speech center. Because of the characteristic language regression and inattentiveness to sound, speech-language pathologists and audiologists are likely to be among the first professionals to evaluate these children. It is imperative that communication specialists be alert to the characteristic symptoms of LKS in order to be instrumental in initiating an appropriate diagnostic and management process. A multidisciplinary approach to identification and rehabilitation is encouraged in order to effectively re-establish communication skills for these children.

Key Words: Acquired epileptic aphasia, central auditory processing, child language, convulsive disorder, epileptic aphasia, Landau-Kleffner syndrome (LKS)

Although it is not unusual for a child to demonstrate delayed initiation of language or to develop language at a slower than average rate, it is unusual for children to begin developing language normally and then demonstrate a disruption in their progress. Such an abrupt interruption is usually caused by

head trauma, cerebrovascular disease, tumors, sudden onset of hearing loss, or a variety of infectious agents. There exists, however, a little known subgroup of children with acquired aphasia for whom there is no overt cause for this interruption in their language progress. This disorder was first described by Landau and Kleffner in 1957 as acquired childhood aphasia with convulsive disorder. Since that initial report, this disorder has become better known as Landau-Kleffner syndrome (LKS), and over 100 cases have been described in the literature (Worster-Drought, 1971; Shoumaker, Bennett, Bray, and Curless, 1974; Deuel and Lenn, 1977; van Dongen and Loonen, 1977; Msall, Shapiro, Befour, Niedermeyer, and Capute, 1986; Cole, Andermann, Taylor, Olivier, Rasmussen, Rabitaille, and Spire, 1988; Blouw-van Mourik, van Dongen, Loonen, and Jannsen, 1989; Penn, Friedlander, and Saling, 1990; Tharpe, Johnson, and Glasscock, 1991).

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BACKGROUND

Although a review of this literature reveals a diverse population with a wide array of presenting symptoms, degrees of impairment, and outcomes, there are some characteristic features that are worth noting. Perhaps the most obvious symptom of this disorder is the often acute and sometimes gradual language regression. An excellent discussion of the language characteristics of these children can be found in Miller, Campbell, Chapman, and Weismer (1984). Briefly, they reviewed 94 cases with diagnosed LKS and found that while these children were initially following a normal course of language development, an onset of language regression occurred between 1.5 and 13 years of age, with the majority of the losses occurring between 3 and 7 years. Most of the language losses were rather rapid, sometimes occurring within hours, while a smaller number were more gradual, slowly regressing over more than 6 months.

The language deficit can encompass both receptive and expressive skills to varying degrees. Reports of receptive language skills have ranged from an ability to follow simple verbal commands to a total inability to understand any verbal input (Worster-Drought, 1971; Deonna, Beaumanoir, Gaillard, and Assal, 1977; Rapin, Mathis, and Rowan, 1977; Msall et al, 1986; Penn et al, 1990; Tharpe et al, 1991). In most cases, expressive language is characterized by a gradual increase in misarticulations and a decrease in sentence complexity. Speech has been noted to decrease to the use of only single words, jargon, or total mutism (Shoumaker et al, 1974; Rapin et al, 1977; Cooper and Ferry, 1978; Msall et al, 1986; Mantovani and Landau, 1980; Holmes, McKeever, and Saunders, 1981; Tharpe et al, 1991).

In some cases, the language regression has been preceded by a decrease in auditory comprehension, which may appear initially as a peripheral hearing loss. Behavioral audiologic findings are sometimes difficult to obtain, depending on the degree of auditory agnosia. Behavioral audiograms can suggest anywhere from normal to profound hearing losses (Worster-Drought, 1971; Shoumaker et al, 1974; Deuel and Lenn, 1977; Rapin et al, 1977; Cooper and Ferry, 1978; van Harskamp, van Dongen, and Loonen, 1978; Msall et al, 1986; Penn et al, 1990; Tharpe et al, 1991); however, when auditory evoked potential testing has been con-

ducted, results have been consistent with normal peripheral auditory systems (Msall et al, 1986; Cole et al, 1988; Penn et al, 1990; Tharpe et al, 1991).

The definitive diagnostic indicator for LKS is a paroxysmal electroencephalogram (EEG) (Mantovani and Landau, 1980). The EEG is most commonly characterized by mid-temporal spike activity, predominantly left sided, and bilateral-synchronous bursts of spikes and slow spike-wave complexes (Deonna et al, 1977; Holmes et al, 1981; Rodriguez and Niedermeyer, 1982; Msall et al, 1986). It should be noted, however, that some children with LKS never develop seizures, although 80 percent ultimately do, usually by 2-3 years after onset of language loss (Shoumaker et al, 1974; Miller et al, 1984; Msall et al, 1986). Forty percent of the seizures will precede the language regression, 40 percent will follow, and 20 percent will occur concurrently with onset of language regression (Miller et al, 1984).

CASE REPORTS

The following case reports are illustrative of findings with LKS.

Case 1

JC is a 6-year-old, black male. JC's parents first became concerned about his speech/language skills when he was approximately 2.5 years of age. At that time, they began to notice a lack of language progress and a slight regression in his language abilities. Concurrently, they noted an increase in adverse behaviors or tantruming. He received a speech/language evaluation at 3 years, 9 months of age and was found to demonstrate severely delayed expressive language skills and a severe phonologic disorder characterized by final consonant deletion, syllable reduction, fronting, and stopping. Receptive language abilities could not be accurately determined, due to adverse behavior. Speech/language therapy was recommended and initiated shortly thereafter.

Audiologically, JC's parents had few concerns. They felt he responded to sound appropriately most of the time, although he seemed to have occasional brief periods of unresponsiveness. JC's hearing had been evaluated in infancy (i.e., 3 and 9 months of age) as a precautionary follow-up to beta streptococcal meningitis contracted at 2 weeks of age and was found to be normal. At the time of his speech/language

evaluation at 3 years, 9 months of age, his hearing was re-evaluated. JC could not be conditioned to play audiometric techniques; therefore, visual reinforcement audiometry was employed. Responses to speech stimuli were obtained at 50 dB HL with no other responses being consistently observed for frequency-specific stimuli. Click-evoked auditory brainstem response testing (ABR) revealed a verifiable wave V down to 20 dB nHL bilaterally. Absolute and interwave latencies were all within normal limits bilaterally. Six months later, at 4 years, 3 months of age, behavioral testing revealed responses to speech and frequency-specific stimuli within normal limits bilaterally. Due to the presence of myringotomy tubes, acoustic reflex measures could not be obtained at any of the test sessions.

At age 4 years, 9 months, JC's hearing was re-evaluated upon the request of his teachers, who expressed concern about his auditory ability. He was then enrolled in a language-impaired classroom in the public school system. Although he was still quite difficult to test due to inconsistent responding, hearing sensitivity was considered within normal limits bilaterally. At that time, however, it was recommended that he be evaluated by a pediatric neurologist in order to rule out LKS.

Physical examination by the neurologist was unremarkable. An EEG was abnormal, showing electrical discharges in the left mid-temporal and posterior quadrant region. A magnetic resonance image (MRI) was also abnormal, showing abnormalities in the parietal occipital white matter that were considered secondary to the meningitis he had when younger and not related to his current problems. He was begun on anticonvulsant medications immediately and, after approximately 1 month, both his mother and teachers felt that he was more attentive to speech. There were periods of time when JC would discontinue his medicine. During those times, his mother and teachers felt that he became less responsive to sound. As a result of a diagnosis of LKS, JC was enrolled in a self-contained classroom for the hearing impaired in the public school system. He began utilizing a total communication approach to which he rapidly adapted. In addition, a mild-gain FM system, coupled with "Walkman" type earphones, was recommended and successfully utilized. At this writing, JC remains on medication, has had no overt seizures, and his oral and signed language skills continue to improve.

Case 2

AD is an 8-year-old, right-handed, Caucasian female first diagnosed with LKS at 41 months of age. As her history has been described in detail elsewhere (Tharpe et al, 1991), only a brief review will be presented here. AD's regression in language ability began at 38 months of age and was characterized initially by an increase in misarticulations, primarily final consonants, and ultimately deteriorated to a few intelligible words. This regression was preceded by an inattentiveness to auditory stimuli. When she was first evaluated audiolgically, AD demonstrated a severe-to-profound hearing loss behaviorally; however, the Sensitivity Prediction by Acoustic Reflex (SPAR) (Jerger, Burney, Mauldin, and Crump, 1974) and ABR findings were consistent with normal hearing sensitivity bilaterally. In addition, absolute and interwave latencies were within normal limits bilaterally, as was the interaural latency difference (IT_v).

Clinical neurologic examination, computerized tomography, and MRI were all negative. An EEG revealed the presence of a left mid-temporal spike focus, and anticonvulsant drug therapy was initiated. AD became slightly more responsive to sound and began to regain a few single words. Four months after starting drug therapy, she had her first seizure and her auditory and language skills again deteriorated. This fluctuation of progress continued for approximately 2 years while her medication and/or medication levels were adjusted.

AD has been enrolled in a total communication program for the hearing impaired in the public school system since 4 years of age. Initially, she used a mild-gain FM system in the classroom setting. The FM system was found to be particularly useful when the classroom background noise was high. Recently, however, she has not seemed to need the FM system, as she consistently attends to auditory stimuli and communicates effectively via a total communication approach.

For almost 2 years, AD has been making slow but steady progress in her communicative skills. She demonstrates a moderate articulation disorder characterized primarily by final deletions of phonemes. Her intelligibility decreases significantly as her rate of speech increases. Receptive language skills are considered mildly impaired. Annual cognitive testing has consistently indicated average or above average intelligence. A recent psychoeducational

evaluation revealed several significant deficits in AD's reading ability. Specifically, deficits were noted in spatial orientation and left-right confusion, and reading comprehension.

AD's seizures have decreased in frequency and now occur only rarely. She has had no overt seizures since 3.5 years of age.

DISCUSSION

At this writing, the pathophysiology of LKS remains unknown, although several possibilities have been suggested (Worster-Drought, 1971; Gascon, Victor, Lambrosco, and Goodglass, 1973; Rapin et al, 1977; Cole et al, 1988; Otero, Cardova, Diaz, Garcia-Tervel, and del Brutto, 1989). Some maintain that the aphasia is the result of the auditory agnosia, implicating a cortical or subcortical dysfunction of the auditory system (Gascon et al, 1973; Rapin et al, 1977; Bishop, 1985; Nakano, Okuno, and Mikawa, 1989).

Prognosis for LKS is unpredictable. Although Miller et al (1984) report that more than 80 percent of the cases they reviewed demonstrated the language disorder for longer than 6 months, few longitudinal studies have been reported in order to ascertain long-term outcome. Complete language recovery has been reported, as well as a range of residual deficits ranging from mild academic difficulties to no verbal communication (Landau and Kleffner, 1957; Worster-Drought, 1971; Gascon et al, 1973; Rapin et al, 1977; Mantovani and Landau, 1980). Recovery of the language deficit may occur rapidly within days or it may take place slowly over a number of years (Deuel and Lenn, 1977; Deonna et al, 1977). Unfortunately, stabilization of the EEG frequently does not guarantee an improvement in the aphasia (Worster-Drought, 1971; Gascon et al, 1973; Rapin et al, 1977; van Harskamp et al, 1978; Holmes et al, 1981). Similarly, there is poor correlation between frequency of the seizure activity and severity of the aphasia.

Several case reports have discussed medical treatment strategies to stabilize the EEG and/or seizure activity (Landau and Kleffner, 1957; Deonna et al, 1977; Deuel and Lenn, 1977; Mantovani and Landau, 1980; Woody, 1990). Less attention has been given to the remaining communication deficit. The reader is referred to Miller et al (1984) for a review of speech/language intervention approaches with this population.

Of the few reports of adults who have completely recovered from the language deficits experienced in childhood, many state that these adults still have difficulty understanding speech in the presence of background noise. In fact, one patient reported difficulty with "hearing distortion" (Mantovani and Landau, 1980). Such reports suggest that an improvement in the acoustic environment of children with LKS may be of benefit. This can be accomplished with the use of low-gain personal or classroom FM systems or by acoustically treating classrooms. FM systems have been used with success by the children described herein. The parents of both children believe that the provision of total communication, although not herein formally assessed, has lessened their children's frustration levels and, therefore, has resulted in improved behavior.

CONCLUSION

Unfortunately, children with LKS can easily be misdiagnosed, since the outstanding features of this disorder may be suggestive of autism, peripheral hearing loss, emotional or behavioral disorder, or other types of acquired aphasia. Penn et al (1990) describe a tragic case of a child with LKS who, after developing normally until 22 months of age, had an onset of seizure activity. She subsequently had a severe regression in her language ability and went through the next 13 years receiving a variety of diagnoses, including brain damage, autism, mental retardation, aphasia, and deafness. Repeated hearing tests yielded variable results. An EEG revealed abnormal activity in the left fronto-temporal areas. She was placed in a school for the deaf at age 4; however, it was an auditory-oral program, and she was never able to develop an effective communication system. It was not until she had an ABR test at age 13 that hearing loss was ruled out.

Although very disheartening, the management of this case is not surprising, given the limited exposure that professionals in the communicative sciences have had to this disorder. It is interesting to note, however, that in both of the cases described herein, the initial referral for determination of the problem was made to a speech/language pathologist or audiologist. Clearly, we must remain alert to the "red flags" of language *regression*, inattentiveness to sound, and partial seizures characterized by staring.

As with all pediatric cases, the wise advice of utilizing the crosscheck principle cannot be overemphasized (Jerger and Hayes, 1976). Reinforcing behavioral auditory assessment with objective measures such as the SPAR and ABR can prevent a crucial error. In addition, a multidisciplinary team approach to diagnosis and management is essential. Ideally, this should include an audiologic/otologic evaluation, speech/language evaluation, psychological examination, and neurologic examination including EEG from appropriate team members.

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