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Neuropsychological characteristics of five children with the Landau-Kleffner syndrome: Dissociation of auditory and phonological discrimination

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Abstract

The Landau-Kleffner Syndrome (LKS) is characterized by acquired receptive aphasia and EEG abnormality with onset between the ages of 3 and 8 years. This study presents neuropsychological assessments in 5 children with LKS. The aims were (1) to specify the neuropsychological deficits characteristic of these children; and (2) to clarify the nature of the receptive aphasia by comparing nonverbal and verbal auditory discrimination. Receptive aphasia was present in all children. Retardation, poor motor coordination, hyperkinesia, and conduct problems were frequent but variable. All children exhibited a dissociation between the discrimination of environmental sounds and phonological auditory discrimination, the latter being more impaired than the former. This suggests that the primary deficit of the receptive aphasia is an impairment of auditory phonological discrimination rather than a generalized auditory agnosia. (*JINS*, 1998, *4*, 566–575.)

Keywords: Landau-Kleffner syndrome, Acquired epileptic aphasia, Continuous spike waves during slow sleep, Specific language impairment

INTRODUCTION

Landau-Kleffner Syndrome (LKS) is a rare disorder characterized by acquired aphasia and EEG abnormalities in the absence of focal lesions, with onset usually between ages 3 and 8 years (Deonna & Roulet, 1995; Landau & Kleffner, 1957; Paquier et al., 1992). About 70 to 80% of the children also exhibit epileptic seizures (Beaumanoir, 1985; Dugas et al., 1982).

The most striking and still unexplained feature in LKS is the acquired aphasia, which frequently renders the child unable to comprehend any language. Impairment of expressive speech varies in pattern and degree from syntactic and dysnomic problems, echolalia, or short-term memory problems in less severe cases, to telegraphic speech, jargon, or complete muteness in severe cases (Paquier et al., 1992; Rapin et al., 1977; Soprano et al., 1994; Van Hout, 1992). Some authors also classify specific expressive disorders related to epileptic discharges as LKS (Dugas et al., 1995; Feekery et al., 1993; Marien et al., 1993). This study is restricted to more typical cases of LKS, characterized by receptive aphasia.

Various nonverbal signs have been associated with LKS. Frequently, agnosia for nonspeech sounds has been reported. The child may appear deaf, yet, audiograms are generally normal (Feekery et al., 1993; Lanzi et al., 1994; Paquier et al., 1992; Tharpe & Olson, 1994). Intellectual capacity deteriorates in some of the children (Lanzi et al., 1994; Paquier et al., 1992; Roulet et al., 1991). Behavioral problems are frequent and may include attention disorders and hyperactivity, aggression, apathy, or depression (Bishop, 1985; Dugas et al., 1982; Soprano et al., 1994), even autistic (Rapin, 1995; Van Hout, 1992) or psychotic regression (Roulet et al., 1991). Manual and oral dyspraxia (Ansink et al., 1989; Bulteau et al., 1995; Rapin et al., 1977)

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and visuospatial problems (Bulteau et al., 1995) have also been reported in some children.

The onset of LKS may be sudden or insidious (Deonna & Roulet, 1995; Soprano et al., 1994). In rare instances precipitating events, such as rubella (Veggiotti et al., 1995) or measles vaccination (Metz-Lutz et al., 1995) have been reported. Fluctuations of signs and EEG abnormalities occur. In a significant number of children with LKS, the language disorder persists, especially in those with early onset (Bishop, 1985; Lanzi et al., 1994; Paquier et al., 1992; Soprano et al., 1994).

The pathogenesis of LKS is not known. To what extent the aphasia is a result of the epileptic activity is controversial (Deonna & Roulet, 1995). In many children sleep EEG recordings have demonstrated spikes (Deonna & Roulet, 1995; Lanzi et al., 1994), sometimes in the form of continuous spikes and waves during slow sleep (CSWS; Hirsch et al., 1995; Paquier et al., 1992; Roulet et al., 1991). Yet, seizure control does not bear any clear relation to recovery from aphasia (Feekery et al., 1993; Van Dongen et al., 1995).

EEG abnormalities involve multiple regions—bitemporal, temporoparietal, and parietooccipital (Beaumanoir, 1985; Cole et al., 1988; Rapin et al., 1977), or they are generalized (Feekery et al., 1993; Nakano et al., 1989). MRI scans are normal in most cases (Bureau, 1995). PET and SPECT studies have demonstrated focal or multifocal changes or perfusion asymmetries predominantly affecting, but not restricted to, the temporal regions (Maquet et al., 1995; Mouridsen et al., 1993; O'Tuoama et al., 1992).

The primary deficit of the receptive aphasia in LKS is considered to be impaired phonological decoding. Expressive disorders are seen as secondary or concomitant to this (Lanzi et al., 1994; Rapin et al., 1977; Soprano et al., 1994). However, agnosia for nonspeech sounds has also been reported. It is therefore possible that the deficit underlying the aphasia may be a generalized auditory agnosia instead of a phonological decoding deficiency. This view was supported in a study by Klein et al. (1995). Young adults with residual LKS and acquired aphasia were shown to have slowed reaction times and a delay of certain components of evoked potentials in response to both nonverbal and phonological stimuli. The result contradicts, to some extent, those obtained by Frumkin and Rapin (1980) and Notoya et al. (1991) of poorer perception of consonants than of vowels. This finding would indicate that there is some degree of selectivity of impairment within the auditory domain.

Most of the studies on children with LKS have presented cases on a descriptive level, and comprehensive, formal assessments have not been performed. Further, the precise nature of the receptive aphasia has not been fully clarified. This study presents a comprehensive, formal assessment of verbal and nonverbal performance in 5 children with LKS. The aims of the study were (1) to specify the neuropsychological deficits characteristic of these children, and (2) to clarify the nature of the receptive aphasia by comparing nonverbal and verbal auditory discrimination.

METHODS

Patients

Five children with a diagnosis of LKS, treated at the Helsinki University Central Hospital during the years 1988 to 1995, took part in the study. The criteria for inclusion were (1) acquired receptive aphasia or dysphasia, (2) no substantial hearing loss by audiometric tests, (3) no evidence of focal damage that could explain the acquired aphasia, and (4) epileptiform activity on EEG recordings.

The age, sex, and clinical data of the children are presented in Table 1. As may be noted in Table 1, 1 child (K.S.) had a premorbid delay of cognitive and motor development but by the age of 5 years, prior to onset of LKS, she spoke fluently. She also had diffuse abnormality on the MRI. Being a somewhat atypical case of LKS she nevertheless fitted the criteria for LKS, as defined above. After the onset of LKS gradual improvement was noted in all children, mainly in the form of improved reaction to sound and speech. However, the aphasia remained severe in all cases.

Assessments

The neuropsychological assessment was performed when the disease had passed its peak and the condition was considered stable, 2 years, 10 months to 9 years after onset.

Comprehensive neuropsychological assessment

Nonverbal intelligence was assessed with the aid of the Wechsler Intelligence Scale for Children–Revised (WISC–R; Wechsler, 1984) Performance scales, when possible. Two children (K.S., M.J.) were unable to perform the WISC–R and were assessed with the Leiter International Performance Scale (Leiter; Leiter, 1969).

Neuropsychological performance was assessed mainly with the aid of the NEPSY Neuropsychological Assessment of Children, which is standardized in Finland for children ages 3 years 6 months to 9 years 6 months (Korkman, 1988a). Detailed descriptions are available in English (Korkman, 1988b, 1988c, 1995). The subtest raw scores of the 1988 NEPSY are converted to standard *z* scores (maximum = +1, minimum = −3), based on the distributions of the appropriate age group. Reliabilities are adequate (.59–.98). An expanded and revised version for 3- to 12-year-olds has been standardized in the U.S (Korkman et al., 1998[1]) and in Finland (Korkman et al., 1997).

Language was assessed using the following three subtests from the 1988 NEPSY: Naming Colors (the child is to name the colors of seven color plates), Digit Span (the child is to repeat digit series forward), Repetition of Words and

[1]The names of the subtests of the 1997 and 1998 versions differ from those of the 1988 version (for comparison see Korkman, in press; Korkman et al., 1998).

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Table 1. Sex, age, and clinical data on the children with LKS

Child	K.S.	V.N.	M.O.	M.J.	V.M.
Sex	F	M	M	F	M
Early development	delayed	normal	normal	normal	normal
Age at onset of LKS (years; months)	6;0	3;0	2;7	2;3	4;0
Age at assessment (years; months)	8;10	12;0	8;6	8;7	8;7
First sign(s)	aphasia	aphasia	aphasia	aphasia, seizures	aphasia, seizures, clumsiness
Loss of reaction to sound	yes/improved	yes/improved	yes/improved	yes/improved	yes/improved
EEG: documented epileptiform activity	yes	yes	yes	yes	yes
Seizures: type	atypical absences	psychomotor or motor	partial motor	atypical absences	atypical absences
Seizures: frequency	periods with daily	1–2/year	occasional	occasional	periods with daily
Effect of medication	good	good	good	good	poor
Documented periods with CSWS[1]	yes	no	yes	yes	yes
EEG at time of assessment	CSWS	normal	spikes	spikes	normal
MRI	mild atrophy	normal	normal	normal	normal
MEG[2]: origin of spikes	left and right Sylvian	left and right Sylvian	left and right Sylvian	left and right Sylvian	left and right Sylvian

[1]Continuous spikes and waves during slow sleep.
[2]Magnetoencephalography (see Paetau, 1994).

Nonwords (an example of the former: *ruuvitaltta* = screwdriver; examples of the latter: *ronk*, *minksakka*).

Nonverbal performance was assessed with the aid of the following three NEPSY subtests: Oral Kinesthetic Praxis (the child is to imitate movements and positions of the oral apparatus, for example, pushing the cheek with the tongue, fluttering lips), Kinesthetic Praxis–Positions of Hands (the child is to imitate positions of the hands and fingers), and Sustained Concentration (the raw score is the average time the child is able to keep working during the testing sessions). The Oral Kinesthetic Praxis subtest was considered nonverbal since it has not been found sensitive to language and/or articulation disorders (Korkman & Häkkinen-Rihu, 1994).

In addition to the NEPSY subtests the shortened version of the Token Test was administered to assess verbal comprehension (DeRenzi & Faglioni, 1978). The Visual–Motor Integration test (VMI; Beery, 1983) was administered to assess copying designs. Finnish norms were developed for these tests in connection with the standardization of the NEPSY.

Clinical observations were recorded, since available tests do not cover all significant aspects of the patient's deficits. Comprehension of everyday speech was evaluated on the basis of expressions that the child had been reported to understand. The child's expressive language was evaluated in the test situation by recording length of utterances and evaluating articulation and pragmatics. Use of sign language was evaluated by the child's speech pathologist.

Motor signs (e.g., paresis, hypotonia) were recorded and motor coordination evaluated (e.g., balance, hopping on one foot, walking along a straight line, sequential finger tapping, diadochokinesis, pointing finger to nose) by a child neurologist. Behavioral signs were considered present if they were clearly disrupting and interfering with everyday activities. Such signs included hyperactivity (excessive motor activity, restlessness), conduct problems (defiant and uncompliant behavior), and poor social contact and gaze avoidance.

Environmental sounds and phonological discrimination tasks

For the assessment of nonverbal auditory discrimination a commercial sound–picture lotto game with tasks of environmental sounds discrimination was used. The material and items are described in the Appendix.

Control data were based on 20 normal Kindergarten students (7 boys and 13 girls, age 5 years 0 months to 7 years 1 month; M = 5 years 10 months; SD = 7.7). This data was collected to (1) elaborate the task psychometrically, and (2) provide normative data for converting raw scores to standard scores. Based on the distribution of the normal children the LKS children's scores were converted to z scores.

Control data from a relatively young age group were preferred in order to facilitate psychometric calculations, because the distribution of the results of older children was expected to be skewed due to a ceiling effect. Age-matched normative data were not considered necessary, because the LKS children were expected to perform more poorly than their own age level, due to their aphasia and history of auditory agnosia.

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Item-to-total correlations, *p* values (percentage of children passing the item), and Cronbach's alpha reliability were calculated. Of the 45 items, those with a correlation less than .30 with the total score were not included in the total score, to improve reliability. Exceptions were six items with lower correlations, which were kept in order to preserve some items with low and some items with high *p* values. Thus, all children, including the children with LKS, performed 45 items but only 29 items were used for the total score. After item selection, reliability was .89. The mean of the normal children was 22.6 ($SD = 5.3$).

Phonological discrimination was assessed with the aid of the Auditory Analysis of Speech subtest from the NEPSY (see Appendix). Norms for children age 5 years 6 months to 6 years 5 months ($N = 49$) from the 1988 NEPSY were used to convert the LKS children's data to *z* scores. This age level was chosen because it corresponded to that of the control data for the environmental sounds discrimination task. The mean of the norm group on the phonological discrimination task was 12.3 (maximum = 16, $SD = 2.0$; Korkman, 1988c).

The distributions of normal children on the two tests of auditory perception were comparable; they were normal although slightly skewed. No ceiling or floor effect appeared. Thus, it was possible to transform the raw scores of the children with LKS to *z* values, based on the results of normal 5-to-6-year-olds and to compare their performance on these two tests.

A testing-the-limits procedure was applied by asking the children to point to the appropriate pictures when the complete words were provided (see Appendix). This task was not psychometrically elaborated since there would be a ceiling effect for normal children, all words belonging to the working vocabulary of normal 5-to-6-year-olds.

All tests were discontinued after four consecutive failures. No opportunity for lip reading was given on verbal tasks. Due to their verbal agnosia, in the absence of these visual cues, the children often did not even attempt to respond to a verbal command or a target word. In these instances the child received a raw score of zero. If the child's attention began to decline the testing was continued at another time. When it was evident that poor attention affected the results, the data were considered missing. This occurred twice on the lengthy environmental sounds task.

RESULTS

Results on the Comprehensive Assessment

Performance IQ, neuropsychological test scores, and behavioral observations are presented in Table 2. Note that V.N.

Table 2. Performance–nonverbal IQ, neuropsychological test results (in standard *z* scores), and additional observations of children with LKS

Child	K.S.	V.N.	M.O.	M.J.	V.M.
Performance–nonverbal IQ	53[1]	91[2]	74[2]	57[1]	60[2]
Verbal test scores					
The Token Test	−3	−3	−3	−3	−3
Naming Colors	−3	−2	−3	−3	−3
Digit Span	−3	−1	−3	−3	−3
Repetition of Words and Non-words	−3	−3	−3	−3	−3
Additional observations					
Speech comprehension (*N* expressions)	none	< 100	< 10	< 10	< 10
Speech production (*N* expressions)	< 10	fluent	none	none	< 10
Length of utterances (*N* words)	1–3	2–6	0	0	1–3
Articulation	poor	poor	—	—	poor
Pragmatics	normal	normal	normal	reduced	normal
Sign language (*N* words)	10–20	none	> 100	10–20	10–20
Nonverbal test scores					
VMI	−3	0	−2	−3	−3
Oral Kinesthetic Praxis	−3	−1	−2	—[3]	−1
Positions of Hands	−3	0	0	−1	—[3]
Sustained Concentration	−3	0	0	0	−3
Additional observations					
Poor motor coordination	yes	no	no	no	yes
Hyperactivity	no	yes	yes	no	yes
Conduct problems	yes	no	yes	no	yes
Poor social and eye contact	no	no	no	yes	no

[1]Leiter.
[2]WISC–R.
[3]Missing observation due to poor cooperation.

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was older than the age range of the NEPSY test norms. Results obtained at earlier ages were comparable to the present ones. At age 8 years, 1 month V.N. had obtained a scaled score of −3 on the Naming Colors test, the Digit Span tests, and the Sustained Concentration test, and a score of −1 on the VMI. The other results were the same as the present results. The results at age 12 years are presented in this study, because the environmental sounds discrimination task was performed at this age only.

On the verbal tests V.N. was the only child who showed any sign of linguistic competence. He obtained seven correct responses on the Token Test. These correct responses were based on his auditory recognition of five color names rather than on a comprehension of the instructions. He was able to name the same five colors on the Naming Colors test, and to repeat a maximum of four digits forward on the Digit Span test. He was not able to repeat any nonword correctly.

Additional observations revealed that, with the exception of K.S., all children were able to comprehend or aurally recognize a few words, but none could understand everyday speech normally. Examples of words recognized by the children were *vaara* (danger; M.O.), *kisse* (kitten; M.J.), and some names of siblings, classmates, or objects. V.N. was able to recognize many everyday expressions when presented in context and with the aid of lip reading (e.g., *You can go to the ward now*); without such cues comprehension was poor. He was able to recognize and write on dictation his own name and the names of three classmates but not single letters (e.g., *O* or *E*).

M.O. and M.J. were completely mute, whereas K.S. and V.M. occasionally uttered short expressions with distorted phonology. V.N. spoke fluently. The verbal expressions of the three latter children were phonologically distorted and largely unintelligible, but intonation was adequate. V.N. was able to provide recognizable names of a few classmates, colors, and numbers from zero to 10.

The result on the nonverbal tasks varied as did the clinical signs observed during the assessment. K.S. and V.M. had widespread impairments whereas V.N. performed normally in most domains (see Table 2). M.J.'s social contact and cooperation had gradually improved but she was still socially withdrawn.

Environmental Sounds Discrimination and Phonological Discrimination

The raw scores, percentage of correct responses, and *z* scores on the environmental sounds discrimination task and the phonological discrimination test are presented in Table 3. As shown in the table, performance on the environmental sounds discrimination task was superior to that on the phonological discrimination test. The difference was significant ($p < .05$) according to nonparametric statistics (Wilcoxon matched-pairs, signed-ranks, two-tailed test). All children were able to identify some environmental sounds; in contrast, only V.N. achieved any correct response on the phonological discrimination test. Providing the whole word did not improve the performance of any child. V.N. achieved the same words correctly when they were presented as syllable blending or word completion tasks and as complete words, which indicated that the correct responses were not obtained by chance.

DISCUSSION

Shared and Variable Deficits

On the comprehensive neuropsychological assessment only deficits related to the aphasia were found in all children, whereas nonlinguistic findings varied between individuals. By definition, all our patients had receptive aphasia. In spite of gradual improvement and an elapsed time of 2 years, 10

Table 3. Performance on tasks of environmental sounds discrimination, phonological discrimination, and comprehension of complete words

Task	Child				
	K.S.	V.N.	M.O.	M.J.	V.M.
Environmental sounds discrimination					
Raw score	11/29	17/23[1]	10/29	19/29	11/17[1]
Percent correct	38%	74%	34.5%	65.5%	65%
z score	−2	0[2]	−2	0	0[2]
Phonological discrimination					
Raw score	0/16	5/16	0/16	0/16	0/16
Percent correct	0%	31%	0%	0%	0%
z score	−3	−3	−3	−3	−3
Comprehension of complete words					
Raw score	0/16	5/16	0/16	0/16	0/16
Percent correct	0%	31%	0%	0%	0%

[1]Observations missing due to loss of interest and discontinuation of test.
[2]Missing observations estimated.

months to 9 years since onset, the aphasia was almost total in 4 children, the only evidence of language comprehension being their ability to recognize a few words or names. A 5th child (V.N.) had improved significantly but was still severely aphasic. He comprehended many expressions when presented in context and as aided by lip reading, but without visual cues his comprehension was very poor, as indicated by the test scores.

Expressive language, when present, consisted of short connected utterances, such as "What's this?", or of one-word expressions, such as "look," as previously described by Paquier et al. (1992). The expressions were phonologically distorted to the degree that longer utterances reminded of jargon aphasia, which suggested that the expressive disorder may be secondary to the receptive. However, in contrast to jargon aphasia, speech tended to be sparse. Two children were completely mute.

Of the nonverbal results only the oral praxis tasks were impaired in all children, to varying degrees. This deficit may be explained by the reduced opportunity for auditory feedback from articulation, which may gradually affect oral motor precision and control. Alternatively, the neural substrate for oral motor control may be anatomically closely related to that of auditory phonological processing and may tend to be concomitantly affected. According to Liberman and Mattingly (Liberman, 1992; Liberman & Mattingly, 1985) the auditory perception of the phonological units of speech are, in fact, based on motor articulation of these units. On this ground it may be argued that the oral dyspraxia may be causatively related to the aphasia. However, it seems intuitively unlikely that oral dyspraxia could account for, or even contribute to, the complete inability to comprehend speech that characterized 4 of the children. In addition, the oral dyspraxia was only mild in 2 cases and moderate in 1. On the basis of these data it cannot be maintained that a significant oral dyspraxia would even be a constant finding in LKS.

Only 1 child (V.N.) had a Performance IQ within the average range. The cognitively impaired children also performed poorly on the complex task of copying designs. Three children had poor manual praxis. Hyperactivity and conduct problems were found in 3 children, poor concentration on tasks in 2. One child (M.J.) exhibited poor social contact and gaze avoidance, suggestive of a disorder at the milder end of the autistic spectrum. These findings indicate that the brain dysfunction in children with LKS is typically not restricted to the substrates for language processing but that other regions may be variably affected. Signs of more generalized involvement were, in fact, present in all of the children. The pattern and degree of the additional neuropsychological signs were, however, variable.

The overall pattern of findings of the children in our study is in accordance with previous reports (e.g., Paquier et al., 1992; Rapin et al., 1977; Soprano et al., 1994). The degree of cognitive impairment found in this study may be regarded as relatively severe, compared to some other studies (Bulteau et al., 1995; Hirsch et al. 1995, Soprano et al., 1994; Van Dongen et al., 1995). Due to the rarity of the LKS and the small number of participants in most studies, some variations in findings are likely to occur.

In contrast to previous reports (Bishop, 1985; Lanzi et al., 1994; Paquier et al., 1992; Soprano et al., 1994) we did not find the severity of the aphasia to be clearly related to age at onset of the LKS. For example, the child with the earliest onset and the child with the latest onset had equally severe aphasia.

Aphasia was also not clearly related to the presence of CSWS—it remained relatively constant even when CSWS and nonverbal signs fluctuated. The possibility still exists that a CSWS type of epileptic activity may have caused permanent damage earlier on to brain structures necessary for receptive language.

To summarize, the only significant impairment that was found in all our cases of LKS was the receptive aphasia and its secondary expressive consequences. In addition, our LKS patients showed variable patterns of nonverbal deficits, the most frequent being oral motor dyspraxia, present to some degree in all children, retardation, including visuomotor problems, manual dyspraxia, hyperkinesia, and conduct problems.

Dissociation of Environmental Sounds Discrimination and Phonological Discrimination

The receptive aphasia was analyzed further in order to determine whether the core deficit was a generalized auditory agnosia or a language-related agnosia; that is, a specific phonological decoding deficit. For this purpose the perception of environmental sounds and of phonological and linguistic stimuli were compared.

There was a striking dissociation between the auditory discrimination of environmental sounds and phonological discrimination. The 4 children with total receptive aphasia were unable to identify any of the test words, yet they correctly identified 34.5 to 65.5% of the environmental sounds (animal sounds, car, shower, etc.). V.N., who had regained some speech, identified only 31% of the test words but 74% of the environmental sounds. The same dissociation appeared when the test scores were converted to z scores based on the distributions of normal 5-to-6-year-olds. The performance of the normal children was similarly distributed on both tasks. Thus, the difference in the results on the environmental sounds and the phonological discrimination tasks was not due to differences in degree of difficulty.

The clinical observations may shed further light on the nature of the receptive aphasia in LKS. The ability of 4 children to recognize a few words seemed to be based on a recognition of some salient auditory features—vowel patterns, sounding consonants, or the tonal gestalt of a word—rather than on an ability to decode speech. For example, M.J. learned to recognized the word *kisse* (kitten) because she seemed to recognized the sound /ss/. V.N.'s production of color and number words was very distorted but the most salient vowel patterns were recognizable. He invariably said,

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for example, "*muuka*" for *musta* (black) and "*kihme*" for *sininen* (blue). In the former word the vowel combination *u–a* and in the latter word *ii–e* were almost correctly reproduced. Thus, in the absence of severe oral apraxia, the basis for his recognition of the color words was probably the recognition of these vowel patterns. A relative facility of discriminating vowels as compared to consonants in LKS was previously reported by Frumkin and Rapin (1980) and Notoya et al. (1991). In the phrases produced by V.N. as well as the other children, prosody and intonation were adequate, which suggested that such auditory patterns were also perceived.

It should be pointed out that the results of the LKS children on the environmental sounds task were below the mean for the normal 5-to-6-year-olds. At the time of the present assessments this impairment was, however, relatively mild—3 children performed less than 1 standard deviation below the mean. Thus, their auditory perception should be sufficient for adequate speech decoding and reception. However, all children had exhibited loss of reaction to sound at onset of the disease and would have scored very poorly on this task during acute phases. Thus, the auditory agnosia may be considered as an indication of the severity of the condition.

To conclude, the deficit underlying the receptive aphasia in LKS seems to be a specific deficit of the decoding of speech rather than a generalized auditory agnosia. These results confirm the views of Rapin et al. (1977) and Lanzi et al. (1994) of verbal auditory agnosia in LKS.

The question still remains whether this deficit in decoding of speech is related to its acoustic characteristics, or to selective impairment of specialized linguistic capacities. According to Tallal et al. (1993) phonemic discrimination may be more demanding than discrimination of environmental sounds, due to the rapid phonemic transitions characteristic of speech. Thus, the deficit underlying the receptive aphasia in the children with LKS could still be an impairment of auditory perception that particularly affects the decoding of speech. In severe cases the impairment might also affect the perception of nonverbal sounds.

Support for the auditory perceptual view was provided in the study by Klein et al. (1995), who found that the perception of both nonverbal and phonological stimuli in patients with residual LKS was slow. Similarly, in the MEG recordings of our patients with LKS, the source of epileptic activity was localized to the Sylvian fissure bilaterally (Table 1; see Paetau, 1994). This finding indicates that the primary auditory cortex would be involved, in support of an auditory perception hypothesis. However, other studies have found sources of epileptic activity in wider, perisylvian regions (speech regions) on intracranial EEG recordings (Cole et al., 1988; Morrell et al., 1995), indicating that a primarily Sylvian abnormality may not be crucial in the pathogenesis of LKS.

The alternative interpretation is that the primary deficit is a linguistic deficit of phonological decoding. This position assumes that there are processes that are selectively specialized in the decoding of linguistic stimuli. An impairment of these processes is not related to the duration of the stimuli, although rapid presentation may put an additional load on the decoding. The disruption of specifically linguistic processes would then produce a primarily aphasic disorder.

There are also arguments for this view. First, language processing includes components that may not be explained by perceptual processes, such as semantic retrieval and comprehension of syntactic structures and logical concepts. In less severe cases of LKS syntactic and dysnomic problems, echolalia, paraphasias, or short-term memory problems have been reported (Paquier et al., 1992; Rapin et al., 1977; Soprano et al., 1994; Van Hout, 1992). It seems logical to assume that receptive language would also be based on a specifically linguistic capacity for decoding speech. Second, sign language acquisition was poorer than expected in most children of our study (see Table 2), which would also point to a linguistic-level disorder. Third, the children were exposed to intense rehabilitation of word recognition. Yet, even the best performing child, V.N., failed to learn to recognize most phonologically simple words, or to write to dictation simple alphabetic sounds (e.g., /O/ or /E/) without visual cues. In contrast, the children were able to discriminate between many relatively similar environmental sounds, such as those of a motorcycle and a helicopter (K.S., V.N., and J.M.), without training. Thus, the evidence in favor of a specific phonological decoding deficit, a phonological agnosia, seems compelling.

Yet, for more conclusive evidence in this question the auditory and the phonological discrimination tasks should be equalized not only with respect to difficulty but also with respect to complexity and duration of individual sounds. The phonological stimuli consisted of relatively complex and dynamic patterns of sounds, characteristic of speech, whereas the environmental sounds were more static. An equalization of the stimuli could be achieved by using more complex auditory stimuli, for example rhythms or melodies, or more static phonological stimuli, for example vowels or sounding consonants. Still, the present comparison and clinical observations undisputedly demonstrated that the children with LKS were able to perceive an astonishing number of environmental sounds, whereas they were almost completely unable to identify word segments, simple words, and vowels that were dictated.

Gathercole and Baddeley (1990) put forward a hypothesis that impaired phonological memory may be a primary deficit in the language disorders of children. We obtained some evidence that this hypothesis might not apply to LKS. V.N. was able to repeat four-digit series, yet he was not able to repeat even one-syllable nonwords. Number words up to 10 belonged to V.N.'s active vocabulary. Evidently, he was able to preserve in working memory words that he clearly perceived. Further, according to normative data (Korkman, 1988a), repeating four-digit series in Finnish corresponds to the level of normal 4-year-olds, and would therefore be sufficient for basic expressive and receptive language. Thus, the results of V.N. do not support a hypothesis of a poor phonological memory as a basis for language impairment in LKS. Indirectly, this finding provides additional support of a phonological agnosia as the basis for receptive aphasia in LKS.

The present results have some practical implications. First, they suggest that it may be a good intervention strategy to build on the perception of salient acoustic characteristics of speech by way of improving recognition of vowel patterns and sounding consonants as well as tonal and prosodic patterns in important everyday expressions. For example, if some capacity for auditory discrimination has been present, we have recommended that parents and teachers present certain standard phrases and words in context, frequently, and by exaggerating a certain intonation, certain vowels or consonants, especially when there is some evidence that the child perceives them. The auditory perception may be aided by lip reading and gestures. The aim has been to teach the children to recognize common phrases and commands. Second, the results support the idea of assessing nonlinguistic auditory discrimination in LKS children to evaluate the possibilities for such intervention. An environmental sounds discrimination test could be developed for this purpose.

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REFERENCES

Ansink, B.J., Sarphatie, H., & Van Dongen, H.R. (1989). The Landau-Kleffner syndrome: Case report and theoretical considerations. *Neuropediatrics*, *20*, 170–172.

Beaumanoir, A. (1985). The Landau-Kleffner syndrome. In J. Roger, C. Dravet, M. Bureau, F.E. Dreifuss, & P. Wolff (Eds.), *Epileptic syndromes in infancy, childhood and adolescence* (pp. 181–191). London: John Libbey.

Beery, K.A. (1983). *Visual–motor integration*. Cleveland, OH: Modern Curriculum Press.

Bishop, D.V.M. (1985). Age of onset and outcome in “acquired aphasia with convulsive disorder” (Landau-Kleffner Syndrome). *Developmental Medicine and Child Neurology*, *27*, 705–712.

Bulteau, C., Plouin, P., Jambaqué, I., Renaux, V., Quillerou, D., Boidein, F., & Dulac, O. (1995). Aphasia epilepsy: A clinical and EEG study of six cases with CSWS. In A. Beaumanoir, M. Bureau, T. Deonna, L. Mira, & C.A. Tassinari (Eds.), *Continuous spikes and waves during slow sleep* (pp. 199–201). London: John Libbey.

Bureau, M. (1995). Outstanding cases of CSWS and LKS: Analysis of the data sheets provided by the participants. In A. Beaumanoir, M. Bureau, T. Deonna, L. Mira, & C.A. Tassinari (Eds.), *Continuous spikes and waves during slow sleep* (pp. 213–216). London: John Libbey.

Cole, A.J., Anderman, F., Taylor, L., Olivier, A., Rasmussen, T., Robitaille, Y., & Spire, J.P. (1988). The Landau-Kleffner syndrome of acquired epileptic aphasia: Unusual clinical outcome, surgical experience, and absence of encephalitis. *Neurology*, *38*, 31–38.

De Renzi, E. & Faglioni, P. (1978). Normative data and screening power of a shortened version of the Token Test. *Cortex*, *14*, 41–49.

Deonna, T. & Roulet E. (1995). Acquired epileptic aphasia (AEA): Definition of the syndrome and current problems. In A. Beaumanoir, M. Bureau, T. Deonna, L. Mira, & C.A. Tassinari (Eds.), *Continuous spikes and waves during slow sleep* (pp. 37–45). London: John Libbey.

Dugas, M., Gerard, C.L., Franc, S., & Lecendreux, M. (1995). Late onset of acquired epileptic aphasia. In A. Beaumanoir, M. Bureau, T. Deonna, L. Mira, & C.A. Tassinari (Eds.), *Continuous spikes and waves during slow sleep* (pp. 143–147). London: John Libbey.

Dugas, M., Masson, M., Le Heuzey, M.F., & Regnier, N. (1982). Aphasie ‘acquise’ de l’enfant avec épilepsie (syndrome de Landau et Kleffner); Douze observations personelles [‘Acquired’ aphasia in the epileptic child (Landau-Kleffner syndrome): Twelve personal observations]. *Revue Neurologique*, *138*, 755–780.

Feekery, C.J., Parry-Fielder, P., & Hopkins, I.J. (1993). Landau-Kleffner syndrome: Six patients including discordant monozygotic twins. *Pediatric Neurology*, *9*, 49–53.

Frumkin, B. & Rapin, I. (1980). Perception of vowels and consonant-vowels of varying duration in language impaired children. *Neuropsychologia*, *18*, 443–454.

Gathercole, S.E. & Baddeley, A.D. (1990). Phonological memory deficits in language disordered children: Is there a causal connection? *Journal of Memory and Language*, *29*, 336–360.

Hirsch, E., Maquet, P., Metz-Lutz, M-N., Motte, J., Finck, S., & Marescaux, C. (1995). The eponym ‘Landau-Kleffner syndrome’ should not be restricted to childhood-acquired aphasia with epilepsy. In A. Beaumanoir, M. Bureau, T. Deonna, L. Mira, & C.A. Tassinari (Eds.), *Continuous spikes and waves during slow sleep* (pp. 57–62). London: John Libbey.

Klein, S.K., Kurzberg, D., Brattson, A., Kreuzer, J.A., Stapells, D.R., Dunn, M.A., Rapin, I., & Vaughan, H.G. (1995). Electrophysiologic manifestations of impaired temporal lobe auditory processing in verbal auditory agnosia. *Brain and Language*, *51*, 383–405.

Korkman, M. (1988a). NEPSU. *Lasten neuropsykologinen tutkimus. Uudistettu versio* [NEPSY. Neuropsychological assessment for children. Rev. ed.]. Helsinki, Finland: Psykologien kustannus.

Korkman, M. (1988b). NEPSY—an adaptation of Luria’s investigation for young children. *Clinical Neuropsychologist*, *2*, 375–392.

Korkman, M. (1988c). *NEPSY—A proposed neuropsychological test battery for young developmentally disabled children. Theory and evaluation*. Published dissertation. University of Helsinki. Helsinki: Yliopistopaino.

Korkman, M. (1995). A test-profile approach in analyzing cognitive disorders in children: Experiences of the NEPSY. In M.G. Tramontana & S.R. Hooper (Eds.), *Advances in child neuropsychology* (Vol. 3, pp. 84–116). New York: Springer-Verlag.

Korkman, M. (in press). Applying Luria’s diagnostic principles in the neuropsychological assessment of children. *Neuropsychology Review*.

Korkman, M., Appelqvist, K., Granström, M-L., Paetau, R., & Lindahl, E. (1996). Auditory–phonemic agnosia in the Landau-Kleffner syndrome. *Epilepsia*, *37*, 29–30.

Korkman, M. & Häkkinen-Rihu, P. (1994). A new classification of developmental language disorders (DLD). *Brain and Language*, *47*, 96–116.

Korkman, M., Kirk, U., & Kemp, S.L. (1997). *NEPSY. Lasten neuropsykologinen tutkimus* [NEPSY. A developmental neuropsychological assessment]. Helsinki, Finland: Psykologien kustannus.

Korkman, M., Kirk, U., & Kemp, S.L. (1998). *NEPSY. A developmental neuropsychological assessment*. San Antonio, TX: The Psychological Corporation.

Landau, W.M. & Kleffner, F.R. (1957). Syndrome of acquired aphasia with convulsive disorder in children. *Neurology*, *7*, 523–530.

Lanzi, G., Veggiotti, P., Conte, S., Partesana, E., & Resi, C. (1994). A correlated fluctuation of language and EEG abnormalities in a case of the Landau-Kleffner syndrome. *Brain and Development*, *16*, 329–334.

Leiter, R.G. (1969). *Leiter International Performance Scale*. Chicago: Stoelting.

Liberman, A.M. (1992). Plausibility, parsimony, and theories of speech. In J. Alegria, D. Holender, J. Junca de Morais, & M. Radeau (Eds.), *Analytic approaches to human cognition* (pp. 25–40). Amsterdam: Elsevier Science Publishers.

Liberman, A.M. & Mattingly, I.G. (1985). The motor theory of speech perception revised. *Cognition*, *21*, 1–36.

Maquet, P., Hirsch, E., & Metz-Lutz, M-N. (1995). Regional cerebral glucose metabolism in children with deterioration of one or more cognitive functions and continuous spike and wave discharges during sleep. *Brain*, *118*, 1497–1520.

Marien, P., Saerens, J., Verslegers, W., Borggreve, F., & De Deyn, P.P. (1993). Some controversies about type and nature of aphasic symptomatology in Landau-Kleffner's syndrome: A case study. *Acta Neurologica Belgica*, *94*, 183–204.

Metz-Lutz, M-N., Guillere, F., Verceuil, L., Hirsch, E., & Marescaux, C. (1995). Five cases of continuous spikes and waves during slow sleep. In A. Beaumanoir, M. Bureau, T. Deonna, L. Mira, & C.A. Tassinari (Eds.), *Continuous spikes and waves during slow sleep* (pp. 205–207). London: John Libbey.

Morrell, F., Whisler, W.W., Smith, M.C., Hoeppner, T.J., Pierre-Louis, S.J.C., Kanner, A.M., Buelow, J.M., Ristanovic, R., Bergen, D., Chez, M., & Hasegawa, H. (1995). Landau-Kleffner syndrome: Treatment with subpial intracortical transection. *Brain*, *118*, 1529–1546.

Mouridsen, S.E., Videbaek, C., Sogaard, H., & Andersen, A.R. (1993). Regional cerebral blood-flow measured by HMPAO and SPECT in a 5-year-old boy with Landau-Kleffner syndrome. *Neuropediatrics*, *24*, 47–50.

Nakano, S., Okuno, T., & Mikawa, H. (1989). Landau-Kleffner syndrome EEG topographic studies. *Brain and Development*, *11*, 43–50.

Notoya, M., Suzuki, S., Furukawa, M., & Enokido, H. (1991). A case of pure word deafness associated with Landau-Kleffner syndrome: A long-term study of auditory disturbance. *Auris, Nasus, Larynx*, *18*, 297–305.

O'Tuoama, L.A., Urion, D.K., Janicek, M.J., Treves, S.T., Bjornson, B., & Moriarty, J.M. (1992). Regional cerebral perfusion in Landau-Kleffner syndrome and related childhood aphasia. *Journal of Nuclear Medicine*, *33*, 1758–1765.

Paetau, R. (1994). Sounds trigger spikes in the Landau-Kleffner syndrome. *Journal of Clinical Neurophysiology*, *11*, 231–241.

Paquier, P.F., Van Dongen, H.R., & Loonen, C.B. (1992). The Landau-Kleffner syndrome or 'acquired aphasia with convulsive disorder.' Long-term follow-up of six children and a review of the recent literature. *Archives of Neurology*, *49*, 354–359.

Rapin, I., Mattis, S., Rowan, A.J., & Golden, G.G. (1977). Verbal auditory agnosia in children. *Developmental Medicine and Child Neurology*, *19*, 192–207.

Rapin, I. (1995). Autistic regression and disintegrative disorder: How important the role of epilepsy? *Seminars in Pediatric Neurology*, *2*, 278–285.

Roulet, E., Deonna, T., Gaillard, F., Peter-Favre, C., & Despland, P.A. (1991). Acquired aphasia, dementia, and behavior disorder with epilepsy and continuous spikes and waves during sleep in a child. *Epilepsia*, *32*, 495–503.

Soprano, A.M., Garcia, E.F., Caraballo, R., & Fejerman, N. (1994). Acquired epileptic aphasia: Neuropsychologic follow-up of 12 patients. *Pediatric Neurology*, *11*, 230–234.

Tallal, P., Miller, S., & Fitch, R.H. (1993). Neurobiological basis of speech: A case for the preeminence of temporal processing. In P. Tallal, A.M. Galaburda, R.R. Llinas, & C. von Euler (Eds.), *Temporal information processing in the nervous system. Special reference to dyslexia and dysphasia. Annals of the New York Academy of Sciences. Vol. 682*. (pp. 27–47). New York: New York Academy of Sciences.

Tharpe, A.M. & Olson, B.J. (1994). The Landau-Kleffner syndrome: Acquired epileptic aphasia in children. *Journal of the American Academy of Audiology*, *5*, 146–150.

Van Dongen, H.R., Meulstee, J., & Paquier, P.F. (1995). Two contrasting cases of children with acquired epileptic aphasia and continuous spikes and waves during sleep. In A. Beaumanoir, M. Bureau, T. Deonna, L. Mira, & C.A. Tassinari (Eds.), *Continuous spikes and waves during slow sleep* (pp. 137–140). London: John Libbey.

Van Hout, A. (1992). Acquired aphasias in children. In S.J. Segalowitz & I. Rapin (Vol. Eds.), F. Boller & J. Grafman (Series Eds.), *Handbook of neuropsychology. Vol. 7. Child Neuropsychology* (pp. 139–161). Amsterdam: Elsevier.

Veggiotti, P., Farinotti, L., Partesana, E., & Lanzi, G. (1995). A case of Landau-Kleffner syndrome with annual relapses. In A. Beaumanoir, M. Bureau, T. Deonna, L. Mira, & C.A. Tassinari (Eds.), *Continuous spikes and waves during slow sleep* (p. 203). London: John Libbey.

Wechsler, D. (1984). *Wechsler Intelligence Scale for Children–Revised käsikirja* [manual, in Finnish]. Helsinki, Finland: Psykologien kustannus.

Appendix

DESCRIPTION OF (1) THE PICTURE-SOUND LOTTO GAME USED TO ASSESS ENVIRONMENTAL SOUNDS DISCRIMINATION, AND (2) THE AUDITORY ANALYSIS OF SPEECH TEST, USED TO ASSESS PHONOLOGICAL DISCRIMINATION

The Picture–Sound Lotto Game

The material is a commercial toy, a lotto game.[1] It consists of 5 wooden plates with 9 pictures arranged in 3 × 3 arrays, 45 small bricks with the same pictures as in the wooden plates, and an audiotape with 45 sounds that match the pictures. The sounds on the audiotape are live recordings. The five plates are placed one at a time together with the corresponding nine bricks in front of the child, the bricks between the picture plate and the child. In each item, the child is to select the brick that corresponds to a sound on the tape and place it on top of the identical picture on the plate. The items are presented below. In parentheses are 16 items that were presented but not included in the total score.

Plate 1	Plate 2	Plate 3	Plate 4	Plate 5
laughter	organ	(dog)	alarm	siren
snoring	violin	cock	(hammer)	(thunder)
crying	guitar	(pig)	phone	ship siren
coughing	base	bird	(dripping tap)	motorcycle
(kissing)	(triangle)	(horse)	shower	(church bell)
whistling	accordion	sheep	typewriter	(wind)
blowing nose	trumpet	cow	vacuum cleaner	car
(singing)	(drums)	(hen)	sewing machine	helicopter
(babbling)	piano	(cat)	(flushing toilet)	train

The Auditory Analysis of Speech Test

The test consists of nine syllable blending and seven word completion tasks. Both types of tasks are preceded by a learning trial. The learning trials and examples of items are given below.

Syllable blending tasks

In the learning trial the child is shown a page with three pictures and is instructed: *Here you see "tukka"* (hair), "*sukka*" (a stocking), *and "kukka"* (a flower). *Which one of them do you think I mean when I say "tuk–ka?"* The phonemes *tuk* and *ka* are said with a pause equal to the time for inserting an *and* (*tuk*-and-*ka*). The child is to point to the appropriate picture. The nine test items consist of similar two-syllable words, presented by separating the syllables by a pause. Each item is introduced by showing a new three-picture page and by giving the full names of the depicted objects. Thereafter, the target word is presented with the two syllables pronounced separately. Examples of the items are as follows:

- Item 1. *Here you see "varvas"* (a toe), "*hammas*" (a tooth) *and "lammas"* (a sheep). *Where is the "lam–mas"*?
- Item 4. *Here you see "torvi"* (a horn), *"torni"* (a tower) *and "korva"* (an ear). *Where is the "tor–vi"?*
- Item 9. *Here you see "tuuli"* (wind), "*tuoli*" (a chair) *and "tuli"* (a fire). *Where is the "tu–li"?*

Word completion tasks

In the learning trial the child is shown a new page with three pictures and is instructed: *Here you see "karhu"* (a bear), "*jalka*" (a foot), *and "parta"* (a beard). *In which word is there a part such as "–hu"?* If the child fails, the task is explained and demonstrated. Each test item is introduced by showing a new three-picture page and by first giving the full names of the depicted objects. Examples of the items are as follows:

- Item 10. *Here you see "lentokone"* (an airplane), *"televisio"* (a television set) *and "elefantti"* (an elephant). *In which word is there a part such as "–ento"?*
- Item 16. *Here you see "jäätelö"* (ice cream), *"käärme"* (a snake) *and "lääkäri"* (a doctor). *In which word is there a part such as "–käri"?*

One point is given for every correctly performed item.

Recognition of complete words

After presenting the Auditory Analysis of Speech test, a testing-the-limits procedure was performed with the children with LKS in order to establish to what extent they were helped by facilitating the tasks. The child was asked to point to one of the three alternative pictures when providing the full name. Examples are as follows:

- Item 1. *Where is the "hammas"* (tooth)?
- Item 4. *Where is the "torni"* (tower)?
- Item 16. *Where is the "jäätelö"* (ice cream?)

[1] *Sound lotto* produced by ILKA Förlagsprodukter AB, Malmö, Sweden.