

# Long-term course of Landau-Kleffner syndrome: visuo-semantic and auditory aspects of comprehension

*Evolução da síndrome de Landau-Kleffner: aspectos visuo-semânticos e auditivos da compreensão*

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## SUMMARY

**Introduction.** Landau-Kleffner Syndrome is characterized by normal speech acquisition followed by epileptic seizures, receptive and expressive language deterioration coupled with agnosia for non-verbal sounds, having variable long-term evolution.

**Case Report.** It is described neurophysiologic and acoustic findings in a patient with Landau-Kleffner Syndrome, and correlate these with the results of a language evaluation carried out 7 years after the acute phase. It is performed Electroencephalography, Immitance Measurements, Basic Audiometry, Auditory Brainstem Response, Middle Latency Response, and P300. Language was evaluated by Boston Diagnostic Aphasia Examination. Electroencephalography was normal and audiologic evaluation revealed normal Immitance Measurements, Basic Audiometry and Auditory Brainstem Response values. An electrode effect was present in the left hemisphere in Middle Latency Response, and bilateral P300 latencies delayed on the right. Language evaluation showed severe receptive and expressive impairment, severe phonemic substitutions, which had an impact on social and academic levels. There were contextual and gestural non-verbal compensations, evidencing intellectual and cognitive domain preservation. **Conclusion.** This case illustrates the specific cerebral areas that can be damaged in patients with Landau-Kleffner Syndrome and which are demonstrable by clinical evaluation and proper neurophysiology studies, showing the importance of neurological, audiological, electrophysiological and language exams in a longitudinal follow up.

**Keywords:** Auditory evoked potentials. Electroencephalography. Hearing disorders. Landau-Kleffner Syndrome. Language. P300 event-related potentials.

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## RESUMO

**Introdução.** A síndrome de Landau-Kleffner é caracterizada pela aquisição normal da fala seguida de crises epiléticas, deterioração da linguagem receptiva e expressiva, juntamente com agnosia para sons não-verbais, havendo uma evolução variável a longo prazo. **Descrição de caso:** Foram descritos os achados audiológicos e neurofisiológicos em um paciente com SLK. Tais achados foram correlacionados com os resultados da avaliação de linguagem realizada 7 anos após a fase aguda. Foram realizados eletroencefalografia, medidas de imitância acústica, audiometria convencional, Potencial Evocado Auditivo de Tronco Encefálico, Potencial Evocado Auditivo de Média Latência e P300. A linguagem foi avaliada por meio do teste *Boston Diagnostic Aphasia Examination*. Os resultados da eletroencefalografia encontraram-se normais e a avaliação audiológica revelou resultados normais para as medidas de imitância acústica, audiometria convencional e Potencial Evocado Auditivo de Tronco Encefálico. Verificou-se efeito de eletrodo no hemisfério esquerdo no Potencial Evocado Auditivo de Média Latência, e atraso nas latências do P300 bilateralmente. Na avaliação da linguagem, verificou-se alteração severa tanto para a linguagem receptiva como expressiva, substituições fonêmicas severas, que tiveram impacto nos níveis social e acadêmico. Foram identificadas compensações gestuais e não-verbais, evidenciando preservação nos domínios cognitivo e intelectual. **Conclusão.** Este caso ilustra áreas cerebrais específicas que podem estar comprometidas em pacientes com SLK e que podem ser demonstradas por meio da avaliação clínica e estudos neurofisiológicos específicos, mostrando a importância das avaliações neurológica, audiológica, eletrofisiológica e de linguagem no acompanhamento longitudinal desses pacientes.

**Unitermos:** Potenciais evocados auditivos. Eletroencefalografia. Transtornos da audição. Síndrome de Landau-Kleffner. Linguagem. Potencial evocado P300.

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**INTRODUCTION**

Landau-Kleffner Syndrome (LKS) is characterized by normal speech acquisition followed by epileptic seizures, receptive and expressive language deterioration coupled with agnosia for non-verbal sounds, having variable long-term evolution. Electroencephalography (EEG) abnormalities are seen in temporal areas, especially during sleep, where verbal language and acoustic information is processed<sup>1-3</sup>.

The clinical picture fluctuates and presents differences over its several phases. Some improvements during development, and after the use of antiepileptic drugs and corticosteroid, are observed. Speech-language therapy, special schools<sup>4</sup> and psychopedagogic management<sup>5</sup> are also indicated. We describe the neurophysiologic and acoustic findings of a patient with LKS, and correlate these with the results of language evaluation 7 years after the acute phase.

**METHOD**

A male right-handed patient presented epileptic seizures, speech difficulties and hyperactivity at the age of 3 years. EEG showed epileptiform discharges in the parietotemporal area. He received antiepileptic drugs and corticosteroids leading to control of seizures, but minimal speech improvement. The subject has been attending regular school since age 3, experiencing learning difficulties, and is currently in the first grade. Improvements in behavior and non-verbal communication in parallel with medical, speech-language and psycho-pedagogic treatment were observed. According to parent information, he reacts to and identifies environment sounds, and is learning Brazilian sign language.

We performed EEG, Immitance Measurements, Basic Audiometry and Auditory Evoked Responses such as brainstem, middle and long latency studies (Auditory Brainstem Response, Middle Latency Response, Cognitive Potential – P300) (Biologic equipment Traveler-Express). Language was evaluated by a number of stimuli from the Boston Diagnostic Aphasia Examination<sup>6</sup>.

**RESULTS**

**Behavioral, Electroacoustic and Electrophysiological Assessments**

a) *EEG*: at onset of the disease, paroxystic activity in the parietotemporal areas; at the latest evaluation, 7 years after, normal in wakefulness and sleep.

b) *Immitance measurements*: type A tympanometric curve with presence of acoustic reflexes in both ears.

c) *Tonal audiometry*: normal hearing thresholds in both ears.

d) *Vocal audiometry*: not performed because of poor cooperation.

e) *Auditory Brainstem Response (ABR)*: presence of waves I, III, V at 80 decibels hearing level for clicks, with absolute and interpeak I-III, III-V and I-V latencies within normal limits in both ears.

f) *Middle Latency Response (MLR)*: absolute latency of the positive wave Pa within normal limits in both ears; significant difference between the amplitude of the wave Pa for the electrode located in C3 (condition C3/A2=2.13 microvolts) when compared with the electrode located in C4 (C4/A2=6.54 microvolts), revealing an electrode effect in the left hemisphere; significant difference between the amplitudes of the wave Pa for the electrode located in C4 ipsilateral (C4/A2=6.54 microvolts) and contralateral (C4/A1=0.86 microvolts), revealing a left ear effect (figure 1).

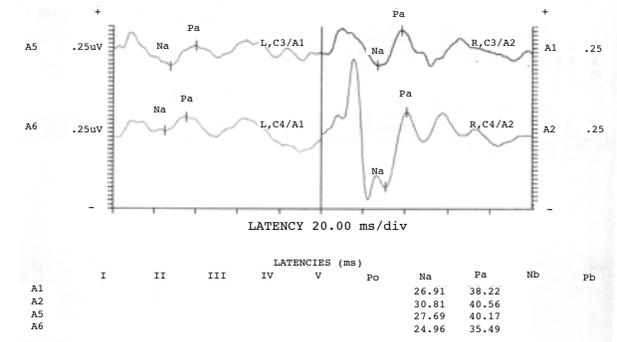
g) *Cognitive Potential (P300)*: present in both ears with prolonged absolute latency (P300=410 milliseconds) in the right ear (figure 2).

**Speech Evaluation**

a) *Conversation and spontaneous speech*

The subject responded to non-verbal communication cues and reacted to gestures and expressions, although he did not understand propositions and did

**Figure 1.** Auditory Middle Latency Response with latency and amplitude values of the Pa wave for the conditions C3/A1, C4/A1, C3/A2, C4/A2.



not respond to verbal calling. When requested to participate in direct interlocution, he produced gestures and unintelligible verbal utterances, which were key words with intense phonetic-phonological alterations, and surprising preservation of the melodic contour, accent and rhythm of his native region.

b) *Word comprehension*

He correctly designated colors and numbers when prompted by acoustic stimuli. Body parts, objects and letters were not recognized, and other complex tests could not be applied.

c) *Oral expression*

The subject repeated isolated words with phonetic-phonological approximations and descriptions of objects (*marrom*/brown = *maku*; *cadeira*/chair = *kinká* – gestures, *kareira*; *que*/that = *te*; and named: *casa*/house = *gaga*; *banco*/bench = *kinká* – *sentar* = to sit).

d) *Written comprehension and expression*

He recognized frequently occurring words (such as *house*, *boll*, *João Paulo*, *kite* and *water*), wrote numerical series (1–11), his own name and simple words. In addition, the subject completed over-trained sentences with errors (*a piscina é azul* = *azul*) and wrote text segments with juxtaposed pronouns and substantives but without syntax, besides unintelligible segments.

## DISCUSSION

D. evolved with a profile akin to the severe cases described in the literature, in which there was a recovery of the capacity to respond to non-verbal sounds of the environment<sup>7</sup>. However, this ability was not enough to achieve the phonemic discrimination that presupposes quick acoustic change perception<sup>8</sup>. The severe impairment in performing auditory-verbal pairing tasks supports our hypothesis that D. presented a disorder in this aspect.

The oral language disorders were severe, where language was limited to the emission of lexi-

cal fragments. In the few repetitions, D. tended to maintain the articulation zones (visual cues support), hindering other phoneme features. Generally, intonation contour preservation is rarely mentioned<sup>9</sup> in the evolution of severe cases with LKS. In the context of damage, in the MLR and P300, traces of improved right hemisphere integrity can be observed where this could explain regional melodic accent preservation in D.'s emissions.

The written language disorder presented by D. has also been verified in cases where the syndrome occurred in the period before the acquisition of this modality<sup>10</sup>.

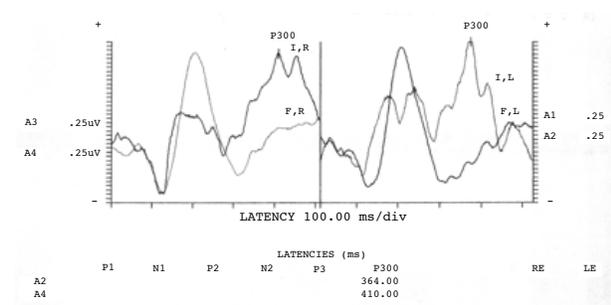
Aphasia with intense reception obliteration retains characteristics of childhood manifestation<sup>11</sup>. The lack of language prevented sophisticated analysis from verifying semantic system integrity. However, reactions to situations involving visual input (gestures and objects) allow us to infer its preservation. Dissociation in the ability to identify stimuli via visual and auditory input was noted: our patient demonstrated a greater facility for identifying colors and numbers, in contrast to identifying other items. Colors and numbers are coded by predominantly visual experiences where this probably took place prior to the disease. Letter coding is similar, although this occurred later, when D. already presented the hearing impairment.

Although it is recognized that the prognostic of childhood aphasia is favorable for majority of cases, Martins *et al*<sup>12</sup> identified the type and location of the aphasia presented by our patient as factors that could have a negative influence on recovery.

In the electrophysiological exams, we were able to verify absence of brainstem auditory pathway damage in the ABR, a finding in agreement with that described by Msall *et al*<sup>13</sup>, along with an alteration in late auditory evoked potentials<sup>14</sup>.

In the present study, alterations in the MLR were found, more specifically an electrode effect in the left hemisphere and a left ear effect, as well as in the cognitive potential, with a prolonged latency of the P300 wave in the right ear. According to Musiek *et al*<sup>15</sup>, patients with central nervous system disorder present altered MLR in the same side of the lesion. This alteration can be observed by the electrode effect, due to the fact that this electrode is closer to the lesion. On the other hand, the same author affirms that the ear effect may be observed *ipsi* and contralaterally to the lesion. These results are compatible with the deficits detected in the language reception and emission tasks in our patient, which, according to the specialized lit-

**Figure 2.** Auditory Late Response – P300 (Cognitive Potential) with the latency values of P300 wave in left and right ears.



erature, may be related to the left hemisphere damage. The electroencephalogram presented alteration over a long period, normalizing after 7 years. The abnormalities observed were seen in the posterior region of the hemispheres (parietotemporal area), which agrees with the other findings.

The results of the electrophysiological and language evaluations are coherent, given that D. presents left hemisphere dysfunction – evidenced by results obtained in the MLR and P300 (left temporal lobe lesion, one of the areas responsible for language and auditory information processing<sup>2</sup>) and also presents speech comprehension difficulties, as well as speech production alteration.

## CONCLUSION

This case demonstrates the importance of neurological, audiological, electrophysiological, and language exams in a longitudinal follow up, in order to relate semiologic data and foresee prognostics, essential requirements to enable precise definitions to be made from a clinical standpoint.

The syndromic etiology, bilateral affection, extension and duration of the lesion, allied with the absence of special education opportunities indicate an unfavorable prognosis. Indeed, this may have limited the development of compensatory language abilities in this child. Functional compensation related to the anterior regions of the brain, such as context management, and those related to posterior regions, such as use of visual cues in phoneme imitation, were also evidenced.

## REFERENCES

1. Landau WM, Kleffner FR. Syndrome of acquired aphasia with convulsive disorder in children. *Neurology* 1957;7(8):523-30.
2. Seri Stefano, Cerquiglioni A, Pisani F. Spike-induce interference in auditory sensory processing in Landau-Kleffner Syndrome. *Electroencephalogr Clin Neurophysiol* 1998;108:506-10.
3. Pablo MJ, Valdizan JR, Carvajal P, Bernal M, Peralta P, Saenz de Cabezón A. Landau-Kleffner Syndrome. *Rev Neurol* 2002;34(3):262-4.
4. Baynes K, Kegl JA, Brentari D, Kussmaul C, Poizner H. Chronic auditory agnosia following Landau-Kleffner Syndrome: a 23-year outcome study. *Brain Lang* 1998;63:381-425.
5. Campos-Castello J. Epilepsies and language disorders. *Rev Neurol* 2000;30(suppl 1):S89-S94.
6. Goodglass H, Kaplan E, Barresi B. The assessment of aphasia and related disorders. Philadelphia: Lippincott Williams & Wilkins, 2001, 85 p.
7. Dugas M, Masson M, Le Heuzey MF, Regnier N. Aphasie acquise de l'enfant avec épilepsie (Syndrome de Landau et Kleffner). *Rev Neurol (Paris)* 1982;138:755-80.
8. Pardo PJ, Makela JP, Sama M, Hari R. Neuromagnetic measurements of hemispheric differences in auditory processing of frequency and amplitude modulations. *Abstr Soc Neurosci* 1994;20:325.
9. Doherty CP, Fitzsimons M, Asenbauer B, McMackin D, Bradley R, King M, *et al.* Prosodic preservation in Landau-Kleffner syndrome: a case report. *Eur J Neurol* 1999;6(2):227-34.
10. Denes G, Balliello S, Volterra V, Pellegrini A. Oral and written language in a case of childhood phonemic deafness. *Brain Lang* 1986;29(2):252-67.
11. Van Hout A. Review of research on the clinical presentation of acquired childhood aphasia. *Acta Neurol Scand* 1997;95(4):253-5.
12. Martins IP, Ferro J. Afasia adquirida na criança: aspectos clínicos e de prognóstico. In: Mansur LL, Rodrigues N (eds). *Temas em Neurolingüística*. São Paulo: Sociedade Brasileira de Neuropsicologia; 1990, pp. 92-102.
13. Msall M, Shapiro B, Balfour PB, Niedermeyer E, Capute AJ. Acquired epileptic aphasia. *Clin Pediatr* 1986;25:248-51.
14. Mel'nichuk PV, Zenkov LP, Morozov AA, Kogan EI, Averyanov YuN. Neurophysiological mechanisms of aphasia in epilepsy. *Neurosci Behav Physiol* 1991;21:380-5.
15. Musiek FE, Baran JA, Pinheiro M. *Neuroaudiology case studies*. San Diego: Singular Publishing Group; 1994, 279 p.