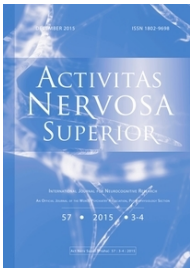

Activitas Nervosa Superior 2015, 57, No. 3-4

ANS: Journal for Neurocognitive Research

Journal Homepage:

www.activitas.org

IDEAS & PERSPECTIVES

LANDAU-KLEFFNER SYNDROME AND SWEARING

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Abstract

Landau-Kleffner syndrome (LKS) is a rare form of epilepsy diagnosed as acquired aphasia alternatively as acquired verbal agnosia co-occurring with epileptic seizures. This article provides an overview of some relevant case studies of Landau-Kleffner patients and also some neuro-measurement studies of the neurophysiology of the disease. Recently there is no evidence whether the epileptic seizures in LKS are located in basal ganglia, limbic or subcortical circuits involved in swear words processing.

Key words: *Aphasia; Epilepsy; Landau-Kleffner syndrome; Swearing*

1. INTRODUCTION

Using swear words has a unique status and the taboo words have clear social, ethical, political and also juridical implications. (Montagu, 1967). Furthermore, swearing has obviously also its prominent status in the field of various language neuropathologies and may be considered as an "automatic speech" (Jackson, 1932; Lancker, 1988; Cole, 1982; Lancker, Bella, 1996; Lum, Ellis, 1994), for example it may be also related to manifestation of the Gille de la Tourette syndrome (Fisarova, 1972) and Landau-Kleffner syndrome, a specific form of epilepsy which appears only in infants and often co-occurs among others with acquired aphasia and verbal agnosia (Landau & Kleffner, 1957).

As an example four reported case studies of LKS patients will be mentioned (Shoumaker, 1974; Rapin et al., 1973; Worster-Dought, 1971, Landau & Kleffner, 1957) with a purpose to study some links of these observations of using swear words in aphasic patients (A. Smith, 1996; O. Zangwill, 1967; Cole, 1982; Lancker & Nicklay, 1992). In those studies, the same brain regions were affected as in the LKS patients.

2. WHAT IS THE LANDAU-KLEFFNER SYNDROME?

Landau-Kleffner syndrome (LKS), also called *acquired epileptic aphasia*, is a rare neurological syndrome. Acquired aphasia and epileptic seizures, and often sleep-activated EEG paroxysms are typical characteristics of Landau-Kleffner syndrome. The seizures are mostly partial, but

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Received July 1, 2015; accepted June 29, 2015; *Act Nerv Super 57(3-4), 122-126; ISSN-1802-9698*

also the tonic-clonic seizures were reported. These symptoms occur in infants mainly in age from three to seven years, more often in boys than girls. This syndrome manifests in critical periods of language acquisition (Deonna, 1991). Although this syndrome causes language disturbances, non-verbal abilities such as drawing, geometrical and spatial reasoning and mathematical computing are usually preserved or less damaged than verbal abilities (Deonna, 1991). Frequently reported disturbances are hyperactivity, aggression and anxiety (Eslava-Cobos, & Meija, 1997). Due to some behavioral abnormalities the LKS sometimes may be wrongly diagnosed as a form of autism. (Chez et al., 2006)

The assumption that epileptic seizures and the acquired speech and comprehension disorders may correlate, is not confirmed. Nevertheless, some researches are thought that the epileptic activity may play a crucial role in the language disorders (Deonna & Roulet-Perez, 2010; Pearl et al., 2001).

3. LANDAU-KLEFFNER SYNDROME, REPORTED CASE STUDIES

Basic overview of the cases reported in studies of the LKS patients is in Table 1 (Shoumaker, 1974; Rapin et al., 1973; Worster-Dought, 1971, Landau & Kleffner, 1957).

Study	Number of patients	Sex		Onset of aphasia	Epileptic seizures preceding aphasia	Co-occurrence of aphasia and seizures	Aphasia preceding epileptic seizures	Reco-very
		M	F					
Landau & Kleffner (1957)	6	2	4	3,5 – 9 years	3	1	2	3
Shoumaker et al. (1974)	4	4	0	5-6 years	2	0	2	1
Worster-Drought (1971)	14	7	7	3-7 years	Not mentioned	Not mentioned	Not mentioned	5
Rapin et al. (1973)	4	4	0	2 years	4	0	0	1

3.1. Seizures

In the majority of cases, the seizures occurred before any difficulties with language. In the Shoumaker's study (1974) is the loss of some communication skills reported 6 months before the first observed seizure and before the seizure the EEG data provided normal results. In the case study made by Rapin et al. (1973) no clinical seizures were reported, but the EEG was abnormal (Table 1).

3.2. Speech and comprehension disabilities

Poor comprehension of language in the majority of studies was the initial observation of the speech disabilities. This symptom is usually followed by difficulties and problems with language expression but some patients were able to comprehend simple language successfully (Worster-Dought, 1971), whereas others became inert to verbal stimuli (Shoumaker et al., 1974; Landau & Kleffner, 1957).

Due to the obliviousness of children to verbal commands, some of them were thought to be deaf which is very important information for differential diagnosis of certain forms of aphasia of Wernicke's type[19] and some patients reported[20] that they were able to understand verbal stimuli were, but he wereable to think the words they would like use. In this context, interesting data repovided Zovari and Choyakh (1997), who measured LKS patients by means of brainstem auditory evoked responses (BAER).[21] In general, the children became partially mute and may produce strange sounds. Some studies also reported children who were later able to recover the speech ability completely (Worster-Drought, 1971, Landau and Kleffner, 1957; Shoumaker et al., 1974; Rapin et al., 1973)

4. NEUROPATHOLOGY OF LANDAU-KLEFFNER SYNDROME

Some studies also reported classification of seizures, EEG findings and clinical outcomes in these patients (Proposal for Classification, 1985; Dugas, 1982; Cole et al., 1988) and these studies frequently reported focal sharp EEG abnormalities.[22] Some data also suggest that magnetoencephalography may enable location of the epileptic focus in these patients (Lewine et al., 1999; Wolff et al., 2005; Neville, 1999). These studies also suggest that and an important role may play superior temporal gyrus.[23] Other data suggest that some of those LKS diagnosed patients had also secondary focal point in the superiortemporal sulcus or the fusiform gyrus which also play a role in the process of recognizing emotional signals of faces (Catani et al., 2005; Vigneau et al., 2006; Leppänen & Nelson, 2009) which consequently may be related to autistic symptoms (Lewine et al., 1999).

One striking example is a patient from the Smit's (1996) and Zangwill's (1997) studies, who got the whole left hemisphere removed. The surgery was done to stop infiltrating the developing tumor into further parts of his brain. The removal included limbic forebrain, all left cerebral lobes and the thalamus. After several months the patient was not able to name any of showed object, he just could repeat with a huge effort some monosyllabic words. The most preserved and spontaneous speech was swearing, namely "*Goddammit"* and "*shit"*.

In a study made by Cole (1982) five aphasic patients were studied who have through a celebrovascular accident affected the interior parietal lobe and Wernicke's territory of their brains. In all of those patients, the swear words and cursing were preserved and the propositional speech connected with swear words was preserved too.

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[19] This is a receptive aphasia in which people with this condition are not able to comprehend language in both, the spoken and written form. In the many cases, the normal syntax, grammar and intonation are preserved, but their expressions are not meaningful. (LaPointe, 2005)

[20] 15 year was the age of the patient when he was post-researched and when his language abilities were returned.

[21] BAER is a test of both the ear and the brain. Brainstem auditory evoked response (BAER) test measures how your brain processes the sounds you hear. The BAER test records your brainwaves in response to clicks or other audio tones that are played for you. (Brainstem Auditory Evoked Responses, 23.6.2015)

[22] The role of EEG, and in particular the focus on focal abnormalities, has evolved over time. In the past, the identification of focal EEG abnormalities often played a key role in the diagnosis of superficial cerebral mass lesions. (Fahoum et. al, 2012) Focal sharp waves are (in the case of epileptiform) the interictal marker of a patient with epilepsy and are the EEG signature of a seizure focus. (Tedrus et al., 2012)

[23] Superior temporal gyrus has is a crucial structure making auditory processing possible. It is rapidly involved in function of language in patients who evince to have impaired vocabulary. (Bigler et al. 2007)

Another study (Lancker & Nicklay, 1992) reported a patient who was diagnosed with global aphasia.[24] Even though the propositional speech was permanently lost, using the swear words have been preserved. Observations in those studies suggest that the swear words may be stored in different parts of human brain than normal speech which is in agreement with certain findings of patients with Gilles de la Tourette syndrome (Darley et al. 1975; Stehlíková, 2001) who frequently manifest coprolalia (Shapiro & Shapiro, 1977).

5. CONCLUSION

In summary, recent findings indicate that patients with Landau-Kleffner syndrome are able to produce swear words even in cases when they acquire speech aphasia or verbal auditory agnosia. There is no evidence that the epileptic seizures are located in basal ganglia, limbic or subcortical circuits involved in swear word processing. Because LKS manifests only in infants there is a question if they process automatic speech and swear words in the same way as adults these findings may provide very important and interesting findings about various dimensions and emotional aspects of the human speech and language processing.

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[24] After a celebrovascular accident. This involved frontal temporal and parietal areas of the left hemisphere.

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