Brain and Cognition 99 (2015) 32–45

Contents lists available at ScienceDirect

Brain and Cognition

journal homepage: www.elsevier.com/locate/b&c

Evidence for normal letter-sound integration, but altered language pathways in a case of recovered Landau–Kleffner Syndrome

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ARTICLE INFO

Article history:
Received 26 September 2014
Revised 2 March 2015
Accepted 14 July 2015
Available online 29 July 2015

Keywords:
Landau–Kleffner Syndrome
fMRI
DTI
Arcuate fasciculus
Auditory cortex
Audiovisual
Multisensory integration

ABSTRACT

Landau–Kleffner Syndrome (LKS) is a rare form of acquired aphasia in children, characterized by epileptic discharges, which occur mostly during sleep. After normal speech and language development, aphasia develops between the ages of 3–7 years in a period ranging from days to months. The epileptic discharges usually disappear after reaching adulthood, but language outcomes are usually poor if no treatment focused on restoration of (non-) verbal communication is given. Patients often appear deaf–mute, but sign language, as part of the treatment, may lead to recovery of communication. The neural mechanisms underlying poor language outcomes in LKS are not yet understood. In this detailed functional MRI study of a recovered LKS patient – that is, a patient no longer suffering from epileptic discharges, audiovisual multi-sensory processing was investigated, since LKS patients are often proficient in reading, but not in speech perception. In the recovered LKS patient a large difference in the neural activation to auditory stimuli was found in the left versus the right auditory cortex, which cannot be attributed to hearing loss. Compared to healthy proficient readers investigated earlier with the same fMRI experiment, the patient demonstrated normal letter-sound integration in the superior temporal gyrus as demonstrated by the multi-sensory interaction index, indicating intact STG function. Diffusion Tensor Imaging (DTI) based fiber tracking in the LKS patient showed fibers originating from Heschl's gyrus that seem to be left-right inverted with respect to HG fiber pattern described in the literature for healthy controls. In the patient, in both hemispheres we found arcuate fibers projecting from (homologues of) Broca's to Wernicke's areas, and a lack of fibers from arcuate left inferior parietal and sylvian areas reported in healthy subjects. We observed short arcuate segments in the right hemisphere. Although speculative, our results suggest intact temporal lobe processing but an altered temporal to frontal connectivity. The altered connectivity might explain observed short-term verbal memory problems, disturbed (speech) sound-motor interaction and online feedback of speech and might be one of the neuronal factors underlying LKS.

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1. Introduction

Landau–Kleffner Syndrome (LKS) was first described in 1957 (Landau & Kleffner, 1957) as a "form of acquired auditory aphasia in children", and it is characterized by either partial or total loss of auditory comprehension or inability of environmental sound discrimination (Steinlein, 2009). Together with aphasia symptoms of epilepsy start to occur. Patients suffer from epileptic discharges occurring in both temporal regions, which become almost continuous during slow-wave sleep (Steinlein, 2009), and LKS is thus considered as a subtype of continuous slow waves during sleep (CSWS). Most epileptic symptoms tend to disappear when the patients enter adolescence.

When symptoms of LKS first arise, the patient seems to be deaf, but audiograms are usually normal (Feekery, Parryfielder, & Hopkins, 1993). The cause of LKS, its pathophysiology and the neural mechanisms behind the language disorders in LKS are not yet understood (Fandiño, Connolly, Usher, Palm, & Kozak, 2011; Hirsch et al., 2006).

LKS children initially acquire speech and language in a normal way. At the age of three to seven years, they develop (semi-) acute aphasia in a period of days to months. Aphasia is mostly receptive (phonological decoding) and develops into expressive impairments at a later stage (Lanzi, Veggiotti, Conte, Partesana, & Resi, 1994).

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http://dx.doi.org/10.1016/j.bandc.2015.07.003
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Auditory agnosia, i.e. the inability to distinguish sounds is also reported in LKS. Currently the language disorders in LKS are therefore classified as verbal agnosia or auditory agnosia (Cockerell, Bølling, & Nakken, 2011). It is unclear whether the inability to distinguish sounds reflects a specific phonological decoding deficiency or whether there is a more general auditory-perceptual processing deficit affecting the analysis of any type of sounds (verbal or non-verbal) (Pedro & Leisman, 2005).

As well as language disorders other higher-order cognitive functions such as memory and attention can be affected as well. These deficits may arise as a consequence of epileptic seizures and/or their associated treatment. These cognitive dysfunctions are often associated with dramatic changes on a behavioral level. Two-thirds of studied LKS patients show signs of behavioral change including aggression, attention and concentration disorders, as well as resistance against behavioral change (disengagement, task switching), echolalia, echopraxia, and even psychotic behaviors (Zivi, Broussaud, Daymas, Hazard, & Sicard, 1990). This might also be correlated to an inability to link sounds to facial expressions and/or body language. Normally, humans are very good in connecting emotional sounds to facial expressions (i.e. sound of laughter with a smiling face (De Gelder & Vroomen, 2000) or in associating body language with emotional sounds (Van den Stock, Grèzes, & de Gelder, 2008)), but if this connection is lost, emotional sounds will not be interpreted correctly and will likely lead to fear in the patient.

Behavioral treatment of any form of audio-visual communication usually leads to a reduction of problematic behavior (Appleton, 1995; Deonna, Prelaz-Girod, Mayor-Dubois, & Roulet-Perez, 2009) and quality of life is greatly improved. The degree of recovery is determined by the time of onset, the response to anti-epileptic medication and the severity of communication problems. To our knowledge there are no reports of full recovery and individual variation is high; while some patients may have permanent language disorder, some regain much of their language capabilities.

Neuro-anatomical abnormalities in LKS patients have been reported based on volumetric MRI analysis. These abnormalities are mainly gray matter volume reduction in bilateral superior temporal areas, foremost in planum temporale and superior temporal gyrus (Takeoka et al., 2004).

This anatomical finding received support by functional studies. Dysfunction of superior-temporal and perisylvian areas, both during the active phase as well as long lasting dysfunction was reported using a single or four word repetition task during positron emission tomography (PET) (Majerus et al., 2003). The tip of the left temporal lobe was also found to be inactive in a LKS case study using resting-state PET (Shiraishi, Takano, Shiga, Okajima, & Sudo, 2007).

Electroencephalography (EEG) studies during the active epileptic phase have shown that auditory information (speech and natural sounds) seems to be processed normally within the brain stem, as the EEG is normal for the early (brain stem related) auditory processing time window (Steinlein, 2009).

With regard to more complex auditory integration, LKS patients in the active and recovered phase suffer from permanent dysfunction of associative auditory cortex as measured in a dichotic listening task during EEG recordings (Metz-Lutz, de Saint Martin, Hirsch, Maquet, & Marescaux, 1999; Plaza, Rigoard, Chevrie-Muller, Cohen, & Picard, 2001; Wioland, Rudolf, & Metz-lutz, 2001). This dysfunction is expressed as unilateral dichotic extinction, contra-lateral to the epileptic focus (Wioland et al., 2001). Furthermore, short-term phonological memory is impaired in LKS, even in patients who have recovered fairly well (Majerus et al., 2003).

It seems that language functions in LKS are reorganized from the left to the right hemisphere, as was demonstrated in a single case follow-up fMRI study (Datta et al., 2013). The initial active epileptic focus for this patient was localized with source EEG in the left fronto-centro-temporal area. The language network detected by fMRI was also left lateralized initially, but seems to have transferred into the right hemisphere as observed by follow-up fMRI.

There is no impairment in the visual domain in LKS; one important observation in the clinics in contact with LKS patients is that many of them show astonishing recovery of communication skills using alternate means, such as non-verbal signing (Deonna et al., 2009; Perez et al., 2001; Stefanatos, 2011). Moreover, the ability to learn and use written language is found not to be impaired (Denes, 1998). The patient described in the latter study had "a flawless performance in lexical decision tasks, as well in the written naming subtest of BDAE (..) and in written naming following semantic cuing" (Denes, 1998).

The question we address here is what is the neural mechanism that allows LKS patients to compensate for their sudden loss of auditory language functions? If the typical route to access meaning about objects/events via spoken language is blocked, how does the brain accommodate the plastic changes needed to recover communication skills?

The present study simultaneously examined auditory, visual and audiovisual processing abilities in LKS using the smallest possible units of spoken and written language (speech sounds and letters). When healthy children learn how to read, a crucial first step is to learn the correspondence between written letters and speech sounds (Van Atteveldt, Formisano, Goebel, & Blomert, 2004). This integration of written letters and letter sounds (multimodal integration) is acquired with high efficiency in a normal developing child (Van Atteveldt et al., 2004) but may be impaired in LKS.

The goal of the present investigation was twofold. Firstly, we aimed to examine the unimodal (only written letters/only letter sounds) and multimodal (simultaneous written letters and letter sounds) integration of letters and sounds using functional magnetic resonance imaging (fMRI). We studied the case of an adult female LKS patient whose communication abilities were largely restored following long-term multi-sensory intervention training (see case description). We compared the results of uni- and multimodal processing found in the patient with the results from an identical experiment in healthy controls ($N = 12$) (Blau, Van Atteveldt, Ekkebus, Goebel, & Blomert, 2009). Second, the language disorders might be related to abnormalities in connectivity of the temporal and associative cortices in the epileptic phase (Hirsch et al., 2006), therefore a second major goal of this paper is to investigate the white matter pathways in temporal and associative cortex in a LKS patient after the active epileptic phase with DTI. Specifically, the connections arising from the primary auditory cortex were reconstructed to investigate how Heschl's gyrus is connected to higher language areas. In addition, we investigated the arcuate fasciculus, as this fiber bundle connects the most relevant functional areas of interest related to language and information integration between temporal lobe and frontal lobe. Specifically we looked into connections between inferior frontal gyrus and superior temporal gyrus/superior temporal sulcus.

2. Materials and methods

2.1. Subjects

2.1.1. Recovered LKS patient

We present a case of a 27-year old female "I.", diagnosed with LKS. At present she lives a full live as an educated married woman with a full time job. In the cause of this investigation she had an audiometric evaluation (Viataal Audiometric Centre NL 2009),

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