Architecture of Human Language from the Perspective of a Case of Childhood Aphasia — Landau–Kleffner Syndrome

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This paper addresses Landau–Kleffner syndrome (LKS), a childhood aphasia, from the perspective of I-language and the critical period for first language acquisition. LKS involves a language disorder and behavioral disturbances resembling autistic spectrum disorders due to electroencephalographic abnormalities with continuous spike-and-waves during sleep over the temporal regions. Comparing LKS with other childhood syndromes, the architecture of language is explored through elucidating the linguistic mechanisms behind the language disorder in LKS on the basis of Hickok & Poeppel's (2007) dual-stream model of speech processing. It is claimed that early onset LKS provides further support for the critical period for first language acquisition and modularity of mind (the faculty of language), and that verbal auditory input during the critical period is most crucial for language recovery and development in LKS. Considering that electroencephalographic abnormalities affect cognitive/motor functions, ameliorating neural dysfunction in the affected brain areas with proper application of transcranial direct current stimulation is recommended.

Keywords: critical period; dual-stream model of speech processing; electroencephalographic (EEG) abnormalities; Landau–Kleffner syndrome (LKS); transcranial direct current stimulation (tDCS)

1. Introduction

Landau–Kleffner syndrome (LKS) is a clinically rare language disorder of acquired childhood aphasia involving epilepsy (with or without clinical seizures) that emerges with epileptiform electroencephalographic (EEG) abnormalities over the

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temporal lobes.[1] The child with LKS first undergoes a period of normal development of language, but usually after the onset of the disorder, the 'language attained' starts regressing.[2] In LKS, typically, both language comprehension and language production acutely or insidiously become virtually impossible, often leading to apparent deafness and mutism in children suffering from it (for more details, see e.g. Gordon 1990; Tharpe & Olson 1994; Kaga 1999, 2011; Pearl et al. 2001). Moreover, the EEG abnormalities cause behavioral and psychiatric disturbances such as hyperactivity, aggressive behavior, impulsivity, and attentional problems, which resemble autism spectrum disorders (Stefanatos 2011; Mikati et al. 2010; see also section 2.1.3).

Ever since Landau & Kleffner's (1957) first report of the syndrome, over 200 cases (Stefanatos 2011) have been reported in the literature (see also Ansink et al. 1989; Tharpe & Olson 1994, and references therein). Specifically, 81 cases were reported between 1957 and 1980, and 117 cases between 1981 and 1991 (Makati et al. 2010). This implies the disorder is rare; but it has become the most frequently described form of acquired aphasia in children (Stefanatos 2011) and many more cases than reported should definitely exist (Mikati et al. 2010). Since LKS is often mistaken for psychiatric or developmental language disorders (Campos & De Guevara 2007), the frequency of the clinical condition is underestimated (Stefanatos at el. 2002).

LKS has been extensively investigated in fields of medicine such as child neurology, and various accompanying symptoms of LKS including the electrophysiological states in patients with LKS have been identified (for reviews, see e.g. Pearl et al. 2001; Stefanatos 2011; Stefanatos & DeMarco 2011). On the other hand, little attention has been paid to LKS in contrast to other language disorders in the field of biolinguistics (see Benítez-Burraco 2013, who briefly mentions this clinical case under the category of 'specific language impairments').[3] There seem to be two main reasons for this state of affairs.

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1 Throughout the discussion, we will use 'childhood aphasia' as a cover term to describe the state of aphasia in children in general. In the literature, the term 'acquired childhood aphasia' is sometimes employed to refer to cases where children sustained language deficits due to some lesions (localized or diffuse) in language areas after they acquired the core of their first language (Van Hout 1997). Thus, in acquired childhood aphasia, there are clear postnatal organic lesions involved in the brain. Generally, children with acquired childhood aphasia have intelligence of normal development except for the language domain. On the other hand, there are cases in which there is no organic lesion that was incurred in the brain, but children suffer from loss of the use of their first language in the process of first language acquisition or after the core of their first language was acquired, presumably due to some congenital brain malfunctioning. This latter type corresponds to what Tuchman (1997) calls 'acquired epileptiform aphasia', as some kind of epileptiform brain activity is typically implicated here. Although the former type of childhood aphasia is quite similar to aphasia in adults (Van Hout 1997), the latter type of childhood aphasia is peculiar to children and LKS is an exemplar (Landau & Kleffner 1957). In what follows, when we refer to acquired childhood aphasia, we will use the term 'ordinary child aphasia' for the sake of simplicity.

2 See Gordon (1997 and references therein) for opposing case reports in which three quarters of LKS patients exhibit language disturbances before the onset of the syndrome. Stefanatos (2011) also points out that LKS can occur with pre-existing language problems as well.

3 See Tsimpli et al. (in press) for detailed systematic discussion on language pathology, which deals with representative language-related pathological conditions other than LKS in the framework of Universal Grammar (UG). See also Benítez-Burraco (2016) for a biolinguistic approach to representative language disorders other than LKS in clinical linguistics.

First of all, LKS itself is a relatively rare clinical syndrome among children (Office of Rare Diseases 2008, cited in Stefanatos 2011). Second, if LKS happens to children, there are many cases where it comes after the onset of the critical period (Lenneberg 1967), with initially normal first language acquisition, followed by the syndrome of childhood aphasia, and then possibly later disappearance of the symptom within the critical period. This corresponds to the case of 'ordinary LKS' (see also fn. 5 and section 2.3). Thus, the state of childhood aphasia looks just temporary and so does not seem to matter much (but see the discussion in section 3.3 about the relevance of ordinary LKS to the concepts of modularity of mind and modularity of the faculty of language).

The primary aim of this paper is, therefore, to bring more attention of the biolinguistic community to this childhood aphasia by investigating it particulary from the perspective of I-language and the critical period hypothesis (Lenneberg 1967).[4] We will focus on what we call 'early LKS'—a sub-type of LKS in which language regression can start as early as at around the age of 18 months before the solid establishment of the core of the first language.[5] Differentiating early LKS from autism spectrum disorders (ASD), particularly autistic regression (AR), is especially significant because it would contribute to avoiding misdiagnosing of patients with LKS as having such developmental disorders as ASD/AR.

Furthermore, early LKS proves to be a quite interesting case in considering the nature of human language, if children with early LKS eventually (re)-start producing their first language while comprehending it at the same time with surprising speed and grammaticality, because the period of childhood aphasia lasts relatively for a long time until recovery, if any. We will submit that the notion of early LKS plays a pivotal role in elucidating the architecture of human language (and cognition) in connection with modularity of mind, modularity of the faculty of language, and a certain version of the critical period hypothesis for first language acquisition (see the discussion in section 3).

This paper is organized as follows. Section 2 attempts to lay out the fundamental characteristics of LKS, while comparing it with other syndromes such as ordinary child aphasia and ASD, especially AR, as well as the age-specific epileptic syndrome called 'benign childhood epilepsy with centrotemporal spikes' and 'continuous spike-and-waves during sleep'. Section 3 addresses some biolinguistic considerations concerning the critical period hypo-thesis for first language acquisition and modularity of the faculty of language as well as modularity of mind, while elucidating the linguistic mechanisms behind verbal auditory agnosia and loss of expressive speech in LKS. Section 4 discusses some implications for biolinguistic research, medical intervention, treatment, and research, and developmental and educational therapy for children with LKS. Section 5

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4 I-language is an abbreviation, where 'I-' stands for individual/internal/intensional, as originally proposed in Chomsky (1986). On this conception, human language is regarded as a brain-internal biological system. We will assume that this conception of human language is fundamentally correct throughout this paper.

5 Given that a child will acquire the core linguistic competence by around 3 years of age (Pinker 1994; O'Grady 2005) and that the beginning age of the peak incidence of LKS is also 3 years of age (see section 2.1.1 for more), as a first approximation, we will define *early LKS* as LKS with its onset before 3 years of age. Just for expository purposes, we will refer to the remaining cases of LKS as 'ordinary LKS' in what follows (see section 2.3 for discussion).

concludes this paper. In a modest attempt in this direction, we will address issues related to the architecture of human language in connection with LKS, while pointing out the significance of studying LKS for the purpose of investigating the biological nature of human language. This in turn could lead to shedding new light on the nature of LKS, and, hopefully, to discovering its cure eventually.

2. Landau–Kleffner Syndrome (LKS)

LKS is a label for the observed symptomatology of a kind of childhood aphasia acquired in the course of development (= ontogeny), presumably caused by more than one etiology with various degrees of deficits and recovery (see e.g. Mikati et al. 2010; Stefanatos 2011; Stefanatos & DeMarco 2011). In addition to an acquired aphasia, LKS has two other main symptoms: EEG abnormalities with continuous spike-and-waves during sleep often accompanied by epileptic seizures, and certain particular behavioral disturbances. We will first explain the cardinal characteristics of this childhood language disorder in light of medical/clinical, linguistic, and behavioral profiles, and later compare LKS with other clinical cases for the sake of more precise understanding of the childhood language disorder to lay out the background for discussion in section 3.

2.1. Fundamental Characteristics of LKS

2.1.1. Medical/Clinical Profile

Although the exact etiology (or etiologies) of LKS still remain unknown, rather diverse and relatively common clinical cases such as encephalitis, hemophilus influenza, and meningitis have been reported as possible causes in the literature (Mikati et al. 2010; see also Pearl et al. 2001 for a review).[6] Both males and females are equally affected by LKS, with the male to female ratio of approximately 2 to 1. Although LKS-affected children with the onset of the disorder from 3 to 8 years old account for 80% of this clinical syndrome (Kaga 2000), its onset ranges from 18 months to 13 years, with its peak incidence between 3 and 7 years (Tharpe & Olson 1994; Temple 1997; Uldall et al. 2000).[7] According to Stefanatos (2011), the recent deviation of the onset is usually between 2 and 7 years of age, ranging from 18 months to 14 years.

Unlike ordinary child or adult aphasia, no consistent organic brain lesion site has been found in children with LKS so far (Gordon 1990; Deonna 1991). Computed tomography and magnetic resonance imaging findings on patients with LKS are normal, while single photon emission computed tomography and positron emission tomography studies show temporal lobe abnormalities in brain

[6] Other clinical cases reported in the literature as possible causes of LKS are the following: abnormal zinc metabolism, toxoplasmosis, neurocysticercosis, temporal astrocytoma, temporal ganglioglioma, subacute sclerosing panencephalitis, inflammatory demyelinating disease, a genetic predisposition, and mitochondrial respiratory chain-complex I deficiency (see e.g. Pearl et al. 2001; Kang et al. 2006, and references therein).

[7] See Uldall et al. (2000) for a case report of LKS with onset at 18 months. Onset as early as 18–22 months and as late as 13–14 years has also been reported (see Stefanatos 2011 and references therein).

perfusion and glucose metabolism—decreased perfusion and hypometabolism, respectively (DaSilva et al. 1997; Pearl et al. 2001, and references therein).[8]

A particularly significant fact is that patients with LKS suffer from abnormal epileptiform electrical activity in the brain occurring particularly during sleep (Patry et al. 1971), which is related to the existence of paroxysmal EEG abnormalities and acquired aphasia in the LKS-affected patients (Pearl et al. 2001 and references therein; see also Stefanatos 2011). Epilepsy is a disorder of electrical activity in the brain consisting of the sudden temporary abnormal hypersynchronous firing of a group of brain cells (neurons) (Deonna 2000).[9] Specifically, the epileptiform EEG abnormalities in LKS are caused by continuous spike-and-waves during sleep or electrical status epilepticus during sleep, during over 85% of the slow sleep period (Gordon 1997 and references therein) over the temporal (and/or parietal) regions (Deonna 1991), and in some studies with magnetoencephalography, the source of the epileptiform activity is more precisely located in the superior temporal gyri and sylvian fissure (Morrell & Lewine 1994; Paetau 1994; Morrell et al. 1995). EEG abnormality findings are the most striking during sleep, but awake EEGs obtained in the early stages of LKS may show isolated or unilateral perisylvian spike discharges, while sleep EEGs show extremely frequent or even constant bilateral electrocerebral seizure activity despite the absence of clinical seizures (Mantovani 2000). Moreover, EEG abnormalities are commonly recorded with the presence of bilateral discharges (Stefanatos 2011), which hinders 'plasticity' of the brain before lateralization and consequently leads to cognitive and behavioral disturbances as well as language deterioration (see also section 2.3 for more discussion).

As for seizures, 70% of LKS patients with the EEG abnormalities result in either clinical or sub-clinical epileptic seizures (Mikati et al. 2010). According to Stefanatos (2011), however, the presence of clinical seizures is not a necessary feature of LKS. Moreover, the clinical seizures are generally infrequent and LKS-related epilepsy can be easily controlled by a single anti-epileptic medication: benzodiazepines such as clobazam (Pearl et al. 2001), valproate, and ethosuximide (Mikati et al. 2010).[10] Since it is well-known that temporal lobe epilepsy is a kind of refractory epilepsy and is generally hard to control by a single anti-epileptic medication (see e.g. Helmstaedter et al. 2003 and references therein),

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8 At least some LKS patients are known to involve metabolic abnormalities, or hypometabolism in the brain (see DaSilva et al. 1997 and references therein), presumably due to malfunctioning of the relevant neuronal circuitry. If Kang et al. (2006) are correct in assuming that some sub-cases of LKS are related to mitochondrial respiratory chain-complex I deficiency, the relevant LKS-affected children would be likely to have metabolic problems and weight problems such as obesity. Taking vitamin substance such as L-carnitine, which helps fat turn into energy in mitchondria and facilitates energy metabolism in neurons (Kang et al. 2006), could be one solution.

9 See Jefferys (2010) for a detailed review of the history of our current understanding of the basic mechanisms of epilepsy and seizures, and Treiman (2001) for a concise explication of GABAergic mechanisms in epilepsy in particular. See also Buzsáki (2006) for detailed discussion on various issues on rhythms/oscillations of the brain.

10 The other pharmacological protocols include corticosteroids, adrenocorticotropic hormones, and intravenous immunoglobulin. Multiple subpial transection as a surgical treatment (Morrell et al. 1995) has also been administered for a subgroup of LKS patients (see Stefanatos 2011; Stefanatos & DeMarco 2011, and references therein).

this pharmacological characteristic could be one of the important clinical markers in making a correct diagnosis of LKS.

Furthermore, seizures in LKS characteristically cease by the beginning of adolescence (Honbolygó et al. 2005), while seizures in other clinical conditions do not necessarily have this property. Moreover, not only the seizures but also EEG abnormalities in LKS tend to disappear between the ages of 8 and 13 years (mean of 10 years) (Massa et al. 2000; Ramanathan et al. 2012), which can be another clinical marker for LKS.

As a consequence of the epileptiform activity over the temporal lobes, language regression, or an acquired aphasia with verbal auditory agnosia and loss of expressive speech, occur. Mikati et al. (2010: 259) explain that the International League Against Epilepsy defines LKS as "childhood disorder in which an acquired aphasia, multifocal spikes and spike and wave discharges are associated". Deonna (2000) also explains that epileptic activity in one or, more often, both cortical auditory areas in the temporal lobes results in an acquired auditory agnosia, or a failure of the brain to decode sounds. Thus, children with LKS are ultimately suspected to have hearing impairment (Mikati et al. 2010).

Originally, Landau & Kleffner (1957: 529) suggested that "persistent convulsive discharge in brain tissue largely concerned with linguistic communication results in the functional ablation of these areas for normal linguistic behavior" (see also Paquier et al. 1992). Recently, Stefanatos (2011: 964) has also expressed the view that "the aphasia is thought to result from a more protracted functional disruption of the neural substrate necessary for normal language caused by the persistent epileptiform discharges evident on the EEG". In this connection, it is to be noted that, as Mikati et al. (2010) report, improvement in EEG is associated with language restoration in LKS.

With regard to the prognosis of language regression in LKS, approximately 50% of patients recover fully while the remaining 50% recover partially or suffer from permanent aphasia/dysphasia (Mikati et al. 2010). This remarkable prognosis compared with other cases such as ASD/AR in terms of language restoration could stem from the fact that the EEG abnormalities, which affect language and other cognitive functions of LKS patients, tend to cease by puberty. Less favorable data show, however, that approximately two-thirds of LKS patients will remain with some persistent language disability and half are severely affected to the extent that they will never regain expressive language, while about one-third can recover from the language disorder (Msall et al. 1986; Paetau et al. 1991, and references therein). Even so, the feature of higher possibilities of perfect or partial language restoration of LKS patients has attracted attention from researchers in the field of medicine, neuropsychology, and child development.

2.1.2. Linguistic Profile[11]

LKS-affected children have language regression in both receptive and expressive linguistic abilities to varying degrees. Now, a question of vital importance is

[11] See Stefanatos & DeMarco (2011) for more detailed linguistic and other cognitive characteristics of a child with LKS based on a variety of neuropsychological evaluation results.

which stage(s) of language processing is/are affected in LKS. Given that approximately half of patients with LKS fully recover from the state of aphasia, it is unlikely that the core central system of language is impaired fatally. It is more likely that the more 'peripheral' system(s) of language could be affected in LKS. In the following, we will discuss what constitutes verbal auditory agnosia in detail and how it causes speech impairment of LKS patients.

Regarding the auditory dimension of LKS, as already mentioned in the last section, children with LKS often appear to have a hearing loss because of their reduced response to speech and even to environmental sounds (McAllister & Greathead 1991). Thus, the first symptom of the receptive language disorder in LKS is an apparent 'word deafness', or verbal auditory agnosia. This auditory agnosia can extend to familiar environmental noises such as ringing bells and phones. As Hurley & Hurley (2009) point out, because children with LKS fail to respond to linguistic and even environmental sounds, quite often, they will be judged to have been suffering from a hearing loss or may be misdiagnosed as having autism or other developmental disorders (see also Tharpe et al. 1991). The degree of the verbal auditory agnosia in LKS can deteriorate from a remaining ability to follow simple verbal commands into a total inability to comprehend any verbal input and total unresponsiveness (Tharpe & Olson 1994 and references therein). As Deonna (2000) remarks, prolonged disruption of the activity of auditory cortex during the critical period of language development can permanently impair some components of auditory function.

Nevertheless, pure-tone audiograms and brainstem auditory evoked responses are normal in children with LKS (see Denes et al. 1986; Paquier & van Dongen 1993; Pearl et al. 2001, and references therein). Furthermore, dichotic listening tasks show one-ear extinction contralateral to the affected temporal cortex due to the epileptic focus during the active phase of LKS, and long-latency auditory evoked potential testing with children having recovered from LKS demonstrates that LKS affects the associative auditory cortex in the temporal lobe (indicated by unilateral voltage reduction involving the N1c peak), while the primary auditory cortex in the temporal lobe remains intact (indicated by the normal N1b peak) (see Wioland et al. 2001 and references therein). Taken together, these facts seem to indicate that, although sequences of sounds reach the primary auditory cortex, they will not be further processed properly in the associative auditory cortex due to the long-term epilepsy-induced dysfunction of those language-related areas in LKS (see Rapin et al. 1977; Matas et al. 2008, and references therein).

Initially, the problematic level of verbal processing in LKS was thought to be the level for decoding of phonemes (Korkman et al. 1998), hence a problem of phonological processes. However, as Deonna (2000) suggests, given the fact that the acute phase of LKS can affect some children in such a way that they can recognize neither linguistic sounds nor non-linguistic environmental sounds (e.g., door bell and phone ringing), it seems that LKS will affect (a) much earlier stage(s) of auditory processing than phonological processing of linguistic input. Deonna (2000) also points out that linguistic sounds are much more complicated acoustically than environmental sounds, which explains why all children with LKS suffer from verbal auditory agnosia, while only sub-groups have difficulties

in recognizing environmental sounds. If this applies to LKS in general, and given the fact that LKS affects articulation at the same time, it seems quite natural to assume that the LKS-affected part for language comprehension is concerned with processes involved with auditory-articulatory phonetics rather than phonology proper. In fact, Vance et al. (1999: 546) note that "a deficit at one level may have consequences for processing at other levels". Thus, if spectrotemporal/auditory analysis of acoustical features of speech sounds is disturbed, the expected correct phonetic analysis of them could not be associated with corresponding phonological units, say, phonemes, which is crucially necessary for speech perception and comprehension. This view is compatible with our proposal that I-language, including phonology (cf. Berent 2013 on I-phonology), remains virtually intact in LKS (see section 3).

There are good reasons to believe that this is indeed the case. Plaza et al. (2001) report the case of a child with LKS with verbal short-term memory impairment and dissociation between efficient phonological ability and verbal auditory deficits. The patient dramatically recovered language and acquired the ability for reading and spelling. They conclude that the patient developed phonological ability from predominantly visual input and that the apparent verbal short-term memory impairment is due to deficits at the level of cortical auditory processing rather than at the level of phonological processing.

In addition, Boyd et al. (1996) examined a child with LKS during a multiple subpial transection operation to the left temporal lobe, by recording intraoperative event-related potentials with respect to the discrimination of phonemes (/ba/ vs. /ga/) in the course of electrocorticography. They found that the child maintains discrimination of phonemes despite the apparent global aphasia. At first blush, this observation seems to be at variance with our scenario about the deficit level in LKS, but it should be noted that the event-related potentials recording for the left temporal lobe in the patient in Boyd et al. (1996) was carried out by inserting an earphone into his right ear under an anesthetized condition. It seems plausible that, without any distraction due to anesthesia and with direct insertion of an earphone into his right ear, the acoustic signals might be more clearly and easily perceived and analyzed spectrotemporally than otherwise. Hence, the observed syllable discrimination between /ba/ and /ga/ based on the more or less successful phonetic analysis seems to be quite expected. If this were to hold, we can still maintain our scenario here.

Although Denes et al. (1986) use the terms such as 'phonemes', 'phonemic discrimination/identification', and 'phonological representations', it seems that malfunctioning of the phonetic system for analysis of acoustic signals rather than the phonological one is what is responsible for what they describe as 'childhood phonemic deafness'. In fact, they even use the term 'the phonetic level' when they explain about their patient's inability to discriminate or identify 'phonemes'. Interestingly, Denes et al. (1986) observe that, although brainstem auditory evoked responses and primary cortical auditory responses are normal, their patient with LKS exhibit an asymmetry with respect to discrimination/identification of segments: While the patient can easily discriminate/identify vowels in linguistic stimuli, he cannot discriminate/identify consonants in them. They claim that this asymmetry can receive a natural explanation in terms of the physi-

cal characteristics of the difference between vowels and consonants, by saying that "while natural vowels usually average 100 to 150 msec, consonants are characterized by rapid frequent changes within the first 40 msec of onset of the stimulus" (p. 264). If this is the case, it would provide a strong reason to believe that what is at stake in the language disorder in LKS is malfunctioning of spectro-temporal analysis of acoustic signals at the phonetic level.

Given that proper phonetic analysis of acoustic signals is a prerequisite for forming proper links with abstract phonological representations corresponding to the acoustic signals, our scenario here is compatible not only with Denes et al. (1986) but also Vance et al. (1999), who argue, on the basis of auditory processing tasks, that phonological representations are highly likely to be inaccurate or insufficiently specified in children with LKS, suggesting that "ongoing auditory processing difficulties, from the onset of LKS, will have inhibited the development of accurate and well-specified phonological representations" (p. 551).

Next, let us turn to the question of language production. If the processes for auditory phonetics are impaired in LKS, it is natural to assume that the processes for articulatory phonetics are also affected because articulation of speech sounds must be carried out via pairing of motor movements and phonetic specifications of each speech sound (see section 3 for details on the mechanism behind language production). With regard to supra-segmental aspects of speech in LKS-affected children, Matas et al. (2008) report a case of a LKS-affected patient with severe receptive and expressive language impairment. The patient "produced gestures and unintelligible verbal utterances, which were key words with intense phonetic-phonological alterations, and surprisingly preservation of the melodic contour, accent and rhythm of his native region" (p. 68). This kind of preservation of prosodic aspects of language shows that LKS does not affect the brain areas related to prosody (see also Landau & Kleffner 1957 for a case in point), as supported by the fact that "traces of improved right hemisphere integrity can be observed" in the patient on the basis of the middle latency response (MLR) and the cognitive potential (P300) (Matas et al. 2008: 69).

With respect to the semantic system, as Matas et al. (2008) point out, although the lack of full expressive language prevents us from analyzing the integrity of the semantic system in a sophisticated fashion, appropriate reactions to situations with visual input such as gestures and objects suggest the preservation of the semantic system (but see also the discussion on the effect of LKS in the thought system in section 3.3). Interestingly enough, the patient with LKS in Denes et al. (1986) maintains the abilities on lexical semantics, which is revealed through reading and writing tasks.

Finally, as for the syntactic system, we would like to suggest that LKS will not eradicate the potentiality of at least the core syntactic mechanism for building up hierarchically structured expressions. Recall that of all LKS-affected patients, approximately 50% recover fully while the remaining 50% recover partially or suffer from permanent aphasia/dysphasia (Mikati et al. 2010). Given this fact, the null hypothesis seems to be that the core syntax is not damaged in LKS but the degrees of manifestation of expressive language depend upon the degrees of availability of lexical items in the mental lexicon and/or proper functioning of the externalization system in the patients with LKS.

2.1.3. Behavioral Profile

Aside from the linguistic characteristics mentioned in the previous section, children with LKS will present associated behavioral disturbances as co-morbidity (see e.g. Landau & Kleffner 1957; Rapin 1995; Tuchman 1997; Pearl et al. 2001; Tharpe et al. 1991), as enumerated in (1):

(1) *Co-morbidity in LKS:*

a. hyperkinesis (= hyperactivity)
b. attention deficit
c. rage outbursts (= tantrums)
d. aggressiveness
e. autistic-like behaviors such as stereotypies (= persistent repetition of an act)
f. apparently poor 'social communication' skills
g. withdrawal
h. clumsiness of fine hand/finger movement (e.g., messy eating)

Such behavioral problems as in (1a–h) are, however, (at least partly) related to the existence of epilepsy (clinical or sub-clinical) in children with LKS (on this point, see Gordon 1990; Deonna 1991; Tharpe et al. 1991; Tuchman 1997). Deonna & Roulet-Perez (2010) actually refer to the possibility, though they do not ascertain, that the 'autistic' behavior is a reaction to the severe receptive language deficit, or an additional developmental comorbidity or the result of an epileptic activity involving not only language but also 'social brain' circuits.

In fact, Stefanatos (2011: 964) notes that LKS has come to be recognized as belonging to the so-called "*epileptic encephalopathies*, in which a deterioration of cognitive, sensory, and/or motor functions results from epileptic activity" (Nabbout & Dulac 2003) and that epileptiform discharges may have deleterious effects on psychological development in some developmental disorders such as ASD (Ballaban-Gil & Tuchman 2000). Stefanatos (2011: 976) also states that "epileptiform abnormalities are often bilaterally synchronous and have disruptive influences on the function of perisylvian cortex in both hemispheres, even if effects are often asymmetric" (see also the remark by O'Hare 2008 in section 2.3 below). He further states that "functional disruption of language cortex in perisylvian regions of temporal lobes might also impede nonlinguistic functions localized in the same areas" (p. 976). Thus, given that children with LKS suffer from expressive language disorder, it can be easily imagined that some motor-related regions of the brain that are relevant to both fine hand/finger movement and articulation/externalization of I-language are affected by LKS. Hence, (1h) can be regarded as resulting from fine motor/praxic difficulties. In fact, it has been suggested in the literature that the opercular syndrome of oromotor dysfunction involving EEG abnormalities is related with LKS (Shafrir & Prensky 1995; Tachikawa et al. 2001; Desal et al. 2013).

As such, it is further expected that all the behavioral problems in (1a–h) would be alleviated or cease to exist along with the disappearance of LKS-related epilepsy/epileptiform EEG abnormalities by adulthood (for discussion of a case report that seems to suggest this possibility, see Ansink et al. 1989).

To sum up, 'autistic' behavioral disturbances such as (1a–h) observed in LKS could take over the clinical manifestation (Campos & de Guevara 2007: 94) and result in key diagnostic dilemmas in clinical practice (Stefanatos 2011). More specifically, early onset of LKS before the solid establishment of the first language, accompanied by various behavioral disturbances as in (1a–h), causes difficulties with clinical diagnosis (Uldall et al. 2000), leading to failure in correctly differentiating patients with LKS especially from those with ASD/AR. In the next section, we will compare LKS with other clinical cases of interest and differentiate the former from the latter.[12]

2.2. Comparison between LKS and Other Clinical Cases

First, we will discuss LKS and ordinary child aphasia in terms of presence or absence of brain lesions. Then, we will compare LKS with *benign childhood epilepsy with centrotemporal splikes* (BECTS) and *continuous spike-wave during sleep* (CSWS) for better understanding of LKS from the perspectives of EEG patterns and other characteristics. Finally, we will highlight crucial differences between (early) LKS and ASD, or more specifically AR, which is extremely important in not only capturing the true nature of (early) LKS but also reducing clinical confusion between the two due to some apparently overlapping features (see e.g. Campos & de Guevara 2007; Penn et al. 1990; Stefanatos 2011; Uldall et al. 2000).[13]

2.2.1. Comparison of LKS and Ordinary Child Aphasia from the Perspective of Brain Lesions

In discussing the particular properties of LKS, it is useful to compare it with ordinary child aphasia. Relevant differences between the two can be summarized as follows (see Pearl et al. 2001 and references therein for more details):

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[12] Tuchman (1997) discusses not only LKS but also what he calls 'disintegrative epileptiform regression' and 'autistic epileptiform regression'. As Rapin (1995) points out, whether or not disintegrative epileptiform regression really constitutes a distinct separate entity from autistic epileptiform regression remains to be seen, so we will not address the dichotomy in question in this paper.

[13] See Rice (2016) for illuminating comparison among specific language impairment (SLI), ASD, attention-deficit/hyperactivity disorder (ADHD), and other conditions. In our comparison, we will not address the clinical condition of SLI because it is typically independent of epilepsy (Ooi 2011:125) and thus in principle readily differentiated from LKS. See Billard et al. (2009) for some discussion on SLI versus LKS. Furthermore, the contrast between LKS and ADHD is relatively clear. Though children with ADHD do not have any particular problems in non-verbal intelligence (Sumi 2015), in interpersonal social communication and pragmatic knowledge including theory of mind (Temple 1997), they are hyperactive, inattentive, and impulsive (Rice 2016), apparently on a par with children with LKS. However, LKS and ADHD are crucially differentiated in that LKS presents as the state of global aphasia, as stated above, but ADHD does not involve any developmental difficulties in linguistic comprehension and production (Redmond 2016). Therefore, since ADHD *per se* as a clinical condition does not involve any language disorder (Redmond 2016), we will not include it as part of our comparative discussion in the text.

(2) *Ordinary Child Aphasia:*

a. The underlying pathology is some organic lesions (localized or diffuse) in the brain.

b. Epileptic brain activity is *not* observed.

c. The general tendency of the prognosis is 'the earlier the onset of the language disorder is, the better its prognosis becomes'.

d. The language disorder occurs after the core linguistic knowledge has been acquired by a child.

e. The aphasic symptoms include receptive aphasia (Wernicke's aphasia), expressive aphasia (Broca's aphasia), anomic aphasia, conduction aphasia, and transcortical aphasia on a par with the case of adult aphasia, depending on which region(s) of the brain has/have been affected.

f. The recovery from the aphasic state is made possible via new formation of a neural network in the hemisphere where the lesion has been incurred or in the other hemisphere.

g. Only the language function is affected and the other cognitive functions remain basically intact.

(3) *LKS:*

a. The underlying pathology is not yet definitely identified, but it is not related to any organic lesions (localized or diffuse) in the brain.

b. Epileptic brain activity is clearly observed.

c. The general tendency of the prognosis is 'the earlier the onset of the language disorder is, the worse its prognosis becomes'.

d. The language disorder occurs either when the core linguistic knowledge has not yet been acquired completely by a child or after it has been acquired by a child.

e. The aphasic symptom is 'verbal auditory agnosia' usually along with reduction of expressive speech eventually to mutism, displaying virtually the state of 'global aphasia'.

f. The recovery from the aphasic state is *not* readily made possible via new formation of a neural network in either hemisphere, presumably, as long as there exist abnormal epileptic discharges generalized over both the hemispheres.

g. Not only the language function but also other cognitive and/or motor functions can be affected (Stefanatos 2011), displaying a particular 'co-morbidity', which is the very reason why children with LKS are likely to be clinically misdiagnosed as having autism.

The most crucial differences between ordinary child aphasia and LKS are the presence or absence of brain lesions and epileptic discharges in the brain. Just as in the case of adult aphasia, ordinary child aphasia involves some sort of

organical brain lesions due to traumas, tumors, or cerebrovascular damages, and does not normally implicate epilepsy. In contrast, as already mentioned in section 2.1.1, children with LKS exhibit particular EEG abnormalities, displaying no organical brain lesions with computed tomography and magnetic resonance imaging scans.

Moreover, unlike aphasia incurred in adulthood, ordinary child aphasia will generally be overcome if it strikes the child early enough in life (Lenneberg 1967, 1969). Considering the case of recovery from aphasia during preteen years, Lenneberg (1969: 639) suggests that such a phenomenon "may partly be regarded as a reinstatement of activities that had never been lost". Curiously enough, however, LKS differs from ordinary child aphasia in that the above-mentioned generalization by Lenneberg does not hold. That is, in LKS, a younger age of the onset of the language disorder is generally related to a gloomy prognosis for recovery from the state of aphasia (see Bishop 1985 and references therein). Thus, the following different patterns emerge for ordinary child aphasia and LKS as the second major difference, as already mentioned in (2c) and (3c), respectively:

(4) *Different Patterns of Prognosis in Ordinary Child Aphasia and LKS:*

a. *Ordinary child aphasia* (= (2c))
The earlier the onset of the disorder is, the better the prognosis will be.

b. *LKS* (= (3c))
The earlier the onset of the disorder is, the worse the prognosis will be.

The pattern of ordinary child aphasia in (4a) seems to be quite expected in the light of plasticity of the child brain in connection with Lenneberg's (1967) critical period hypothesis (see section 3.1 for discussion). In (4a), if the onset of the language disorder is earlier, relevant language functions would be relocated or compensated for by the use of other parts of the brain to the extent that the child is still within the critical period. This means that in the case of (4a) the child could overcome the aphasic state by appealing to plasticity of the neural network under development in the brain before full maturity of the neural network is attained.

The question therefore arises: Why does LKS behave differently from ordinary child aphasia (4b)? One possibility that immediately comes to mind is that, unlike ordinary child aphasia, LKS displays EEG abnormalities (Denes et al. 1986), due to epileptiform discharges typically with spike activity over the temporal (and/or parietal) regions (Deonna 1991). As Gordon (1997) clearly states, the main problem of LKS lies in the presence of epileptiform activity, or more precisely, the presence of CSWS discharges during slow-wave sleep, as reflected in the abnormal EEG. In the next section, we will compare LKS with other clinical conditions from the perspectives of EEG abnormalities and other characteristics.

2.2.2. Comparison of LKS, BECTS, and CSWS from the Perspectives of EEG Patterns and Other Characteristics

In the first place, recall from section 2.1.3 that LKS is a particular clinical instance of a newly defined class of *epileptic encephalopathies*, in which "a deterioration of cognitive, sensory, and/or motor functions results from epileptic activity" (Stefa-

natos 2011: 964; see also Nabbout & Dulac 2003). Hirsch et al.'s (2006: 244–245) review of neurophysiological and neuroimaging studies of LKS also summarizes the recent view on LKS by saying "LKS is an acquired aphasia secondary to an epileptogenic disturbance affecting a cortical area involved in verbal processing". Accordingly, as discussed in section 2.1.3, the apparent 'language disorder' and 'developmental disorder' in LKS are secondarily derived epiphenomena.

Although the ultimate etiology/etiologies of the paroxysmal EEG abnormalities in LKS *per se* still remain unclear, recall that the EEG abnormalities will usually disappear by puberty in LKS (Massa et al. 2000; Ramanathan et al. 2012). Thus, in principle, LKS is a curable disease to the extent that the EEG abnormallities can be removed and therefore the acquired aphasia (verbal auditory agnosia and loss of expressive speech), accompanied by behavioral disturbances and possibly clinical seizures, are ultimately derived from the presence of paroxysmal EEG abnormalities, as illustrated in Figure 1:

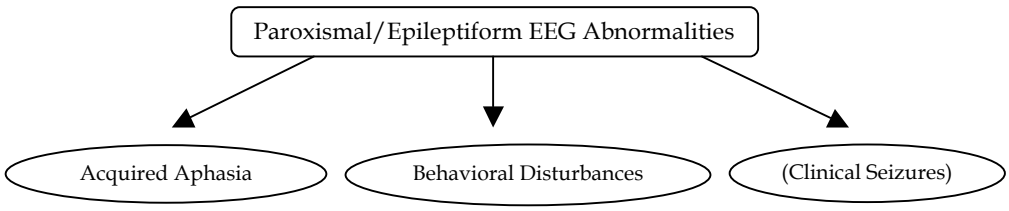

Figure 1: The causal relations in LKS.

The two clinical cases of BECTS and CSWS are to be differentiated from LKS: BECTS shares with LKS the general EEG patterns and severity but not the location of EEG abnormalities; CSWS, on the other hand, shares with LKS the general EEG patterns but not the location and the severity as well as the frequency of EEG abnormalities.[14]

Deonna & Roulet-Perez (2010) state that the generally accepted view is that "LKS constitutes one severe end of the continuum of cognitive manifestations that can be observed in idiopathic (genetic) focal epilepsies of childhood which may start quite early in development, the benign end being represented by Rolandic epilepsy" (where Rolandic epilepsy is another term referring to BECTS). Based on the acknowledgement that (the aphasia in) LKS is of epileptic origin and that it occupies the rare and severe end of a spectrum in idiopathic focal epilepsies of childhood with the more frequent typical BECTS at the other end (Deonna & Roulet-Perez 2010), LKS is to be compared with BECTS.

Deonna & Roulet-Perez (2010: 748) present the similarities between BECTS and LKS as follows (with some inconsequential modifications in wording in (5)):

[14] One caveat is in order here: The term CSWS is, strictly speaking, ambiguous between 'CSWS as a particular electrographic pattern' and 'CSWS as a clinical syndrome' on a par with electrical status epilepticus during sleep. Van Hirtum-Das et al. (2006) use 'electrical status epilepticus during sleep' and CSWS for referring to the particular electrographic pattern and the clinical syndrome, respectively. However, in the following discussion, CSWS is used in either sense, depending on the content.

(5) *Similarities between BECTS and LKS:*

a. Cases of BECTS can develop verbal auditory agnosia typical of LKS.
b. Cases with LKS and remission have later onset of Rolandic seizures.
c. Cases of BECTS who develop persistent oromotor deficits (anterior opercular syndromes) later remit like LKS.
d. Cases of BECTS with subtle acquired reversible language disturbances (oral and written) often have preexisting auditory-verbal and written language deficits (= 'mild forms of LKS').
e. Seizure semiology and course of epilepsy in BECTS and LKS are similar (= benign).
f. EEG findings: Focal sharp waves, increased by sleep, disappear with age in BECTS and LKS.
g. There are families described with one sibling having BECTS and the other LKS.

On the other hand, CSWS is one of the two epileptic syndromes that are associated with the EEG pattern of electrical status epilepticus during slow wave sleep, an electroencephalographic pattern in which the epileptiform discharges increase during sleep (Patry et al. 1971), the other being LKS (Tuchman 2009). There are differences in the frequency and severity of epilepsy between patients with CSWS and LKS, with children with CSWS having more severe and frequent and difficult-to-treat seizures than those with LKS (Jayakar & Seshia 1991; Smith & Hoeppner 2003).

Furthermore, BECTS exhibits minor developmental cognitive and behavioral problems, and some children with BECTS undergo deterioration in these domains (usually temporary), which are called 'atypical' forms of the syndrome. The severity and types of deterioration correlate with the site and spread of epileptic spikes within the perisylvian region, and CSWS frequently occurs during the period of the epileptic disorder. Some of these children have more severe preexisting communicative and language developmental disorder. If early stagnation or regression occurs in these domains, presumably it can be assumed to reflect epileptic activity in the networks outside the perisylvian region, that is, those involved in social cognition and emotions.

Table 1 depicts the main differences among LKS, BECTS, and CSWS. Note that the term CSWS in the first row in the right-most column is CSWS as a clinical syndrome rather than a particular electrographic pattern (see the caveat in fn. 14):

	LKS	BECTS	CSWS
EEG features	• focal or multifocal epileptiform discharges consistently enhanced during sleep; CSWS occurs in 80% of cases (Mikati et al. 2010)	• focal epileptiform abnormalities (Levisohn 2004) or focal sharp wave increased by sleep (Deonna & Roulte-Perez 2010) • CSWS occurs in occasional cases (Mikati et al. 2010) • a spike-and-wave index greater than 85% is not observed (Tassinari et al. 2000)	• in the waking state, focal and/or multifocal, and/or generalized diffuse spike wave activity (Mikati et al. 2010) • during sleep, continuous bilateral and diffuse slow spike wave activity through all or most (>85%) of the slow sleep stages (in all cases) (Mikati et al. 2010)
Location of EEG Abnormalities	superior temporal regions (Honbolygó et al. 2005)	centro-temporal regions (Tassinari et al. 2000)	fronto-central regions (Stefanatos 2011)
Linguistic Condition (difficulties in comprehension/ production)	• severe disturbance of auditory language comprehension (verbal auditory agnosia) (Stefanatos 2011) • substantial disruption of expressive language (Stefanatos 2011)	• milder (Nevill 1999), though verbal auditory agnosia is possible (Deonna & Roulte-Perez 2010) • regression is not normally verbal auditory agnosia (Mikati et al. 2011)	• expressive aphasia (Ekinci et al. 2012) • difficulties with lexical and syntactic skills (Ekinci et al. 2012) • language comprehension generally spared (Ekinci et al. 2012)
Non-linguistic Conditions	• non-verbal IQ and other cognitive functions can be affected with behavioral problems (attentional deficits, impulsivity, distractibility, hyperactivity, aggressiveness) (Deonna & Roulte-Perez 2010; Stefanatos 2011)	• most patients keep normal global intellectual efficiency, but some may suffer from oromotor dysfunction, neuropsychological deficits, or attention deficits with learning disorders (Mikati et al. 2010)	• widespread behavioral regression (decreased IQ, apraxia, memory loss, deficits of spatial and temporal orientation, psychiatric disturbances) (Stefanatos 2011)
Prognosis	• 50% of patients recover fully, while the remaining 50% recover partially or suffer from permanent aphasia (Mikati et al. 2010)	• most patients have good long-term outcome (Mikati et al. 2010)	• in adulthood, 50% of patients suffer from speech abnormalities and behavioral problems (Mikati et al. 2010)

Table 1: Comparison among LKS, BECTS, and CSWS.

2.2.3. Comparison of (Early and Ordinary) LKS and AR from the Perspectives of Language Disturbances and Other Characteristics

Although the exact current demographic data on the prevalence of each of the two clinical syndromes of LKS and AR are not available, and the figure on the prevalence rate of LKS is unclear at this point, ballpark figures on the basis of the relevant literature are listed in Table 2:

	LKS[15]		AR
	Early	**Ordinary**	
Occurrence Rate in Population	• 10% before 3 years of age (Bishop 1985)	• unclear	• one third of children with ASD (Trevathan 2004) • ~30% of children with ASD (Tuchman 1997)
Linguistic Condition (difficulties in comprehension/ production)	• severe (= global aphasia) (Landau & Kleffner 1957; Stefanatos et al. 2002)		• severe (Mantovani 2000)
Cognitive deficit	• non-verbal IQ and other cognitive functions can be affected (Deonna & Roulte-Perez 2010; Stefanatos 2011)		• severe (Mantovani 2000)
Difficulties in Pragmatics/Social Communication	• no (Temple 1997)		• severe (Nass & Devinsky 1999; Stefanatos et al. 2002)
Behavioral Characteristics: • **Hyperactive** • **Inattentive** • **Impulsive**	Tuchman (1997) Yes Yes Yes		Rice (2016) Yes Yes Yes
EEG abnormalities	• always present (Deona 2000) • CSWS in bitemporal or diffuse in most active phase (Deonna & Roulte-Perez 2010)		• not significantly frequent (Tuchman 1997; Deonna & Roulte-Perez 2010) • only 20% (Tuchman 2009) • epileptiform discharges in the centro-temporal regions (Tuchman 1997) • significant correlation with children without clinical seizures (Mantovani 2000)
EEG patterns	• focal epileptiform abnormalities are similar to BECTS (Levisohn 2004)		• no differences in localization (centrotemporal or other) of EEG discharges seen in children with epilepsy (Tuchman & Rapin 1997; Levisohn 2004)
Seizures	• 70% (20% do not have clinical seizures) (Neville 1999) • simple or complex partial seizures and/or atypical absence seizures • Rolandic seizures possible with LKS and remission (Deonna & Roulte-Perez 2010)		• 31% (Kobayashi & Murata 1998; Trevathan 2004)

Table 2: Comparison between (early and ordinary) LKS and AR.

[15] More than 160 cases of LKS have been reported from 1957 to 1990 (Paquier et al. 1992), but the prevalence is unclear (Pearl et al. 2001). In a recent study based on a questionnaire sent to all Japanese hospitals (3,004 hospitals as of March 2009), Kaga et al. (2014) conducted the first epidemiological estimation of LKS in Japan and found that the incidence of children with LKS aged 5 to 14 years is about 1 in a million (978,000) and the prevalence of children with LKS aged 5 to 19 and under medical care is 1 in 302,147 to 407,420 in Japan.

The comparison between LKS and ASD/AR seems to be the 'trickiest' and at the same time the most crucial. The two clinical cases may not be so easy to tease apart, because both children with LKS and those with ASD/AR have similar characteristic behaviors of being hyperactive, inattentive, and impulsive (Tuchman 1997; Rice 2016) and because LKS could affect the non-verbal intelligence on a par with the case of ASD (Deonna & Roulet-Perez 2010 and Stefanatos 2011; see also Great Ormond Street Hospital 2010 and Kimata et al. 2014). Thus, a majority of children with LKS (70–80%) exhibit clinically significant behavioral disturbances, and the combination of the profound communication disorder and severe behavioral abnormalities can approximate to the typical characteristics of non-high-functioning ASD (Ansink et al. 1989; Denes et al. 1986; Roulet-Perez et al. 1991; Roulet-Perez 1995; Stefanatos et al. 2002).

Especially since children with AR develop the impairments of autism after initial normal development (Mantovani 2000), AR attracts particular interest due to overlapping clinical and EEG features with LKS. Among children with ASD, at least 30% develop normally or nearly normally during the first year or two of life before developmental skills regress (Mantovani 2000). According to Mantovani (2000), their regression is not limited to language but also includes dramatic deterioration of social interaction and cognitive abilities, which usually begins between 18 and 24 months of age acutely or insidiously. The pathophysiology remains unknown, but electrophysiological disruption of normal brain development could be a contributing cause of AR (Mantovani 2000), in light of the fact that autistic children without clinical seizures have a significant correlation between AR and EEG abnormalities (Tuchman & Rapin 1997). Given the similarities between LKS and AR, we can suspect that a subgroup of the children diagnosed with AR, especially, those with epileptiform EEG abnormalities, could actually have suffered from early LKS.

In fact, Deonna & Roulet-Perez (2010) also suspect that "some children with an autism spectrum disorder, especially those who have a history of regression, which always involves language, and who have epileptiform EEG abnormalities, could actually have suffered an early form of LKS" (p. 746). They continue: "In several children finally diagnosed as early LKS, autism had been the initial diagnosis, but on closer look, the language deficit was clearly predominant" (p. 749). Moreover, Stefanatos (2011) has criticized the fact that the traditional clinical descriptions and boundaries of LKS have remained largely unchanged since their original formulation and suggested greater cross-disciplinary communication to enhance better diagnostic evaluation. Thus, clearer distinction between early LKS and AR becomes necessary.

The most crucial landmark is the differing rate of the presence of EEG abnormalities between LKS and AR. Patients with LKS always have EEG abnormalities with CSWS, while the rate of AR patients having EEG abnormalities is not significantly high. McVicar et al. (2005) have examined whole-night EEG records of 149 children with language regression and found that those with isolated language regression had a higher frequency of epileptiform abnormalities and seizures than children with both language and autistic (i.e., social and behavioral) regression (see also Deonna & Roulet-Perez 2010). Language regression (with or without autistic features) associated with epilepsy and paroxysmal EEG

abnormalities may represent early LKS in light of the fact that children with a history of autistic regression did not have significantly higher rate of EEG abnormalities than those who did not have autistic regression (Tuchman & Rapin 1997 and Baird et al. 2006; see also Deonna & Roulet-Perez 2010). Thus, EEG abnormalities are likely to be part of the underlying pathophysiology for LKS, whereas these are much less clear in the group with AR.

As for EEG patterns, although centrotemporal spikes in autistic children without language regression, independent of the presence of epilepsy, are prominent, no differences in localization of EEG discharges are seen in children with AR and epilepsy; whereas focal epileptiform abnormalities with CSWS, similar to BECTS, are obvious in LKS (Tuchman & Rapin 1997; Levisohn 2004). In other words, CSWS with autistic regression is a rare occurrence (Tuchman 2009). Moreover, as explained earlier, EEG abnormalities as well as seizures in LKS are likely to disappear between the ages of 8 and 13 years (Massa et al. 2000), which can be another clinical marker for LKS.

Secondly, another important landmark differentiating LKS from AR is pragmatic or social function. Mantovani (2000) identifies the pragmatic or social function as the most important differing feature because children with LKS retain their social awareness, use of gestures, and cognitive abilities measured on standardized tests of non-verbal skills. Deonna & Roulet-Perez (2010) also point out that, while LKS involves absent verbal communication, withdrawal, and stereotypies, lack of play and lack of understanding of social situations are clearly not in the forefront of LKS. Typically, children with ASD in general have severe difficulties in interpersonal social communication, due to abnormal development of pragmatic function, including theory of mind (Baron-Cohen 1995, 1998; Temple 1997; Pearl et al. 2001; Matsui 2010).[16] Bishop (2000) also specifies difficulties of pragmatically appropriate use of language as additional impairments of autistic children in addition to their difficulties in mastering syntax and semantics. On the other hand, LKS-affected children, who develop proper attachment to their parents and caregivers, do not have particular problems in interpersonal social communication and can develop pragmatic knowledge, including theory of mind (Temple 1997). Mikati et al. (2010) clearly state that "problems in reciprocal social relatedness and limited stereotypical forms of interests and behaviors that are associated with autism are not manifested in LKS patients" (pp. 259–260). As Deonna & Roulet-Perez (2010) explain, "if the epileptic process is restricted to the perisylvian cortex like in LKS, specific features of developing verbal language are expected to be lost, but not global social interaction like seen in children with primary autism who regress" (p. 748). Since the reciprocal social relatedness in social interaction is related to pragmatic function, its intactness is a clear clinical marker for LKS.

The third landmark is the differences in language restoration patterns between LKS and AR. Nearly three quarters of LKS-affected children (spontane-

[16] Somewhat contradictory views are expressed in the literature, though. See, among others, Tager-Flusberg & Joseph (2005) and Tager-Flusberg (2007) for the view that "autism involves delays and deficits not only in the development of a theory of mind but also in additional aspects of social affective information processing that extend beyond the traditional boundaries of theory of mind" (Tager-Flusberg 2007: 311).

ously) restore language skills completely or partially by adolescence (Mikati et al. 2010), whereas AR patients usually retain severe language deficits (Tuchman 1997). As Deonna & Roulet-Perez (2010) claim, it is true that "the proof that these children really suffered from an early form of LKS can only be brought convincingly if they improve significantly in correlation with the suppression of the EEG discharges or if they show definite relapses and remission in their language and other communicative abilities, a course which is not seen in a developmental condition" (p. 749) (see also Deonna & Roulet-Perez 2005). Nevertheless, the fact remains that approximately three quarters of LKS patients (spontaneously) restore language skills completely or partially by adolescence as the EEG abnormalities diminish and disappear (Mikati et al. 2010), which can serve as yet another clinical marker for LKS.

In summary, we believe that the most confusing case of comparison is between early LKS and AR, because both cases apparently involve grave language deficits for both comprehension and production. However, as pointed out above, by closely examining whether or not the relevant child has a particular pattern of EEG abnormalities (CSWS), has already developed an appropriate ability for interpersonal social communication with proper pragmatic knowledge, and can restore language skills with the disappearance of the characteristic EEG abnormalities, in theory, it seems to be possible to differentiate children with early LKS from those with AR.

2.3. Significance of LKS for Linguistic Investigation

Recall from fn. 5 that a child will acquire the core linguistic competence by around 3 years of age in the normal course of first language acquisition (Pinker 1994; O'Grady 2005). Recall also from section 2.1.1 that 80% of LKS have the onset ranging from 3 to 8 years of age, but the earliest onset of LKS falls on 18 months (Uldall et al. 2000). With those facts in mind in addressing LKS in the context of biolinguistics, we would like to propose to divide LKS into two gross sub-types. We refer to the early onset LKS as *early LKS* and all other cases of LKS as *ordinary LKS*, as defined in (6) on the basis of the differences of the onset of the language disorder in relation to the degrees of the state of I-language acquisition/growth at the time of its onset, with the first approximation dividing line being specified as 3 years of age, as already briefly mentioned in fn. 5:

(6) *Two Sub-types of LKS:*

a. *Early LKS*
Early LKS has the onset before 3 years of age, when the I-language of the affected child has not yet acquired the core linguistic competence sufficiently.

b. *Ordinary LKS*
Ordinary LKS refers to all other cases of LKS.

In this paper, we will focus on early LKS rather than ordinary LKS, which can be more easily diagnosed with the obvious establishment and disappearance of the first language. In fact, the term 'early LKS' is not novel, as Deonna &

Roulet-Perez (2010) use the term basically in the same sense, although they do not mention the notion of I-language. Actually, 12%–14% of children with LKS undergo language regression before three years of age (Bishop 1985; Dugas 1991; Tuchman 1997) and even a case of LKS with its onset at 18 months has been reported in the literature, as mentioned in the previous discussion.

Since LKS is not caused by any lesions of anatomically identifiable specific substrate in the brain, unlike adult aphasia or ordinary child aphasia, nor has any particular gene been linked to it so far (Benítez-Burraco 2013 and references cited therein),[17] investigation into LKS in the context of biolinguistics has significant virtues. Particularly, early LKS is extremely important from two perspectives. One is clinical improvement, as mentioned above, in terms of preventing misdiagnosis of LKS with ASD or AR, since early LKS with the onset before the solid establishment of the first language, accompanied by behavioral disturbances, is hard to distinguish it from these other developmental disorders (Uldall et al. 2000). If the onset of LKS in children were to be around 18 months, as in the case of early LKS, their first language acquisition and development of other cognitive and motor skill functions would still be at early and immature stages. Under these circumstances, it is highly likely that quite a lot of children with early LKS would be misdiagnosed as ASD or AR with severe retardation because of the overlapping co-morbidity, and would not be treated properly.

The other is linguistic investigation and analysis into the nature of human language and first language acquisition in connection with the critical period, modularity of mind, and modularity of the faculty of language (I-language). To the extent that LKS is not directly caused by any identifiable specific gene defects (but see the caveat in fn. 18), we can also assume that the genetic endowment (whatever it may be) responsible for emergence of UG remains virtually intact in patients with LKS, based on the fact that 50% of the patients recover fully and 50% of the remaining patients recover partially (Mikati et al. 2010) after a certain period of time.[18] Rather, given the lack of any identifiable organic lesions in the brain, we can naturally assume that at least some neuronal-level mechanism(s) in the brain, but not the lack or deficits of I-language, would be responsible for the language disorder observed in LKS, as suggested by the following remark:

> Neurophysiological techniques such as magneto-encephalography can also now help explain why children [with LKS] have limited potential to relocate the devastated language area as there is bilateral involvement of the cortex. It appears that the likely 'pacemaker' for the electrical disruption of the language arises from the intrasylvian cortex but spreads to the contralateral sylvian cortex. (O'Hare 2008: 724)

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17 Although a particular genetic cause for LKS has not been identified so far, a number of recent studies have suggested that *GRIN2A* (16p13.2) mutations may underlie familial and sporadic cases of LKS (see Conroy et al. 2014 and references therein). We are grateful to a reviewer for pointing out this fact.

18 While the patient with ordinary LKS would be highly likely to recover from the state of aphasia in a relatively short period of time, the patient with early LKS would either recover from such a state after a relatively long period of time or not recover from it. This description is based on the observation in Bishop (1985), which does not express absolute correlations but just tendencies (see Deonna et al. 1977 for the varied prognosis of LKS depending on factors other than the onset of the disorder).

If this is the case, as long as epileptic electrical disruption of the relevant neural network of the language areas continues bilaterally in the brain, which interferes with plasticity of the brain functioning for language development, the state of aphasia observed in LKS would not cease to exist. Landau & Kleffner (1957) themselves do not specify what "brain tissue largely concerned with linguistic communication" and "the functional ablation of these areas for normal linguistic behavior" refer to, so identifying the relevant brain areas for language and the mechanisms as well as the culprit of the EEG abnormalities is clinically of great importance. If the EEG abnormalities of LKS patients can be controlled, it is highly likely that language might re-emerge or be restored (see Figure 1). In the following section, we will closely examine the phenomena of LKS in terms of Lenneberg's critical period hypothesis for first language acquisition, Chomsky's first language acquisition model, and his views on modularity.

3. Some Biolinguistic Considerations

3.1. The Critical Period Hypothesis for First Language Acquisition

The notion of a 'critical period' for (first) language acquisition was entertained by Penfield & Roberts (1959) (see also Lenneberg 1960) and was clearly formulated by Lenneberg (1967) (see also Lenneberg 1969), considering a variety of cases of child language acquisition (both normal and handicapped).[19] Lenneberg (1967) hypothesizes that the critical period for first language acquisition corresponds to the time span from around 2 years of age to around 12 or 13 years of age, and that during this period children can acquire their mother tongue on a biologically determined course of language development, given appropriate linguistic input from their environment.[20]

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19 The critical period hypothesis has been discussed extensively and revised dramatically in the literature over the past five decades (see e.g. Weber-Fox & Neville 1996; Locke 1997; Hyltenstam & Abrahamsson 2003; Knudsen 2004; Michel & Tyler 2005; Meisel 2013; Balari & Lorenzo 2015). The notion of 'sensitive period(s)' has come to be used instead of the critical period in order to reflect the relative plasticity of our brain handling first/second language acquisition rather than a sudden halt, even after the end of Lenneberg's (1967) original specification of such a period.

Furthermore, unlike a single critical period in Lenneberg (1967), multiple different sensitive periods are assumed to exist in relation to various 'components' of language such as phonetics, phonology, morphology, syntax, semantics, and so forth, or several clusters of 'sensitive phases' in such multiple sensitive periods are postulated to account for the development of different subcomponents of grammar. While we fully acknowledge the significance of these various refinements of the critical period hypothesis, we would like to invoke Lenneberg's (1967) original version of the critical period hypothesis in the following discussion on LKS for a certain reason to be clarified later.

20 It is also to be noted that the exact onset and end of the critical period are controversial in the literature. Thus, the onset of the critical period may well be much earlier than two years of age (see e.g. Mayberry & Lock 2003; Dettman et al. 2016), and it may end much earlier or later than 12–13 years of age, say, somewhere between the ages of 6–7 and 16–17 (see DeKeyser 2000 and references therein for the latter). Although we will keep to Lenneberg's original specification of the onset of the critical period as 2 years of age in the following discussion, it would be more appropriate to set an onset, depending on systems in I-language, say, at the perinatal or even the prenatal period, particularly with respect to the development of the sound system (see, e.g., Werker 1989; Kuhl 1993).

Lenneberg's critical period is related to the putative steady state of I-language attained by the relevant neuronal circuitry within the brain. In fact, with regard to the critical period, Lenneberg (1969) also remarks that "it is interesting that the critical period coincides with the time at which the human brain attains its final state of maturity in terms of structure, function, and biochemistry (electroencephalographic patterns slightly lag behind, but become stabilized by about 16 years). Apparently the maturation of the brain marks the end of regulation and locks certain functions into place" (p. 639).[21]

It is to be noted that, as already mentioned, since LKS affects both hemispheres due to secondary generalization of a focal epilepsy, lateralization of the language function in the brain does not result in the employment of the contralateral brain regions for the language function, unlike in the case of ordinary child aphasia (see the remark by O'Hare 2008 in section 2.3 above). Therefore, linguistic input would be practically unavailable to the child with LKS to the extent that verbal auditory agnosia due to the EEG abnormalities exists in the child. However, if linguistic input somehow were to become available to the child again along with the disappearance of the EEG abnormalities during the critical period, re-start of acquisition of the first language and re-emergence of language would be possible in principle.

Morrell et al. (1995) put forth a hypothesis that the presence of epileptiform activity within the relevant circuits for language in LKS may hinder pruning of inappropriate cells and axons for the optimal network of language, and that those circuits may become permanent if the critical period has passed. Accordingly, it can be assumed that the full-fledged acquisition/growth of I-language in patients with LKS would become virtually impossible or extremely hard to achieve, unless the epileptiform activity as reflected in the EEG abnormalities would be removed before the critical period ends.

At this point, it is in order to correctly understand the original version of the critical period hypothesis put forth by Lenneberg (1967) in connection with our proposal in this section. First of all, Lenneberg's critical period is only concerned with first language acquisition and he does not say anything clearly about second/foreign language acquisition. Furthermore, although the term has been commonly used in the broad notion of 'first language acquisition' in the literature, which encompasses linguistic input and output, Lenneberg's original

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The following discussion is not affected much as long as the onset of the critical period is before 3 years of age, which is the age for differentiating between early LKS and ordinary LKS. We are grateful to a reviewer for raising our attention to recent research on cochlear implanting in children (Dettman et al. 2016) and on deaf signers (Mayberry & Lock 2003) in connection with the critical period hypothesis.

21 For a recent study on the maturation of components of event-related potentials as measured with EEG and event-related fields as measured with MEG in connection with auditory processing, see Ruhnau et al. (2011). They demonstrate that a mature N1 can be observed in children of 9 to 10 years of age on a par with the one in adults and reveal that the source of N1m in children and adults is mainly located in primary auditory cortex on the basis of source localization of the MEG data. Their result is in support of Ponton et al.'s (2002) findings based on dipole source modeling that brain areas underlying early auditory processes are mature in children at around 9 to 10 years of age. We are grateful to a reviewer for bringing our attention to Ruhnau et al. (2011) in relation to the maturation of the brain and auditory processing.

version of the critical period for first language acquisition only applies to linguistic input. It crucially claims that linguistic output/externalization, say, by articulation is *not* subject to such a critical period (see Lenneberg 1967: 158). As such, even if a child is suffering from childhood aphasia, it is predicted that, in principle, there should be a case where *externalization* of I-language could happen *after* the critical period, once the deficit in the neural system for articulatory motor skills of externalization is removed or somehow disappear—on the condition that acquisition of the mental lexicon and language-particular morpho-phonology, syntax, and semantics should become possible in time for the completion of the critical period.

Thus, Lenneberg's critical period hypothesis is of great significance in considering the case of early LKS, in which the re-start of I-language acquisition would be rendered possible if linguistic input became available within the critical period. Since the development of the system of articulatory motor skills is not subject to the critical period, according to Lenneberg (1967), the child with early LKS could become capable of producing speech even after the critical period ends in accordance with gradual redevelopment of such an externalizing sensori-motor (SM) system. In a nutshell, Lenneberg's critical period is only concerned with the linguistic input, so the linguistic output is outside of its domain.

Consequently, the pattern examples of the two sub-types of LKS, ordinary LKS and early LKS, can be schematically illustrated as follows in Figure 2:

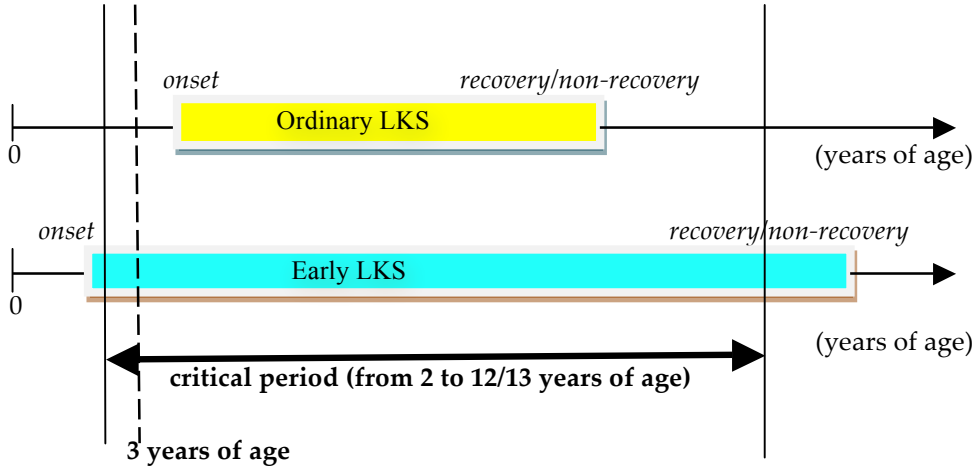

Figure 2: Pattern examples of ordinary LKS, early LKS, and the critical period.

Nonetheless, there is a serious issue concerning the end of the critical period and the time of termination of EEG abnormalities. As stated in Massa et al. (2000: 89), EEG abnormalities as well as seizures in LKS patients could disappear between the ages of 8 and 13 years (mean of 10 years), after being controlled by, say, anti-epileptic drugs and/or corticosteroids. Suppose the EEG abnormalities alleviate and disappear by around 14 years (considering margins of error): Then, there is a temporal gap (1–2 years) between the end of the critical period (12/13 years of age) and the end of the EEG abnormalities. If the intake of linguistic input for language development becomes possible after controlling the EEG abnormalities at around 14 years of age, is it too late for language acquisition since it is over the critical period?

It seems, however, that EEG could improve gradually, not suddenly, by around 14 years of age (Massa et al. 2000; Robinson et al. 2001; Deonna & Roulet-Perez 2010). Thus, it is quite natural to assume that verbal auditory input would become possible gradually well before 14 years of age and the quality of linguistic input would concurrently improve during the process of gradual amelioration of the EEG status in LKS. In any case, one cannot stress enough the importance of offering the child with LKS the opportunities to secure linguistic input within the critical period.

3.2. Primary Linguistic Data (PLD) for First Language Acquisition and LKS

In the tradition of generative grammar, the relevant process of first language acquisition has been abstractly characterized as follows (see Chomsky 1967 for an earlier and Chomsky 2004a, among others, for a more recent version):[22]

Figure 3: Generative model of language acquisition.

On this model of first language acquisition, the universal properties of syntax and semantics (and morpho-phonology) of I-language are biologically given, or more appropriately determined, and are not learned ontogenetically and only the language-particular aspects of linguistic knowledge pertaining to the *primary linguistic data* (PLD) must be learned in the course of first language acquisition.[23] Therefore, Lenneberg's critical period hypothesis should only apply to the acquisition of lexical items along with the language-particular dimensions of syntax, semantics, and morpho-phonology of I-language on the basis of the PLD.[24]

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[22] The content of LAD used to be regarded as virtually UG as a genetic endowment, but the role of UG has been radically reduced to the bare minimum while the role of interface conditions and that of a 'third factor' has been emphasized in the context of the Minimalist Program (see Chomsky 2005). Here, we are only concerned with the general conception of Chomsky's first language acquisition model, without delving into the debate on the content of LAD, including UG. Although we believe the line of proposals on the content of LAD in Boeckx & Leivada (2014) and Boeckx & Theofanopoulou (2014) is biolinguistically on the right track, we will use the original term LAD with this caveat in mind. Furthermore, the 'instantaneous model of language acquisition' conceived in the tradition of generative grammar, as illustrated in Figure 3, is an idealized model, abstracting away from actual stages of language development or growth in children, as emphasized in Lorenzo & Longa (2009), who propose a new model of language acquisition from a developmentalist point of view in the framework of the Minimalist Program. Although we fully recognize the importance of interface conditions and third factor principles (Chomsky 2005) along with the minimized role assumed by UG and the role of individual linguistic experiences, we will keep to the label LAD without going into such an elaborated model of first language acquisition in this paper (see also Locke 1997; Longa & Lorenzo 2008), since our main point in this section is on the role of PLD in connection with LKS.

[23] See also Guasti (2002) for detailed explication of various aspects of language acquisition in the framework of generative grammar.

[24] Note that, although not mentioned here, pragmatics/pragmatic knowledge should constitute part of the system of interpretation together with semantics/semantic knowledge (see

Therefore, if there is a situation where the PLD were to be unavailable for the *language acquisition device* (LAD) in a child, acquisition of lexical items would become impossible and as a result the child would not be able to expand the domain of words (and other linguistic expressions). Furthermore, the universal aspects of syntax and semantics (and morpho-phonology) of I-language would remain at least potentially intact.[25] On the other hand, if the PLD should become available again somehow within the critical period, re-start of acquisition or growth of I-language, including the mental lexicon, would become possible, even if the child would be in a situation where he/she could not speak his/her first language while understanding it. Thus, if the critical period hypothesis is on the right track, in principle, a child with LKS could re-start acquiring his/her I-language to the extent that the PLD becomes available again as linguistic input to the LAD in the sense of Chomsky's model of first language acquisition somewhere within the critical period, even in the case of 'covert language acquisition', or language acquisition without involving any expressive speech.[26] More specifically, if the EEG abnormalities in LKS were to be gradually suppressed within the critical period, it is expected that the quality of the PLD for the LAD should become better, leading to re-starting of I-language acquisition in time before the end of the critical period.

Furthermore, if the externalization/articulation in the SM system, which is not subject to the constraint of Lenneberg's (1967) critical period, could be restored in LKS somehow (see our concrete proposal toward this goal in section 4.2), even a child with early LKS could re-start producing speech at some point with a surprising speed of language development, compared with the one of normal language development, after regaining an ability to comprehend speech, because of the existence of potentiality of I-language even without its externalization. This would give the impression that a 'linguistic big bang' could occur in a child with early LKS. Accordingly, if such a linguistic big bang should happen, the case of early LKS would dramatically demonstrate the validity of Lenneberg's version of the critical period hypothesis.[27]

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e.g. Chomsky 1980/2005 for the concept of pragmatic competence). However, pragmatics, by nature, encompasses 'non-linguistic contexts' such as intentions of others independently of PLD. Given that pragmatics, including theory of mind, can be dissociated from I-language, as observed in ASD and LKS (see the discussion in sections 2.2.3 and 3.3.3), it develops as a separate system in the mind. Since we will be only concerned with I-language and PLD *per se* in discussing LKS in this paper, we will not include pragmatics here.

25 If both lexical items and syntactic structures are equally generated by Merge, as Merge-α in the anti-lexicalist approach (Fujita & Matsumoto 2005; Fujita 2014; Boeckx 2015; see also Marantz 1997; Borer 2005.), Merge should be potentially ready for acquisition of lexical items even in the face of unavailability of the PLD in LKS (see also Nasukawa 2015 for a Merge approach to the lexical structure of morphemes in intra-morphemic phonology). This might explain why a 'linguistic big bang' could occur in LKS (once a sufficient amount of PLD becomes available again due to (gradual) amelioration of EEG abnormalities) (see the discussion below).

26 Lenneberg (1962) reports an interesting case in which an eight-year-old boy who had a congenital language disorder developed language comprehension ability without ability to speak, arguing that this kind of case argues against the view that speech production is crucial to the development of speech comprehension.

27 Deonna et al. (2009) argue that learning a sign language will not delay or prevent oral language recovery in children with LKS but possibly even facilitate the recovery process by

The image of language development of the normal child and that of the early LKS child could be roughly illustrated as follows in Figure 4 (note that these are just images, not exact graphs showing actual language development in the two groups of children):

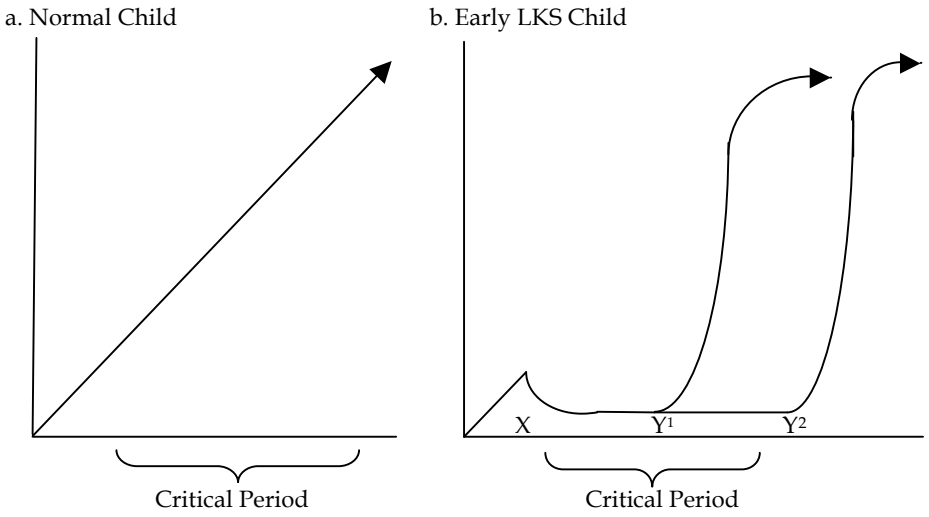

Figure 4: Images of language development in the normal child and recovery of expressive language in the early LKS child.

While normal children will develop their first language steadily within the critical period on the basis of the biologically determined course, children with early LKS would first begin developing their first language normally but suddenly start regressing at a point indicated by X before 3 years of age, possibly before the critical period starts. From that point on, some degree of lexical acquisition, if it ever exists, might occur on the basis of the PLD of poor quality. Then, at some point in time within the critical period, epileptic abnormal brain activity will be suppressed and the quality of the PLD will become better, boosting up the process of first language acquisition again. Finally, 'normalization' of the neural circuitry in the SM system for externalization of I-language should happen at some point in time either within the critical period, as shown by Y^1, or after the end of the critical period, as shown by Y^2, respectively (it is to be noted that the period from X to Y^1/Y^2 is a 'virtually silent period' and the PLD would become available again sometime before the critical period ends).

In this way, as mentioned earlier, children with early LKS would be able to experience something like a linguistic big bang. As a matter of fact, Uldall et al. (2000) observe that their patient with early LKS (with onset at 18 months) sped up language acquisition in his 'catch-up periods' in such a way that he acquired vocabulary that would have normally taken one whole year in just 3 months after the age of 5 years. Uldall et al. (2000) remark that "the normal spurt of vocabu-

stimulating the 'functionally connected core language networks', resulting in being bilingual. This clearly indicates that, even if children with LKS are in the state of verbal auditory agnosia and do not produce any utterances, the neural circuitry of I-language and its externalization system potentially remain in the brain, albeit with some deficiency in externalization.

lary usually seen at the age of 17–19 months seemed to have been blocked until it was 'released' by the prednisone course at the age of 5 years" (p. 85). In sum, as long as I-language is established before the critical period ends, externalization of I-language is not affected by the critical period, and thus it would become possible even later in life, in principle.

3.3. Modularity and LKS

We regard I-language itself—or more strictly, the FL[28]—as composed of independent but interacting sub-systems, or sub-modules (see Chomsky 1980/2050, 1981, 1984, 1986, 1995, among others; for more recent views on FL, see also Hauser et al. 2002; Chomsky 2016; Berwick & Chomsky 2016). Specifically, we take for granted the following basic design of FL that has been assumed as standard in the current Minimalist Program:

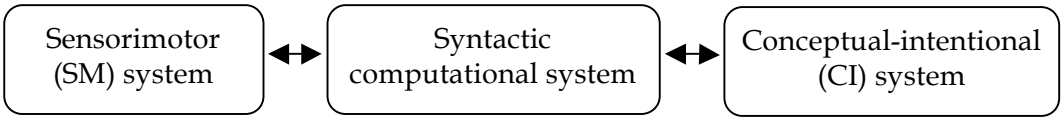

Figure 5: Basic design of FL.

Given the nature of LKS that we have discussed in the previous sections, our focus in theorizing the language-related mechanisms behind LKS is the SM system and the 'mapping' between the syntactic computational and the SM systems, including the phonological system. In this connection, recall from section 2.1.1 that Landau & Kleffner (1957) originally proposed a 'functional ablation' view on LKS: "[P]ersistent convulsive discharge in brain tissue largely concerned with linguistic communication results in the functional ablation of these areas for normal linguistic behavior" (p. 529). Then the linguistically significant question is: What does the phrase 'these areas' in the above quote refer to? The literature in the past generally mentions the temporal(-parietal) and perisylvian cortices as relevant areas responsible for the language disorder in LKS, *viz.* verbal auditory agnosia and loss of expressive speech.

The main purpose of this subsection is to zero in on these linguistically relevant areas of the brain, putting forth a concrete hypothesis on the LKS-affected linguistic function and its related cortical areas involved in verbal auditory agnosia and loss of expressive speech in LKS. Since Chomsky himself does not articulate the content of the SM system in neurophysiological terms, we would like to consider the general architecture of language in LKS in the context of speech processing, drawing on a recent study on the cortical organization of speech processing in Hickok & Poeppel (2007).[29]

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[28] For an alternative to the traditional FL, see Balari & Lorenzo's (2015) new concept of language as a 'gradient' proposed in a dynamic developmental perspective.

[29] See also Friederici (2011) for a comprehensive detailed discussion on the structural and functional neural network in the brain underlying sentence processing.

3.3.1. *Hickok & Poeppel's (2007) Dual-Stream Model of Speech Processing*

Hickok & Poeppel (2007) put forth the *dual-stream model of speech processing*, as roughly illustrated in Figure 6, on the basis of a wide range of empirical observations such as basic perceptual processes, aspects of speech development and speech production, linguistic and psycholinguistic facts, verbal working memory, task-related effects, sensorimotor integration circuits, and neuropsychological facts (e.g., patterns of sparing and loss in aphasia). It is to be noted that the numbers from 1 to 7 in Figure 6 are not included in the original but are assigned by us for expository purposes, and that they do not indicate temporal ordering or sequencing of processing. In Figure 6, based on our interpretation of Hickok & Poeppel (2007), we inserted a blue dotted enclosure and a black dotted enclosure to indicate the portion dedicated only to speech production and the one shared by speech comprehension and production, respectively, which are not included in their original chart.

In order to help the reader to visually grasp the approximate anatomical locations of the dual-stream model components and their interconnections, a rough illustration of the left-hemisphere of the brain is provided in Figure 7 with the same colors used for the corresponding relevant components in Figure 6. In the following discussion, we will not be concerned with the conceptual network and its interconnections with the lexical interface and the articulatory network.

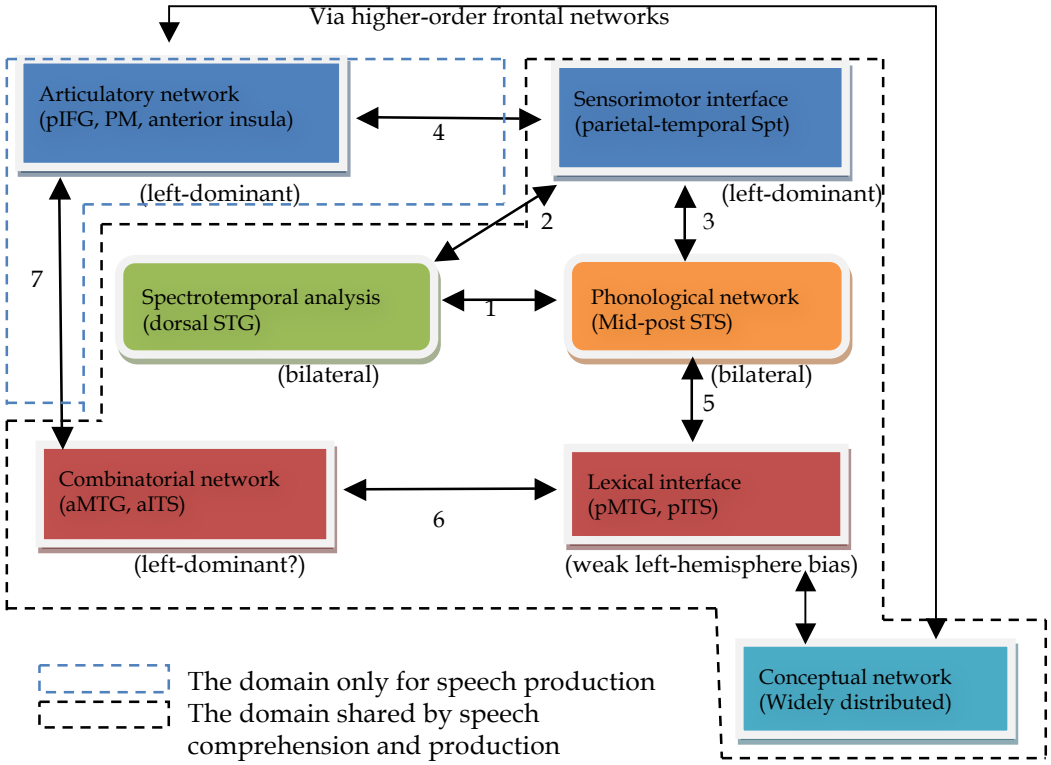

Figure 6: The dual-stream model of speech processing, based on Hickok & Poeppel's (2007) figure 1a with some simplification and adaptation. Though the original 'input from other sensory modalities' is not depicted here due to space constraints, the sensorimotor interface component is supplied with input from other sensory modalities.

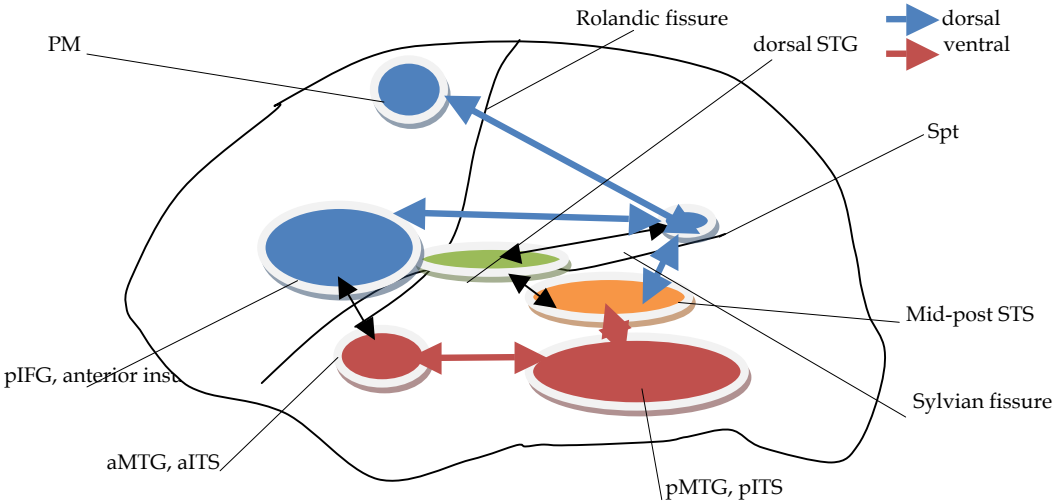

Figure 7: The relevant brain areas in the dual-stream model of speech processing, based on Hickok & Poeppel's (2007) figure 1b with some simplification and adaptation. Unlike in the original, only the left hemisphere is depicted, and the dorsal stream and the ventral stream are highlighted by the blue connection and the pink connection, respectively.

Now, let us briefly go over the whole process of language comprehension in this model. First of all, the earliest stage of cortical speech processing involves some form of spectrotemporal analysis of acoustic signals, or auditory phonetic analysis of sequences of speech sounds, which is carried out in the auditory cortices bilaterally in the supratemporal plane, i.e., in the superior temporal gyrus (STG) (see e.g. Zatorre & Belin 2001 for details on spectrotemporal processing in the human auditory cortex). Then, the result of the spectrotemporal analysis is transmitted to the bilateral phonological network, accompanied by the feedback from the latter to the former, in the middle to posterior portions of the superior temporal sulcus (STS) (= 1), which is responsible for carrying out phonological-level processes and creating phonological representations.

Subsequent to the phonological system, the information flow diverges into two broad streams. The dorsal stream, which is strongly left-hemisphere dominant, maps phonological representations onto articulatory motor representations in the sensorimotor interface at the Sylvian-parietal-temporal (Spt) area located within the planum temporale (PT) (= 3), with the feedback from the sensorimotor interface to the phonological network as well. Furthermore, the articulatory motor representations are handed over to the articulatory network in the posterior inferior frontal gyrus (pIFG) involving Broca's region,[30] the premotor cortex (PM), and the anterior insula (= 4), again accompanied by the feedback from the articulatory network to the sensorimotor interface. Note, incidentally, that the spectrotemporal analysis component and the sensorimotor interface could be directly interrelated as indicated by the two-way arrow (= 2).

[30] See Yusa (2012, 2016) and references therein, including Grodzinsky & Amunts (2006), for detailed discussion on the fine-grained architecture of Broca's region (BA 44, 45). In this paper, we will not delve into this issue while acknowledging its theoretical importance ultimately in connection with LKS as well in the biolinguistic context.

In contrast, in the ventral stream, which is largely bilaterally organized with a weak left-hemisphere bias, phonological representations are associated with lexical conceptual representations in the lexical interface (= 5), which is with weak left-hemisphere bias, in the posterior middle temporal gyrus (pMTG) and the posterior inferior temporal sulcus (pITS), with the feedback from the lexical interface to the phonological network. Then, an array of lexical conceptual representations (linked with corresponding phonological representations) are handed over to the combinatorial network (= 6), which is assumed to be left-dominant, to generate post-lexical conceptual and semantic representations (with corresponding phonological representations), in the anterior middle temporal gyrus (aMTG) and the anterior inferior temporal sulcus (aITS), accompanied by the feedback from the combinatorial network to the lexical interface, and the interaction between the combinatorial network and the articulatory network as well.

In Hickok & Poeppel's (2007) dual-stream model of speech processing, within the whole process of speech comprehension, *speech perception* of pre-lexical stages (such as segmental/phonemic identification and supra-segmental identifycation like syllabification) are to a greater extent handled by the dorsal stream, while *speech recognition*, including processing of lexical/post-lexical stages (such as word identification and hierarchical syntactic structure identification), relies more on the ventral stream.[31] In section 4.2, we will make use of the term *verbal auditory comprehension* in discussing several recovery patterns of language disorder in LKS. It is to be kept in mind that the term verbal auditory comprehension implicates both speech perception and speech recognition in the sense of Hickok & Poeppel (2007), because LKS typically incurs *verbal auditory agnosia*, which refers to a situation where not only sublexical-level but also lexical-level and phrasal-level processing is disrupted in a severe period of the disorder.

Although Hickok & Poeppel (2007) themselves do not explicitly describe the concrete processes of speech production unlike for speech comprehension in the dual-stream model of speech processing,[32] we assume that at least the initiation of speech production does not involve the spectrotemporal analysis component, whereas the subsequent processing of speech production will employ the spectrotemporal analysis component along with the phonological network so that the speaker can monitor his/her own speech. In the case of speech comprehension, the interactions between the articulatory network and the sensorimotor interface and between the articulatory network and the combinatorial network as indicated by 4 and 7 may not be required (see the case of Lenneberg 1962 in fn. 27), but in the case of speech production those interactions are absolutely

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31 Hickok & Poeppel (2007: 394) define the three terms *speech processing*, *speech perception*, and *speech recognition* as follows: (i) "speech processing refers to any task involving aurally presented speech"; (ii) "speech perception refers to sublexical tasks (such as syllable identification)"; and (iii) "speech recognition (auditory comprehension) refers to the set of computations that transform acoustic signals into a representation that makes contact with the mental lexicon".

32 Hickok (2012) proposes a hierarchical state feedback control (HSFC) model of speech production. Since the purpose of this section is to consider verbal auditory agnosia and loss of expressive speech from the perspective of the dual-stream model of speech processing, we will not incorporate Hickok's (2012) model of speech production in the following discussion, leaving the task to another occasion.

necessary. Given these assumptions, it seems natural to suppose that verbal auditory comprehension of sentences, which involves accessing to hierarchically structured expressions, must comprise the components of spectrotemporal analysis, phonological network, sensorimotor interface, lexical interface, and combinatorial network along with the interactions indicated by 1, 2, 3, 5, and 6. In the case of verbal production (speech production) of sentences, it seems to be natural to assume that, in addition to those components and interactions, the articulatory network is also involved via interactions with the sensorimotor interface and the combinatorial network, as indicated by 4 and 7, respectively.

We also presume that basically the same asymmetries between the dorsal stream and the ventral stream hold in the case of speech production as well with all the various interactions/feedbacks illustrated in Figures 6 and 7: (i) the dorsal stream is strongly left-hemisphere dominant, while the ventral stream is largely bilaterally organized, with a weak left-hemisphere bias; (ii) the dorsal stream is mainly for the processing of pre-lexical units such as phonemes and syllables, whereas the ventral stream is to a greater extent for the processing of lexical/ post-lexical units such as words and phrase structures.

3.3.2. Input and Output Problems in LKS

We would like to propose that LKS is a language disorder that involves two major problems regarding the FL, which would be ultimately ascribed to some deficiencies in the SM system, as depicted in Figure 8:

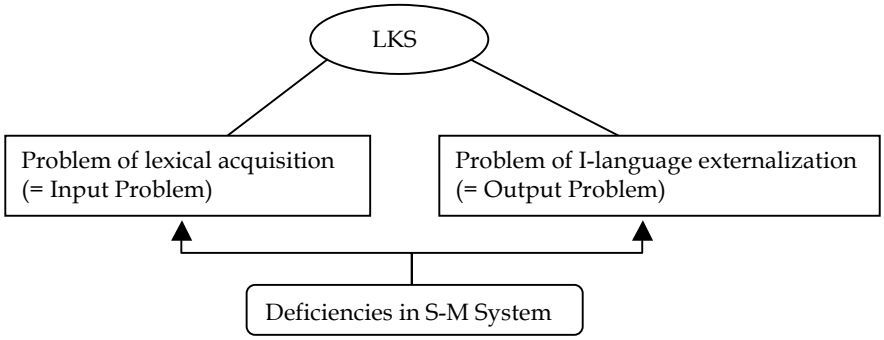

Figure 8: Language disorder in LKS caused by deficiencies in SM system.

The first problem is with acquisition of lexical items (the 'input problem') and the other with the externalization of I-language, say, by articulation (the 'output problem'). Let us first consider the input problem of LKS.

At first, it is of vital importance to identify the core deficiencies underlying the input problem of LKS. In view of the linguistic profile of LKS discussed in section 2.1.2 and Stefanatos's (1993) insight into LKS as "an apperceptive disturbance in which there is primary impairment of processes subserving the auditory analysis of acoustical features [amplitude modulation (AM) or frequency modulation (FM)] necessary for speech perception" (p. 412), we propose to analyze LKS as affecting the system for spectrotemporal analysis located bilaterally in the dorsal superior temporal gyrus (STG) (and possibly the routes connecting the

system for spectrotemporal analysis and other relevant systems).[33,34] Recall from section 2 that children with LKS suffer from spike-wave discharges predominating over the superior temporal regions activated by sleep and secondarily generalized to both hemispheres. Given this state, it is quite natural to imagine that such abnormal brain wave activity will disrupt proper working of the system for spectrotemporal analysis bilaterally.

Note that in Hickok & Poeppel's (2007) model, the dorsal STG for spectrotemporal analysis, the posterior half of the superior temporal sulcus (STS) for phonological processing, and the parietal-temporal Spt for sensorimotor interface processing are interconnected with each other bidirectionally (see Figures 6 and 7). Crucially, this implies that if the system for spectrotemporal analysis were impaired in LKS, it would be expected to yield deleterious effects on both the ventral stream and the dorsal stream, as clinically observed as verbal auditory agnosia and loss of expressive speech in LKS.

More specifically, if acoustic signals of sequences of speech sounds cannot be properly analyzed spectrotemporally in the dorsal STG (bilaterally), the phonetic sound sequences cannot be correctly linked with appropriate abstract phonological units, even if the phonological system in the mid-post STS *per se* remains intact. As a result, the supposed lexical items cannot be formed/identified at the lexical interface in the pMTG and pITS, presumably due to the lack of appropriate pairing of <P, S> (where P stands for a phonological representation including specification of distinctive features, and S for a semantic representation).[35,36] Consequently, there would be no proper input of lexical items for the combinatorial network in the aMTG and aITS to form/identify hierarchically structured expressions (i.e., phrases and sentences). Hence, the 'input problem' of LKS, or the state of verbal auditory agnosia in LKS, emerges.

As such, if no correct *P* is available to the child with LKS, normal acquisition of lexical items would not be possible as long as the child with LKS is suffering from the state of verbal auditory agnosia. However, given the fact that comprehension will be regained in due course in accordance with the amelioration of the EEG abnormalities in LKS (Massa et al. 2000) (after anti-epileptic medication), the input problem of LKS will more or less disappear eventually.

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33 The PET results in Zatorre & Belin (2001) indicate that "(i) the core auditory cortex in both hemispheres responded to temporal variation, while the anterior superior temporal areas bilaterally responded to the spectral variation; and (ii) responses to the temporal features were weighted towards the left, while responses to the spectral features were weighted towards the right" (p. 946).

34 Tsuru & Hoeppner (2007) suggest the possibility that deficits in Wernicke's area (= post-STG) and the supramarginal gyrus are involved in LKS on the basis of Iwata (1996). Their suggestion is not exactly the same with our proposal, but seems to partly overlap with it.

35 Phonological features (e.g., [+voiced]) and semantic features (e.g., [+artificial]) of each lexical item will become part of a phonological representation and a semantic representation, respectively. See Chomsky (1965, 1995 *et seq.*) for discussion on different kinds of features in lexical items.

36 In the framework of Distributed Morphology, *P* (phonological features) will be inserted later in the derivation in the post-syntactic Morphology component (e.g., Halle & Marantz 1993). Even if this is the case, the fact remains that the two feature bundles (*P* and *S*) have to be 'lumped together' somehow to guarantee Saussurean arbitrariness in a coherent lexical item. See Harley (2014) for recent developments of the framework, in which indices are employed as a device for this purpose.

Next, let us turn to the output problem of LKS. If acoustic signals of sequences of speech sounds cannot be properly analyzed spectrotemporally in the dorsal STG, the correct information on the phonetic sound sequences (and the correct phonological analysis of them in the phonological network) cannot be transmitted to the sensorimotor interface at the parietal-temporal Spt, which in turn would lead to failure in transmitting appropriate relevant sensorimotor information to the articulatory network in the pIFG, PM, and anterior insula for articulation/externalization of the expected phonetic sound sequences corresponding to the 'intended' hierarchically structured expressions supplied by the combinatorial network in the aMTG and aITS. Hence, the 'output problem' or the state of loss of expressive speech in LKS appears.[37] Consider Figure 9:

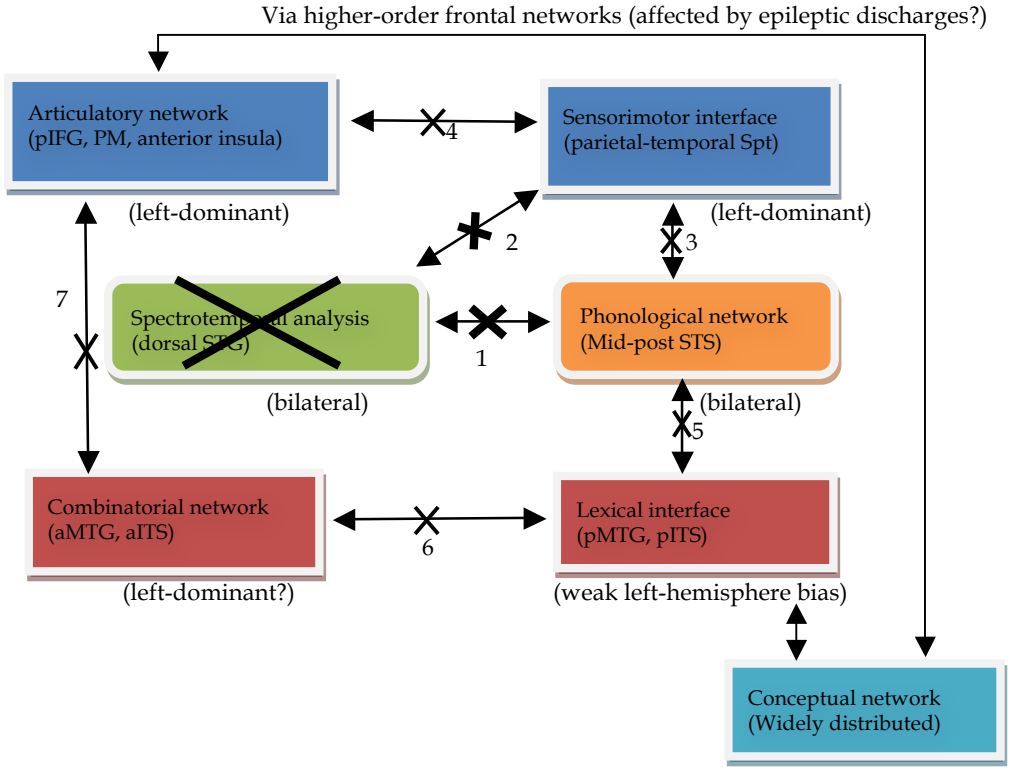

Figure 9: 'Domino effect' in LKS in the dual-stream model of speech processing. The deficiencies of the system of spectrotemporal analysis are indicated by a large, relatively thick cross, and the 'direct disruption' between the component of spectrotemporal analysis and that of phonological network or that of sensorimotor interface is depicted by small, relatively thick crosses, while the 'indirect disruption' between the other relevant systems is represented by small, relatively thin crosses.

The figure summarizes the 'domino effect' behind the 'input problem' and the 'output problem' in LKS that we discussed in the framework of Hickok &

[37] Pulvermüller et al. (2006) demonstrate, using event-related fMRI, that speech perception activates motor circuits responsible for corresponding speech production, without any speech production. If this is the case, it is plausible to assume that children with LKS who have become capable of comprehending speech to some extent, due to the improvement of the system of spectrotemporal analysis, might be able to activate the phonological network and the sensorimotor interface, even without any overt speech production.

Poeppel's (2007) dual-stream model of speech processing (although we will not discuss the higher-order frontal networks, note that (part of) the networks may be affected by the characteristic epileptic discharges particularly in the brain of the children with LKS suffering from non-linguistic cognitive dysfunction). In proposing the dual-stream model of speech processing, Hickok & Poeppel (2007) make an interesting claim that the dorsal auditory-motor circuitry offers the basic neural mechanisms for phonological short-term memory. Given the domino effect in LKS depicted in Figure 9, it is expected that a child with LKS would suffer from phonological short-term memory disturbances due to the deficiencies related to spectrotemporal analysis in the STG. This prediction seems to be borne out. Majerus et al. (2003) report that there is a correlation between the quality of phonological working memory and the degree of activity in the STG (PET data) in their patients with LKS with varying prognosis.

If the reasoning above is basically on the right track, we would reach the following hypothesis about LKS (both early and ordinary) in (7):

(7) *Hypothesis on LKS:*

LKS only affects the neuronal-level mechanism(s) in the SM system for spectrotemporal analysis of acoustic signals of sequences of speech sounds, which will in turn result in failures to acquire further lexical items and to externalize I-language in the wake of the domino effect upon the dorsal stream and the ventral stream, although the potentiality of phonological, syntactic and semantic components in I-language *per se* remains virtually intact.

Accordingly, if our hypothesis in (7) is correct, the apparent 'disconnection' (Tsuru & Hoeppner 2007) in LKS should result from the dysfunction of the spectrotemporal analysis component in the SM system and the disruption of its relevant interconnections with other components due to the deleterious domino effect, as illustrated in Figure 9.

Finally, Berwick et al. (2013) emphasize that "regarding the neural mechanisms of human language, research should focus on distinguishing neural networks supporting the externalization of language from those engaged in core syntactic computations, such as 'merge'" (p. 96). The hypothesis in (7) is in line with this suggestion; so, if it is on the right track, LKS seems to be conducive to research in such a direction. Furthermore, it is to be recalled that, although children with LKS are highly likely to be incapable of producing speech, they will become capable of comprehending speech once the relevant neuronal-level mechanism(s) in the SM system start(s) to function properly. This point is important in understanding children with LKS in the context of the hypothesis in (7).

3.3.3. Modularity of Mind

Another fundamental assumption adopted in this paper is 'modularity of mind' (see e.g. Chomsky 1980/2005, 1981, 1984, 1986, 1995). From this point of view, I-language functions as an independent system, interacting with other modules such as the vision system, the number system, the memory system, the pragmatic

system (including theory of mind), the system of general knowledge, the sensorimotor system, and the thought system among others in the mind.[38] In connection with Chomsky's view of modularity of mind, a caveat seems to be in order. He clearly states that the faculty of language (FL) is "a subcomponent of (mostly) the brain that is dedicated specifically to language" (Chomsky 2004b: 104), but he also clearly defines the subcomponent as "a system, that is, its elements might be recruited from, or used for, other functions" (p.124). Accordingly, Chomsky's version of modularity of mind does not presuppose the existence of what Marcus (2006) calls '*sui generis* modularity' and is compatible with Marcus' description of '*descent with modification* modularity' (see also Marcus et al. 2013).[39] In fact, the modularity of mind is empirically supported by a variety of clinical symptoms of dissociations among cognitive sub-systems (e.g., Curtiss 1977, 1981; Yamada 1990; Smith & Tsimpli 1991, 1995; see also Jenkins 2000 for a concise review).[40,41]

As mentioned in section 2.2, children with ASD/AR differ significantly from children with LKS with regard to development of theory of mind. Generally, children with LKS can develop theory of mind as part of their pragmatic competence and proper attachment to their parents and caregivers; whereas, children with ASD/AR characteristically cannot or have difficulties to develop them (for LKS, see Pearl et al. 2001; for ASD, see Baron-Cohen 1995; Matsui 2010; Baron-Cohen et al. 2013). Thus, the contrast between LKS and ASD/AR suggests a dissociation between the module of I-language and the module of theory of mind (pragmatics) in a much clearer fashion.

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38 See Pinker (1994) and Jackendoff (1996), among others, for discussion on the independence of the thought system from I-language.

39 Marcus (2006) points out that recent neuroimaging results seem to support this view of modularity (for relevant evidence, see Crosson 1992; Lieberman 2002; Poeppel & Hickok 2004). Although this view is not explicitly stated by Lenneberg (1967), his conception of language and cognition in the context of biolinguistics is also to be considered as a precursor and is in line with Chomsky's view of modularity of mind. See Boeckx & Longa (2011) for recent discussion on the correct interpretation of Lenneberg (1967); see also Fujita (2016) for an interesting proposal on modularity of mind and FL, which is in line with Marcus (2006).

40 Fodor (1983) also proposes his view of modularity of mind, which differs from Chomsky's. Unlike Chomsky, for Fodor, modules are 'informationally encapsulated' without directly interacting with each other and the 'language module' is regarded as only an input system. This view of modularity of mind, to which Marcus (2006) refers as '*sui generis* modularity', clashes with the clinical findings: Generally, complete dissociation is very rare and co-occurrence of multiple cognitive deficiencies quite common, as argued by Marcus (2006; see also Marcus 2004). At the same time, it is to be noted that any non-modular domain-general view of the mind is also at variance with clinical cases that show symptoms of dissociation (even if not a complete one) among cognitive systems in the first place.

41 Karmiloff-Smith (2009, 2010) argues for what she calls 'neuroconstructivism', which rejects the notion of innate, genetically pre-determined modules in the mind, taking issue with the Fodorian modularity and claiming that human intelligence, including language, is an emergent property over developmental time as a result of dynamic and multidirectional interactions between genes, brain, cognition, behavior, and environment. It is to be noted that, unlike Fodor's notion of modularity, Chomsky's notion of modularity is *not* incompatible with Karmiloff-Smith's neuroconstructivism, especially in the context of the Minimalist Program, which deemphasizes innately specified domain-particular genetic endowment while emphasizing the interactions of environment and the 'third factor' (including developmental paths), giving rise to the Chomskyan system of modularity (Chomsky 2005); for discussion of the non-gene-centric nature of the Minimalist Program in the context of evo-devo, see Benítez-Burraco & Longa (2010).

This suggestion is quite significant for considering modularity of mind in connection with the apparent co-morbidity in LKS. It opens up the possibility that all relevant modules of the mind in the child with LKS develop as different systems, while they are simultaneously affected by spreading of the LKS-related epileptic discharges to various brain regions involved in functioning of these modules. Deonna (2000) also remarks that the loss of language in LKS does not necessarily mean a sign of global mental deterioration (dementia). This view can naturally account for the fact that some children with LKS suffer from only language disorder, while the other cognitive functions remain relatively intact, even though they may look apparently severely mentally handicapped due to the lack of verbal production.

Furthermore, if Jackendoff (1996) is right in claiming that 'inner speech' (i.e., the phonological output of I-language in the mind) aids us to articulate our thought by providing a 'handle' for attention, the dysfunction of the phonological network in LKS as a result of the 'domino effect' in Figure 9 suggests that the child with LKS would not have access to propositionally complex articulated thought associated with appropriate phonological forms, presumably until the recovery of the system of spectrotemporal analysis and the proper working of the mechanism(s) for phonological processing in the phonological network. This might account for the co-morbidity of deteriorated cognitive function of thinking among children with early LKS.

4. Broader Implications

4.1. Implications for Biolinguistic Research

First of all, identifying a group of children with early LKS would bring a benefit to investigation into the nature of human language in the field of biolinguistics. Note that, unlike in the case of ordinary LKS, children with early LKS stop acquiring their first language in the middle of its acquisition due to unavailability of linguistic input derived from malfunctioning of the SM system (see Figure 8). However, if the availability of linguistic input should come back some time before the end of the critical period, thanks to the success of epilepsy control and amelioration of the EEG abnormalities, for instance, one can theoretically expect the children to experience a 'linguistic big bang', with a modular reinstatement of the properly functioning SM system of I-language. Namely, children with early LKS would suddenly display an ability to produce syntactically complex sentences via Merge in their first language. Thus, this kind of linguistic big bang would reveal that the core computations of syntax and semantics are virtually innately determined, as assumed in the current theorizing of the Minimalist Program (see e.g. Chomsky 2004b, 2005, 2010, 2016 and Berwick & Chomsky 2016).

Furthermore, a linguistic big bang would demonstrate that, if linguistic input should become available (again) within the critical period in the sense of Lenneberg, externalization of I-language would still be possible even after the end of the critical period (see Figure 4). Thus, we can assume that, in theory, as long as linguistic input becomes possible within the critical period, language

development would occur later even in children with early LKS. However, in order to realize this theory-based conjecture, we have to deal with the problem of neural dysfunction in the SM system that hampers externalization of I-language, making smooth speech difficult in LKS (see also Tsuru & Hoeppner's 2007 'disconnection' view of LKS). To do this, we would like to suggest the use of transcranial direct current stimulation (Nitsche & Paulus 2000) as one of the possible non-invasive medical interventions using external devices, a subject we turn to in the next subsection.

4.2. Implications for Medical Intervention/Treatment/Research

4.2.1. tDCS Treatment

First of all, recall from section 2 that there are three different patterns in the recoverability prognoses of LKS: Approximately 50% of patients recover fully, while the remaining 50% recover partially or suffer from permanent aphasia/dysphasia (Mikati et al. 2010). Given this situation, it is imperative to consider effective ways of medical intervention on behalf of the remaining LKS-affected patients with partial or no recovery of (expressive) language ability.

Transcranial direct current stimulation (tDCS) is a non-invasive stimulation technique for inducing polarity-dependent focal changes in cortical excitability, modulating spontaneous neuronal network activity; anodal stimulation increases and cathodal stimulation decreases the excitability of the cortical areas underneath the active electrode (see Brunoni et al. 2012 and references therein). Thus, the former has an excitatory effect, while the latter has an inhibitory effect. This neuromodulation technique has been clinically employed for treatment of neuropsychiatric disorders such as major depressive disorder, chronic and acute pain, or drug addiction, as well as for rehabilitation of stroke, including stroke-induced aphasia, among others (see Brunoni et al. 2012; Fiori et al. 2011, and references therein).[42]

Also, tDCS has been applied to patients with LKS in an attempt to improve their clinical conditions. Although Varga et al. (2011) failed to demonstrate the efficacy of cathodal tDCS with an inhibitory effect in reducing the epileptiform activity in children, including children with LKS (age at tDCS: 6; 1 and 7; 2), they have at least shown that this non-invasive neuromodulation technique can be safely applied to children with epilepsy (see Varga et al. 2011 for details). On the other hand, Faria et al. (2012) successfully demonstrate that cathodal tDCS is not only safe but also possesses "enough cortical polarization power to modulate epileptic activity focally" (p. 424) in patients with epilepsy, including a patient with LKS (age at tDCS: 7;0), for whom approximately 50% reduction of the paroxysmal activity was observed.

One difference between Varga et al. (2011) and Faria et al. (2012) is that tDCS was applied to patients who were awake in the former and asleep in the

[42] Interestingly, Fiori et al. (2011) demonstrate that application of anodic tDCS (20 min, 1 mA) over Wernicke's area of patients with stroke-induced aphasia significantly improves word retrieval in the aphasics with a long-term effect on recovery of their anomic disturbances.

latter. Faria et al.'s (2012) success of tDCS application to LKS patients can be justified, because LKS patients usually have EEG abnormalities during sleep. Other differences between the two methods were a more precise localization of the epileptogenic foci, a more focal tDCS application, and quantified epileptiform EEG discharges during and immediately after tDCS when applied to patients who were asleep. These results show that epileptiform EEG abnormalities in LKS can be technically reduced by tDCS if it is applied to patients with LKS whilst they are asleep, and with a precise localization of the foci and a sufficient focal stimulation supported by simultaneous EEG recording. To the extent that Faria et al.'s (2012) approach is on the right track, EEG abnormalities in patients with LKS can be controlled to a significant degree by tDCS.

Notice that, even though LKS-affected children's epileptic clinical seizures can be readily suppressed by anti-epileptic medication, typically they still have EEG abnormalities until around 15 years of age (Ramanathan et al. 2012). However, as attested by Varga et al. (2011) and Faria et al. (2012), among others, tDCS may be safely applied to the affected areas of the brain before a patient's EEG has become normalized. This may also solve the problem of the time lag between the end of the critical period (say, 12–13 years of age) and the termination of EEG abnormalities (15 years of age) for the sake of providing linguistic input within the critical period for fully establishing I-language in time.

Given the safety and efficacy of tDCS for LKS, we would like to suggest that this neuromodulation technology be focally applied in a careful manner not only for targeting the epileptogenic origin to alleviate EEG abnormalities but also for targeting Hickok & Poeppel's (2007) 'linguistic neuropathways', including the ventral stream and the dorsal stream in the brain, in the hope of ameliorating the deficiencies in the SM system and directly resolving both the input problem and the output problem of LKS shown in Figure 8. In doing so, of course, application of tDCS should be as careful as possible, and the correct identification of the target areas of the brain is to be done as precisely as possible by neuro-imaging techniques such as EEG (Faria et al. 2012), MEG (Sobel et al. 2000), PET (Kang et al. 2006), and SPECT (O'Regan et al. 1998). Therefore, if EEG abnormalities of LKS patients can disappear by puberty (Massa et al. 2000; Ramanathan et al. 2012), it may be ideal to apply tDCS when the EEG abnormalities are controlled to some extent and when the focus of the epileptiform discharges can be detected more precisely.

4.2.2. tDCS Application to Various Language Recovery Patterns in LKS

Let us now consider theoretically how tDCS could contribute to linguistic improvement in children with LKS. To start with, in connection with Figure 9, we can make the following speculations for the theoretically conceivable three patterns of recoverability from LKS: full, partial, and no recovery. In considering this issue, it is imperative to define what these recovery patterns refer to.

Notice that the term 'recovery' comprises two components: recovery of verbal auditory comprehension and recovery of verbal production. Accordingly, we have to define each category of the three patterns of recovery in LKS in a more refined way in terms of the two components. We will summarize logically

possible patterns of linguistic recovery in LKS on the basis of three degrees of recovery in the ability of verbal auditory comprehension and verbal production, along with the recommended loci for tDCS application (which we will discuss later) in Table 3.

	Verbal auditory comprehension	Verbal production	Recommended Loci for tDCS Application
Full recovery	✓	✓	n.a.
Partial recovery (I)	??	✓	STA,1, 2
Partial recovery (II)	✓	??	AN, 4, 7
Partial recovery (III)	??	??	STA, PN, SMI, LI, CN, AN, 1, 2, 3, 4, 5, 6, 7
Partial recovery (IV)	Φ	??	STA, PN, SMI, LI, CN, AN, 1, 2, 3, 4, 5, 6, 7
Partial recovery (V)	✓	Φ	AN, 4, 7
Partial recovery (VI)	??	Φ	STA, PN, SMI, LI, CN, AN, 1, 2, 3, 4, 5, 6, 7
No recovery	Φ	Φ	STA, PN, SMI, LI, CN, AN, 1, 2, 3, 4, 5, 6, 7
Partial recovery (VII)	Φ	✓	n.a.

Key: STA = spectrotemporal analysis, PN = phonological network, SMI = sensorimotor interface, AN = articulatory network, LI = lexical interface, CN = combinatorial network, and numbers 1 to 7 correspond to the numbers for the interconnections in Figures 6 and 9.

Table 3: Logical possibilities of linguistic recovery patterns in LKS and tDCS application loci. ✓, ??, and ϕ stand for virtually complete recovery, incomplete/deficient recovery, and virtually no/extremely poor recovery, respectively.

First, the case of full recovery, shaded in blue, corresponds to a situation in which both verbal auditory comprehension and verbal production have recovered virtually completely (for this kind of case, see Landau & Kleffner 1957; Worster-Drought 1971; Deonna et al. 1977; Mantovani & Landau 1980; Dugas et al. 1991; Paquier et al. 1992; Kaga 1999). In this case, the system of spectro-temporal analysis can be assumed to have regained its proper function, and as a result, appropriate phonological representations can be formed in the phono-logical network system. The phonological representations can then be transduced to motor instructions in the sensorimotor interface system, which transmits the motor instructions to the articulatory network system, and this works in co-operation with the combinatorial network system for externalization, via the dorsal-stream pathway. In the ventral-stream pathway, then, the phonological representations can be associated with the proper semantic representations in the lexical interface system, and the formed lexical items will be sent to the combinatorial network system for constructing phrases and sentences, which will be externalized through the articulatory network system. Note that it must be assumed that no particular damage as a result of the domino effect is found in any of the components of the dorsal stream and the ventral stream in this case. Thus, obviously, no application of tDCS is necessary here.

The case of partial recovery (I), also shaded in blue, looks like full recovery superficially on verbal production-based prognosis but can only be categorized as partial recovery under our criteria: The system of spectrotemporal analysis has not recovered 100%, and the still defective but sufficient information could flow into both the dorsal stream and the ventral stream, which have not suffered any damage from the domino effect. So, I-language has been externalized verbally in a fluent manner, possibly with some degree of mis-articulation due to the deficiency of spectrotemporal analysis. This case, which is clearly reported in Paquier et al. (1992) and Kaga (1999), may imply that although the quality and quantity of verbal auditory input is not perfect, verbal externalization could be possible as long as I-language is established without any domino effect damage on the dorsal and the ventral stream; thus it is speculated that a modicum of input might be enough to trigger functioning of I-language. The two cases in blue account for ca. 50% of LKS patients, according to Mikati et al. (2010). In the partial recovery pattern (I), bilateral application of tDCS to the dorsal STG and the connecting routes (indicated by 1 and 2) between the dorsal STG and the mid-post STS and between the dorsal STG and the parietal-temporal Spt, respectively, is recommended to improve the function of spectrotemporal analysis and its interconnections with the two systems, as specified in Table 3 (see Figure 9 for reference).

The next three cases (II), (III), and (IV), shown in orange in Table 3, are partial recovery on verbal production-based prognosis, which accounts for 25 % of LKS patients, based on Mikati et al.'s (2010) data. In the case of partial recovery (II), verbal auditory comprehension has become virtually normal, while verbal production has still remained defective (see Mantovani & Landau 1980), presumably because of the domino effect on at least the articulatory network system in Figure 9. In (II), the virtually normal verbal auditory comprehension seems to suggest that all the components and interconnections other than the articulatory network and its interconnections (4 and 7) with the sensorimotor interface and the combinatorial network have recovered and are functioning properly. Hence, tDCS should be applied to pIFG, PM, anterior insula, and the connecting routes 4 and 7, as indicated in Table 3 (see Figure 9 for reference).

Although precise identification of the cause of such damage from the domino effect in the partial recovery pattern in (II) awaits further investigation, with respect to the dorsal stream at least, it seems reasonable to imagine the following as one possibility. If the articulatory network is not employed for a relatively long period of time in LKS, due to persistent dysfunction of the interconnection between the sensorimotor interface and the articulatory network, the strength of the neural connection between the two systems would become weakened in the LKS patient, leading to difficulties in recovering expressive speech.

Another partial recovery pattern (III), on the other hand, constitutes a case where both verbal auditory comprehension and verbal production have stayed defective (for this kind of case, see Worster-Drought 1971; Deonna et al. 1977; Mantovani & Landau 1980; Ansink et al. 1989; Dugas et al. 1991; Paquier et al. 1992; Penn et al. 1990; Kaga 1999; Kimata et al. 2014). Such a state of affairs implies that both the dorsal-stream and the ventral-stream pathways have sustained some damage from the domino effect triggered by the disruption of the spectro-

temporal analysis system. Therefore, tDCS should be applied to the cortical areas related to all the relevant components and interconnections to improve verbal auditory comprehension and verbal production, as specified in Table 3.

Furthermore, the partial recovery pattern in (IV) (see Worster-Drought 1971; Deonna et al. 1977; Dugas et al. 1991) might strike us as a bit odd. Since the system of spectrotemporal analysis has stayed defective in this case, new spectrotemporal analysis of streams of sounds should be extremely difficult or virtually impossible. However, suppose that the phonological network system and both the dorsal stream and the ventral stream were to be free from any serious damage from the domino effect in Figure 9. Suppose also that, before the onset of LKS, some degree of first language acquisition has been carried out, with a certain amount of lexical items being stored in the mental lexicon. Then verbal externalization of I-language should be partially possible, albeit with some degree of defective articulation due to the deficiency of the system of spectrotemporal analysis. This should be, at any rate, a rare case, probably not easily seen among ordinary LKS children. With respect to tDCS application in this case, similarly to the case in (III), the cortical areas responsible for all the relevant functions and interconnections must be properly targeted to ameliorate verbal auditory comprehension and verbal production, as recommended in Table 3.

As for 'apparent no recovery' on verbal production-based prognosis, indicated in red, two partial recovery cases in (V) and (VI) are included in addition to no recovery. First of all, in the case of no recovery (for this kind of case, see Worster-Drought 1971; Deonna et al. 1977; Dugas et al. 1991), presumably due to the severity of the damage to the system of spectrotemporal analysis, appropriate auditory phonetic information cannot be linked with phonological representations in the phonological network system. As a result, neither the dorsal-stream pathway nor the ventral-stream pathway would be able to function due to the lack of input of proper information.

Note, however, that the case of no recovery in Table 3 should not be taken as suggesting 'no I-language'. To the extent that other modalities such as visual linguistic input in a sign language are available within the critical period to the LKS child with no recovery of verbal auditory comprehension and verbal production, the child could still acquire a sign language as his/her mother tongue (see e.g. Bishop 1982; Deonna 2000; Roulet-Perez et al. 2001; Deonna et al. 2009 for discussion on the effectiveness of use of sign language learning in LKS). Note, incidentally, that the fact that LKS patients can acquire a sign language with the proficiency that equals that of an individual with congenital deafness (Roulet-Perez et al. 2001) clearly shows that "higher-order linguistic representational processes are relatively spared in LKS" (Stefanatos 2011: 969). In addition, theoretically, there remains a possibility that application of tDCS is still effective even in the case of no recovery. If the cortical areas related to all the relevant components and interconnections are targeted, as indicated in Table 3, both the functions of verbal auditory comprehension and verbal production in LKS-affected children might be ameliorated in this category.

The partial recovery pattern in (V) is often mistaken for no recovery because of the lack of verbal production. But in fact it is a case in which verbal auditory comprehension has virtually recovered completely, indicating that the

system of spectrotemporal analysis has been sufficiently reinstated and all the relevant components and interconnections in the dorsal stream and the ventral stream have recovered enough and are functioning properly, except for the articulatory network and its interconnections with the sensorimotor interface and the combinatorial network (4 and 7). As such, on a par with the partial recovery pattern in (II), tDCS should be applied to pIFG, PM, anterior insula, and the connecting routes 4 and 7, as shown in Table 3 (see Figure 9 for reference).

In the partial recovery case of (VI) (see Landau & Kleffner 1957), in which verbal auditory comprehension has recovered incompletely/deficiently and verbal production has remained virtually nil due to the incomplete recovery of the system of spectrotemporal analysis and some serious damage from the domino effect at least on the dorsal stream, verbal externalization of I-language will be impossible. Unlike the pattern in (V) but similarly to the patterns in (III) and (IV), tDCS must be applied to the cortical regions in charge of all the relevant functions and interconnections properly to improve both functions of verbal auditory comprehension and verbal production, as specified in Table 3.

Note, incidentally, that the partial recovery patterns in (V) and (VI) as well as no recovery may likely lead to simple 'no recovery' prognosis, which might in turn lead to misdiagnosis of LKS patients in the red zone in Table 3 as having ASD/AR, and as a result, impede proper medical treatment of them.

The final case of partial recovery pattern (VII), shown in purple in Table 3, is not attested as LKS but corresponds, so to speak, to 'pure Wernicke's aphasia'. In this case, verbal auditory comprehension is supposed to have remained virtually nil, while verbal production is supposed to have recovered virtually completely. The non-existence of this recovery pattern in LKS seems to suggest that the output problem cannot be resolved, at least in theory, unless the input problem can be resolved to some extent.

In addition to tDCS application to the language-related brain regions, if the neuromodulation technique could be equally successfully applied to the relevant brain regions responsible for the co-morbidity listed in (1) and other related cognitive dysfunctions, such non-linguistic disturbances could be alleviated as well. Thus, it might be applied to the pre-motor/motor cortex for improvement of fine motor skills and the perisylvian cortex including the STG, STS, and insula for amelioration of an array of 'autistic behavioral disturbances' (see Stefanatos 2011 and references therein for the point that deficits in the perisylvian cortex are responsible for such autistic behaviors). Given that tDCS was invented and has been widely employed in treating various motor and cognitive disorders, this move for treatment of LKS seems to be quite natural (see, e.g., Hummel & Cohen 2006 for application of tDCS to rehabilitation of stroke patients).[43]

[43] Bludau et al. (2014) show that the human frontopolar cortex is made up of two cytoarchitectonically and functionally distinct areas called lateral frontopolar area 1 (Fp1) and medial frontopolar area 2 (Fp2) and that Fp1 is involved in cognition, working memory and perception while Fp2 is responsible for affective processing and social cognition. If the EEG abnormalities in LKS also affect the frontopolar cortex, deficiencies in these functions would be expected, and thus possibly these areas in the frontopolar cortex might be considered as relevant targets for application of tDCS in some cases of LKS and ASD/AR as well.

4.2.3. 'Risk Markers' of LKS

Just as important as selection of effective medical interventions for LKS patients is to identify correctly potential LKS patients among the vast ASD-diagnosed population (Tharpe et al. 1991) and differentiate them especially from AR patients. In order to avoid misdiagnoses, we should pay careful attention to the following 'risk markers'.

First, it is to be recalled that epileptic seizures (clinical or subclinical) in patients with LKS can be characteristically quite readily controlled with a single anti-epileptic medication such as benzodiazepines, in contrast to other cases of epileptic seizures in children or adults, which often require the use of more than one kind of anti-epileptic medication (Pearl et al. 2001). Thus, this criterion can be the first risk marker for LKS. If the children in question fit into this characterization, they should be suspected of having LKS as a first approximation.

The second risk marker for LKS is concerned with the presence of the EEG abnormalities with CSWS over the temporal (or perisylvian) regions. The occurrence of CSWS during non-REM sleep and its location over the brain regions can be essential for diagnosis. Although the disappearance of EEG abnormalities of LKS-affected children requires us to wait until puberty, as already mentioned, the EEG abnormalities will generally disappear by/around 15 years of age (Ramanathan et al. 2012), while children with ASD do not necessarily suffer from epileptiform EEG abnormalities, which can be infrequent and intermittent, if any (McVicar 2005).

Finally, although this is rather a psychiatric criterion, as discussed in section 2, one prominent characteristic of children with LKS is that they can develop pragmatic ability including theory of mind and can enter into interpersonal social communication without serious problems, in contrast to children with ASD/AR. As such, if a child in question has this characteristic, he/she should be counted as a possible candidate for LKS rather than ASD/AR. Thus, it is recommended to take EEG of all children with language regression during the entire time that they are asleep, including non-REM sleep, as conducted in McVicar et al. (2005), to discover potential LKS patients, who can be somewhat different from typical ASD/AR children in terms of pragmatic competence.

4.3. Implications for Developmental and Educational Therapy

In discussing problems with behavior therapy, which aims to 'train' children with developmental disorders, Konishi (2011) remarks that, although behavior therapy may be helpful to some degree toward severely autistic children who lack speech, caution must be exercised in using such a therapy toward children with Asperger's syndrome and those with developmental disorders who have come to acquire language. He points out that such a mechanical training in behavior therapy will cause too much burden on the children and their parents/caregivers and have emotionally negative impact on the children. We believe that the same holds with respect to children with LKS who have regained verbal auditory comprehension without (sufficient) verbal production.

Given that they have functioning I-language without externalization, they should be put in a natural environment where their parents, caregivers, therapists, and peers communicate with them by using natural languages rather than artificial communication systems such as artificial gestures or pictures used in developmental therapy. Note that some children with LKS can normally regain the ability of language comprehension (but not usually the ability of language production) in due course under anti-epileptic medication. This clearly indicates that they have I-language without externalizing it. Therefore, to increase language input, natural language is better suited for stimulating children with LKS, which would help boost their language comprehension.

However, we should pay attention to the tendency that children with LKS would not have access to propositionally complex forms of thought associated with appropriate phonological forms, before the recovery of the mechanisms for spectrotemporal analysis and phonological processing (see section 3.3.3). Therefore, the use of short, simple sentences with clear phonetic articulation in natural language contexts is recommended when addressing the children with LKS.

It is also to be noted that, as discussed in section 2, children with LKS are capable of developing and maintaining pragmatic cognitive functions, unlike the quintessential case of ASD/AR, and can socially communicate with others appropriately, even if non-verbally, by reading the minds of others without any problems. Given this nature of LKS, it is important to create environments or design educational settings where children with LKS can interact closely and form emotional bonds with their parents, caregivers, educators, peers, and therapists. The children can then maximize their pragmatic cognitive ability by using their natural language. Given that it normally takes approximately four years for theory of mind to fully develop in children (Wellman et al. 2001), parents and caregivers of a child with early LKS might give up trying to foster communicative interactions by appealing to the child's own pragmatic ability, including theory of mind, under the misjudgment or misdiagnosis of their child as ASD/AR. The parents and caregivers might misunderstand the child's behavioral disturbances, verbal auditory agnosia, and loss of expressive speech caused by EEG abnormalities as merely 'autistic' symptoms.

Moreover, the preserved pragmatic ability and willingness to communicate in LKS-affected children could possibly contribute to the restoration of their output abilities. Deonna (2000) warns that a prolonged disruption of the activity of auditory cortex can permanently impair some components of auditory functioning, and this could be applied to reproducing speech acts as well. Since LKS-affected children have longer absence of output experiences, they may give up externalizing I-language in spite of their potential abilities, unless they have a strong desire to listen to and communicate with others including parents and caregivers, demonstrating a 'dysbulia of speech' (Stefanatos 2011: 140). With ample developmental connection with others and willingness to communicate, the final stage of intake of verbal auditory input to connect with sensorimotor skills for articulation would accelerate and stimulate the emergence of speech production. Otherwise, LKS-affected children without recognition of the meaning of language and communication would finally be doomed to mutism. With a belief in LKS-affected children's hidden abilities of comprehending linguistic

input and with a hope of their being able to externalize I-language, parents and caregivers should continue to engage the children in natural daily conversations and show them the joy of communicating with others.

Finally, children with LKS show fluctuations with respect to the degree of linguistic and cognitive recovery, which often frustrates them and their parents/caregivers. Accordingly, it is also vital for them to be raised and provided with therapy in a stress-free setting. Unfortunately, there is no established special therapy currently available for children with LKS (see Jansing 2007 and references therein).[44] Accordingly, there is no special educational institution designed for them either (see e.g. Penn et al. 1990). Thus, it is urgently hoped for linguists, doctors, therapists, educators, and parents/caregivers of children with LKS to collaborate closely in creating a better educational condition in the near future (see Gordon 1990).

5. Concluding Remarks

This paper has examined the so-called Landau–Kleffner syndrome (Landau & Kleffner 1957), particularly from the perspective of I-language and the critical period hypothesis. We argued that this childhood language disorder provides further empirical foundations to the critical period for first language acquisition and modularity of mind as well as modularity of FL, while elucidating the linguistic mechanisms behind the language disorder in LKS by invoking the framework of Hickok & Poeppel's (2007) dual-stream model of speech processing. It was also claimed that the concept of what we called *early LKS* holds a key to differentiating children with LKS from those with ASD/AR.

From a medical perspective, we first emphasized the importance of discovering potential LKS-affected children from the vast ASD-diagnosed population by paying close attention to the three 'risk markers': (i) whether or not epileptic seizures (clinical or subclinical) in the patient can be readily controlled by a single anti-epileptic medication; (ii) whether or not the EEG abnormalities with CSWS exist over the temporal (or perisylvian) regions during non-REM sleep and can be normalized by around 15 years of age; and (iii) whether or not the patient can develop pragmatic knowledge, including theory of mind, to the extent that he/she can engage in interpersonal social communication, even non-verbally.

Much more careful scrutiny is urgently called for in diagnosing such children with early language disorder and other cognitive dysfunctions. Especially, the number of autistic children has been dramatically increasing for the last few decades (see Sumi 2015 and references therein), so it can be presumed that children with early LKS are included in the large population. This implies that more early-LKS patients might exist than are being reported, given the possibility

[44] But see Hurley & Hurley (2009), who report a case study of auditory remediation for a patient with LKS, which employs two distinct auditory training programs (Fast ForWord® and dichotic interaural intensity difference (DIID) training). They argue that the improvement of the patient's auditory system as a result of the two training programs suggests "the plasticity of the central auditory nervous system" and provide "a viable auditory remediation therapy" for LKS patients. See Hurley & Hurley (2009) for details.

of mixing the loss of the early-stage language development in LKS with the congenital lack of language development in addition to developmental cognitive and co-morbidity problems seen in other disorders like ASD/AR. Unfortunately, the necessity for revision of the original definition of LKS has not been well understood and shared by all medical specialists (see Stefanatos 2011). If any LKS-characteristic EEG abnormalities can be detected correctly at an early stage and (potential) epilepsy can be controlled with appropriate anti-epileptic medicine, clinical interventions would become possible to regain the language development.

Furthermore, from a linguistic viewpoint, we first analyzed the mechanisms behind the verbal auditory agnosia and loss of expressive speech in LKS on the basis of Hickok & Poeppel's (2007) dual-stream model of speech processing, and then emphasized the importance of eliminating the EEG abnormalities with the use of appropriate anti-epileptic medication and the intake of vitamin substance to improve the function of mitochondria in neurons (see fn. 8) in order to facilitate language input internally and establish I-language before the critical period ends. With I-language establishment in time in terms of the critical period hypothesis, language restoration becomes possible theoretically as long as linguistic input has been processed properly before the end of the critical period. In addition, in order to solve the input problem on lexical acquisition and output problem on I-language externalization of LKS patients, as a promising protocol for medical intervention, we suggested the possible loci for application of tDCS to seven recovery patterns of LKS patients as external medical intervention, as summarized in Table 3, based on Hickok & Poeppel's (2007) dual-stream model of speech processing. We also claimed that the EEG abnormalities are the culprit of LKS and that the language disorder and concomitant developmental cognitive and behavioral disturbances are secondary epiphenomena, suggesting that the restoration of the language function as well as other cognitive and sensorimotor functions would be possible by resolving the neural dysfunction and disruption among the relevant brain regions with proper application of tDCS. In sum, as explained above, using both internal and external medical intervention is highly recommended to treat LKS patients.

In addition, it is extremely important to discover children with early LKS, and closely observe and analyze the patterns of language and other cognitive development after they have recovered from LKS and re-started externalizing I-language. This would lead to providing further empirical evidence for Lenneberg's critical period hypothesis for first language acquisition and for Chomsky's modularity of mind and modularity of FL, as discussed in section 3. As our final speculation, let us touch upon LKS in connection with the issue of evo-devo on human language based on our assumption of early-LKS patients' possible 'linguistic big bang' in first language acquisition. Chomsky (2010) speculates that the human language capacity evolved as a result of some genetic mutation, which led to some neural re-wiring of the brain around 50,000 years ago in Africa and that externalization of I-language took place at some point subsequent to the evolutionary event. Even though it is surely impossible to pin down the cause of externalization of I-language at the moment, LKS seems to suggest one possible scenario. Recall that we characterized LKS as a case where externalization of I-

language is hampered by neural dysfunction caused by epileptiform abnormal electrical discharges as reflected in the EEG abnormalities. Suppose that some Homo sapiens individual who had I-language without its externalization was attacked by a series of epileptic seizures (clinical or subclinical) for some reason, which led to neurally connecting unconnected parts of the brain, resulting in externalization of I-language.[45] If this speculation is not widely off the mark, a patient with LKS might well be regarded, so to speak, as a 'living fossil' or more correctly a 'living proof' of reflecting the state of I-language in our ancestors before its externalization in the evo-devo context. Although this is a mere speculation, it might be compatible with Chomsky's (2010) view that the I-language externalization problem "may not have involved an evolutionary change—that is, genomic change" (p.61).

Last but not least, we would like to emphasize the importance of investigating LKS from both 'bottom-up' and 'top-down' perspectives in a collaborative and systematic way, so that we can discover the real cause(s) of the clinical symptoms and gain more understanding of the underlying mechanisms behind language and other cognitive functions in the brain. On one hand, the bottom-up approach to LKS has been extensively attempted in the field of medicine, accumulating relevant data on LKS from patients, as has been cited in this paper. On the other hand, the top-down approach to LKS has not been seriously undertaken thus far, and this is exactly where the field of biolinguistics can play a pivotal role and make a great contribution. They can provide a theoretical model of language and related cognition in light of biology and linguistics. It is our sincere hope that the present study will serve to facilitate further collaboration among professionals, including linguists, biologists, cognitive neuroscientists, medical doctors, developmental therapists, educators, parents/caregivers, and so forth in discovering more children with (early) LKS and zeroing in on the ultimate cause(s) and cure for the disease.

Landau concludes his remark with the following hope:

> Just as Schilder's disease has become a more intellectually gratifying illness called *adrenoleukodystrophy*, Frank Kleffner and I hope that an organized research effort may spare the next generation of pediatric neurologists from the useless chore of recalling our names.
>
> (Landau 1992: 353)

It is also our desire that 'Landau–Kleffner's dream' will come true in the near future, with the top-down and bottom-up approaches converging on a concerted enterprise and endeavor for this grand dream.

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45 For discussion of the effects of epilepsy on neuronal circuits in the brain, see e.g. Holmes (1991), Holmes & Ben-Ari (2001), Lynch et al. (1996). See also Benítez-Burraco & Murphy (2016) for discussion of the oscillopathic nature of language deficits in ASD. Although we will leave investigation into the oscillopathic nature of language and cognitive deficits in LKS to another occasion, we believe that detailed oscillopathic comparative study between ASD/AR and LKS from the perspective in Benítez-Burraco & Murphy (2016) will shed new light on the evo-devo issue and discovery of new protocols for ASD/AR and LKS as well.

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