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The assessment of auditory function in CSWS: Lessons from long-term outcome

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Summary

In Landau-Kleffner syndrome (LKS), the prominent and often first symptom is auditory verbal agnosia, which may affect nonverbal sounds. It was early suggested that the subsequent decline of speech expression might result from defective auditory analysis of the patient's own speech. Indeed, despite normal hearing levels, the children behave as if they were deaf, and very rapidly speech expression deteriorates and leads to the receptive aphasia typical of LKS. The association of auditory agnosia more or less restricted to speech with severe language decay prompted numerous studies aimed at specifying the defect in auditory processing and its pathophysiology. Long-term follow-up studies have addressed the issue of the outcome of verbal auditory processing and the development of verbal working memory capacities following the deprivation of phonologic input during the critical period of language development. Based on a review of neurophysiologic and neuropsychological studies of auditory and phonologic disorders published these last 20 years, we discuss the association of verbal agnosia and speech production decay, and try to explain the phonologic working memory deficit in the late outcome of LKS within the Hickok and Poeppel dual-stream model of speech processing.

KEY WORDS: Aphasia, Auditory agnosia, Speech processing, Phonology, Short-term memory.

Deafness versus Receptive Aphasia

Already in the seminal description of acquired aphasia with convulsive disorder, impaired speech comprehension was mentioned as the most prominent and often first aphasic symptom (Landau & Kleffner, 1957). Contrary to childhood aphasia related to structural lesions, in which the receptive disorder typically spares the phonologic level of processing, thus allowing the use of verbal repetition for remediation of lexical and morphosyntactic abilities, a severe disorder in auditory comprehension is considered a distinctive feature of the acquired epileptic aphasia. The progressive decay of verbal expression as a consequence of impaired auditory verbal processing leads in many cases to muteness.

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Does Auditory Dysfunction in CSWS Occur without Aphasia?

In the literature, auditory dysfunction in continuous spike-and-wave during slow sleep (CSWS) was investigated in Landau-Kleffner syndrome (LKS), either as an aphasic receptive disorder or an auditory verbal agnosia (Bishop, 1985; Denes et al., 1986; Beaumanoir, 1992). Impaired phonologic decoding is considered the primary deficit of the receptive aphasia in LKS (Rapin et al., 1977; Soprano et al., 1994), and the use of visual speech (lip-reading or cued speech) or sign language has served as a basis for remediation of verbal communication. Agnosia for nonspeech sounds has been reported in some cases, but in addition to the impairment of verbal processing. A comprehensive neuropsychological investigation of auditory processing in five children with LKS showed a clear dissociation between phonologic auditory and environmental sound discrimination (Korkman et al., 1998). Many authors have suggested that abnormalities in phonologic production or the decay of verbal expression leading in many cases to muteness result from abnormal auditory experience (Landau & Kleffner,

1957; Rapin et al., 1977; Bishop, 1985; Denes et al., 1986).

Landau and Kleffner related the presence of language disorders directly to a dysfunction in the cortical areas dedicated to verbal processing as a consequence of epileptic discharges (Landau & Kleffner, 1957). The similar effect of right temporal epileptic discharges on verbal processing has been explained as resulting from bilateral spreading of spike-and-wave discharges (Maquet et al., 1995; Morrell et al., 1995). Indeed, the bilateralization of discharges and their generalization in CSWS covering almost 80–90% of the slow-wave sleep time; the homotopic temporal cortex in the opposite hemisphere is no more available as a functional area for auditory and verbal processing (Morrell, 1995; Morrell et al., 1995). This may also explain why auditory disorders limited to nonverbal sounds have not been described in CSWS.

Neurophysiologic Investigations of Auditory Dysfunction in the Presence of CSWS

Children with LKS behave as deaf, despite normal hearing levels and normal brainstem auditory evoked potentials (BSAEPs). Almost all studies investigating the auditory system observed a normal functioning at least up to the level of the thalamus (Paetau et al., 1991). One investigation of the auditory and visual system in four LKS children with generalized CSWS predominant over the left temporal region, found that high-amplitude spikes generated within the temporal region altered the late, not only auditory but also visual evoked potentials (Mel'nichuk et al., 1990). These findings were discussed within Luria's conception of the integrative role of the temporal cortex essential to the understanding of speech: ''the convergence of auditory, visual, and modally non specific afferentation in the temporal cortex play[s] an important role in the understanding of speech'' (Luria, 1966). Data from recent neuroimaging studies provide evidence for the role of the superior temporal sulcus multisensory area in integrating auditory and visual information, not only about speech but also about objects and other behaviorally relevant stimuli (Beauchamp et al., 2004a,b). Using magnetoencephalography (MEG), Paetau et al. (1991) localized the epileptiform activity in the non-primary auditory cortex and showed that a unilateral focus altered the late auditory evoked potentials bilaterally (Paetau et al., 1991; Paetau, 1994). Other neurophysiologic data suggest that the primary epileptic focus in LKS involves the superior bank of the sylvian fissure in the area of the planum temporale (Morrell et al., 1995, Sobel et al., 2000, Paetau et al., 1999). Paetau et al. (1999) also showed that unilateral spike activity originating in the intrasylvian cortex could spread to the contralateral sylvian cortex and operate like a pacemaker with secondary spikes in the ipsilateral temporooccipital and parietooccipital areas. Such a sequence of events may explain the neuropsychological deficits associated with auditory dysfunction in LKS, involving speech production and behavior.

Impaired Verbal Abilities Subsequent to Phonologic Processing Impairment

Phonologic processing is essential for a number of cognitive abilities, and its impairment may compromise verbal comprehension, phonologic short-term memory, new word acquisition, and the acquisition of reading and writing skills. It has a pivotal role in working memory through the phonologic loop system specialized in storing verbal material. According to Baddeley's model of working memory, the phonologic loop system is composed of two subsystems: the phonologic store and the subvocal rehearsal system. The phonologic store receives directly and mandatorily all auditory verbal information and stores it into sound-based code, whereas the subvocal rehearsal system maintains the information within the store and serves to register visual information within the store, provided the items can be named (Baddeley, 2003; Baddeley et al., 1998). The phonologic loop system has been shown to play a crucial role in the acquisition of new words (Baddeley et al., 1998). According to the cognitive structure of the phonologic loop, the phonologic short-term store can be fed only with auditory information that has passed the phonologic analysis, which is also required prior to the rehearsal process. Therefore, a deficit involving only the phonologic analysis would disable the whole phonologic loop system and impede all verbal processing relying on it. If receptive language impairment, arrest in lexical growth, acquisition of novel morphosyntactic abilities, and further difficulties in the development of reading skills typically associated with auditory dysfunction in CSWS may be explained by a single impairment in phonologic processing, the decline of speech production subsequent to auditory verbal agnosia remains unexplained.

Speech Production Decline Following Auditory Verbal Agnosia

The relationship between verbal agnosia and speech production disorders in LKS may be accounted for within the recent model of functional neuroanatomy of speech processing proposed by Hickok and Poeppel (2007). This model attempts to resolve some paradoxical clinical

findings, such as deficits of speech production instead of auditory comprehension following a lesion of the left superior temporal gyrus, or deficits in discrimination or identification of speech consecutive to damage in the left frontal or inferior parietal areas. It posits that the early cortical stages of speech perception involve auditory fields in the superior temporal gyrus bilaterally (although asymmetrically) and that the cortical processing system then diverges into two broad processing streams, one involved in mapping sound onto meaning (the ventral stream), and another involved in mapping sound onto articulatory-based representations (the dorsal stream). According to this model, the first step of speech processing involving both hemispheres includes a spectrotemporal analysis in the dorsal superior temporal gyrus and a phonologic processing in the middle to posterior portions of the superior temporal sulcus. The second step of processing includes a left dominant sensory motor interface in the dorsal stream and a lexical interface in the ventral stream. The sensorimotor interface, located in a left lateralized region at the parietotemporal boundary of the posterior sylvian fissure—area Spt—is considered part of a larger auditory-motor integration circuit, which would play an important role in speech development and continue to support speech production and verbal working memory in adults (Hickok et al., 2003). This model may account for the main features of auditory dysfunction and their relationship with impaired speech production in LKS. Indeed, according to this model, left as well as right temporal focus spike-and-wave (SW) activity would similarly hamper the first steps of verbal auditory processing, spectrotemporal analysis, and phonologic processing, and consequently impair verbal auditory comprehension. Moreover, as SW activity spreads to the left posterior sylvian fissure, one may conjecture that its inhibitory effect on area Spt might also imply the anterior part of the dorsal stream, the posterior inferior frontal gyrus (IFG), thus hampering speech production. This model is also helpful in accounting for the impairment of phonologic short-term memory in the late outcome of LKS. An alternative explanation of impaired speech production in LKS could be related to progression or spread of the epileptic abnormality from the sylvian fissure to Broca's area through the arcuate fasciculus.

Auditory Function and Phonologic Memory in the Late Outcome of LKS

With the exception of rare cases with chronic auditory agnosia or aphasia (Baynes et al., 1998; Sieratzki et al., 2001), verbal auditory comprehension and expressive language recover with the normalization of the electroencephalography (EEG) findings at the beginning of adolescence. Comprehensive evaluation of auditory function consistently evidenced a one-ear dichotic listening extinction contralaterally to the focus of SW activity localized during the active phase of epilepsy in the posterior temporal area. A similar one-ear dichotic listening extinction was described in temporal or parietotemporal lesions (Kimura, 1961; Damasio & Damasio, 1980) and in the selective disruption of the geniculocortical pathway (Damasio & Damasio, 1979). Observed transitorily in temporal lobe epilepsy, it was related to continuous epileptic discharges, suggesting an interference of epileptic activity with the processing and transmission of simultaneously presented auditory information (Roberts et al., 1990). In five children who recovered from LKS, the comparison of auditory evoked potentials with those of five control children matched for age and gender provided arguments for a dysfunction in the associative auditory cortex contralateral to the dichotic extinction (Wioland et al., 2001). Poor performance in sentence, word, and nonword repetition in children who recovered from LKS has been related to a verbal short-term memory (STM) deficit, but a recent study investigating three children with LKS evidenced a selective impairment of phonologic STM with normal lexicosemantic STM (Majerus et al., 2007). This phonologic STM deficit may also be explained in the light of the model of cortical organization of speech processing, which proposes that the basic neural mechanisms for phonologic STM is supported by the dorsal auditory–motor stream including the posterior superior temporal gyrus.

Lessons for the Future

Several issues regarding the specification of auditory and verbal dysfunction during the active phase of and after recovery from CSWS remain unsolved. To improve our understanding of language disorders in LKS, neuropsychological investigations during the active epileptic period should tap the different levels of auditory verbal processing quite exhaustively. A comprehensive evaluation of the audiovisual, visual-motor, and auditory-motor integration may be useful to orientate rehabilitation. Indeed, auditory dysfunction arises from an SW focus in the temporal lobe, the posterior part of which is involved in multimodal integration along with phonologic processing. Consequently, audiovisual speech processing (e.g., cued speech) would be less appropriate than sign language when the integration of visual and auditory information is impeded by the epileptic activity.

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