APHASIA AND AUDITORY AGNOSIA
IN CHILDREN WITH LANDAU-KLEFFNER SYNDROME

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1. DELINEATION OF THE SYNDROME

Initially described by Landau and Kleffner\textsuperscript{12} as an “acquired aphasia with convulsive disorders”, this rare form of childhood aphasia occurs in the age range of three to eight years old. Seizures are infrequent but may precede, coincide with, or follow the language symptoms. Often of a generalized motor type, they may even not occur at all.\textsuperscript{19} EEG is abnormal, with uni- or bilateral 1 to 3 Hz spikes and waves predominating in the temporal regions. The more prominent and early manifestation of the language disturbances is an acquired word deafness (loss of comprehension for words). Receptive disorders may become so pronounced as to evoke peripheral deafness and may develop into auditory agnosia (loss of comprehension even for the environmental sounds). Expressive language deteriorates usually later on. These language manifestations fluctuate and are aggravated in the early stages at least, by the occurrence of seizures. Although they are considered, even in the princeps description, as a direct consequence of the EEG disorders, a clear relationship has not been demonstrated between the course of the seizures (which often subside easily with anticonvulsive therapy), the course of the standard EEG, and the course of the language deficit. Neurologic and ENT examinations are usually normal and no causal etiology has ever been clearly demonstrated either by CT scans, MRI or biopsy. Although a rare disorder, more papers have been devoted to its description than to acquired childhood aphasias of other etiologies together. Major clues for the explanation of this puzzling entity have appeared within the last few years from findings on sleep EEG that link the condition to ESES (epileptic status epilepticus induced by sleep),\textsuperscript{17} and from the demonstration of a specific cortical activation pattern on PET scan.\textsuperscript{13}
2. SYMPTOMS

As stated by De Wyngaert and Gommers\textsuperscript{2} “little attention has been given to the specific nature of the language disorders” in this syndrome, which is very strange, particularly in view of the recent discoveries in acquired childhood aphasia\textsuperscript{25}. The variability and fluctuations of symptoms might explain the relative paucity of linguistic data for this entity. Developmental milestones are usually normal in those children, in particular for language, although rare reports have indicated the contrary.\textsuperscript{18} In most cases at onset, the children become unresponsive to calls (“word deafness”): they develop a progressive lack of attention to language, with an ineffectiveness of voice raising (owing to the fluctuations of the symptoms, the troubles may be confused with attention deficit disorder as the “startle” response to loud noises is also usually suppressed). Soon after, the child frequently fails to react even to environmental sounds, thus disclosing “auditory agnosia”. Incipient peripheral deafness is evoked but audiometry remains in the normal limits. This striking initial receptive deficit has been particularly emphasized by authors who consider these severe comprehension disorders as the basis of the Landau-Kleffner syndrome and suggest that it be called” verbal auditory agnosia in children”.\textsuperscript{18} In some children, the deterioration of language affects reception only during some episodes, and, in rare instances, expressive impairment precedes the auditory disorders.\textsuperscript{20} Expressive symptoms consist of a difficulty in finding the name of well-known objects with auto-corrections resembling stuttering\textsuperscript{12} and a progressive loss of vocabulary. Phonemic or semantic verbal substitutions (paraphasias) are observed and even neologistic jargon (use of words of which at least half the phonemes are substituted so that they become meaningless).\textsuperscript{18,19} Sentences are short often in a “telegrammatic style” (use of non-inflected verbs) or with a reduction of “mean length utterance” (mean number of words pronounced in a given unit of time). Within a few days or weeks, the child may become mute, losing both his/her receptive and his/her expressive language abilities. Even in cases with initial jargon, in opposition to adult receptive aphasia where fluency usually does increase, expression soon becomes limited with phonological disturbances. However, the brevity of reports on those phonological disturbances does not always allow to decide if the authors refer to phonemic substitutions (the sounds, although incorrectly located, are well articulated) or to a disorder of articulation \textit{per se} with distortion of sounds pronunciation. In some cases, both conditions may co-occur or appear in succession. There can be a “return” to babbling with perseverative utterances. As with acquired deafness in very young children, this limitation in expressive speech in auditory agnosia has been interpreted in function of Geschwind’s hypothesis: “the child’s Broca area has not adequate practice to run as freely as that of adults with auditory agnosia”\textsuperscript{,1} An alternative explanation has been proposed by Van de Sandt-Koenderman \textit{et al.},\textsuperscript{21} who observed that, during acute speech breakdowns, precisely when more neologisms were produced, the children were less “inclined” to speak and had a reduced mean length of utterances, together with periods of production of grammatically complex sentences. In these cases, the reduction in speech utterances seems to be part of an overall hypospontaneity of speech, as often occurs in childhood aphasia.\textsuperscript{23} However, symptoms that are usually associated with motor aphasicas have been also described in Landau-Kleffner syndrome (dropping of words endings, consonants substitutions, dysprosodia)\textsuperscript{18} and purely expressive symptoms with mutism or syntactic deficits have occasionally been reported; but only in one case in ten are there no apparent receptive disorders.\textsuperscript{22} Children with late-onset Landau-Kleffner syndrome have been described by Gérard \textit{et al.}\textsuperscript{8} as a rule, there is a mixture of paraphasias and syntactic disorders. For these children, aged 9 and older, the