LENNOX-GASTAUT SYNDROME ASSOCIATED WITH DYSGENESIS OF CORPUS CALLOSUM

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Introduction: Lennox-Gastaut syndrome (LGS) is an electro-clinical syndrome composed of the triad of mental retardation, multiple seizure types, and the characteristic generalized slow spike-wave complexes in the EEG. In this article, we report on two patients with LGS whose brain MRI showed dysgenesis of corpus callosum(CC). We review the literature and stress the role of CC in the genesis of secondary bilateral synchrony (SBS).

Method: This was a clinical study conducted at King Khalid Hospital.

Results: The EEG was consistent with LGS in patient 1 and unilateral slow spike-wave complexes in patient 2. The MRI showed hypoplasia of the splenium of CC in patient 1, and global hypoplasia of CC combined with Joubert syndrome in patient 2.

Conclusion: Based on the data, we proffer the following hypotheses: 1-Hypoplasia of CC interferes with functional integrity of this structure. 2-The genu of CC plays a pivotal role in the genesis of secondary bilateral synchrony. 3-Electrodecremental seizures in LGS emanate from pacemakers generated in the brain stem, in particular the mesencephalon projecting abnormal signals to the cortex via thalamic nuclei. 4-Unilateral slow spike-wave complexes in the context of mental retardation and multiple seizure types may represent a variant of LGS, justifying neuroimaging studies.