IS THERE A CLINICAL SPECTRUM WITHIN LENNOX GASTAUT SYNDROME (LGS)?

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INTRODUCTION

Lennox Gastaut syndrome is one of the devastating childhood epilepsy syndrome that has been associated with poor treatment outcomes.

It is characterized by a triad of multiple seizure types, characteristic electroencephalogram (EEG) findings, and intellectual impairment.

OBJECTIVES

The study set out to characterize the seizure semiology, interictal EEG features, etiology and developmental outcomes of children with LGS to ascertain if there is a spectrum within LGS that can be defined by EEG and clinical features

METHODS

The records of all patients with slow spike wave complexes were retrieved and reviewed. Patients who met the authors' criteria for definition of LGS were recruited. LGS was defined by the authors as 1.) The presence of slow spike wave complexes greater than 3 seconds with a burst of fast rhythms and slow polyspikes during wakefulness, and paroxysmal fast in drowsiness and slow wave sleep, 2.) Multiple seizure types including generalized tonic seizures, and 3.)Cognitive impairment. All the video EEG/ medical records were reviewed, documenting the seizure semiologies, EEG features, possible etiologies and the patient's developmental outcomes.

RESULTS

Clinical -15 participants out of 36 patients with slow spike wave complexes met the inclusion criteria. About half (8, 53%)of the participants had a combination of 3 seizure types, 3 (20%) had 4 seizure types, while 4 had a combination of 2 seizures. The most frequent seizure in addition to tonic seizures were myoclonic (9, 60%), clonic(7, 46%) and atonic (6, 40%). Tonic-myoclonic-clonic and tonic-myoclonic –atonic were the commonest seizure combinations. The upper limbs were the most commonly involved body part in the tonic seizures. Ten (63%) had their first seizure in the first year of life.

Interictal findings - All patients had slow spike wave complexes and paroxysmal fast in sleep/drowsiness. Other common findings include spikes(13, 87%), generalized continuous slow (11, 73%) . Uncommon findings include background slow, asymmetry and hypsarrythmia.

Etiologies of the LGS among the participants include genetic/syndromic (8, 53%)e.g. Rett syndrome, chromosome 6q deletion, acquired e.g. periventricular leukomalacia following birth injury. Etiology was unclear in 5(33%) Fourteen (93%) had significant cognitive impairment with speech being most frequently affected.

Ten (63%) had their first seizure in the first year of life. Majority (8,53%) had 3 different seizure semiology combination. The upper limbs were the most commonly involved body part in the tonic seizures

CONCLUSIONS

Patients with LGS in this study had multiples seizure types most commonly myoclonic and clonic seizures with spikes and generalized continuous slow waves on EEG. Majority was of genetic origin and speech was the most common impairment

There were no significant differences in the etiology, seizure semiology and EEG finds of patients with LGS and no clear spectrum could be described

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