Infantile epileptic spasms syndrome in Chinese cohort Southern China: the clinical practice based on the 2017 and 2022 ILAE classification

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Background

Infantile spasm (IS) has been a knotty problem for pediatricians since Dr. West first reported it in 1841. International League Against Epilepsy (ILAE) Commission first defined IS as an epileptic syndrome (west syndrome, WS) with "the epileptic spasms (ES), a characteristic electrographic pattern of hypsarrhythmia and psychomotor developmental disorders" in 1989[1]. The ES are unmistakable seizures with unique characteristics, formally named in 2006[2], and then become an independent form of seizures. They consist of brief axial and rhizomelic contractions lasting usually less than one second, and often associated with a slow wave transient and an after going period of attenuation.

Introduction

In 2022, ILAE defined the infantile epileptic spasms syndrome (IESS) as a developmental and epileptic encephalopathy with the onset ES in early age, regardless of hypsarrhythmia and developmental regression[3]. IESS is the most common refractory epilepsy syndrome in pediatrics, with the estimated incidence of 30/100 000 liveborn infants, despite a range of specific treatments may be effective, including steroid hormone.

For IESS, usually three etiologic groups including structural, genetic, and unknown group be classified clinically according to the ILAE classification guidelines in 2017[4].

Current study based on the clinical practice according to 2017 and 2022 ILAE classification, focusing on the relationship between etiology, neurodevelopment and outcomes in children with IESS.

Materials and Methods

The children with IESS were diagnosed according to the following diagnostic criteria: 1) typical epileptic spasms were witnessed by ictal-EEG; 2) the age of ES onset be less than 2 years old.

The exclusion criteria were as follows: 1) the normal EEG when recording the clinical events of suspected spasms; 2) no regular follow-up. The clinical characteristics were collected, including demographic data, age of seizure onset, seizure types, hypsarrhythmia types, and neurodevelopmental status prior to seizure onset. All recruited cases were follow-up more than 6 months.

All cases were retrospectively studied and classified into west syndrome (WS) and non-WS group, with respectively etiological classification of genetic, structural or unknown etiology and therapeutic categorization of anti-seizure medicines (ASMs) only, steroid hormone and ketogenic diet (KD) group.

Results

228 cases were recruited, with etiological distribution as structural (n=73), genetic (n=69), and unknown (n=86). 184 cases belonged to WS group, though other 44 in non-WS group. Between WS and non-WS, only significant difference on electroencephalogram (EEG) outcomes (p=0.042) were found. Significantly etiological correlation to seizure, EEG and neurodevelopmental outcomes in WS group was revealed, but non-WS group wasn't. The seizure control outcome was similar for both WS and non-WS group on ASMs
only. In WS, steroids hormone treatment had an effective trend for seizure control for patients with unknown etiology, and had more effective trend for patients having structural etiology than those with genetic etiology. There was no significant correlation between etiology and outcomes in KD group. No stratified analysis was performed between outcome and etiology in non-WS due to limited cases accepted steroid (n=1) or KD treatment (n=6).

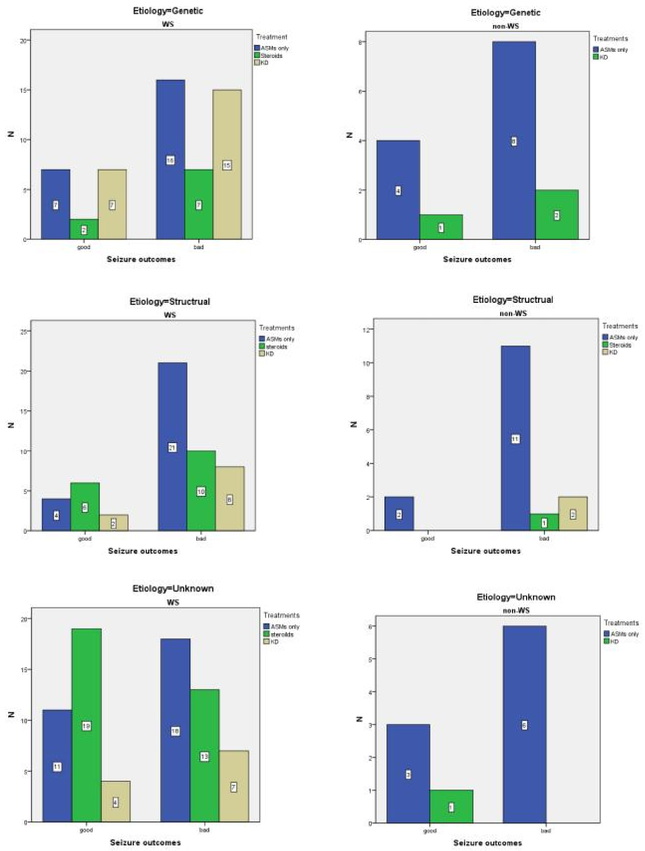

Chart 1. The seizure outcomes and treatment under different etiology in WS and non- WS group

Conclusions

For IESS patient without hypsarrhythmia, if delayed standardized treatment i.e. steroid hormone therapy, there may be no better seizure control or neurodevelopmental outcomes. Etiology and early treatment are both the important factors determining the prognosis of IESS, especially for those with non-WS based on 2022 ILAE classification.

References

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