

A0039: Use of Early Childhood Epilepsy Severity Scale (E-Chess) in Classification and Prognostication of Children with West Syndrome: A Study from Tertiary Care Pediatric Neurology Centre

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Background: West syndrome is a type of pediatric epilepsy syndrome often associated with a grave prognosis. The aim of this study was to evaluate clinicoradiologically cases of West syndrome, to use E-chess scoring, and classify and use it for prognostication.

Materials and Methods: Prospective observational clinical study for 1 year in the pediatric neurology out patients department of S.C.B. Medical College, Cuttack, Odisha. Patients were included as a case of West syndrome when they met all three criteria: (1) developmental plateau or regression, (2) epileptic spasm, (3) hypsarrhythmia on electroencephalography (EEG) who came to our center for first time (with/without previous treatments) after informed consent. They were classified into three groups by E-chess scoring according to severity of disease (→ **Table 1**).

Results: The total number of children included in the study was 13. Mean age of presentation was 9.4 months. Maximum patients were males. Mean duration of the disease was 3 months. Most of the patients were having hypoxic ischemic sequel in MRI. Twenty-three percent patients were categorized into type-III, 31% into type-II, 46% into type-I. Types II and III were drug-resistant epilepsy with poor response.

Conclusion: West syndrome is one of the infantile epilepsy syndromes with grave prognosis. E-chess scoring is a good and useful scoring system for classification and prognostication. This can be used in OPD basis for categorization of West syndrome. The types II and III are drug-resistant varieties with poor response to drugs. They should be planned for surgical therapy.

A0040: De Novo Mutations in TUBB2A Cause Infantile-Onset Epilepsy and Mental Retardation and Literature Review

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Objective: To investigate the clinical features of infantile-onset epilepsy and mental retardation caused by de novo mutations in TUBB2A and review the literature.

Methods: The clinical manifestations of two children with TUBB2A mutations in Pediatrics of Peking University, First Hospital were analyzed. Literature from Wanfang, Weipu, CNKI, and PubMed databases (self-built to August 2018) were searched and analyzed with the words “TUBB2A” and “epilepsy or convulsion” “epilepsy or convulsion.” Summary features included clinical presentation, EEG, imaging, treatment response, and genetic mutations.

Table 1 Classification of patients by E-chess scoring according to severity of disease

Types	E-chess score
I	6–9
II	10–12
III	13–15

Results: Two cases of de novo mutations in TUBB2A were females, with infantile-onset epilepsy and global developmental delay. Case 1 is a new hybrid heterozygous mutation: c.728C > T(exon4), p.P243L(NM_001069.2), MRI images showed white matter stunting, and the left frontal angle is slightly wider; case 2 is a newborn heterozygous mutation: c.743 (exon4) C > T, p.A248V (NM_001069.2), brain MRI showed bilateral anterior humeral gyrus with large cerebral gyrus, considering the possibility of giant gyrus deformity, bilateral frontal lobe abnormal signal, corpus callosum dysplasia. Literature retrieval was related with four references (in English), six cases were reported de novo mutations in TUBB2A, including this study has eight cases, three cases were p.A248V mutation. The subjects were infantile-onset epilepsy (spasms), with different levels of global developmental delay. MRI images were gyri developmental deformities, bad myelination and corpus callosum dysplasia. The seizure in p.P243L mutated was easy to control, and cerebral structural change was less severe.

Conclusion: TUBB2A mutations can cause infantile-onset epilepsy and global developmental delay, and different degree of brain malformations. The p.A248V mutation may be “hot spots” mutations. The p.P243L mutation caused a slight change in brain structure, and seizures were easily controlled.

A0041: Status Epilepticus in Pediatric Patients Severity Score (STEPSS): A Clinical Score to Predict the Outcome of Status Epilepticus in Children

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Purpose In adults, Status Epilepticus Severity Score (STESS) is a good predictor of clinical outcome and treatment response. We devised a pediatric modification of this score: Status Epilepticus in Pediatric patients Severity Score (STEPSS) and evaluated it in children with status epilepticus.

Methods: In this prospective study, children aged 1 month to 18 years presenting with seizure duration of at least 5 minutes or actively convulsing to the emergency were enrolled. The parameters noted for scoring STEPSS included: level of consciousness, age, type of seizure, and previous history of seizures. Outcomes included death, Pediatric Overall Performance Category (pOPC) at discharge, and treatment response. The primary outcome variable was the predictive accuracy of STEPSS score for determining unfavorable outcome (death or OPC ≥ 3; indicative of moderate disability or more).

Results: One hundred forty children (mean age = 5.8 years, 94 boys) were enrolled. Overall 15 children had an

unfavorable outcome, while 7 died. At a cutoff of 3, STEPSS had 93.3% sensitivity, 80.8% specificity, and negative predictive value of 99% for unfavorable outcome. The negative predictive values for death and treatment failure (refractory or super-refractory status epilepticus) were 99% and 98%, respectively. At a cutoff of 2, STEPSS had 100% sensitivity, 60.6% specificity for unfavorable outcome, and 100% negative predictive value for unfavorable outcome, death, and treatment failure. The predictive accuracy was comparable to that of STESS in adults.

Conclusion: The STEPSS, a simple bedside clinical score was found to be useful to predict the outcome and treatment response in children with status epilepticus.

A0042: Role of Pyridoxal Phosphate in Modulation of Endogenous Kynurenic Acid Synthesis Associated with Hippocampal Sclerosis

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Objective: Hippocampal sclerosis (HS) is the most common form of drug-resistant epilepsy where temporal lobe structures are responsible for unprovoked seizures. The hallmark of HS is enhanced glutamatergic excitatory neurotransmission. Pyridoxal 5-phosphate (pLP) is an active form of vitamin B6 and plays an important role as a cofactor of various enzymes including kynurenine aminotransferase II (KAT II), which catalyzes kynurenic acid (KYNA) synthesis from kynurenine within cortical astrocytes. KYNA, a tryptophan metabolite, is the only natural inhibitor of glutamate receptors. The present study was designed to test the hypothesis that in HS, altered PLP concentration is responsible for reduced endogenous kynurenic acid synthesis in hippocampus.

Methods: Hippocampus from HS patients ($n = 33$) were used for this study. Tissues resected from tumor margin during brain tumor surgery of seizure-free patients as nonepileptic control ($n = 14$) were used. To determine total KYNA and PLP concentration, tissues were kept in perchloric acid at -80°C . Tissues were homogenized, centrifuged, and supernatants were collected and estimated using HPLC with fluorescence detection (pLP excitation 300 nm, emission 400 nm; KYNA excitation 344 nm, and emission 404 nm).

Results: KYNA concentration was significantly less in HS hippocampus (0.0244 ± 0.0219 ng/mg of protein) compared with nonepileptic controls (0.2117 ± 0.1332 ng/mg of protein). PLP concentration was also significantly less in HS patients (1.77 ± 3.96 mg/mg of protein) compared with controls (10.91 ± 11.34 ng/mg of protein).

Conclusion: PLP concentration is reduced which, we suspect, may consequences in decreased KYNA due to dysfunctional machinery to synthesize kynurenic acid from kynurenine in HS patients.

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A0043: A Home-Based, Primary-Care Model for Epilepsy Care in Low and Middle Income Countries: Basis and Design

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Objective: A cluster-randomized trial of home-based care using primary care resources for people with epilepsy has been set up to optimize epilepsy care in resource-limited communities in low- and middle- income countries. The primary aim is to determine whether treatment adherence to antiepileptic drugs is better with home-based care or with routine clinic-based care. Secondary aims are to compare the effects of the two care pathways on seizure control and quality of life.

Methods: The home-based intervention comprises epilepsy medication provision, adherence reinforcement, and epilepsy self-management and stigma management guidance provided by an auxiliary nurse-midwife equivalent. The experimental group will be compared with a routine clinic-based care group using a cluster-randomized design in which the unit of analysis is a cluster of 10 people with epilepsy residing in an area cared for by a single accredited government grass-root health worker. The primary outcome is treatment adherence as measured by monthly tablet counts and two self-completed questionnaires. The secondary outcomes include monthly seizure-frequency, time to first seizure (in days) after enrolment, proportion of subjects experiencing seizure freedom for the duration of the study and quality of life measured by the "Personal Impact of Epilepsy Scale," all assessed by an independent study nurse.

Results: The screening phase and neurological evaluations and randomizations have been recently completed and follow-up is currently underway.

Conclusion: The Results of the trial are likely to have substantial bearing on the development of governmental policies and strategies to provide coverage and care for epilepsy in resource-limited countries.

A0044: Clinical and Demographic Profile of Epilepsy Patients in Rural Rajasthan

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Objective: The present study attempted to explore the clinical and demographic profile of epileptic patients, and pharmacological management in these patients.

Methods: The prospective study was done in center situated at Ratannagar, a Community Health Centre in district Churu under the auspicious of Epilepsy Care and Research foundation, an NGO involved with epilepsy work. It included 6,993 epileptic patients who visited the center from majority