

BECTS (BENIGN EPILEPSY OF CHILDHOOD WITH CENTROTEMPORAL SPIKES) - CLINICAL PROFILE, PROBLEMS IN SCHOLASTIC PERFORMANCE AND ELECTROENCEPHALOGRAPHIC CHARACTERISTICS

Paediatrics

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ABSTRACT

Background Benign childhood epilepsy with centrotemporal spikes (BECTS) is the most common and most characteristic form of idiopathic focal epilepsy. There is only scant data available from India on BECTS and on its impact on the cognitive and academic functions in the affected children.

Methodology The cases which satisfied the inclusion criteria were selected for the study. They were evaluated according to a standard protocol. Problems in scholastic performance was considered if there were failures in the examinations or if the child was repeatedly getting low grades in the examinations. Detailed neuropsychological evaluation was done in all.

Results A total of 73 children were included in the study. Male to female ratio was 41:32 (1.3). The age of onset of seizures ranged from 5.5 – 12 years. Majority of children 35(48%) had bilateral centrotemporal discharges. 45 (61%) children had parentally complained problems in scholastic performance, 44 children had neuropsychological impairment under any one domain or in combination.

Conclusion The salient observation in this study was that more than half (60%) of the children with BECTS had neuropsychological impairment with normal intelligence. Most of them (91%) had problems in scholastic performance showing a statistically significant association with neuropsychological impairment

KEYWORDS

BECTS (Benign Epilepsy of childhood with centrotemporal spikes), neuropsychological impairment

Background

Epilepsy is a disease of the brain defined by any of the following conditions

1. At least two unprovoked seizures occurring >24 h apart
2. One unprovoked seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
3. Diagnosis of an epilepsy syndrome

Epilepsy is considered to be resolved for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure-free for the last 10 years, with no seizure medicines for the last 5 years [1]

Epilepsy in children has wide spectrum of severity. Conditions like benign infantile convulsions, benign familial neonatal convulsion, and benign epilepsy of childhood with centrotemporal spikes have favourable outcome. On the other hand malignant syndromes like Ohtahara syndrome, West syndrome, febrile infection-related epilepsy syndrome (FIRES), Lennox Gastaut syndrome, Landau Kleffner syndrome, Malignant migrating partial seizure of infancy and Continuous spike-and-wave during slow wave sleep syndrome (CSWSS) have malignant evolution [2]. Precise classification of seizure type and epileptic syndrome is a prerequisite for appropriate management.

Benign rolandic epilepsy (BRE), first described in 1597 by Martinus Rulandus, is a form of partial idiopathic epilepsy, according to the 1989 ILAE Commission of Classification and Terminology classification. Many epileptic syndromes (including BRE) contain in their name benign or self-limited, which refers to the syndromes good treatment response and self-limiting nature. In these forms of epilepsy, there may be associated cognitive and behavioral co-morbidities, psychiatric illness, migraine, or sudden death syndrome; and thus, the term benign is not appropriate in these cases. Recent proposals for classification recommended the use of the term self-limited instead of benign. The characteristics of self-limited epileptic syndromes are as follows: a defined age of onset; seizures that are self-limited, meaning that the spontaneous remission has been reached; and the consequences of seizures, if they exist during the active phase of the disease, are not debilitating. However, it did not exclude an increased risk for cognitive or behavioral disorders of moderate intensity prior to the onset of seizures, during or beyond the active phase of the disease. The last classification proposal of epileptic syndromes includes benign rolandic epilepsy as a self-limited epileptic syndrome. [3]

Benign epilepsy of childhood with centrotemporal spikes was initially described as a benign epilepsy syndrome, but a number of studies later showed that a significant number of patients had some degree of

neuropsychological impairment. The evolution of benign epilepsy of childhood with centrotemporal spikes into electrical status epilepticus in slow wave sleep (ESES) as well as into Landau-Kleffner syndrome (LKS) has been described [4]. There is only limited data available from India on BECTS (Benign epilepsy of childhood with centrotemporal spikes) clinical profile and on its impact on the cognitive and academic functions in the affected children.

Methodology

Objectives of the study is to characterize the clinical profile, problems in scholastic performance, EEG and prevalence of neuropsychological impairment in children (<16 yrs) with BECTS presenting to M O S C Medical college, Kolenchery from March 2014 to March 2015

Study Design: Hospital based cross sectional study

Duration of Study: March 2014 – March 2015

Sample Size: Calculated sample size 81 (Using formula $4PQ/L_2$, Prevalence of educational problem in rolandic epilepsy/BECTS, $P \sim 55\%$ [5])

Study population: All the cases of rolandic epilepsy, which presented during March 2014 – March 2015 study period as outpatient or inpatient in department of paediatrics and department of neurology

Study Setting: M.O.S.C Medical College, Kolenchery. Department of Neurology, Department of Paediatrics.

INCLUSION CRITERIA:

Clinical criteria [6]

1. Brief partial mainly motor seizures that are stereotyped in semiology
2. Onset of seizures after 2 years
3. Frequent nocturnal occurrence and activation of epileptiform activities during sleep
4. No gross neurological defects

EEG Criteria [7]

1. Normal background
2. High amplitude biphasic spike followed by prominent slow waves in midtemporal (T3, T4) and central areas (C3, C4)
3. Occasional generalized spike wave discharges

“Benign epilepsy of childhood with centrotemporal spikes (BECTS) - clinical profile, problems in scholastic performance and electroencephalographic characteristics” – a cross sectional study conducted in MOSC medical college, Kolenchery during march 2014 – march 2015 after ethical clearance from institutional review board. All the cases of rolandic epilepsy, which presented during study period as outpatient or inpatient were screened by inclusion criteria. Those who satisfied the clinical and electroencephalographic inclusion criteria were selected

for the study .They were evaluated according to a standard protocol. Detailed history including seizure characteristics, age of onset, perinatal events, development and family history were obtained. Neurological examinations were carried out. EEG details including background activity and spike characteristics were included

In neuropsychological assessment attention, memory, learning disabilities were tested according to the age of the children using NIMHANS BATTERY for children .The IQ was assessed using the Malins intelligence scale for Children (MISC), which is a standardized Indian adaptation of the Wechsler intelligence scale for Children (WISC). Perceptual motor ability was tested using Bender gestalt test (BGT), Benton visual retention test (BVRT) and social functioning was assessed by Vineland social maturity scale (VSMS). Scholastic performance of the child was assessed by a standard parental interview. Problems in scholastic performance was considered if there were failures in the examinations or if the child was repeatedly getting low grades in the examinations. Datas collected were entered in to windows excel sheet and statistical analysis was done using chi-square test

Results

In this cross sectional study conducted in M O S C Medical College, Kolenchery (March 2014 –March 2015) a total of 73 children were included in the study. Male to female ratio was 41:32 (1.3:1). The age of onset of seizures ranged from 5.5 – 12 years (mean age 7.6 years).The average age of study population was 9.8 years.12 children (16.4%) had history of febrile seizures before the onset of epilepsy. 17 children (23%) gave a family history of epilepsy. Adverse perinatal events, like prematurity, minimal birth asphyxia and neonatal jaundice were reported, in 13 children (18%). None of the children had significant developmental delay; but delayed language development (DLD) was seen in 19 children (26%).

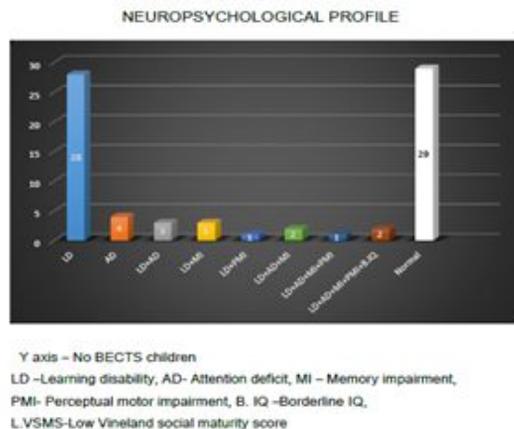
Most of the cases had less than five episodes of total number of seizures. 22 (30%) cases had only one episode of seizure. Majority children with rolandic epilepsy (41 cases [56%]) had one to five episodes of seizures. Five to ten episodes of seizures were observed in 7 (10%) cases. More than ten episodes of seizures were observed in three (4%) cases with atypical evolution .Children with rolandic epilepsy predominantly had [34 (46%)] nocturnal seizures, 10 (14%) children had only daytime seizures and the rest of them had combination of the two 29 (40%).

All patients had at least one EEG (electroencephalograph) typical of BECTS (benign epilepsy of childhood with centrotemporal spikes). Majority of children 35(48%) had bilateral centrotemporal discharges .Left sided centrotemporal discharges were seen in 21 (29%) and right sided discharges were seen in 16 (22%). One case initially had bilateral centrotemporal spikes later turned out to be continuous spike and wave during slow wave sleep syndrome (CSWS).

Overall analysis showed majority of children [63 (85%)] had less than five episodes of seizures which were well controlled by antiepileptic drug therapy. But few cases showed atypical evolution with severe neuropsychological impairment.

Among 73 children evaluated 45 (61%) children had parentally complained problems in scholastic performance, 97% (71) children had normal intelligence. Learning disability was found in 40/73 (55%), 16 (22%) children showed features of attention deficit, 8 (10.9%) had memory impairment and 4 (5.4%) had perceptual motor impairment. None of them had low social score by VSMS

The graph below shows that most common neuropsychological impairment was LD (Leaning disability) 40 (55%). Learning disability alone was found in 28 children. Rest of neuropsychological impairment include learning disability and attention deficit (3), learning disability and memory impairment (3), learning disability and perceptual motor impairment(1), learning disability, attention deficit and memory impairment(2), learning disability, attention deficit, memory impairment and perceptual motor impairment(1) and learning disability, borderline IQ, memory impairment and perceptual motor impairment (2). Attention deficit alone was found in 4 children



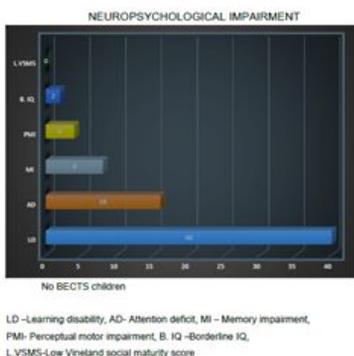
There were 73 children included in the study, in which 44 children had neuropsychological impairment under any one domain or in combination. So the prevalence of neuropsychological impairment in this study was 60%

In 44 children with neuropsychological impairment, 40 children (91%) had problems in scholastic performance based on parental interview whereas only 5 children without neuropsychological impairment had parentally complained problems in scholastic performance. Significant association was observed between neuropsychological impairment and parentally complained problems in scholastic performance (P value <0.05).Among children with seizure number more than five (10), 9 (90%) children had neuropsychological impairment. There was a significant correlation between total number of seizure episodes (>5) and neuropsychological impairment [P value - 0.003].Even though the prevalence of neuropsychological impairment in patients who were already on treatment was 66% (41/62), it was not statistically significant when compared with the prevalence of neuropsychological impairment [45 % (5/11)] in newly diagnosed case during study period, who were assessed before starting treatment (P value- 0.736). No significant correlation was found out between the presence of neuropsychological impairment and adverse perinatal events (P-0.532), delayed language development (P value-0.286), family h/o epilepsy (P value-0.57) or h/o febrile seizure (P value-1).

Conclusions

Benign epilepsy of childhood with centrotemporal spikes (BECTS) has been classically described as a well-defined paediatric epileptic syndrome with an excellent seizure outcome and free of neurological or neuropsychological deficits. The affected children generally have normal intelligence and normal neurological examination. [8]. In the current study majority of children (85%) had less than five episodes of seizures which were well controlled by antiepileptic drug therapy. Only three (4.1%) cases had atypical evolution, in which one case turned out to be CSWSS (continuous spike and wave during slow wave sleep syndrome). Neuropsychological examination revealed majority of children (97%) had normal intelligence, only two of them (3%) had border line intelligence quotient The salient observation in this study was that more than half (60%) of the children with BECTS had neuropsychological impairment with normal intelligence. Most of them (91%) had problems in scholastic performance showing a statistically significant association with neuropsychological impairment.

Recommendations: State dependent educational problems are observed in benign epileptic syndromes and are potentially reversible. It may be possible to identify the children at risk for development of scholastic problems by careful neuropsychological evaluation at the first visit itself. These children may need special evaluation and management by a multidisciplinary team.



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