

Is Benign Childhood Epilepsy with Centrotemporal Spikes Always Benign?

How to Cite This Article: Saeed M, Azam M, Shabbir N, Qamar ShA. Is Benign Childhood Epilepsy with Centrotemporal Spikes Always Benign? Iran J Child Neurol. 2014 Summer;8(3): 38-43.

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Received: 1-Sep-2013
Last Revised: 20-Mar-2014
Accepted: 4-Apr-2014

Abstract

Objective

To determine the prevalence of associated behavioral problems and prognosis with Benign Childhood Epilepsy with CentroTemporal Spikes (BCECTS). Descriptive, Cross Sectional study that was conducted from October 2009 to April 2013 in the Department of Pediatric Neurology, the Children's Hospital Taif, KSA.

Material & Methods

This study was conducted after approval from the Ethics Committee of the Children's Hospital Taif, Saudi Arabia. Thirty-two patients from the age of 3 to 10 years old were recruited from the pediatric neurology clinic over a period of 4 years. All the patients were selected based on history, EEGs, and neuropsychological and neurological examinations.

EEGs were performed for all the patients while in awake and sleep states. Those who had centrotemporal discharges were included in the study. All the patients also underwent a brain MRI. Only two patients had mild cortical atrophy but developmentally they were normal.

Results

In our study, prevalence of BRE is 32/430 (7.44%). Among the 32 cases, 24 were male and eight were female. Six cases out of 32 indicated a family history of BRE. Twenty-eight cases had unilateral right sided centrotemporal discharges and four had bilateral discharges.

Conclusion

It is possible that for BECTS, a high number of seizures might play an important role in the development of mild cognitive impairment and/or behavior disturbances.

Keywords: Benign childhood epilepsy with centrotemporal spikes (BECTS); EEG (Electroencephalography); Treatment; Behavioral problems

Introduction

Benign Rolandic epilepsy (BRE) or benign childhood epilepsy with centrotemporal spikes (BECTS) is the most common condition of idiopathic focal epilepsies in children. It is known to be benign because of the absence of neurological deficits, infrequent focal somatosensory or motor seizures predominantly occurring during sleep, reasonable responses to the medication, and spontaneous resolution before the age of 15 to 16 years.

BRE is the most common epileptic disorder in childhood with an incidence of

approximately 21 in 100000 of 5-20 years of age, which is characterized with brief repetitive unprovoked attacks (1).

BRE is an alternate name for BECTS. Genetic transmission is as an autosomal dominant trait and it represents 9.6–10.3% of all childhood epilepsies determined at presentation and 2 years later (2).

Although this disorder was once thought of as a universally benign syndrome, increasing evidence suggests that a subpopulation of children may present with recent impairment of overall cognitive functioning, or difficulties with visual perception, concentration, and short-term memory (3). Like all forms of epilepsy, BRE results in seizures. The seizures are usually mild. They typically begin in the face and can take a variety of forms, face or cheek twitching, tingling, numbness, or unusual sensations in the tongue or face, difficulty speaking, and drooling due to inability to control the mouth muscles (4,5).

In about one out of every two children with it, seizures spread from the rolandic area to the rest of the brain. When this happens, the seizure is called a secondarily generalized seizure. They are also called tonic-clonic seizures.

Typically, the seizures occur during sleep. For this reason, they may not be noticed at all. Other times, parents witness a seizure after investigating nighttime noises in their child's room.

Some children with BRE may also have learning difficulties and behavioral problems. These children may need additional attention and treatment.

Recently, several studies have demonstrated that children with BRE are at a higher risk for cognitive, behavioral, or emotional difficulties in association with frontal lobe dysfunctions (6,7,8).

The syndrome was not described and defined until the 1950s. The EEG features were first reported by Gastaut (1952), who called the discharges as pre-Rolandic and Gibbs and Gibbs (1952) entitled them mid-temporal (9). Nocturnal seizure accounts for 20–25% of all childhood epilepsies (10).

The present study assesses behavioral and other neuropsychological aspects without treatment in children who are newly diagnosed.

Material & Methods

This study was conducted in the Department of Neurology at the Children's Hospital in Taif after approval from the ethics committee of the Hospital from October 2009–April 2013.

During this period, 32 patients were consecutively recruited from the pediatric neurology clinic. Based on detailed history, clinical features, and EEG findings, a diagnosis of BRE was made by the neurologists and assessment of behavioral problems before and after treatment by the Childhood functional assessment rating scale, Childhood behavioral checklist.

These patients were included in the study and had experienced two or more episodes of seizures over the past year. Exclusion criteria included primary generalized seizures and partial epilepsy of a symptomatic etiology. Each patient went through comprehensive neurologic examinations and a battery of tests to evaluate various aspects of mental functions, including cognition, behavior, emotion, and attention both by a neurologist as well as a child psychiatrist.

The results of these patients were recorded on the prescribed Performa for statistical analysis.

Results

This study was conducted in 32 children with partial seizures who had rolandic discharges on their interictal EEGs. Prevalence rate of Rolandic epilepsy in our study was 32/430 (7.44%).

All patients were evaluated within the framework of follow up in the neurology clinic and intellectual ability, cognition, and behavior were assessed by a child neuropsychologist.

Among them 24 cases were male (75%) and eight cases were female (25%) (Fig 1). Six cases had a positive family history that included two cases with the same epileptic attacks in one of parent and four cases had one affected sibling. All of the children were developmentally normal.

In assumption, a positive family history was 18.75%. Out of 32 cases, twenty-four cases had only simple partial seizures (75%) and eight cases (25%) had secondary generalization.

Most of our patients had seizures that occurred nocturnally and became secondary generalized and the

events were associated with Rolandic epilepsy include anarthria, hemiclonic, or tonic posturing with excessive drooling, twitching of one side the face, and paresthesia of the face, gums, or inner cheeks.

All children underwent both a waking and a sleep EEG and patients were selected on clinical as well as on EEGs findings. EEG results are distinctive and diagnostic in benign focal epilepsy with centrotemporal spikes. Twenty-eight of the patients, (87.5%) had unilateral dominantly right sided discharges and four out of thirty-two patients, (12.5%) had bilateral discharges. Background was normal in all of our cases.

Neuroimaging MRIs were performed on all patients, in 30 patients, the results were unremarkable, but two patients revealed cortical atrophy.

Two of our cases had Todd's paralysis, (9.37%) and two patients admitted with status epilepticus, (6.25%). Eighteen of our cases, (56.25%) had seizures after falling asleep, six cases has seizures exclusively on awakening, (18.75%) and eight among them had both sleep and awake state, (25%) (Fig 2)

Twenty six patients observed seizures between the age of 4–6 years and among them eighteen cases were school going, (56.25%) and ten cases were under school age, (31.25%). All of our patients had more than two attacks of seizures and we counseled the parents about the treatment. Eighteen parents, (56.25%) agreed for management and fourteen cases, (43.75%) refused treatment but we continued follow up of all these patients to observe the cognitive, behavioral, and language problems. All patients who were treated with Carbamazepine at the start showed excellent results after three months of treatment.

Nine cases out of fourteen untreated patients during their follow up showed behavioral, cognitive problems, and their parents also noted same behavioral problems. Half of parents who agreed for the treatment also gave a history behavioral problems before starting treatment. The most common behavioral disturbances noted by parents were hyperactivity, aggressiveness, and lack of concentration. Parents who refused the treatment were counseled again for management and, eventually, agreed. All the patients were treated with Carbamazepine for two years. Among these patients, two out of fourteen did not respond well to CBZ. Levetiracetam was added

consequently all patients showed excellent response along with cure of behavioral problems. None of the all patients showed any significant side effects of medicines. In our study drug of choice considered was CBZ, all of our patients showed excellent response with CBZ and Levetiracetam in treating the seizures as well as behavioral problems.

Discussion

Despite the presumed benign nature of BRE, some evidence now exists that patients with this disorder may have cognitive, behavioral, emotional, and other neuropsychological deficits (6,7,11). These studies have demonstrated that full scale IQ is within the normal range, but lower scores have been noted on language related tasks, some executive functions, attention, memory, auditory and verbal learning tasks, and a variety of behavioral and emotional difficulties (12-15). It is challenging to investigate the complex links between BRE and neuropsychological dysfunction, but the present study focused on evaluating the behavioral and emotional status in newly diagnosed patients with BRE without treatment.

Most publications regarding childhood epilepsy report a clear predominance of cases affecting males, especially between the ages of 5 and 10 (16). This is also valid for BRE cases, which affects boys more than it does girls. In the present study, 75% of the cases were boys. We noticed a greater coincidence between the electrographic and clinical classification of girls, and a higher percentage of discordant cases among boys.

We also observed a higher percentage of "non-benign" cases among boys, both clinically and Electrographically. This predominance for males is probably a consequence of the BRE prevalence in boys as described by Holmes (17).

Despite the fact that, by definition, this type of epilepsy is considered benign, recent atypical evolution has been recognized. In some large studies, the percentage of atypical features (including diurnal seizures, screaming as a seizure component, aura, and Todd's paralysis) ranged from 10 to 50% (18,19). Our experience is in agreement with these authors and suggests that among patients, a small proportion might have learning disabilities and/or behavioral disturbances.

The main reasons to treat children with antiepileptic drugs is to avoid bodily injury from seizures and treatment of psychosocial behavioral problems. Results from several studies have indicated that injury is uncommon with most seizure types in children. Peter et al described 79 children with BRE who together had more than 900 seizures over an 8.5-year period (20). In contrast, in our study, no one had any bodily injuries but most had multiple episodes of seizures, behavioral problems, and daytime seizures.

It would be reasonable to imagine that unilateral rolandic spikes are more prone to be associated with CNS pathologies, whereas bilateral or unilateral spikes that do not remain fixed in one specific side would have a more dysfunctional character (21-22).

In the present study, we examined specifically the behavioral and cognitive functions in children with BECTS. These findings are in accordance with previous studies (23,24).

Children with BRE experienced elevated rates of cognitive, behavioral, and affective problems. Frequent epileptiform spike discharges may impair behavioral functioning (25).

Jones et al. indicated that 53 patients with recent-onset childhood idiopathic epilepsies (45.3%) of the children met the criteria for a DSM-IV Axis I disorder. However, the aforementioned studies analyzed patients with new-onset seizures and new onset idiopathic childhood epilepsies, but were not restricted to patients with BECTS (26).

Austin et al. with the first recognized seizure found higher behavioral problems compared with the controls with 32.1% being in the clinical practice in childhood epilepsies. They found that children with seizures had significantly higher scores in the subscales of attention problems, thought problems, and somatic complaints compared with their nearest-in-age healthy siblings (27). In our study, lower grades in mathematics in the group of the untreated children with BECTS were observed. These findings support the hypothesis of the impact of the active phase of epilepsy on cognition. Bhise et al. found evidence for an intrinsic weakness in attention associated with epilepsy (28). Kavros et al. in a systematic review of attention impairment in BECTS reported that attention systems were impaired in children with active centrotemporal spikes, but these impairments resolved on EEG remission (29).

In our study, 28 patients had unilateral right sided centrotemporal discharges and only four had bilateral discharges, which is consistent with the above-mentioned study. Neuroimaging indicated that only two patients had mild cortical atrophy but developmentally they were normal. However, Beaussart M reported that few patients were refractory to AEDs; however, in our study, all patients responded well to AEDs (30).

Our findings show that behavioral problems in the patients with BECTS and who were in the early course of the disease were not as obvious as in other forms of epilepsy and after treatment, they showed significant improvement.

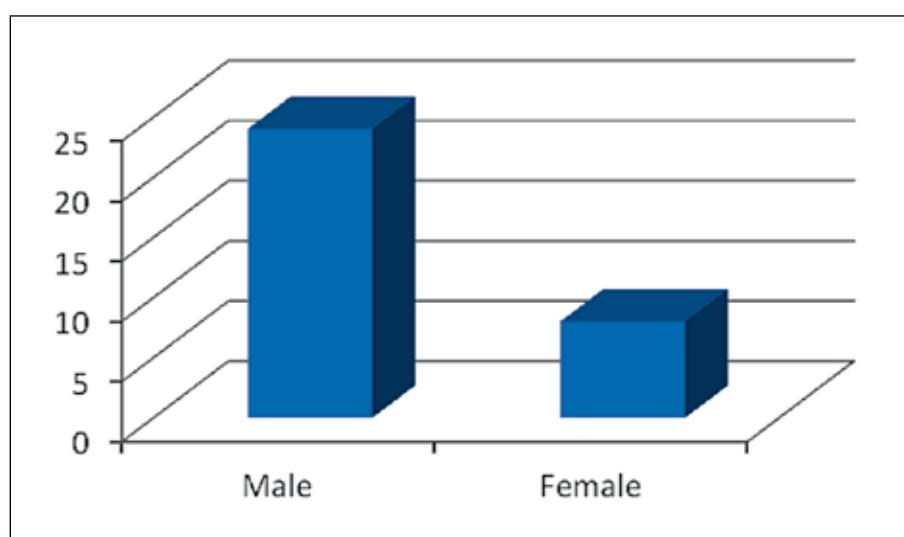


Fig 1. Frequency of Sex in patients.

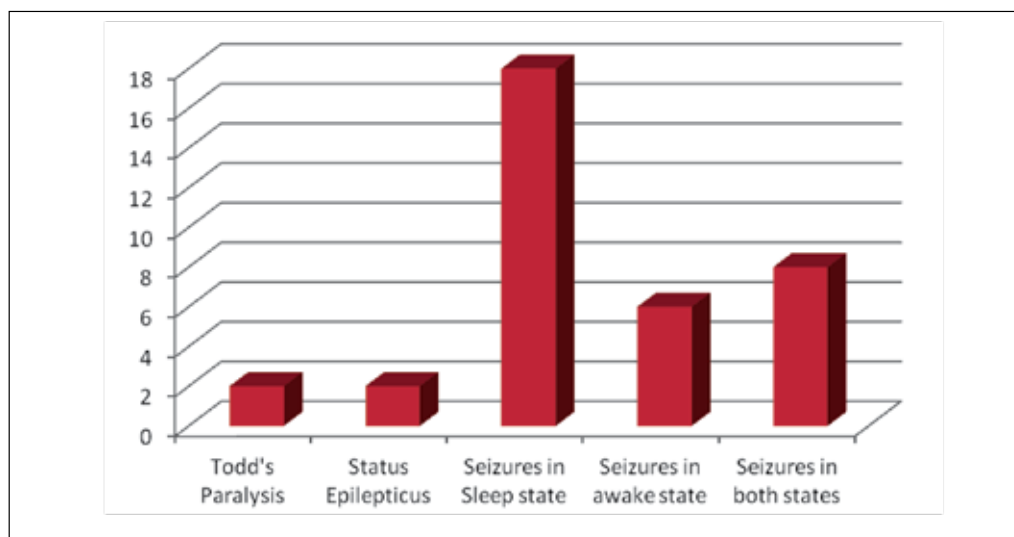


Fig 2. Diagram of Different types of seizures.

This study is limited by its small sample size and the heterogeneity of its subjects. As a result, a large scale, more objective study is required in the future.

Based on our results and the data of other researchers, we can conclude that comorbidity of BECTS and behavioral problems does exist.

Our findings alert clinicians to the possibility of behavioral problems and attention to behavioral problems as important in the clinical management of children with BECTS.

However, future studies are needed to evaluate deeper behavioral problems and associated factors in children with BECTS and to elucidate the influence of the disease and treatment on them.

In conclusion, the data have shown that for children with BECTS, it appears benign at the onset but may there is a risk for cognitive, behavioral, and other psychiatric disorders during the active phase of epilepsy and these problems might even outlast it. Therefore, we recommend all patients to be treated with close follow-ups and regular neuropsychological assessments.

Acknowledgments

We are grateful to all the children and their parents who participated in our study. We also thank the staff of Children's Neurology Department, the Children's Hospital Taif, Affiliated with Taif Medical University, and the EEG team for their contribution to the data collection.

Author's contribution

Dr Azam, Dr Nadeem, and Dr. Qamar, providing me up to date material and data analysis

Dr Saeed: Writing the manuscript

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