Epileptic Disorders

The Educational Journal of the ILAE International League Against Epilepsy

Benign childhood epilepsy with centro-temporal spikes (BCECTS): early onset of seizures is associated with poorer response to initial treatment

Volume 8, numéro 4, December 2006

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Article reçu le 24 Mai 2006, accepté le 26 Août 2006

According to the ILAE classification benign childhood epilepsy with centro-temporal spikes (BCECTS) is a form of idiopathic partial epilepsy characterized by typical, brief, unilateral motor seizures preferentially involving the face and the oropharyngeal musculature. Secondary generalization is observed in almost 20% of the patients. Interictal EEG findings include slow, diphasic, high voltage, centro-temporal spikes that are activated by sleep. The seizures, which usually appear in the first decade of life and disappear during the second, typically occur in otherwise healthy children who show no evidence of cerebral lesions. BCECTS is considered as having a genetic etiology, although to date no genes have been identified. The EEG marker is probably inherited as an autosomal dominant trait with age-related penetrance, although only about 10% of children inheriting this trait actually have recognized seizures (Kramer et al. 2002). Thus, whether or not a child develops BCECTS depends on various other factors, some of which may be inherited.A minority of affected children however, present with atypical features (Aicardi 2000, Hahn 2000, Saint-Martin et al. 2001, Verrotti et al. 2002). Among the atypical features, frequent seizure recurrence (Kramer et al. 2002), refractoriness to medical treatment (Ong HT and Wyllie E, 2000, Deonna et al. 1986, AL-Twajri and Shevell 2002, Blom et al. 1982), cognitive decline (Yung et al. 2000, Welgage et al. 1997, Baglietto et al. 2001, Staden et al. 1998), and episodes of status epilepticus (Fejerman and Di Blasi 1987) have been reported. These features appear to occur in children with an earlier age-at-onset of seizures.Most children have only rare seizures and, when treated, the response to antiepileptic drugs is good, indicating a low epileptogenicity of the focus which remains clinically silent in over 90% of cases (Arzimanoglou et al. 2004, Dalla Bernardina et al. 2005). However, in a poorly identified subset of children with BCECTS, more than one AED is required for an effective control of seizures. Only a few studies are available on factors associated with initial poor response to AEDs (Kramer et al. 2002). We therefore compared two groups of children with BCECTS, one group using one AED and a second group that required more than one AED, in an effort to determine the factors associated with the use of multiple AEDs for seizure control.

METHODS

Patients were selected by reviewing all EEG reports in the Division of Pediatric Neurology, Asan Medical Center (a tertiary care referral epilepsy center), between March 1, 1995, and April 30, 2002.
Patients were included in the study when: 1) they presented with typical EEG features containing either spikes in one or both centro-temporal regions or spikes with typical rolandic morphology; 2) they had more than two attacks of seizures with typical BCECTS semiology; 3) they had been followed up at least every three months and had at least one EEG every year, for a minimum of two years. Other AEDs were added when secondarily generalized seizures were refractory to initial AED treatment. Patients with any simultaneous features suggesting another epilepsy syndrome were excluded. Patients were divided into two groups according to the number of AEDs used for effective seizure control. In Group A we included children that received only one AED, and in Group B patients that have been administered more than one AED (table 1(*Table 1*)).

The Mann-Whitney U test was used to compare the two groups with respect to mean age-at-seizure onset, number of seizures experienced before treatment, and interval between seizure-onset and initiation of AED.

Chi-square analysis was used to compare the two groups with respect to sex; type of AED initially used; unilateral or bilateral EEG abnormalities; occurrence of secondarily generalized seizures; and family history or past history of epilepsy or febrile seizure. Statistical significance was assumed for $p < 0,05$.

Data processing and analysis were performed by SPSS 13.0 version.

Table 1 Dermographic data of 144 patients with BCECTS.

		Group A	Group B
Number		119	25
Male/female		64/55	11/14
Age at seizure-onset (years)		7.6 ± 2.2	5.1 ± 1.9
Duration of follow-up (years)		4.5 ± 3.2	4.8 ± 2.1
Family history of epilepsy or febrile seizure	Epilepsy	2	0
Febrile seizures	6	0	
Past history of seizures or febrile seizures	Neonatal seizures	3	0
Febrile seizures	6	1	
Antiepileptic drugs	Carbamazepine	40	13
Oxcarbazepine	72	8	
Valproate	3	4	
Lamotrigine	2	0	
Topiramate	2	0	

RESULTS

The entire study cohort consisted of 144 patients with BCECTS, 75 males and 69 females, with mean age at disease onset of 7.2 ± 2.3 years (range, 2.1-14.3 years). Of the 144 patients, 119 were taking one AED (Group A), and 25 were taking more than one AED (Group B). There were no significant differences between the two groups with regard to the female-to-male ratio; prescribed AEDs; number of seizures before treatment onset; interval between seizure-onset and start of treatment, the presence of secondarily generalized seizures, the presence of bilateral epileptiform activity or the duration of follow-up. The two groups differed, however, as regards the mean age of onset, which was 7.6 ± 2.2 years in Group A and 5.1 ± 1.9 years in Group B ($p < 0.05$). The two groups also differed in the percentage of patients with seizure-onset below three years old ($p < 0.05$, table 2(*Table 2*)), and the distribution of seizure-onset age (table 3(*Table 3*)).

Table 2 Age-at-onset of seizures in Group A and Group B.

Age at seizure-onset (years)	**Group A**	**Group B**
< 3	1	3
≥ 3	118	22

Table 3 Age distribution of seizure-onset.

Seizure-onset age (years)	**Group A**	**Group B**
2 ≤ 3	1	3
3 ≤ 4	4	2
4 ≤ 5	10	5
5 ≤ 6	14	8
6 ≤ 7	16	4
8 ≤ 9	21	1

Seizure-onset age (years)	Group A	Group B
9 ≤ 10	25	0
10 ≤ 11	8	2
≤ 12	20	0

DISCUSSION

The long-term prognosis for children with BCECTS is usually excellent, in that, when treated, they respond well to AEDs, even those who initially experience frequent and troublesome seizures, with almost all patients achieving long-term remission by mid-adolescence.
In our study, we considered as atypical an evolution that necessitated the use of more than one AED for the control of seizures, a marker of initial medical refractoriness. Of the 144 children studied, 25 (17.4%) were taking more than one AED. There was a significant difference in mean age-at-onset between this group and the group of 119 children taking one AED. There was also a significant difference in the percentage of children in each group with age at seizure-onset below 3 years old, suggesting an association between earlier seizure-onset and medical refractoriness, a finding confirming previous results (Al-Twajri and Shevell 2002, Loiseau et al. 1988). Our findings suggest that, although BCECTS is an age-related epilepsy syndrome, following its own evolution and tending to disappear at a certain age regardless of age at onset, children who experience earlier seizure-onset are likely to be more refractory to initial medication whereas those who experience later seizure-onset are likely to be controlled by initial medication.

These features may be partially explained by the multifactorial pathogenesis of BCECTS (Doose and Baier 1989, and Doose et al. 1996). Thus, the complexity of causal factors may account for a wide spectrum of epileptic and non-epileptic conditions, ranging from mild, selective performance deficits to complex psychomental retardation, and from simple BCECTS to severe epilepsies with minor seizure and bioelectrical status. These conditions are not strictly syndromes, but sets of variably weighted symptoms of a complex pathogenetic background. In addition, these features may be explained by genetically different mechanisms between early-onset BCECTS and late-onset BCECTS.

This being a retrospective study, based on an EEG data selection of the patients, some limitations need to be underscored. Our data on seizure frequency were based on patients' reports, and we cannot exclude that seizures with secondary generalization were more systematically reported as compared to a minor, sensorimotor event. Patients with a very benign evolution may have been lost from follow-up and consequently not included. A control group of patients with BCECTS that did not receive any treatment would be of value. A future prospective study addressing refractoriness of BCECTS, should take into consideration these remarks.

Although BCECTS with early seizure-onset may prove to be refractory to initial medical treatment, the long-term prognosis for BCECTS is excellent, with almost all patients achieving long-term remission by mid-adolescence. However, minor atypical clinical and electrographic features are not uncommon. Thus, in children with early-onset of rolandic seizures, the physician should inform the patient and the family that a relatively refractory course may be observed, which should not necessarily be interpreted as a reason to question the diagnosis of BCECTS and the overall satisfactory evolution.

Request PDF | Benign childhood epilepsy with centrotemporal spikes (BCECTS): early response to treatment with regard to age at seizure onset and long term outcome. | Purpose: TofollowtheclinicalcourseofchildrenwithBCECTStreated with antiepileptic drugs (AEDs), estimate the percentage of early | Find, read and cite all the research you need on ResearchGate. However, there are conflicting data as to whether the CC or TT genotype of the 3435C>T polymorphism is associated with drug resistance. Methods and results: We investigated the association between this [Show full abstract] polymorphism in drug-resistant childhood epilepsy by comparison with drug-responsive patients. Benign childhood epilepsy with centrotemporal spikes (BCECTS) is the most common pediatric focal epilepsy syndrome and typically has positive clinical outcomes. However, a few patients experience recurrent seizures, and therefore, require treatment with antiepileptic drugs (AEDs). This study aimed to identify risk factors associated with poor response to initial AED therapy in BCECTS patients. Methods: We retrospectively reviewed the files of 57 patients who were diagnosed with BCECTS between January 2008 and September 2013. Patients not being treated with AEDs have been excluded. We placed th Juvenile Absence Epilepsy. Benign Epilepsy of Childhood with Centrotemporal Spikes. Benign Partial Epilepsy of Childhood with Occipital Paroxysms. Benign Epilepsy with Affective Symptoms (Benign Psychomotor Epilepsy). Show All. These epilepsies are presented according to the age of onset, starting from the neonatal period. Benign epilepsies in the neonatal period. The benign partial epilepsies include benign partial epilepsy of childhood with centrotemporal spikes, benign occipital epilepsy, and benign epilepsy with affective symptoms. Go to Epilepsy and Seizures for a general overview, and see the following articles for information on these specific disorders