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BENIGN FOCAL EPILEPSY OF CHILDHOOD WITH CENTROTEMPORAL SPIKES (BECTS)

Clinical characteristics of seizures according to age at first seizure

Carmen Silvia M.G. Miziara[1], Maria Luiza G. Manreza[2]

ABSTRACT - BECTS is characterized by the presence of simple partial motor seizures in the face and/or oropharynx, with or without sensory symptoms and often with secondary generalization. These seizures tend to occur more often during sleep or drowsiness. According to some authors, generalized seizures prevail over other types particularly among children aged five or less. The purpose of this study is to determine the characteristics of the first epileptic episode among children with BECTS, grouped by age as of their first epileptic seizure, as well as to analyze how such seizures change over the course of clinical evolution. A total of 61 children were examined, 16 of whom below the age of 5 and 45 above. With regard to the first and recurrent epileptic episodes, our final assessment showed that partial seizures occurred more frequently than generalized tonic-clonic seizures in both groups. Although no conclusive relation could be established between the type of partial seizure (i.e. simple versus complex) and the children's age as of their first epileptic episode, it was possible to correlate the type of epileptic seizure with their clinical evolution, in which case simple partial seizures proved to be more frequent than complex partial seizures. It should be noted that the number of children under the age of five was relatively small, which evinces the need for further studies. It should also be borne in mind that the reported frequency of generalized seizures in these children's first epileptic episode may be due to their parents' lack of attention and familiarity with this pathology and their attendant difficulty in characterizing its clinical symptoms.

KEY WORDS: childhood benign focal epilepsy with centrotemporal spikes, orofacial epileptic seizure, nocturnal epileptic seizure, oropharyngeal epileptic seizure, rolandic epilepsy.

Epilepsia focal benigna da infância com ponta centrotemporal (EBIPCT): características clínicas de acordo com a idade na primeira crise epiléptica

RESUMO - EBIPCT é caracterizada pela presença de crises focais motoras simples envolvendo a musculatura da face e da orofaringe, com ou sem sintomas sensitivos e, muitas vezes com generalização secundária. Estas crises epilépticas tendem a ocorrer durante o sono ou a sonolência. Alguns autores referem o predomínio de crises generalizadas, especialmente nas crianças com menos de 5 anos de idade. O objetivo deste estudo é avaliar a característica da primeira crise epiléptica em crianças com EBIPCT com ponta centrotemporal de acordo com a idade no primeiro episódio epiléptico, assim como estudar as transformações das crises epilépticas durante a evolução clínica. Foram avaliadas 61 crianças, sendo 16 menores de 5 anos de idade e 45 maiores. Em relação à primeira crise epiléptica e a recorrência, a avaliação final mostrou nos dois grupos uma frequência maior de crise parcial. No entanto, não foi possível verificar uma relação entre o tipo da primeira crise parcial (simples ou complexa) de acordo com a idade da criança na época deste evento, mas sim uma variação do tipo de crise epiléptica com a evolução clínica, quando foi observado predomínio de crise parcial simples em relação à crise parcial complexa. Deve-se observar que o número de crianças menores que 5 anos foi pequeno, mostrando a necessidade de estudos mais amplos. Questiona-se, ainda, se a frequência de crises generalizadas no primeiro episódio apresentado pela criança não seria devido a dificuldade de caracterização das manifestações clínicas pelos pais ainda pouco atentos e familiarizados com a patologia.

PALAVRAS-CHAVE: epilepsia focal benigna da infância com ponta centrotemporal, crise epiléptica orofacial, crise epiléptica noturna, crise epiléptica orofaríngea, epilepsia rolândica

Department of Neurology, Clinicas Hospital, University of São Paulo, São Paulo SP, Brazil: [1]MD, Child Neurology Post Graduate Student; [2]MD, PhD, Medical Supervisor for the Child Neurology Service.

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Dra. Carmen Silvia M.G. Miziara - Rua Teodoro Sampaio 352/85 - 05405-000 São Paulo SP – Brasil. E-mail: csmgm@uol.com.br

Benign focal epilepsy of childhood with centrotemporal spikes (BECTS), also known as rolandic epilepsy, is the most common and widely studied form of all benign childhood epilepsies[1]. It accounts for 24% of all epilepsies among children aged five to fourteen[2], and for 16.5% of such disorders among children under fifteen years of age[3]. The initial age of seizures ranges from three to thirteen years, peaking at seven to eight years[4]. Nocturnal seizures occurring among children with no neurological or cognitive disorders after they fall asleep or upon awakening are quite typical[5, 6]. Gender prevalence in this form of epilepsy is 60% males and 40% females[7]. In 1989, BECTS was classified as an age-dependent partial idiopathic epilepsy, namely a genetic focal epilepsy without defined structural wounds, yet with excellent prognosis[8]. The remission of seizures takes place spontaneously during the teen years[9].

BECTS is a relatively well-defined disorder, with typical clinical and electroencephalographic (EEG) features. The symptoms observed during the fits may vary considerably from child to child and, in some cases, from one episode to another[9]. Clinically, it is characterized by simple partial seizures (SPS), including motor, hemifacial and rapid spasms that generally occur during sleep. In addition, such SPS may be accompanied by sensory symptoms. With regard to its EEG features, BECTS is characterized by sudden, high-voltage centrotemporal spikes, often followed by sleep-activated slow waves tending to spread out or shift from side to side. These spikes are unilateral in about 60% of all cases and bilateral in about 40%. They may be mono- or even multifocal[5,10,11]. However, background activity remains normal. The fact that BECTS seizures often involve facial and oropharyngeal muscles suggests that epileptic discharges originate from the inferior rolandic cortex, bilaterally to rolando's fissure and in juxtaposition to Sylvius' fissure[12, 13]. The presence of motor symptoms or upper limb sensitivity in some children during such seizures will depend on the involvement of the higher rolandic area. According to some authors, BECTS features, particularly those of clinical nature, change from one age group to another. Beaussart and Loiseau claim that very young children with BECTS show a higher incidence of generalized hemiepilepsy, which rarely changes sides or progresses into secondary generalized epilepsy[5-7].

The objective of this study was to analyze the clinical characteristics of the first epileptic seizure among children with BECTS below or above the age of five, as well as how such features change over the course of clinical evolution.

METHOD

Throughout 1999 and 2000, the Child Neurology Department of Clinicas Hospital, assisted by the medical faculty of the University of São Paulo (USP), evaluated 61 children who had been diagnosed with BECTS according to the clinical and EEG features of this disorder. The subjects were separated into two groups according to the age at which the symptoms had first occurred. Group 1 included 16 children who had had their first epileptic seizure at age five or under, and Group 2 comprised 45 children whose first epileptic episode had occurred after the age of five.

Data were collected through informal interviews with the subjects' mothers and then checked for accuracy by means of extensive questionnaires designed to ensure that no earlier episodes had been overlooked. Further interviews were carried out with other family members who had witnessed such episodes, in order to verify the data gathered in the previous steps. Finally, the subjects themselves were interviewed to determine their sensitivity to the symptoms reported during the episodes.

The following criteria were presented to clarify our characterization of partial crises:

1. Simple focal seizure with oropharyngeal involvement: consciousness is not impaired and the child responds to external stimuli during the seizure; unilateral clonus often accompanied by commissural twitches or tonic contractions causing the mouth to be lopsided.
2. Simple focal seizures involving the hands: alteration of motor signals in the muscular system or the presence of clonic twitches; hypersensitivity, and particularly a tingling sensation.

Determining the exact location of the discharges helps to classify epileptic seizures and to rule out the possibility of structural wounds. The following procedures were carried out:

1. EEG during sleep and wakefulness monitoring according to the international system 10-20;
2. Computerized tomography and/or magnetic resonance imaging;
3. Single photon emission computed tomography.

Seizures were classified according to the specifications of the International League Against Epilepsy[4] and parametric tests were used for statistical data evaluation.

RESULTS

Our evaluation of the 61 subjects indicated a predominance of seizures among males (Table 1) and during sleep monitoring (Table 2).

The subjects' age as of their first epileptic episode ranged from 2 years and 5 months to 11 years and 4 months, averaging out to approximately 6 years and 5 months. Most subjects experienced their first seizure before the age of 11 (Table 3). Group 1

Table 1. Distrubuition according to gender.

Gender	Nº	%
Masculine	34	55.75
Feminine	27	44.25

Table 2. Frequency of epileptic seizures during monitoring or sleep.

	Nº	%
Sleep	46	75.4
Wakefulness	15	24.6

Table 3. Distribuition of patients according to age at the first epileptic seizure.

Age (years)	Nº	%	% Accumulated
< 3	5	8.20	8.20
3 – < 5	11	18.04	26.23
5 – < 7	18	29.50	55.74
7 – < 9	19	31.15	86.89
9 – < 11	7	11.47	98.36
11 - < 13	1	1.64	100

consisted of 16 children under the age of 5 years, while Group 2 comprised 45 children aged 5 years or older.

Our analysis of the characteristics of the first epileptic phase in all the children showed a predominance of partial seizures (PS). Among these, there was a higher frequency of simple partial seizures (SPS) (Fig 1).

Our evaluation of the characteristics of the first epileptic phases in both Group 1 and Group 2 showed a higher frequency of SPS. In relation to the children's age at the time of their first seizure, no statistically significant differences were found between Groups 1 and 2 ($p>0.5$) (Fig 2).

Six children from Group 1 and 15 from Group 2 showed changes in the characteristics of their epileptic seizures over the course of clinical evolution. The difference between the two groups was not statistically significant ($p>0.5$). It should be pointed out that one child from Group 1 and four children from Group 2 experienced only one seizure. An analysis of the changes in epileptic seizure type during the

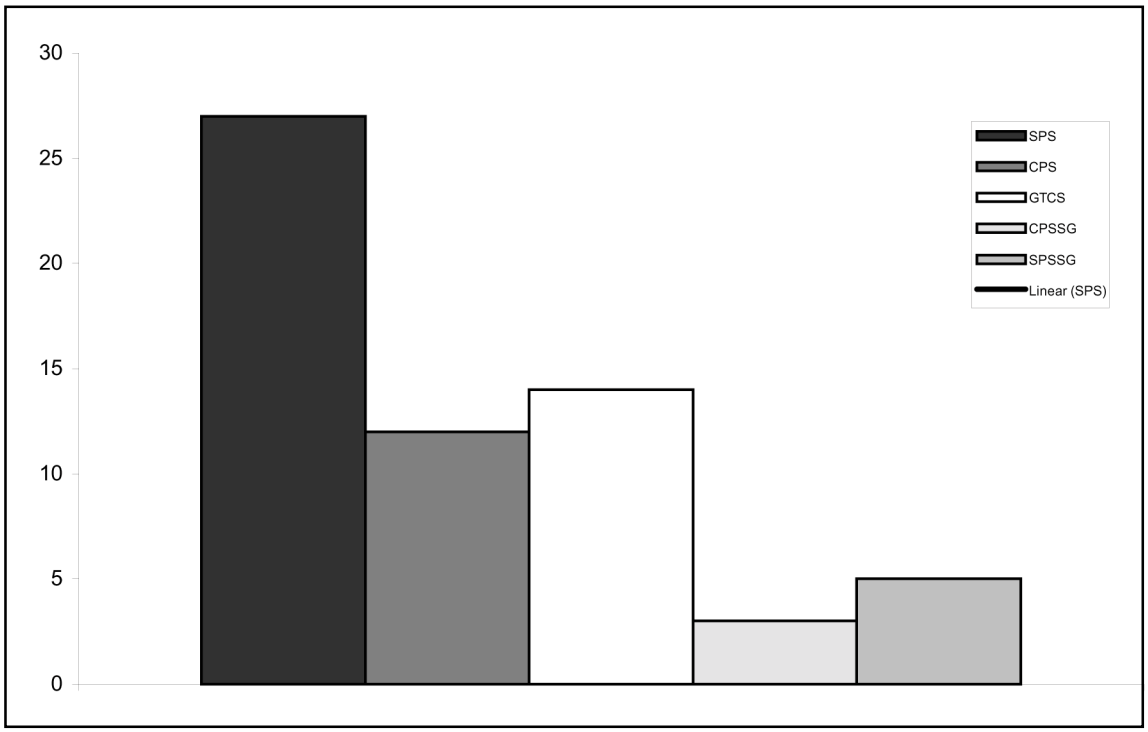

Fig 1. Distribuition of different types of epileptic seizures in total of 61 children.
SPS, simple partial seizure; CPS, complex partial seizure; GTCS, generalized tonic-clonic seizure; CPSSG, complex partial seizure with secondary generalization; SPSSG, simple partial seizure with secondary generalization.

clinical evolution of children in Group 1 revealed that GTCS gave way to CPS, while all the four cases of complex partial seizure (CPS) evolved to SPS (Table 4).

As for Group 2, it was possible to evaluate the changes in epileptic seizures in 15 children during the course of their clinical evolution, for these subjects experienced recurrent episodes. Most of the children with initial GTCS (4 out of 5) evolved to SPS (Table 5).

All patients demonstrated centrotemporal spikes activated by sleep in their EEGs. The focus was on the right side for 26 patients, and on the left side for 25 patients. In the remaining 10 patients, the discharges occurred independently bilaterally.

DISCUSSION

Epileptic seizures in patients with BECTS are usually described in literature as SPS involving facial and oropharyngeal muscular tone, as well as brief and not too frequent. Other types of seizure may occur less frequently. For some authors, however, this standard varies according to the age at which seizures occur, whereby generalized seizures or generalized hemiepilepsies would be more frequently observed among children under 5 years of age.

In this study, PS and GTCS were found to be statistically similar in terms of frequency between chil-

Table 4. Changes in the type of epilepsy seizure during the clinical evolution of children from Group 1 (Total: 6 patients)

Characteristics of 1st seizure	Change in characteristics	Nº
CPS	SPS	4
CPS	GTCS	1
GTCS	CPS	1

SPS, simple partial seizure: CPS, complex partial seizure; GTCS, generalized tonic-clonic seizure.

Table 5. Change in the type of epileptic seizure during the clinical evolution of children from Group 2.

Characteristics of 1st seizure	Change in characteristics	Nº
CPS	SPS	5
GTCS	SPS	4
SPS→GTCS	SPS	2
GTCS	SPS→GTCS	1
SPS→GTCS	GTCS	1
CPS→GTCS	CPS	1
SPS	CPS	1

SPS, simple partial seizure; CPS, complex partial seizure; GTCS, generalized tonic-clonic seizure; SPS→GTCS, simple partial seizure with secondary generalization; CPS→GTCS, complex partial seizure with secondary generalization.

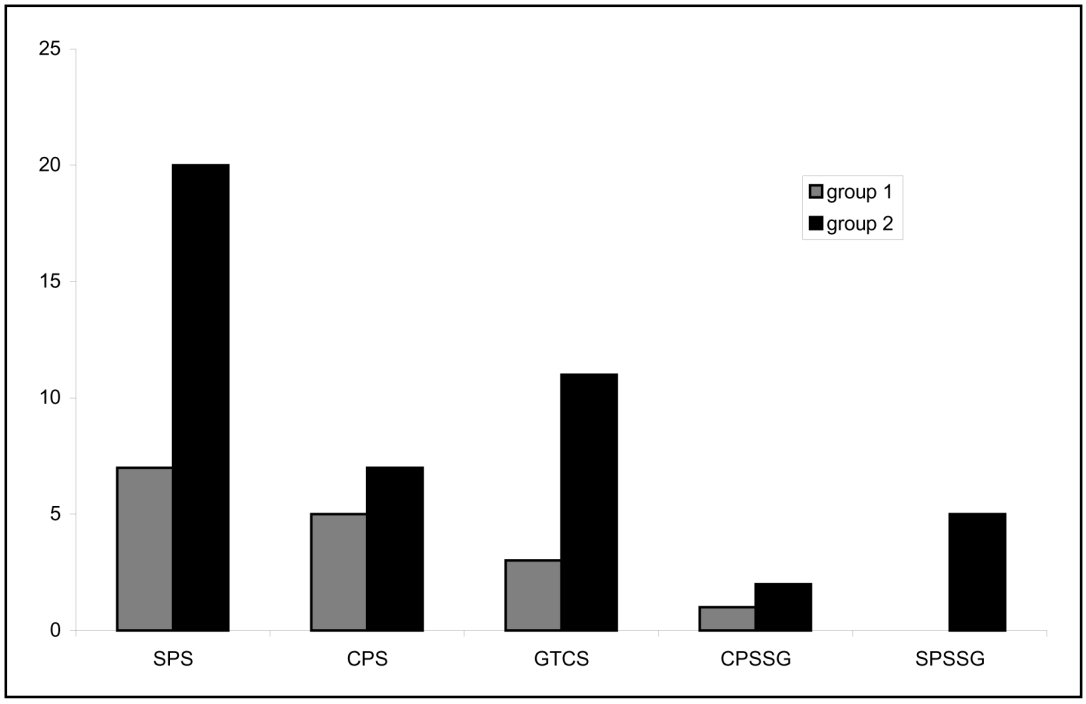

Fig 2. Distribuition of different types of seizures according to the age.
SPS, simple partial seizure; CPS, complex partial seizure; GTCS, generalized tonic-clonic seizure; CPSSG, complex partial seizure with secondary generalization; SPSSG, simple partial seizure with secondary generalization.

dren above and below 5 years of age. It appears that age has no influence on clinical evolution, for one change was noted in standard epileptic seizures in about one third of the children, regardless of the age group analyzed. Our findings indicate a clear predominance of PS in relation to GTCS. It is important to emphasize that the seizures described as GTCS could be PS (simple or complex) with secondary generalization, principally among children under the age of five.

In Group1, one child with GTCS evolved to CPS and four with CPS evolved to SPS, whereas only one with CPS changed to GTCS. In Group 2, four of the five children with GTCS evolved to SPS. A closer study showed that all the twenty children with SPS during the initial phase maintained this standard over the course of their clinical evolution. Our results demonstrate a high incidence of SPS among children with BECTS, regardless of their initial symptoms. Furthermore, the predominance of SPS in both groups during the evolution seems to strengthen the hypothesis that the determining factor behind standard epileptic seizures among children with BECTS is their clinical evolution rather than age.

The conclusion arrived at herein may be questioned because this study involved a relatively small number of subjects below the age of five. Thus, further studies may and were therefore unaware of the initial manifestations of such critical events have to be performed with a larger number of patients. Another point to bear in mind is that most references to GTCS were made in relation to first seizures, when the subjects' parents knew little or nothing about the features of this disorder,

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