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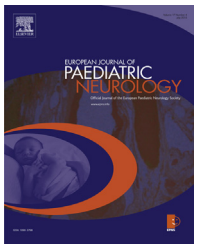

Original article

Early classification of childhood focal idiopathic epilepsies: Is it possible at the first seizure?

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ABSTRACT

Purposes: To evaluate the possibility of early syndrome classification of idiopathic partial epilepsies in children at the first seizure.
Patients and methods: In this observational study we prospectively evaluated 298 patients, aged between 1 month and 17 years and consecutively referred for the first unprovoked focal seizure. The whole cohort included 133 patients; the final analysis was carried out on 107 (59 males) individuals. Age at the first seizure ranged between 2.3 and 13.0 years. Clinical and EEG data of all patients were independently reviewed by two medical doctors. Patients were followed-up for at least 5 years, with a mean period of follow-up of 6.9 years.
Results: After the first seizure, a specific syndrome could be diagnosed in eighty (74.7%) children. In particular, Childhood Epilepsy with Centro-Temporal Spikes (CECTS) 42.9% of cases, Panayiotopoulos Syndrome (PS) 28.9%, idiopathic childhood occipital epilepsy of Gastaut (ICOE-G) 2.8%. Unclassified cases were 25.4%. At the end of the follow-up, the diagnosis was confirmed in 72 of 80 children (90%): BCECTS 89% of patients, PS 90% and ICOE-G 100%: among the unclassified cases, in 11 patients (40.7%) the diagnosis did not change, whereas 16 patients (59.3%) evolved into other syndromes or into atypical forms.
Conclusions: At the onset an initial diagnosis is possible in the majority of cases; epilepsy syndromes can be identified at the time of the initial diagnosis and at follow up this diagnosis has not to be revised in 90% of the cases.

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1. Introduction

There is still no definite answer concerning the diagnostic uncertainty and, consequently, the accuracy of diagnosis of the various epileptic syndromes.[1–4] Most studies focus on incorrect diagnosis and many do not recognize that some diagnostic uncertainty is unavoidable. In some cases, even after complete clinical evaluation, it may be difficult even for an experienced specialist to make a precise diagnosis at the onset. Hospital-based studies in children with epilepsy have highlighted the diagnostic uncertainty at the onset.[2,5] In addition, misdiagnosis is a common problem in epileptic patients with up to 30% of diagnostic mistakes reported in clinical practice.[2,5–7]

The purpose of the study was to assess the 5- year follow-up of a cohort of patients with idiopathic partial epilepsy admitted to a Child Epilepsy Center and to compare the early syndrome classification with the definite diagnosis.

2. Patients and methods

This prospective observational study included a series of consecutive patients referred for a first focal epileptic seizure at 'G. Gaslini' Child Institute (Genoa, Italy) from 1 January 2000 to 31 December 2008. The study was approved by the ethics committee of the hospital and informed consent to participate in the study was obtained from the parents or caregivers.

Among a total of 1512 children, 298 (19.7%) children presented with of the first focal unprovoked seizure (i.e., occurring in absence of any known immediate precipitating event). Inclusion criteria were: a) age between 1 month and 17 years; b) at least one afebrile focal seizure; c) normal psychomotor development and neurological examination. We excluded patients with: 1. a lesional, infective, or metabolic cause; 2. signs and symptoms of epileptic encephalopathy; 3. major neuropsychiatric diseases (e.g., schizophrenia, attention-deficit/hyperactivity, tic disorders delirium, severe mood disorder).

Seizures and epilepsy syndromes were defined according to international recommendations and the following revisions.[8–10]

At inclusion definition of the putative epilepsy syndrome was attempted at the onset of the first seizure based on clinical and EEG criteria. An awake and sleep EEG recording was obtained in all children within 24 h from the seizure. A complete medical assessment (detailed general and neurological history—obtained from the patients, from the parents and at least one reliable eyewitness- and clinical and blood examinations) has been carried out within the first 24 h from the hospital admission.

Patients with apparently generalized seizures, nonspecific or conflicting clinical and EEG features (e.g., orofacial seizures and occipital EEG abnormalities) were considered as unclassified. Children with generalized or multifocal continuous spike waves (SW) activity (SW index = 50–80%) were also considered as unclassified at the onset.

One hundred thirty-three patients were recruited to enter the 5-years follow-up observation. Family history, detailed medical history and possible provoking factors were collected. All children underwent physical and neurological examination, intelligence quotient, awake and sleep EEG, and neuroimaging (CT scan or brain MRI). At least 60′ digital recording including awake and sleep segments was performed using the international 10–20 system electrodes placement. Routine activation procedures included hyperventilation, eye closure, and photic stimulation. At the first evaluation, sleep EEG was performed in all patients. EEG recordings were independently analyzed by two medical doctors board-certified experts in neurophysiology, both blinded for clinical information. Follow-up evaluation visits were carried out at interval of 6 months: at each control clinical evaluation, awake and sleep EEG, 18 h ambulatory EEG (in the cases with severe EEG abnormalities during sleep) were performed.

No therapy after the first seizure was started, except for patients presenting with status epilepticus. Antiepileptic therapy was prescribed in all cases after the second seizure. First choice drug was valproic acid (VPA); in non responders or in cases of side effects, a different antiepileptic drug was introduced. Treatment was stopped within 18–24 months from the last seizure.

A classification of the seizures and a syndrome definition were assessed at the onset of the disease and, at the end of the follow-up, any information that came to light that might

Table 1 – Demographic and clinical parameters of the patients studied.

	n = 107(%)
Gender	
Males	59 (55.1)
Seizures type	
Tonic-clonic	28 (26.1)
Oro-facial	30 (28.2)
Partial-motor	26 (24.3)
Occipital	17 (15.8)
Other partial	6 (5.6)
Presence of vomit	24 (22.4)
Seizure duration < 5 min	53 (49.5)
First seizure during sleep	90 (83.3)
Status epilepticus at the onset	10 (9.3)
Additional seizures after the first episode	85 (79.4)
First diagnosis	
BCECTS	46 (42.9)
PS	31 (28.9)
ICOE-G	3 (2.8)
Unclassified	27 (25.4)

	Mean [SD]	Median [min–max]
Age at first episode of seizure (years)	6.9 [2.6]	6.8 [2.3–13.0]
Age at last episode of seizure (years)	9.6 [3.3]	9.3 [3.5–18]
Age at study visit (years)	14.1 [2.9]	14.0 [8–20]
Time between 1st and 2nd seizure (months)	7.0 [9.4]	3.0 [1.0–38.1]

Figures in round parentheses are percentages calculated over the total number of subjects reported at top of the column; figures in squared parentheses are standard deviations [SD] or minimum and maximum values [min–max]. Status epilepticus is defined as seizure lasting more than 30 min. BCECTS: Benign childhood epilepsy with C T spikes; PS: Panayioutopoulos syndrome; ICOE-G: Idiopathic Childhood Occipital Epilepsy 'Gastaut-type'.