Seizure 2001; **10**: 379–381
doi:10.1053/seiz.2000.0526, available online at http://www.idealibrary.com on IDEAL®

Clinical correlations of electroencephalographic occipital epileptiform paroxysms in children

ŽARKO MARTINOVIĆ

Department for Epilepsy, Clinical Neurophysiology and Cognitive Neurosciences, Institute for Mental Health, Belgrade, Yugoslavia

Correspondence to: Professor Žarko Martinović, Department for Epilepsy, Clinical Neurophysiology and Cognitive Neurosciences, Institute for Mental Health, Palmotićeva 37, 11000 Belgrade, Yugoslavia.
E-mail: martinovic@sezampro.yu

A longitudinal prospective approach was used to investigate clinical correlations of interictal occipital paroxysms with or without fixation-off sensitivity (FOS). Occipital paroxysms were recorded in the electroencephalograms (EEGs) of 76 children with heterogeneous clinical conditions including seizures in 39 patients. Occipital paroxysms with FOS (42 patients) were only fractionally more frequent than non-FOS (34 patients) and were not specific of any clinical condition. Although present and FOS-related in all 11 children with benign childhood epilepsies with occipital paroxysms (CEOP), they were also frequently encountered in symptomatic occipital epilepsy[6]. The differentiation of CEOP from other syndromes established on clinical grounds could also be aided by the analysis of background EEG activity that was frequently significantly more abnormal in symptomatic than CEOP. Clinical characteristics and ictal seizure semiology as well as follow-up clearly distinguish two type of idiopathic CEOP syndromes: (1) early onset type or Panayiotopoulos syndrome characterized by excellent prognosis and rare, prolonged nocturnal seizures with tonic deviations of the eyes and vomiting, and (2) late onset or Gastaut type showing a common ictal visual symptomatology, co-occurrence of migraine, diurnal complex partial seizures and less favourable EEG-clinical prognosis.

Key words: idiopathic childhood epilepsies with occipital paroxysms; fixation-off sensitivity; Panayiotopoulos syndrome.

INTRODUCTION

Over the last 20 years, there has been continuing significant interest and debate on electroencephalographic (EEG) occipital paroxysms and their clinical correlations[1–5]. This was generated particularly by the Commission's recognition of 'childhood epilepsy with occipital paroxysms' (CEOP)[1] that is now accepted to encompass two distinctive clinical syndromes:[3] Panayiotopoulos type (early onset CEOP) that is common and benign[2,4–6] and Gastaut type (late onset CEOP) that is rare and of uncertain prognosis[2,4].

Occipital paroxysms are interictal EEG spike and slow-wave repetitive discharges that often activate by elimination of central vision and fixation (fixation-off sensitivity (FOS))[2–5]. A recent report[3] has confirmed the heterogeneity of clinical associates with occipital spikes but did not examine FOS and it is unclear whether the two clinical syndromes of CEOP were investigated[3].

To address these issues, we utilized a longitudinal prospective approach and investigated clinical correlations of occipital paroxysms with or without FOS.

PATIENTS AND METHODS

During a 9-year period, all patients aged 1–15 years who showed occipital paroxysms in their EEG recording were prospectively studied. A detailed EEG analysis of all patients, also performed during a longer than 5-year follow-up period, included hyperventilation, photic stimulation and testing of visual reactivity with exclusion of central vision[2]. Two types of occipital paroxysms were defined: (1) FOS activated by exclusion of central vision and inhibited by eye opening and (2) visually unrelated (non-FOS) that were not reactive. Children with additional EEG abnormalities, extraoccipital focal or generalized, were included.

1059–1311/01/050379 + 03 $35.00/0

Table 1: Main features of 76 subjects with occipital paroxysms.

Demographic and clinical data	Type of occipital paroxysms		*P* values
	Fixation-off sensitive (FOS)	Visually unrelated (non-FOS)	
Age at occurrence (years)			
Range	3–8	1–11	
Mean ± standard deviation	4.4 ± 2.1	3.7 ± 5.6	<0.100[c]
Girls/boys[a]	21/21	19/15	<0.900[d]
Referral diagnoses[a]			
Epilepsy (total 39)	25	14	
All other diagnoses (total 37)[b]	17	20	<0.250[d]
Epilepsy types[a]			
Benign occipital epilepsy	11	0	0.0008[e]
Early onset	7	0	
Late onset	4	0	
Benign Rolandic epilepsy	3	2	0.3635[e]
Childhood absence epilepsy	2	1	0.9191[e]
Symptomatic partial epilepsy	6	9	0.6156[e]
Symptomatic generalized epilepsy	3	2	0.3635[e]

[a] Number of subjects; [b] ophthalmological disorders in 12, mild mental retardation in eight, migraine in five, behaviour disorder in five patients, four children with febrile convulsions and three of their siblings; [c] *t* test for two small samples; [d] chi-square-test; [e] Fischer's exact test.

All patients had appropriate neurological, psychiatric and other clinical evaluations. Brain neuroimaging was performed when indicated. The epileptic syndromes were determined according to the criteria of the International classification[1, 3].

The EEG characteristics and clinical semiology of patients with various epileptic syndromes were compared and their significance for the differential diagnosis was tested by nonparametric statistical methods.

RESULTS

Occipital paroxysms were recorded in the EEGs of 76 children with heterogeneous clinical conditions (Table 1). They were seen equally in children with (39 patients) and without (37) seizures. Occipital paroxysms with FOS (42) were only fractionally more frequent than non-FOS (34) and were not specific of any clinical condition. However, in all 11 children with CEOP these were FOS-related though they were also frequently encountered in symptomatic occipital epilepsy[6]. Thus, occipital paroxysms *per se* could not differentiate CEOP from other syndromes. It was only the background activity that was frequently significantly more abnormal in symptomatic than CEOP (Fisher exact test, $P = 0.0095$).

On follow-up, EEG normalization was the rule in Panayiotopoulos syndrome, Rolandic epilepsy and childhood absence epilepsy. Though comparatively delayed, EEG normalization in Gastaut-type CEOP occurred more frequently than in symptomatic partial epilepsy. Three of 25 patients with FOS occipital paroxysms at the age 3–5 years later developed Rolandic spikes.

Clinically, of seven patients with Panayiotopoulos syndrome, seizure semiology including the nocturnal appearance of tonic deviation of the eyes and vomiting was characteristic. However, in two patients lacking the history of these ictal signs, the clinical distinction from other forms of epilepsy was not initially attainable. Clinical follow-up of all seven patients was characterized by rare, nocturnal seizures (with a median value of only four) beginning at pre-school age and excellent prognosis. Conversely, Gastaut-type CEOP was less frequent (four patients), and displayed late onset at ages of 8 years or greater, and visual seizures were frequent, sometimes daily, with postictal migraine headache. The EEG background was slow and prognosis was much less favourable than in Panayiotopoulos type or other syndromes of idiopathic childhood epilepsy.

DISCUSSION

This study demonstrates that similarly to occipital spikes[2, 3], occipital paroxysms with or without FOS, are an age-related EEG abnormality that may occur in heterogeneous seizure or non-seizure disorders. Also, this is the first study to document that occipital paroxysms with FOS, though more frequently associated with CEOP, also occur in symptomatic and other epilepsies. Therefore, their significance is only as a confirmatory EEG abnormality of clinical manifestations that are of more importance[2–6].

Regarding CEOP, this study also confirms[2,4,5] that the Panayiotopoulos type is more frequent and benign than that of the Gastaut type. As both types show occipital paroxysms with FOS, their differentiation should be based on seizure semiology,[2,3,5,6] associated post-ictal migraine headaches (occurring frequently and only in Gastaut type)[2] and the normal background EEG activity (always present only in Panayiotopoulos syndrome). Panayiotopoulos type, a homogenous and benign epilepsy syndrome, should be recognized by seizure semiology and occipital paroxysms with FOS. The deviation of the eyes is an occipital symptom occurring in only 70–80% of cases[2,4,5], while other symptoms such as autonomic manifestations with ictal vomiting, behavioural disturbances and convulsions are extra-occipital[2,4,5]. If the latter occur in isolation, or when ictal semiology is not characteristic from the history, the diagnosis of Panayiotopoulos syndrome should be suggested by recording occipital paroxysms with FOS on normal background activity. Occipital paroxysms with FOS in Panayiotopoulos syndrome usually outlast the date of last seizure for some years. In this study, three of 25 patients with FOS occipital paroxysms later developed Rolandic spikes which is less than the 30.3% value reported in a recent series of 66 patients[6]. This type of evolution does not occur in Gastaut type. Thus, in children showing occipital paroxysms with FOS, a prospective analysis of clinical and EEG findings during follow-up are the best method to determine accurate diagnosis and prognosis of the epilepsy syndrome.

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