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Late-onset, "Gastaut Type", childhood occipital epilepsy: an unusual evolution

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ABSTRACT – We report on two girls and one boy with clinical and electroencephalographic features of late-onset childhood epilepsy with occipital paroxysms of the "Gastaut type", showing an unusual evolution. Neurological examination and brain imaging were normal in all three. At the age of 7.5 years, eight years and ten years respectively, the three children presented with episodes of visual symptoms when awake, and in one of them, the seizures were occasionally followed by oculocephalic deviation. The interictal EEG showed bilateral occipital spike-wave activated by eye closing. In two patients, the occipital seizures had been immediately followed by typical absences, since onset; in the other patient, five months after onset. The ictal EEG showed irregular bilateral occipital spike-wave discharges during the visual symptoms, followed by generalized spike-wave activity during the typical absences. The typical absences were activated by hyperventilation; the EEG did not show continuous spikes and waves during slow sleep. These three patients, with typical electroclinical features of "Gastaut type", childhood occipital epilepsy, demonstrated an evolution which, to our knowledge, has not been previously described. We investigated whether this unusual, age-dependent evolution was due to secondary bilateral synchrony or if these electroclinical features represent two types of idiopathic epileptic syndromes in the same patients.

Key words: idiopathic, occipital seizures, secondary bilateral synchrony, typical absences, Gastaut type occiptal epilepsy

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The latest proposed diagnostic scheme for people with epilepsy, the Classification of Epilepsies and Epileptic Syndromes of the International League Against Epilepsy (ILAE 2001), includes among the idiopathic focal epilepsies in childhood: benign childhood epilepsy with centro-temporal spikes (BCECTS), early-onset benign childhood occipital epilepsy ("Panayiotopoulos type") and late-onset childhood occipital epilepsy ("Gastaut type") (Engel 2001).

Childhood occipital epilepsy as described by Gastaut (1982) is rare, of uncertain boundaries and often of unpredictable prognosis. This "Gastaut type" of childhood occipital epilepsy is characterized by brief seizures with mainly visual symptoms such as elementary visual hallucinations, illusions or amaurosis, followed by hemiclonic convulsions. Post-ictal migraine headache occurs in half of the patients, and age at onset is around 8-9 years. The EEGs show occipital spike-wave paroxysms that attenuate when the eyes are opened (Panayiotopoulos 1999).

Atypical evolutions of BCECTS have been repeatedly reported (Fejerman *et al*. 2000). Atypical evolutions of child-

hood occipital epilepsy have also been recognized (Tenembaum *et al*. 1997, Caraballo *et al*. 2003, Eriksson *et al*. 2003). In an earlier paper (Caraballo *et al*. 2001), we reported two patients who initially presented with typical clinical and EEG features of childhood occipital epilepsy Panayiotopoulos type" that evolved atypically. In "Gastaut type" childhood occipital epilepsy, children may exceptionally develop status epilepticus and continuous spike-waves during slow sleep (CSWS).
In the present study, we report three cases with "Gastaut type" childhood occipital epilepsy, who presented occipital seizures followed by typical absences.

Case report 1

The patient is a 19-year-old girl, whose father had a history of febrile seizures. Pregnancy and delivery were normal, as was the child's early development. There is no family history of migraine.
At the age of 7.5 years, she started having frequent, often daily episodes of elementary, visual hallucinations. These consisted of brilliant colored spots in front of her right visual field. Their duration was from seconds to as long as one minute. Occasionally, visual seizures were followed by right oculocephalic deviation and headaches. The interictal EEG showed bilateral high-amplitude occipital spikes, predominantly on the left, and mainly when the eyes were closed. Neurological examination and routine laboratory investigations were normal. Brain CT scan and MRI were normal. Carbamazepine was prescribed at 15 mg/kg/day.
When she was eight years old, the occipital seizures were followed by typical absences with rhythmic, clonic, eyelid twitching. Carbamazepine was discontinued and sodium valproate was administered, with good seizure control. The interictal EEG did not change. Occipital paroxysms were activated during sleep. CSWS were not found. The response to IPS was normal and closing of the eyes did not activate absences. The EEG during hyperventilation showed generalized spike-wave activity at 3-2.5 Hz during typical absences, predominantly in anterior regions. Her ictal EEG showed irregular, bilateral occipital spike-wave discharges during simple visual hallucinations followed by generalized spike-wave activity during typical absences (*figure 1*). At the onset of the seizure, the patient had her eyes open.
All seizures stopped and she remained free of seizures. The EEG normalized at age 12 and treatment was withdrawn at the age of 13 years. Since the age of 13, she has experienced frequent episodes of migraine.
The girl is now 19 years old and presents a normal neurological examination and a normal EEG.

Case report 2

The patient is a 9-year-old girl, with healthy, nonconsanguineous parents. Personal history was unremarkable. Her older brother and mother had episodes of migraine.
At the age of eight years, she started having visual hallucinations characterized by bright, multicolored, circular lights followed by typical absences with rhythmic clonic eyelid seizures while awake. These episodes occurred

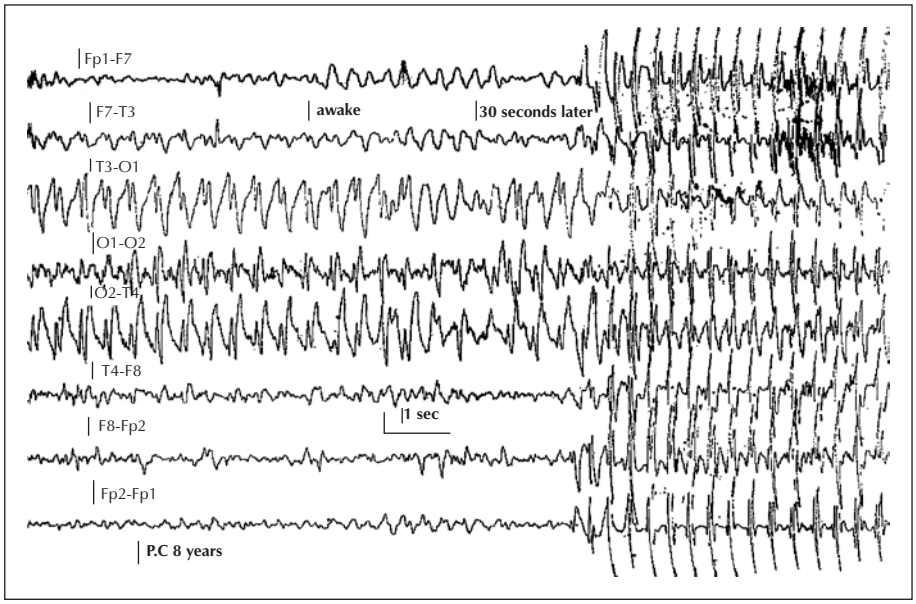

Figure 1. The ictal EEG shows irregular bilateral occipital spike-wave discharges during simple visual hallucinations immediately followed by generalized spike-wave activity during typical absences.

daily, lasted no more than one minute, and were not accompanied by headache or vomiting. The interictal EEG showed bilateral occipital spikes activated by eye closing and generalized polyspike-waves, predominantly in the right hemisphere. Occipital paroxysms were activated during sleep. CSWS was not found. The response to IPS was normal and closing of the eyes did not activate absences. The EEG during hyperventilation showed generalized polyspike-waves and spike-wave discharges at 3 Hz during typical absences. The ictal EEG showed bilateral occipital irregular spike-waves during visual hallucinations followed by generalized polyspikes and spike-wave activity during typical absences (*figure 2*). The patient had her eyes open at the onset of the seizure. Physical and fundus examinations were normal, no focal neurological signs were found and routine laboratory investigations, as well as brain CT scan and MRI were normal. Sodium valproate was prescribed, at a dose of 40 mg/kg/day.

At her last examination, when the girl was nine years old, she presented sporadic visual seizures. Her interictal EEG showed only bilateral occipital spikes.

Case report 3

The patient is a 10-year-old boy, whose brother had a history of febrile seizures. Pregnancy and delivery, as well as the child's early development were normal. One simple febrile seizure occurred when the boy was three years old. His father experienced episodes of migraine.

At the age of 10 years, he started having seizures that were characterized by brilliant, multicolored spots and circles lasting for a few seconds. There was no loss of consciousness. On two occasions, his vision went black. Postictally, he was pale and had headache. Three months later, when he visited our service, he was still experiencing the same type of seizures that were now followed by typical absences with automatisms and atonic components with autonomic signs. His interictal EEG showed bilateral occipital spike-waves activated by eye closing, occasionally propagated to the anterior region. Occipital paroxysms were activated during sleep. CSWS was not found. The response to IPS was normal and closing of the eyes did not activate absences. The EEG during hyperventilation showed generalized spike-waves activity at 3-2.5 Hz during typical absences. The ictal EEG showed rhythmic occipital spike-wave during visual seizures followed by generalized spike-wave activity during absences (*figure 3*). The patient had his eyes open at the onset of the seizure. Treatment with sodium valproate at 35 mg/kg/day was started. Physical and neurological examinations, as well as brain CT scan and MRI were normal.

At his last examination, at the age of 10 years, he was seizure-free but the EEG showed frequent bilateral occipital spikes activated by eye closing.

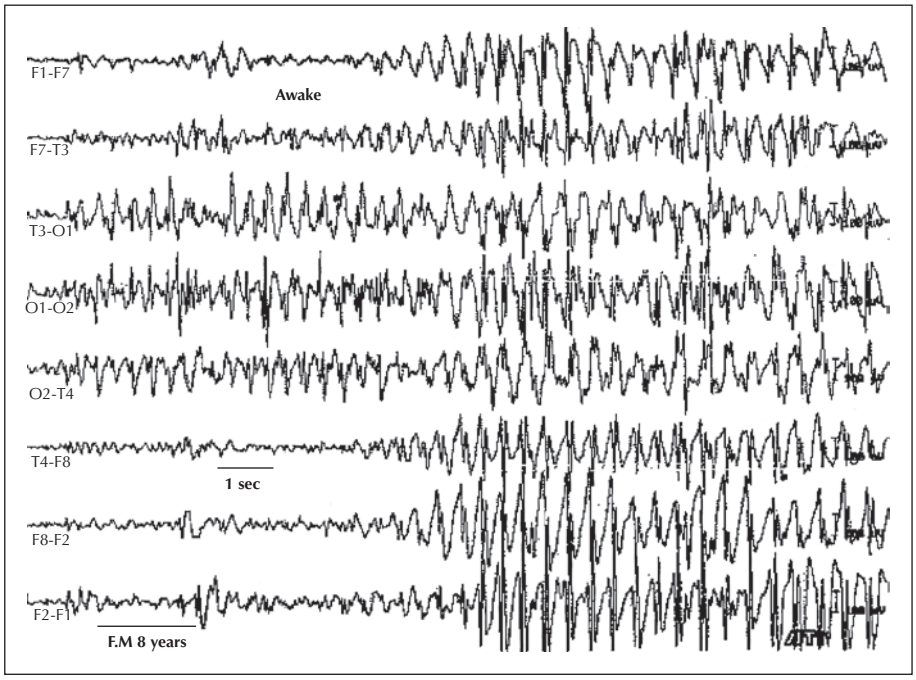

Figure 2. The ictal EEG shows bilateral occipital spike-wave paroxysms during brief visual symptoms immediately followed by generalized spike-wave activity during typical absences.

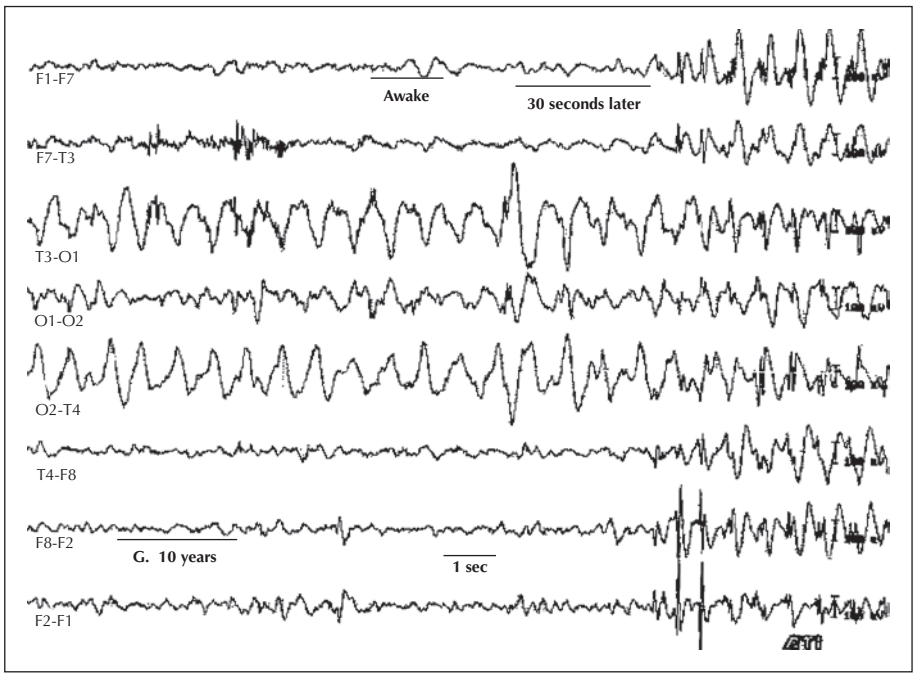

Figure 3. The ictal EEG shows irregular bilateral occipital spike-wave discharges during simple visual hallucinations followed by generalized spike-wave paroxysms during typical absences.

Discussion

All three patients presented electroclinical features compatible with "Gastaut type" childhood occipital epilepsy, with seizures starting mainly with visual symptoms (occipital seizures), immediately followed by typical absences. One of the patients has been reported by us earlier (Caraballo *et al.* 2004). Remarkably, these three patients had their eyes open when the seizures started and none of them presented CSWS.

Two patients presenting dementia associated with continuous spike-waves on the EEG in the course of "Gastaut type" childhood occipital epilepsy have been reported, and one of them had remaining neuropsychological impairment (Tenembaum *et al.* 1997).

In a previous report (Caraballo *et al.* 2004), we described idiopathic occipital and absence epilepsies appearing in the same child. Three of the children presented both syndromes at the same time while the other three had typical absences after the seizures and their EEGs were compatible with the diagnosis of childhood occipital epilepsy. Five of them presented the "Gastaut type" childhood occipital epilepsy and the other one "Panayiotopoulos type" childhood occipital epilepsy (Caraballo *et al.* 2004).

This association has been previously reported for patients with rolandic seizures and centrotemporal spikes by Beaumanoir *et al.* (1974), who found a high incidence of absences amongst 26 children with rolandic seizures. These authors had exhaustively investigated their patients with all types of EEG during wakefulness and sleep. Six patients had clinical and EEG evidence of typical absence seizures. In the same report, 11 out of 24 patients, adults and children with typical absences had unilateral or bilateral centrotemporal spikes.

According to Gastaut and Zifkin (1987), in 19 of 50 children (38%), occipital paroxysms were associated with generalized, bilaterally synchronous spikes and waves or polyspikes and waves, characteristic of idiopathic generalized epilepsy or with central or mid-temporal spikes characteristic of other types of idiopathic focal epilepsy. Beaumanoir *et al.* (1983) found asymptomatic generalized discharges of polyspike or spike and slow waves in three out of 11 patients with "Gastaut type" childhood occipital epilepsy that underwent 24 hours EEG monitoring. One patient also had absence seizures. A combination of typical absence seizures with occipital paroxysms, but no clinical evidence of "Gastaut type" childhood occipital epilepsy is considered by Panayiotopoulos (1999), to be suggestive of a poor prognosis.

However, in the present report we describe children with "Gastaut type" childhood occipital epilepsy who manifested occipital seizures followed by typical absences; we believe this may be due to secondary bilateral synchrony. A thalamocortical mechanism may be involved (Tenenbaum *et al.* 1991). In order to determine whether absences and generalized spikes and waves are secondary bilateral synchrony, we should estimate small, interhemispheric

time differences during the seizures using coherence and phase analysis (Kobayashi *et al*. 2000).

Ictal single photon emission computed tomography (SPECT) in absence seizures may reveal the neuronal mechanisms involved, indicating a different origin (Iannetti *et al*. 2001). The focal seizures and absences may be combined so that when the basal ganglia are activated, focal or generalized seizures may arise, and when the thalami are activated, absence seizures may be triggered (Iannetti *et al*. 2001).

The occurrence of typical generalized 3 Hz spike-and-wave discharges has led to speculation about a neurobiological and genetic continuum between childhood absence seizures and benign focal epilepsies (Gelisse *et al*. 1999). To date, it is still not clear if childhood absences epilepsy and idiopathic focal epilepsies have a different genetic marker (Gelisse *et al*. 1999). An autosomal dominant pattern for the EEG abnormalities, with age-dependent expression and variable penetrance of the seizure disorder has been proposed (Kuzniecky and Rosenblatt 1987).

The interictal EEG in patients with COE shows repetitive occipital paroxysms that are reactive to eye opening. In routine EEGs in an illuminated room, occipital spikes are mainly or exclusively observed when the eyes are closed and are inhibited when the eyes are opened. If occipital spikes or paroxysms persist with the eyes open, these can be eliminated by asking the child to fixate on a visual target (the tip of a pencil will do). In our patients the response to IPS was normal.

In 1998, Panayiotopoulos reported that the effect of eye closing on occipital spikes is due to the elimination of central vision and fixation, a phenomenon for which he coined the term fixation-off sensitivity (FOS). Activation and inhibition of the occipital paroxysms usually occur immediately after or within a second of closing or opening of the eyes. The underlying mechanisms of FOS are not known.

Most reports on ictal EEGs of occipital seizures show occipital paroxysms followed by ictal fast spikes/rhythms (Panayiotopoulos 1999). In all three patients, the ictal EEG showed bilateral rhythmic occipital spike-waves during visual hallucinations. These ictal EEG manifestations were difficult to differentiate from interictal occipital paroxysms.

The electroclinical picture of an atypical evolution in children with "Gastaut type" COE is different from the one presented here. The present unusual evolution may be due to a secondary bilateral synchrony mechanism. Secondary bilateral synchrony implies an initial diffusion of discharges through the corpus callosum, with or without intervention of centro-encephalic structures in their generalization (Kobasashi *et al.* 1994).

Conclusion

To our knowledge, no other case of "Gastaut type", childhood occipital epilepsy with this unusual evolution has been reported in the literature. It needs to be determined if this age-dependent evolution is due to a secondary bilateral synchrony, or if these electroclinical features represent two types of idiopathic epileptic syndrome in the same patients. □

References

Beaumanoir A, Ballis T, Varfis G, *et al.* Benign epilepsy of childhood with Rolandic spikes: a clinical and electroencephalographic study. *Epilepsia* 1974; 15: 301-15.

Beaumanoir A. Infantile epilepsy with occipital focus and good prognosis. *Eur Neurol* 1983; 22: 43-52.

Caraballo RH, Astorino F, Cersósimo RO, *et al.* Atypical evolution in childhood epilepsy with occipital paroxysms (Panayiotopoulos type). *Epileptic Disord* 2001; 3: 157-62.

Caraballo R, Sologuestua A, Grañana N, *et al.* Idiopathic occipital and absence epilepsies appearing in the same children. *Pediatr Neurol* 2004; 30: 24-8.

Engel J. A proposed diagnostic scheme for people with epileptic seizures and with epilepsy: report of the ILAE Task Force on Classification and Terminology. *Epilepsia* 2001; 42: 796-803.

Eriksson K, Kylliainen A, Hirkonen K, *et al.* Visual agnosia in a child with non-lesional occipito-temporal CSWS. *Brain Dev* 2003; 25: 262-7.

Fejerman N, Caraballo R, Tenembaum S. Atypical evolutions of benign localization-related epilepsies in children: are they predictable? *Epilepsia* 2000; 41: 380-90.

Gastaut H. A new type of epilepsy: Benign partial epilepsy of childhood with occipital spikes-waves. *Clin Electroencephalogr* 1982; 13: 13-22.

Gastaut H, Zifkin BG. Benign epilepsy of childhood with occipital spike and wave complexes. In: Andermann F, Lugaresi E, eds. *Migraine and epilepsy*. Boston, MA: Butterworths, 1987: 47-81.

Gelisse P, Genton P, Bureau M, *et al.* Are there generalised spike waves and typical absences in benign rolandic epilepsy? *Brain Dev* 1999; 21: 390-6.

Iannetti P, Spalice A, De Luca F, *et al.* Ictal single photon emission computed tomography in absence seizures: apparent implication of different neuronal mechanisms. *J Child Neurol* 2001; 16: 339-44.

Kobayashi K, Nishibayashi N, Ohtsuka Y, *et al.* Epilepsy with electrical status epilepticus during slow sleep and secondary bilateral synchrony. *Epilepsia* 1994; 35: 1097-103.

Kobayashi K, Maniwa S, Ogino T, *et al.* Myoclonic seizures combined with partial seizures and probable pathophysiology of secondary bilateral synchrony. *Clin Neurophysiol* 2000; 111: 1813-6.

Kuzniecky R, Rosenblatt B. Benign occipital epilepsy: a family study. *Epilepsia* 1987; 28: 346-50.

Panayiotopoulos CP. *Benign childhood partial seizures and related epileptic syndromes*. London: John Libbey & Company Ltd, 1999.

Tenembaum S, Deonna TW, Fejerman N, *et al.* Continuous spike-waves and dementia in childhood epilepsy with occipital paroxysms. *J Epilepsy* 1991; 10: 139-45.