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Epilepsy classification and additional definitions in occipital lobe epilepsy

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ABSTRACT – *Aim*. To evaluate epileptic children with occipital lobe epilepsy (OLE) in the light of the characteristics of Panayiotopoulos syndrome and late-onset occipital lobe epilepsy of Gastaut (OLE-G).
Methods. Patients were categorized into six groups: primary OLE with autonomic symptoms (Panayiotopoulos syndrome), primary OLE with visual symptoms (OLE-G), secondary OLE with autonomic symptoms (P-type sOLE), secondary OLE with visual symptoms (G-type sOLE), and non-categorized primary OLE and non-categorized secondary OLE according to characteristic ictal symptoms of both Panayiotopoulos syndrome and OLE-G, as well as aetiology (primary or secondary). Patients were compared with regards to seizure symptoms, aetiology, cranial imaging, EEG, treatment and outcome.
Results. Of 108 patients with OLE (6.4±3.9 years of age), 60 patients constituted primary groups (32 with Panayiotopoulos syndrome, 11 with OLE-G, and 17 with non-categorized primary OLE); the other 48 patients constituted secondary groups (eight with P-type sOLE, three with G-type sOLE, and 37 with non-categorized sOLE). Epileptiform activity was restricted to the occipital area in half of the patients. Generalized epileptiform activity was observed in three patients, including a patient with Panayiotopoulos syndrome (PS). Only one patient had refractory epilepsy in the primary groups while such patients made up 29% in the secondary groups.
Conclusion. In OLE, typical autonomic or visual ictal symptoms of Panayiotopoulos syndrome and OLE-G do not necessarily indicate primary (*i.e.* genetic or idiopathic) aetiology. Moreover, primary OLE may not present with these symptoms. Since there are many patients with OLE who do not exhibit the characteristics of Panayiotopoulos syndrome or OLE-G, additional definitions and terminology appear to be necessary to differentiate between such patients in both clinical practice and studies.

Key words: epilepsy classification, epilepsy syndrome, occipital epilepsy, children, occipital lobe epilepsy of Gastaut, Panayiotopoulos syndrome

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Occipital lobe epilepsy (OLE) accounts for 5-10% of all epilepsies (Adcock and Panayiotopoulos, 2012). It is aetiologically genetic or secondary to structural/metabolic or unknown causes. Idiopathic OLE comprises 20-25% of benign partial epilepsies of childhood (Panayiotopoulos *et al.,* 2008; Adcock and Panayiotopoulos, 2012). Classification of epilepsy has a key role in the evaluation of patients with epilepsy. In both the 1989 classification of epilepsy and the electroclinical syndromes proposed by the ILAE in 2010, there are two epilepsy syndromes which are related to the occipital lobe: Panayiotopoulos syndrome (PS) and late-onset childhood occipital epilepsy (Gastaut type) (Panayiotopoulos *et al.,* 2008; Berg *et al.,* 2010; Adcock and Panayiotopoulos, 2012). Since these were described in the 1980s by Gastaut and Panayiotopoulos, studies have shown that these conditions are separate disorders despite the fact that both predominantly originate from the occipital lobe (Covanis *et al.,* 2005; Panayiotopoulos, 2010). PS generally starts at the age of 1-14 years and is characterized by autonomic seizures, such as ictal emesis (nausea, retching, or vomiting), cardiac or breathing changes, sweating and pallor. The seizures beginning with autonomic manifestations are usually followed by tonic eye deviation and impairment of consciousness and may terminate with hemiclonic or generalized convulsions. Two thirds of seizures start during sleep. Syncope-like manifestations occur in at least a fifth of seizures (Panayiotopoulos, 2010; Adcock and Panayiotopoulos, 2012) and autonomic status epilepticus may also even occur (Ferrie *et al.,* 2006). Interictal EEG is characterized by shifting and multiple foci with occipital predominance. However, it can occur without occipital spikes in a third of the patients, and the epileptogenic focus may also be extraoccipital. As for the Gastaut-type occipital lobe epilepsy (OLE-G), onset age is later, at 3-16 years old. Seizure usually begins with an elementary visual hallucination or sometimes ictal blindness (Caraballo *et al.,* 2008). Visual seizures with eyelid blinking may evolve into hemiconvulsions and secondary generalized tonic-clonic seizures (GTCS). Postictal headache is frequent.

As the classification of epilepsy plays a central role in both understanding epilepsies and managing patients, it has been increasingly used in clinical practice as well as researches. Besides this, as new data and experience accumulate, it is inevitable that classifications of epilepsy require revision and new insight with time (Udani and Desai, 2014). When descriptions and terms being used for electroclinical epilepsy classification are inadequate, new definitions and terminology are necessary to differentiate such patients.

In this study, we evaluated clinical and laboratory findings of our patients with OLE. Since we noticed during this study that many of the OLE patients did not completely match the characteristics of PS and OLE-G, additional definitions were created and patients were then categorized according to these new definitions as well as PS and OLE-G. Based on this study on the clinical implementation of epilepsy syndrome classification in a large cohort of patients with OLE, we aimed to present the challenges in classifying patients with OLE and to share our proposals on this issue.

Materials and methods

After approval by the local ethics committee of the Medical School, Istanbul Medeniyet University, the study was conducted on patients with occipital epileptiform activity, recruited at our department of paediatric neurology between September 2014 and January 2015. After patients with occipital epileptiform activity were reviewed, along with clinical and laboratory data based on the medical records, as the first stage, ictal symptoms were obtained again by interviewing the children and parents in order to ascertain the seizure semiology as much as possible. Patients were included in the study regardless of ictal symptoms when epileptiform activity in all interictal EEG recordings was (1) localized to the occipital lobe only or (2) diffuse but predominantly located in the occipital region. The other patients who were included in the study were those who had multifocal epileptiform activity, providing that (1) occipital epileptiform activity was the prevailing epileptiform activity on all EEG records, and (2) ictal symptoms indicated the occipital lobe only (such as ictal vomiting or ictal blindness) or were not characteristic of any brain region (such as hypomotor seizures, blank stare or generalized convulsions). Thereafter, in the light of the demographic, clinical and laboratory data, including seizure semiology, psychomotor/mental status, and brain MRI findings, patients were categorized according to aetiology (idiopathic [*i.e.* genetic] cases as primary OLE and cases with structural, metabolic or unknown aetiology as secondary OLE) and initial seizure symptoms (according to the typical initial symptoms of PS and OLE-G). Patients with no abnormality regarding both clinical and laboratory findings, including neurodevelopmental/cognitive status and neuroimaging data, were referred to as primary OLE (pOLE); otherwise, secondary OLE (sOLE). Patients with pOLE were classified as PS or OLE-G when seizures were reported to start with one of the typical initial autonomic symptoms of PS or visual symptoms of OLE-G. Patients with sOLE were categorized into either the group of Panayiotopoulos-type secondary OLE (P-type sOLE) or the group of Gastaut-type secondary OLE (G-type sOLE) when their seizures started with any typical autonomic or visual symptoms of PS or OLE-G.

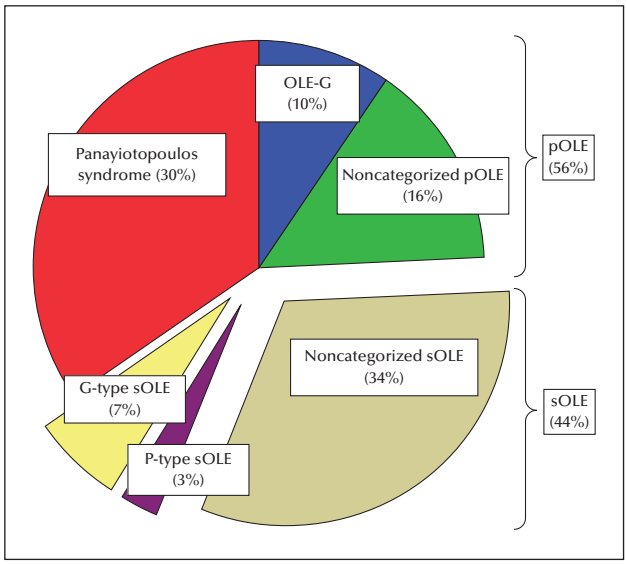

Figure 1. Distribution of the entire study group according to seizure semiology and aetiology. OLE-G: occipital lobe epilepsy of Gastaut; p: primary (idiopathic or genetic); s: secondary (structural/metabolic or unknown aetiology).

As for the patients whose seizures started with neither autonomic nor visual symptoms, these were classified as either non-categorized pOLE or non-categorized sOLE, depending on the aetiology (*table 1)*. Patients were compared with regards to seizure symptoms, aetiology, cranial imaging, EEG, treatment and outcome. Statistical tests were performed using the SPSS v18.0. The findings were compared using a chi-square test or student-*t* test, with a value of $p<0.05$ considered to be significant. Numerical values are shown as average±standard deviation or percentage (%).

Results

We evaluated 108 children with OLE (mean age: 6.4±3.9 years; male/female: 1.12) in this study. The patients were categorized into six groups, as presented in *tables 2 and 3* and *figure 1*.

– With regard to the ictal symptoms, the most common seizure symptoms were hemiconvulsion/GTCS (reported by 65% of all the patients) and impairment of consciousness (40%). The other ictal symptoms were oculomotor (36%), autonomic (37%), or visual symptoms (13%). Visual symptoms were elementary visual hallucinations (flashes and phosphenes) and, less commonly, negative visual symptoms (ictal blindness). Initial visual or autonomic symptoms were followed by another ictal symptom in most of the patients, especially in the cases with secondary OLE. Autonomic status epilepticus was found in 5 of 32 patients with PS and 2 of 8 patients with P-type sOLE. Ictal headache was reported by none of the patients, but postictal headache was reported by 18 patients.

– Classification of the patients according to seizure symptoms and aetiology is presented in *figure 1*.

– Epileptiform activity was restricted to the occipital area in half of the study group, in two thirds of the patients with pOLE, and in a third of those with sOLE. Occipital paroxysms were bilateral in half of the patients. Generalized epileptiform activity consisted of bilaterally synchronized paroxysms of 3.5-4-Hz spike-and-waves in the patient with PS but morphologically irregular spike-and-waves in the two patients with sOLE. The parents of the PS patient denied any symptoms suggestive of dialeptic or hypomotor seizures.

– Cranial imaging studies demonstrated various abnormal findings, such as brain atrophy, leukomalacia, agenesis of corpus callosum, heterotopias, subependimal nodula, and giant cell astrocytoma. Abnormal findings were not limited to the occipital region in most patients regardless of seizure semiology. Widespread brain lesions were observed to be more frequent in the non-categorized symptomatic/cryptogenic groups (*table 3*).

– The mean age at seizure onset was younger in the patients with sOLE (5.1±3.5 years) than the patients with pOLE (7.6±3.7 years) ($p<0.001$). The mean age of each primary group was younger than that of the corresponding secondary groups (PS vs P-type sOLE; OLE-G vs G-type sOLE; non-categorized pOLE vs non-categorized sOLE), but was found to be statistically significant between non-categorized pOLE vs non-categorized sOLE only ($p=0.04$).

– There were 29 symptomatic cases, with a wide range of aetiology. The medical history and clinical findings of 14 patients (five with P-type sOLE, one with G-type sOLE, and eight with non-categorized sOLE) suggested that sequelae brain lesions on cranial imaging were secondary to perinatal events (hypoxia, hypoglycaemia, and intracranial bleeding) and meningitis/encephalitis. Five patients had an inborn error of metabolism (phenylketonuria [two patients], mucopolysaccaridosis type III, non-ketotic hyperglycinaemia and Gaucher disease) and four patients a neurocutaneous syndrome (tuberous sclerosis [two patients], Sturge-Weber syndrome, and Gricelli syndrome), in addition to the patients with Angelman syndrome, 3p duplication, or isolated cortical dysplasia (four patients). Two patients had Celiac disease and reported neither autonomic nor visual ictal symptoms, and no abnormal findings were reported based on either clinical examination or brain imaging. Of the 29 symptomatic patients, 20 patients had non-categorized sOLE, seven had P-type sOLE, and two had G-type sOLE. Seven patients had a medical history of West syndrome (five with unknown aetiology and two with symptomatic West syndrome).

Table 1. Demographic findings and categorization of patients according to initial seizure symptom and aetiology.

Initial Ictal Symptom	**Autonomic symptom**		**Visual symptom**		**Neither autonomic nor visual symptom**	
Aetiology	Primary	Secondary	Primary	Secondary	Primary	Secondary
Category	*P-syndrome*	*P-type sOLE*	*Gastaut OLE*	*G-type sOLE*	*Non-categorized pOLE*	*Non-categorized sOLE*
N	32	8	11	3	17	37
Age (mean±SD) years	6.7 ± 3.4	4.9 ± 2.3	10.6 ± 3.5	8.6 ± 3	7.2 ± 4.3	4.8 ± 3.8
Gender (M/F)	10/22	8/0	5/6	0/3	11/6	23/14
Seizure symptoms						
(A) Autonomic symptom at seizure onset						
• Autonomic seizure only	10 (8 IE, 4 IS)	1 (IS)				
• Autonomic symptom plus	22 (18 IE, 6 IS)	7 (7 IE, 2 IS)				
○ Oculomotor	11 (ED)	7 (6 ED, 1 EF)				
○ Head turning	2	3				
○ Hemiconvulsion/GTCS	9	4				
(B) Visual symptom at seizure onset						
• Visual ictal seizure only			2 (1 EH, 2 B)	0		
• Visual ictal symptom plus			9 (7 EH, 2 B)	3 (1 EH; 2 B)		
○ Oculomotor			5 (5 ED, 3 EF)	2 (2 ED, 1 EF)		
○ Head turning			3	0		
○ Hemiconvulsion/GTCS			7	2		
(C) Seizures without autonomic or visual onset					17	37
• Oculomotor (with or without head turning)					4	10 (8 ED, 2 EF, 1 IN)
• Hemiconvulsion/GTCS					16	32
*Others						
• Ictal impairment of consciousness	28	6	4	0	1	5
• Postictal headache	9	1	5	2	1	0
• Status epilepticus	9 (5 autonomic; 4 convulsive)	3 (2 autonomic; 1 convulsive)	0	0	2 (convulsive)	6 (3 convulsive, 3 non-convulsive)

Abbreviations: B: Blindness; EH: Elementary visual halucinations; ED: Eye deviation; EF: Eyelid fluttering, IE: Ictal emesis (ictal nausea, retching or vomiting); IN: Ictal nystagmus; IS: Ictal syncope; s/c: Symptomatic/cryptogenic; G-type: Gastaut-type; P-syndrome: Panayiotopoulos syndrome; P-type: Panayiotopoulos-type; pOLE: primary Occipital Lobe Seizures; sOLE: secondary Occipital Lobe Seizures.

Table 2. Findings on cranial imaging, EEG and treatment response.

Category	*Panayiotopoulos syndrome*	*P-type s/cOLE*	*OLE of Gastaut*	*G-type s/cOLE*	*Non-categorized iOLE*	*Non-categorized s/cOLE*
N	32	8	11	3	17	37
Cranial MRI findings						
• Normal	32	0	11	1	17	4
• Abnormal	-	8	-	2	-	33
○ Occipital finding only		1		-		-
○ Extraoccipital finding only		2		-		6
○ Multilobar including occipital lobe		5		2		27
Epileptiform activity on EEG						
• Occipital area only	19 (bilateral in 10)	4 (bilateral in 3)	8 (bilateral in 3)	1 (bilateral)	10 (bilateral in 4)	18 (bilateral in 11)
• Wider area including occipital area	11 (bilateral in 6)	3 (bilateral in 3)	3 (bilateral in 2)	2 (bilateral in 1)	3 (unilateral in 3)	19 (bilateral in 8)
• Occipital and extraoccipital areas (independent foci)	1 (parietal)	1 (temporal)				
• Occipital and generalized discharges	1					2
Response to antiepileptic drug treatment						
• No seizure after drug treatment was stoped	2	1	0	0	2	3
• No seizure with 1 or 2 antiepileptic drugs	29	5	8	0	12	13
• No seizure with 3 or more AEDs	1	1	2	0	3	11
• No response to antiepileptic drugs	0	1	1	3	0	10

Abbreviations: P-syndrome: Panayiotopoulos syndrome; G-type: Gastaut type.

– Seventeen per cent of the entire study group was not seizure-free in spite of three or more antiepileptic drugs. Most of the patients with pOLE (59/60) were seizure-free for more than six months. However, one third of the patients with sOLE were not seizure-free despite antiepileptic drug treatment. The rate of seizure freedom with two or less antiepileptic drugs was 88% in primary cases but 46% in secondary cases ($p<0.001$). Furthermore, the patients who had sOLE, without autonomic seizures, responded to drug treatment to a lesser extent than those with sOLE and autonomic symptoms (13% vs 32%; $p=0.04$).

Discussion

There are two types of epilepsy syndromes that can be utilized to classify patients with OLE: PS and OLE-G.

Table 3. Epilepsy syndrome classification in OLE and proposal additional definitions for patients who are neither PS nor OLE-G (OLE: Occipital lobe epilepsy; PS: Panayiotopoulos syndrome; OLE-G: Late-onset childhood occipital epilepsy of Gastaut; pOLE: Primary OLE; sOLE: Secondary OLE).

(A) Patients with primary OLE
- Patients who have typical autonomic or visual seizures of PS or OLE-G → PS or OLE-G
- Proposal definition for patients having seizures that do not start with typical autonomic or visual seizures of PS and OLE-G → Noncategorized pOLE

(B) Proposal definitions for patients with secondary OLE, that is, those with OLE related to structural/metabolic or unknown etiology
- Patients who have typical autonomic or visual seizures of PS or OLE-G → P-type sOLE or G-type sOLE
- Proposal definition for patients having seizures that do not start with typical autonomic or visual seizures of PS and OLE-G → Noncategorized sOLE

(C) Proposal definitions for patients with OLE and under investigation for etiology
- Depending on seizure semiology → P-type OLE, G-type OLE, noncategorized OLE

In this study, we evaluated 108 children with OLE (6.4±3.9 years) in the light of the characteristics of these epilepsy syndromes (*tables 2 and 3*, *figure 1*). The patients with sOLE constituted 44% of the entire study group. This ratio can be expected to be smaller in the general population since there are likely to be more patients with symptomatic epilepsy in referral epilepsy centres. Regarding the age at seizure onset, children with sOLE were younger than those with pOLE (7.6±3.7 vs 5.1±3.5 years; $p<0.001$) and the patients with noncategorized sOLE had the youngest average age. Tata *et al*. (2014) reported the age at seizure onset to be younger in patients with sOLE than in those with PS (3.4 vs 5.6 years). While idiopathic OLE usually starts in childhood, symptomatic OLE may start at any age depending on underlying disorders. When the congenital and perinatal causes in paediatric cases are considered, it is not surprising that symptomatic cases with OLE present at younger age compared to patients with pOLE.

OLE manifests with a wide range of ictal symptoms, from visual or oculomotor symptoms to autonomic symptoms (Bien *et al*., 2000; Panayiotopoulos, 2010). Besides these symptoms, different ictal spread patterns of initial occipital discharge may give rise to various non-occipital ictal symptoms and even obscure the initial occipital symptoms. Therefore, focal sensory or motor seizures, hemiconvulsions, secondary generalized seizures and temporal seizures with complex partial semiology may be the presenting symptoms. Likewise, hemiconvulsion/GTCS and impairment of consciousness, which were found in 65% and 40% of all the patients with OLE, respectively, were the most common ictal symptoms in our study. Oculomotor symptoms were noted in 36% of our patients with OLE. These rates underline the importance of conventional ictal symptoms (such as GTC, impairment of consciousness, and oculomotor symptoms) in OLE. As for the typical occipital ictal symptoms, patients with typical autonomic or visual symptoms of PS and OLE-G constituted half of the patients with OLE. It was remarkable that a quarter of the patients with pOLE had no typical initial autonomic or visual ictal symptoms of PS or OLE-G. In addition, a quarter of the patients with sOLE had seizures beginning with either autonomic or visual ictal symptoms of these two syndromes.

Onset and progression of ictal symptoms during seizures form the basis of the description of PS and OLE-G. In the literature, studies on childhood OLE have indicated that many patients could not be classified as either PS or OLE-G. Genizi *et al*. (2007) reported that half of their patients with occipital paroxysms could not be segregated into either specific syndrome. Kivity *et al*. (2000) pointed out that 28% of their patients showed mixed signs and symptoms of both syndromes. In our study, 28% of the patients with pOLE, 20% of the patients with the typical autonomic symptoms of PS, and 21% of the patients with the typical visual symptoms of OLE-G could not be classified as either PS or OLE-G. With regard to the classification of OLEs, the most important factor in the literature appears to be the method of study. PS and OLE-G have distinct ictal semiology characterized by not only ictal symptoms but also the order of appearance of the ictal symptoms (Covanis, 2008). It is very important to use appropriate definitions for seizure semiology because of their central role, not only in appropriately classifying patients with epilepsy but also in making a correct decision on the ictal-onset zone and symptomatogenic zone. In this respect, categorization of localization-related epilepsy is as important as electroclinical epilepsy syndrome classification. In our study,

we classified the patients depending on both their ictal symptoms and the order of appearance of the ictal symptoms, which is in contrast to some studies in the literature. In addition, we have made a further categorization (*table 1*), which has not previously been reported, since many patients with OLE did not present the characteristics of PS or OLE-G *(figure 1*). For instance, 20% of the patients with OLE who had the characteristic autonomic or visual ictal symptoms of PS or OLE-G had structural/metabolic or unknown aetiology. In addition, 28% of the patients with pOLE reported that their seizures started with no autonomic or visual symptom. We defined the OLE patients with structural or unknown aetiology as P-type sOLE or G-type sOLE when they had one of the typical ictal symptoms of either PS or OLE-G, respectively. As for the patients who had no characteristic ictal symptoms of PS or OLE-G, we formed two groups in addition: non-categorized pOLE and non-categorized sOLE. Using this approach, we were able to distinguish and present the features of the patients with OLE who did not present the characteristics of PS or OLE-G *(figure 1*).

The most common cause of sOLE in our study was perinatal events such as hypoxia/ischaemia, hypoglycaemia and intracranial bleeding. Occipital brain damage resulting from perinatal hypoxic or metabolic insults and its clinical consequences have been increasingly underlined in recent years. It is also remarkable that some patients with such perinatal events show normal neurodevelopment during infancy but develop occipital epilepsy later (Gil-Nagel *et al.,* 2005; Oguni *et al.,* 2008). In addition to the perinatal causes, our study showed a wide range of aetiology including inborn errors of metabolism and Celiac disease. Celiac disease has been reported to be frequent in OLE compared to the other epilepsies in the literature. Epilepsy associated with Celiac disease is highly variable with regards to seizure semiology and prognosis, ranging from benign syndromes to intractable epilepsy (Licchetta *et al.,* 2011). Likewise, the patients with Celiac disease in our study showed no prevailing ictal symptom or epilepsy syndrome and responded to antiepileptic drug treatment well.

As for the interictal EEG abnormalities, EEG showed generalized epileptiform activity additionally in four patients, one of whom was a PS patient. Studies have reported that EEG may show generalized discharges in 10% to 50% of patients with pOLE; occipital epileptiform discharges with occipital seizures may also occur in children with typical absence seizures (Caraballo *et al.,* 2004; Verrotti *et al.,* 2010; Adcock and Panayiotopoulos, 2012).

Antiepileptic drug treatment for OLE is similar to that for any other type of focal seizure, and is usually effective. However, some patients may require surgical intervention (Salanova *et al.,* 1992). In our study, 100% of the patients with PS, 91% of those with OLE-G, and 87% of the entire study group were found to be seizure-free with antiepileptic drug treatment. While 98% of the patients with pOLE responded to antiepileptic drug treatment, 71% with sOLE responded. Of the patients with sOLE, those with autonomic symptoms (*i.e.* those with P-type sOLE) responded to antiepileptic drugs better than other sOLE patients (p=0.04).

In this study, we interviewed the patients and parents about seizure symptoms in order to reveal ictal symptoms as accurately as possible. This method appeared to be both an advantage and a limitation of this study. We would have found more non-categorized patients if the study had been based on past medical records only. On the other hand, some symptoms may have been forgotten by the parents and patients, as the interviews were performed several months or years after initial admission of the patients. Besides this, there are some additional factors that challenge the collection of data on ictal symptoms. Some symptoms at seizure onset may be overlooked when mild, and not witnessed when they occur in sleep. Children, especially those with intellectual deficit, may fail to report symptoms. Observing occipital symptoms can be difficult due to fast ictal spread to other brain areas (Salanova *et al.,* 1992). All these factors make it difficult to collect data on ictal symptoms, which is an issue for all clinical studies that are not based on objective ictal monitoring. Therefore, our study group did not include occipital epilepsy patients with no interictal occipital epileptiform activity on EEG. In addition, some of the patients with multifocal epileptiform activity predominantly in the occipital region may not have had occipital lobe epilepsy even though they lacked non-occipital ictal symptoms. However, all these limitations are inevitable obstacles unless ictal video-EEG monitoring is performed.

In conclusion, children with OLE should be carefully evaluated for underlying causes because a significant proportion of patients are not idiopathic, especially children at younger age and those requiring three or more antiepileptic drugs. Non-autonomic and non-visual symptoms are no less important than autonomic and visual symptoms in OLE. On the other hand, seizures beginning with typical autonomic or visual symptoms of PS or OLE-G in a patient with OLE does not necessarily mean that this patient has either PS or OLE-G. Many patients with OLE in our study, 60% of patients with OLE and 28% of those with pOLE could not be classified as PS or OLE-G. In the light of these findings, our study demonstrates that additional definitions and terminology are necessary alongside the descriptions that are used in epilepsy syndrome classification. All the patients in this study, not classified as either PS or OLE-G, would have otherwise

been lumped into a single group. For instance, symptomatic occipital epilepsy patients who have seizures that start with vomiting would have to be categorized together with patients with occipital epilepsy related to genetic aetiology who do not have typical seizures of PS or OLE-G. The literature is not helpful on this issue at all. Our proposals presented here consider both the categorization of localization-related epilepsy and classification of electroclinical epilepsy syndrome. We hope that our findings and proposed definitions and terminology are helpful in differentiating between patients with the different features OLE in clinical practice and research. □

Supplementary data.
Summary didactic slides are available on the www.epilepticdisorders.com website.

Disclosures.
None of the authors have any conflict of interest to disclose.

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TEST YOURSELF

(1) What are the electroclinical epilepsy syndromes related to the occipital lobe?

(2) Most of the children with occipital lobe epilepsy have autonomic or visual ictal symptoms; is this correct?

(3) An occipital epilepsy patient whose seizures begin with autonomic or visual ictal symptoms has either Panayiotopoulos syndrome or late-onset occipital lobe epilepsy of Gastaut. Is this correct?

(4) Are all cases with occipital lobe epilepsy classified under the two epilepsy syndromes related to the occipital lobe (*e.g.* Panayiotopoulos syndrome or late-onset occipital lobe epilepsy of Gastaut)?

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com, under the section "The EpiCentre".