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Original Article

Childhood Epilepsies with Occipital Discharges: Evaluation of 84 Patients

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ABSTRACT

Aim: Childhood epilepsy with occipital discharges encompasses various subtypes, including childhood occipital visual epilepsy (COVE), self-limited epilepsy with autonomic seizures (SeLEAS), photosensitive occipital lobe epilepsy, symptomatic epilepsy, and unclassified cases. The primary aim of this study was to analyze the clinical characteristics of pediatric epilepsy patients with occipital discharges based on their etiological classification and to compare any differences between these subgroups. Additionally, this study sought to identify prognostic factors by comparing patients who achieved remission within 36 months (Group 1) with those who did not respond within the same period (Group 2).

Materials and Methods: This study included 84 children diagnosed with occipital discharge-related epilepsy. A comprehensive review of their medical records was conducted, assessing their demographic data, ictal symptoms, neurological examination findings, electroencephalography and magnetic resonance imaging results, family history, febrile seizures, and treatment responses.

Results: Of the total cohort, 32% (n=27) were classified as Group 1, while 68% (n=57) were in Group 2. Structural brain abnormalities were significantly more prevalent in Group 2. The age at diagnosis was significantly younger in Group 2 compared to Group 1 (p=0.003), and the rate of intellectual disability was higher in Group 2 (p=0.05). The presence of systemic diseases and the use of multiple anti-epileptic drugs were significantly more frequent in Group 2 (p=0.021, p=0.018). The duration of epilepsy follow-up was notably longer in Group 2 (p<0.001). COVE and SeLEAS were more commonly found in the early remission group (p=0.012, p=0.034), while no cases of symptomatic occipital epilepsy achieved remission within the first 36 months (p=0.001).

Conclusion: The majority of children with occipital epilepsy did not achieve remission within 36 months. Younger age at onset and the presence of intellectual disability were associated with longer periods of non-remission. COVE and SeLEAS were more likely to achieve early remission, whereas symptomatic occipital epilepsies showed no remission within the first 36 months. These findings underline the importance of early diagnosis and highlight the potential impact of structural brain abnormalities and cognitive impairments on the prognosis of childhood occipital epilepsy.

Keywords: Occipital epilepsy, children, prognostic factors, remission

Introduction

Occipital lobe epilepsies (OLE) represent a diverse group of epileptic syndromes with distinct clinical features, etiologies, and prognostic outcomes. These epilepsies, which are relatively rare in children, can be broadly categorized into idiopathic and symptomatic types. Idiopathic OLE include syndromes such as childhood occipital visual epilepsy (COVE), self-limited epilepsy with autonomic seizures

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(SeLEAS), and photosensitive occipital lobe epilepsy (POLE), whereas symptomatic occipital epilepsies are associated with structural brain abnormalities, metabolic disturbances, or other underlying conditions.

The classification of occipital lobe epilepsy is essential in order to determine treatment approaches and predict prognosis. According to the International League Against Epilepsy (ILAE), the major subtypes of occipital lobe epilepsy in children are: COVE, SeLEAS, POLE, and symptomatic occipital lobe epilepsy. COVE and SeLEAS are typically characterized by self-limited, benign courses, with seizure onset often occurring between the ages of 3 and 16 years. POLE, in contrast, is marked by photosensitivity and focal seizures originating in the occipital lobe. Symptomatic occipital epilepsies, which are associated with identifiable brain lesions or systemic conditions, tend to have worse prognoses (1).

Despite significant progress in the understanding of occipital lobe epilepsies, identifying prognostic factors for treatment response remains a clinical challenge. It is known that the prognosis for children with occipital lobe epilepsy varies considerably depending on the subtype and the presence of structural abnormalities. Previous studies have shown that idiopathic occipital epilepsies, such as SeLEAS, typically exhibit remission within the first two years, whereas symptomatic occipital epilepsies have a much lower rate of remission and often require long-term management (1). Additionally, factors such as age at onset, the presence of cognitive impairments, and the use of multiple anti-epileptic drugs (AEDs) have been shown to influence the outcome (2).

While there is a growing body of literature addressing pediatric occipital epilepsy, studies focusing specifically on the comparison of clinical features and treatment outcomes across different subtypes are limited. Understanding these differences is crucial for developing more tailored treatment strategies and improving outcomes for affected children. Moreover, identifying early prognostic markers could guide clinicians in predicting the likelihood of remission and customizing treatment regimens accordingly.

Interestingly, prior pediatric case series involving occipital discharges have reported that remission is commonly observed within 1-2 years, and no patients exhibited symptoms after the age of 12 years (3,4). In contrast, adult studies have shown significantly lower rates of remission, highlighting the importance of age-dependent differences in the disease course (5). Additionally, differences in prognosis and clinical characteristics between subtypes such as COVE and SeLEAS, despite both being classified as idiopathic, have also been emphasized in the literature (3,6).

In this study, we aimed to analyze the clinical and imaging characteristics of 84 children diagnosed with occipital discharge-related epilepsy. We also sought to identify factors associated with early remission and treatment failure, comparing patients who achieved remission within 36 months (Group 1) with those who did not (Group 2). By examining the relationship between these factors, we hope to provide more insights into the management of pediatric occipital lobe epilepsy and improve our understanding of the factors influencing treatment response.

Materials and Methods

This retrospective cohort study included a total of 84 children aged 0-17 years who were diagnosed with occipital epilepsy between 2018 and 2023. Cases with a follow-up period of less than 6 months, those with generalized discharges on electroencephalography (EEG), those with dominant abnormal discharges detected in areas other than the occipital lobe, and cases with irregular follow-up exceeding one year or those non-compliant with treatment were excluded. Patients who were diagnosed with occipital epilepsy but did not receive anti-epileptic drug therapy were also excluded from this study.

This study was approved by the İzmir Bakırçay University Non-Interventional Clinical Research Ethics Committee (approval no.: 1056, date: 05.24.2023).

The clinical and laboratory features, EEG, and magnetic resonance imaging (MRI) results of the patients were analyzed. The demographic data of the patients (age at epilepsy diagnosis, current age, gender, parental consanguinity), family history of epilepsy, history of febrile convulsions, presence of other systemic diseases, AEDs used and their number, the duration of seizure control with anti-epileptic drug therapy, EEG findings, subtype of occipital lobe epilepsy (COVE, POLE, SeLEAS, symptomatic, unclassified), duration of epilepsy follow-up, MRI findings, additional neurological findings, and psychiatric disorders were recorded.

Patients with occipital pathological discharges on EEG (high-amplitude sharp and/or spike discharges) who met the clinical criteria for occipital epilepsy syndromes, including COVE, POLE, SeLEAS, and symptomatic occipital epilepsy, were classified under separate diagnoses according to the ILAE classification system and included in this study as part of the idiopathic epilepsy group.

Those patients with occipital lesions and occipital discharges on EEG were diagnosed with symptomatic occipital epilepsy.

Patients with occipital lobe discharges but not meeting the clinical features of any defined occipital lobe epilepsy syndrome were classified as unclassified epilepsies.

In our study, those patients who had not experienced seizures within the prior six months or who had normal EEG findings despite receiving one or more anti-epileptic drugs, or those who were treated for epilepsy and had completed treatment with drug withdrawal within 36 months, were classified as responders to treatment (Group 1). Patients were classified as non-responders to treatment (Group 2) if they met at least one of the following criteria: (1) they had never experienced a seizure-free period longer than six months; (2) they had experienced a seizure-free period longer than six months, but occipital discharges persisted on EEG; or (3) both of these conditions were present (Group 2).

Statistical Analysis

Statistical analyses were performed using Jeffreys's Amazing Statistics Program and Jamovi software. Descriptive statistics are presented as frequencies (percentages) for categorical variables and medians (minimum-maximum) for numerical variables. Pearson's chi-square, Fisher's exact, or Fisher-Freeman-Halton tests, and the Mann-Whitney U test were used, and p-values were calculated.

Results

The total group of 84 patients was divided into two groups based on their characteristics. Group 1 (n=27) included patients with a shorter duration of epilepsy follow-up, while Group 2 (n=57) had a longer follow-up period. Statistical analysis revealed significant differences between the groups in several variables (Table I).

The epilepsy follow-up duration was significantly shorter in Group 1 [27.0 months (8.0-54.5)] compared to Group 2 [63.3 months (37.0-210.49)], with a p-value of <0.001. The age of epilepsy onset was also significantly different between the groups, with Group 1 having an average age of 5.88±3.88 years, while Group 2 had a lower average age of 4.10±3.80 years (p=0.024).

There was no significant difference between the groups in terms of sex distribution (p=0.308), family history of

Table I. Patient characteristics and group comparison based on treatment response

Variable	Total Group (n=84)	Group 1 (n=27)	Group 2 (n=57)	p-value
Follow-up duration (months)	49.3 (8.0-210.4)	27.0 (8.0-54.5)	63.3 (37-210.4)	<0.001
Age of epilepsy onset (years)	5.07±4.16	5.88±3.88	4.10±3.80	0.024
Sex female/male	36/48	26/31	10/17	0.308
Family history of epilepsy n (%)	20/84 (23.8)	6/27 (22.2)	14/57 (24.5)	0.523
Automatisms, n (%)	21 (25)	4 (14.8)	17 (29.8)	0.225
Visual symptoms, n (%)	34 (40.5)	12 (44.4)	22 (38.6)	0.391
Intellectual disability n (%)	8 (9.5)	0 (0)	8 (14.03)	0.038
Neurological findings, n (%)	13 (15.5)	3 (11.1)	10 (17.5)	0.535
Longest seizure-free period (months)	24.0 (2.0-60.0)	16.5 (2.0-60.0)	24.0 (4.0-60.0)	0.030
MRI abnormalities, n (%)	18 (21.4)	1 (3.7)	17 (29.8)	0.015
Use of two or more anti-epileptic drugs, n (%)	29 (34.5)	4 (14.8)	25 (43.9)	0.018
Febrile seizures, n (%)	16 (19.0)	2 (7.4)	14 (24.6)	0.116
Occipital lobe epilepsy subtypes, n (%)				
- COVE	11 (13.1)	5 (18.5)	6 (10.5)	0.012
- POLE	5 (6.0)	2 (7.4)	3 (5.3)	0.424
- SeLEAS	26 (31.0)	13 (48.1)	13 (22.8)	0.034
- Symptomatic	11 (13.1)	0 (0.0)	11 (19.3)	0.001
- Unclassified	31 (36.9)	7 (25.9)	24 (42.1)	0.322

MRI: Magnetic resonance imaging, COVE: Childhood occipital visual epilepsy, POLE: Photosensitive occipital lobe epilepsy, SeLEAS: Self-limited epilepsy with autonomic seizures

epilepsy (p=0.523) or automatisms (p=0.225). Similarly, no significant difference was found in visual symptoms (p=0.391) or neurological findings (p=0.535).

The longest seizure-free period was shorter in Group 1 [16.5 months (2.0-60.0)] compared to Group 2 [24.0 months (4.0-60.0)], with a significant difference (p=0.030). MRI abnormalities of the patients were significantly more common in Group 2 (29.8%) than in Group 1 (3.7%) with a p-value of 0.015. Only one patient in the group who achieved remission within the first 36 months (Group 1) showed bilateral ventricular enlargement. In contrast, among those patients who did not achieve remission within 36 months (Group 2), MRI abnormalities were detected in 17 cases. These included 4 cases of cortical dysplasia, 6 cases of periventricular leukomalacia, 2 cases of occipital hypoglycemia sequelae, and 5 cases of non-specific ventricular enlargement. The use of two or more AEDs was more prevalent in Group 2 (43.9%) compared to Group 1 (14.8%), with a statistically significant difference (p=0.018).

Regarding febrile seizures, the occurrence was higher in Group 2 (24.6%) compared to Group 1 (7.4%), but this difference did not reach statistical significance (p=0.116).

In terms of epilepsy subtypes, there were notable differences between the groups. The frequency of occipital lobe epilepsy subtypes was significantly different between the groups. COVE was more frequent in Group 1 (18.5%) than Group 2 (10.5%) (p=0.012), while SeLEAS was more prevalent in Group 1 (48.1%) than in Group 2 (22.8%) (p=0.034). Symptomatic epilepsy was significantly more common in Group 2 (19.3%) than in Group 1 (0.0%) (p=0.001). No significant differences were found for POLE (p=0.424) and the unclassified subtypes (p=0.322).

These findings highlight key differences in clinical characteristics, seizure control, and epilepsy subtypes between the two groups.

Discussion

OLE consist of idiopathic (POLE, COVE, SeLEAS), symptomatic (cortical dysplasia, hypoglycemia, infarction, etc.), and unclassified groups. It has been reported that 1.2-2.6% of newly diagnosed epileptic patients have occipital epilepsy (7). In pediatric patients, one of the most common questions after clinical diagnosis is when the disease will improve. In search of an answer to this question, we aimed to identify some prognostic factors which could assist at the time of diagnosis. We compared the characteristics of those patients who achieved remission within the first 36 months with those who did not.

We set the limit for early remission at 36 months in this study. It was found that those patients who did not achieve remission had an average treatment duration of 63.3 months. This finding suggests that identifying prognostic factors and making predictions from the time of diagnosis are crucial for both the patient's expectations and the patient's trust in the physician.

Early onset of epilepsy has been found to be a negative prognostic factor for early remission and recovery. In a similar study, early diagnosis age in OLE was related to abnormalities in brain MRI findings (8). Although that study did not identify robust predictors for seizure outcome, our data suggest that early MRI abnormalities and younger age at onset may also be linked to poorer long-term seizure control. Together, these findings underscore the clinical importance of integrating neuroimaging and semiological features when evaluating prognosis in pediatric occipital epilepsy. Symptomatic epilepsy is often associated with earlier onset, and given its generally poorer prognosis compared to idiopathic epilepsy, early onset itself emerges as a risk factor for poor treatment response. The presence of MRI abnormalities, early age at seizure onset, and symptomatic etiology appear to be interrelated factors, collectively contributing to the clinical significance of the observed outcomes. The greater the extent of structural abnormality on brain MRI, the higher the likelihood of drug-resistant occipital lobe epilepsy. A systematic review and meta-analysis reported that the presence of a lesion on preoperative MRI increases the odds of medical treatment failure by approximately 3.24-fold in OLE (9). Our findings align with this literature: Group 2 demonstrated a significantly higher rate of MRI abnormalities (29.8%, p=0.015) and correspondingly poorer remission outcomes.

In our study, the frequency of automatisms was found to be 25%. There are only a limited number of studies in the literature reporting on the prevalence of automatisms in occipital lobe epilepsies. Salanova et al. (10) reported that approximately 50% of patients with occipital lobe epilepsy exhibit automatisms resembling those seen in temporal lobe epilepsy. Similarly, Wong et al. (11) emphasized that the presence of automatisms may indicate temporal spread of epileptic activity. In our cohort, those patients in Group 2 were older, and although not statistically significant, automatisms were more frequent in this group. Oguni (12) showed that occipital EEG foci tend to evolve with age,

shifting toward the centro-parietotemporal and frontopolar regions, with this transformation continuing up to the age of 12-16 years. Considering that automatisms are primarily associated with temporal lobe activity, the older age and persistent occipital discharges observed in Group 2 suggest that the clinical presentation in these children may have gradually transitioned from occipital to a temporal lobe epilepsy semiology.

In a prospective study by Ko and Holmes (13) involving 343 pediatric patients, intellectual disability was shown to be a significant risk factor for drug-resistant epilepsy. In our study, 14% (8 patients) of the patients in Group 2 had intellectual disability, and the difference between the groups was statistically significant (p=0.038).

In pediatric patients, if the appropriate drug and dose are administered, seizure control is achieved with monotherapy in 65-70% of cases (14). Although polytherapy is generally avoided, it has been reported that 30-50% of cases require polytherapy (15). In a study by Datta and Wirrell (16), it was reported that the number of AEDs negatively affects prognosis. In our study, 29 patients (34.5%) were on polytherapy, with 25 of them (43.9%) being in Group 2 and 4 of them (14.8%) being in Group 1. A statistically significant difference was found between the groups in terms of the likelihood of using two or more AEDs (p=0.018).

The median value of the longest seizure-free period was 24 months for patients in Group 2. The longest seizure-free period in Group 2 was significantly longer than in Group 1 (p=0.030). Patients in Group 2 had not been seizure-free for more than six months or continued to show abnormal occipital discharges on EEG after 36 months of treatment. Interestingly, there were patients who had been seizure-free for 48 months, but their EEGs remained pathological.

In a study by Gherpelli et al. (17), the presence of sharp waves, spikes, multiple spikes, and spike-slow wave patterns on EEG before drug withdrawal was found to increase the risk of seizure recurrence. In another meta-analysis, the relative risk between abnormal EEG patterns prior to drug withdrawal and recurrence was found to be 1.45 (18). Interictal EEG abnormalities are directly proportional to the presence of clinical seizures and serve as an indicator of poor prognosis before treatment termination. Therefore, a careful decision must be made regarding treatment withdrawal in patients with persistent EEG abnormalities (19). In our clinic, recovery is defined as both seizure freedom and complete resolution of EEG abnormalities. As a result, our recovery times are longer compared to the literature.

In our study, there were no patients in the symptomatic group who achieved remission. The SeLEAS group was the most promising group with the quickest recovery, with 48% of patients achieving remission within the first 36 months of treatment. These results are consistent with the literature, which reports symptomatic OLE and POLE subtypes as the groups with the longest time to response and the highest recurrence rates (20).

Study Limitations

This study has several limitations which should be acknowledged. First, its retrospective design inherently limits the ability to establish causality between clinical features and treatment outcomes. Second, although the study cohort included a relatively large sample of pediatric patients with occipital lobe epilepsy, the subgroup sizes (e.g., SeLEAS, symptomatic, POLE) were modest, which may limit the statistical power of certain comparisons and restrict the generalizability of subtype-specific conclusions. Third, follow-up durations varied across patients, and the minimum follow-up period of six months might have been insufficient to detect long-term remission or relapse in some cases. Fourth, EEG interpretation was not centrally reviewed and may have been subject to inter-observer variability. Prospective, multicenter studies with standardized imaging protocols and extended follow-ups are needed to validate and expand upon these findings.

Conclusion

This study provides valuable insights into the clinical and prognostic factors of OLE in pediatric patients. The findings suggest that early onset of epilepsy and the presence of symptomatic epilepsy are associated with a less favorable treatment response. In particular, those patients with early onset epilepsy, as well as those with MRI abnormalities or symptomatic forms of epilepsy, tend to have longer treatment durations and more persistent seizure activity. Conversely, idiopathic epilepsies, such as SeLEAS, show better outcomes, with a higher rate of remission within the first 36 months of treatment.

Additionally, the use of polytherapy and the presence of cognitive impairments, such as intellectual disability, were found to be significant factors contributing to poor prognosis in this cohort. These results emphasize the importance of the early identification of prognostic factors which can help guide treatment strategies and improve the management of pediatric occipital lobe epilepsy.

In conclusion, understanding the clinical and imaging characteristics associated with different OLE subtypes

is crucial in predicting patient outcomes and tailoring individualized treatment plans. Further research is needed to identify additional factors influencing treatment response and to develop more effective therapeutic approaches for pediatric patients with occipital lobe epilepsy.

Ethics

Ethics Committee Approval: This study was approved by the İzmir Bakırçay University Non-Interventional Clinical Research Ethics Committee (approval no.: 1056, dated: 24.05.2023).

Informed Consent: Informed consent was obtained from the patients and their parents.

Footnotes

Authorship Contributions

Surgical and Medical Practices: H.G.T., Concept: H.G.T., Design: H.G.T., Data Collection or Processing: D.Ö., Analysis or Interpretation: H.G.T., Literature Search: D.Ö., Writing: H.G.T., D.Ö.

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