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Clinical Correlates of Occipital Intermittent Rhythmic Delta Activity (OIRDA) in Children

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Summary: *Purpose:* The clinical significance of occipital intermittent rhythmic delta activity (OIRDA) on the electroencephalogram has not been fully established. Recent studies suggest that this pattern occurs almost exclusively in children and is probably of epileptic origin in most cases. We sought to characterize the electrographic features and clinical correlates of occipital intermittent rhythmic delta activity.

Methods: A review of 697 consecutive pediatric electroencephalograms detected occipital intermittent rhythmic delta activity in 24 studies. Mean patient age was 7.96 years.

Results: Recent convulsions and absence seizures constituted the main indications for the study. Concomitant, independent epileptiform activity was noted in half of the cases. This activity was focal in all but one case. Conversely, in most cases of absence seizures, epileptiform activity intermixed with occipital intermittent rhythmic delta activity. Furthermore, the frequency of the occipital rhythmic discharges in studies of children with absences was generally faster (3–4 Hz) than in localization-related epilepsy (2–3 Hz). Most patients were awake when occipital intermittent rhythmic delta activity occurred. Chronic encephalopathy was seen in one child only. Analysis of neuroimaging studies in eight cases revealed no structural pathology associated with occipital intermittent rhythmic delta activity.

Conclusions: Occipital intermittent rhythmic delta activity is probably an epileptiform pattern, although it is noted occasionally in encephalopathic children. Its electrographic characteristics appear to differ between localization-related epilepsy and primary generalized epilepsy, particularly absence seizures.
Key Words: Occipital intermittent rhythmic delta activity—Absence seizures—Encephalopathy.

The significance of intermittent delta activity in the electroencephalogram (EEG) has intrigued neurologists, epileptologists, and electroencephalographers for decades. Throughout the years, the range of clinical interpretations of this pattern by researchers varied from being nonspecific (Sharbrough, 1987) to suggesting a metabolic (Fariello et al., 1982; Watemberg et al., 2002), structural (Cobb, 1945; Watemberg et al., 2002), or infectious etiology (Uysal et al., 2001; Buoni et al., 2005), and even epilepsy (Riviello and Foley, 1992; Di Gennaro et al., 2003). Furthermore, it has become evident that three distinct forms of intermittent rhythmic delta activity exist, with different clinical importance: frontal (FIRDA), occipital (OIRDA), and temporal (TIRDA).

Frontal intermittent rhythmic delta activity was initially attributed to deep or midline central nervous system lesions (Cobb, 1945). However, more recent studies have shown that it occurs during metabolic derangement in adults with previous brain insults (Watemberg et al., 2002) and as a rare finding of unclear significance (Watemberg et al., 2003) in children. Ipsilateral temporal intermittent rhythmic delta activity appears to be indicative of temporal epilepsy (Di Gennaro et al., 2003).

Occipital intermittent rhythmic delta activity occurs almost exclusively in children. It has been associated with epilepsy, particularly with primary generalized syndromes, such as childhood absence (Loiseau et al., 1983; Riviello and Foley, 1992). These findings were supported by the recent work by Gullapalli and Fountain (Gullapalli and Fountain, 2003).

The aim of our study was to analyze further the clinical significance of occipital intermittent rhythmic delta activity in children. Particular attention was given to the neurologic background of the patients whose EEGs depicted this pattern.

METHODS

Pediatric EEGs depicting OIRDA between April 2000 and March 2005 were retrieved from the database of the EEG laboratory at Wolfson Medical Center. This laboratory serves inpatients as well as community patients from the Southern Tel Aviv area referred for routine

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TABLE 1. *Main clinical and electrographic features of EEG records depicting OIRDA*

Indications for EEG study	
Suspected convulsions/postictal status	15
Follow-up for absence seizures	4
Suspected absence seizures	3
Patient's state when OIRDA occurred	
Awake	16
Awake and asleep	6
Asleep	2
EEG background activity in 22 awake children	
Resting occipital alpha rhythm	17
Theta	5
Frequency of OIRDA discharge (22 cases)	
1–2 Hz	1
2–3 Hz	16
3–4 Hz	5

EEGs and children from around the country referred for prolonged video-EEG studies. The international 10-20 electrode placement system is applied to all cases. Recordings were performed on the mobile Ceegraph IV BioLogic system (BioLogic Systems Corporation, Mundelein, IL, U.S.A.). Charts of in- and outpatient records of patients with OIRDA were reviewed. Compiled clinical data included demographics, perinatal and medical history, neurodevelopmental status, specific genetic syndromes or metabolic diseases, and previous central nervous system insults. Regarding the EEG study, we recorded its indications, background activity, the presence of delta activity other that OIRDA, the type and location of epileptiform activity, if any, and the patient's mental state when OIRDA occurred. The frequency and duration of OIRDA bursts were also noted, as well as any clinical changes that may have taken place when this rhythm appeared. All EEG studies were analyzed by the same interpreter (N.W.), who was not blinded as to the indications for obtaining the test. The presence of OIRDA and its characteristics was already noted during the initial EEG reading. The characteristics of OIRDA were recorded by visual analysis based on 1-s intervals.

Epilepsy history and epilepsy syndrome diagnosis were noted for patients with epilepsy. Epilepsy syndrome diagnosis was made, when applicable, according to the International League Against Epilepsy (ILAE: epileptic syndromes and related conditions, www.ilae-epilepsy.org/Visitors/Centre/ctf/syndromes.cfm, 2006). The presence or absence of concomitant illness also was recorded. When available, laboratory findings and recent or concomitant neuroimaging findings also were included in the data gathering.

RESULTS

OIRDA was detected in 24 records of 17 pediatric patients, of 697 EEG studies. The total number of EEG records reflects those studies performed on an outpatient basis and on inpatients, except for cases of real-time status epilepticus and other tracings obtained in severely ill children (no OIRDA cases were detected among these patients). All three patients whose EEG indications were related to follow-up for absence seizures had repeated EEGs depicting this delta rhythm; one of them underwent three studies within a 3-year period, all showing OIRDA. Sex distribution was 1:1. Mean patient age was 7.96 years (range, 3 to 16 years; median, 8 years). Main indications for EEG and the principal electroclinical characteristics of the EEG records are presented in Table 1. The timing of the EEG record with respect of seizure occurrence for the 15 patients with convulsions was as follows: <12 h in one case and 12–24 h in seven. Data for the remaining seven children were not available, but these were all outpatients, suggesting that at least 12–24 h had elapsed since seizure occurrence.

Table 1 also depicts the data on the frequency of the occipital rhythm delta discharge, which was available for 22 of the 24 cases (two studies in which OIRDA frequency was not originally stated could not be retrieved for technical reasons). Patients with absence epilepsy were more likely to depict faster occipital delta: all five cases with OIRDA frequency of 3–4 Hz (of a total of seven absence seizures–related EEGs) corresponded to children with absence epilepsy, whereas all cases with localization-related epilepsies showed an OIRDA frequency of 2–3 Hz (Fig. 1). Most occipital rhythmic delta discharges (16 of 24) lasted <5 s. No correlation was found between occipital rhythmic delta frequency or duration and patient state (awake or asleep).

Concomitant, independent epileptiform activity was present in 12 of the 24 studies. Although no particular pattern of distribution was noted (Table 2), the epileptiform discharges were focal in all but one case. Spike–wave complexes constituted the most common pattern, irrespective of their location. Epileptiform activity intermixed with the occipital rhythmic delta in six cases; five of them belonged to children with absence epilepsy; the sixth study pertained to a boy with Angelman syndrome and also depicted frontal intermittent rhythmic delta activity. In two studies, the occipital rhythmic delta appeared only during hyperventilation. This procedure also was associated with enhancement of the rhythmic delta in three further cases. Occipital intermittent rhythmic delta activity was bilateral and synchronous in 22 of the 24 cases. Frontal intermittent rhythmic delta activity was detected in three cases: a 7-year-old girl with acute absence exacerbation and enlargement of the right sylvian fissure, an 8-year-old girl with a right white matter lesion of undetermined origin, and a 4-year-old boy with Angelman syndrome.

No patients had acute illness. Eight patients had concomitant computed tomography (CT) or magnetic resonance imaging (MRI), most depicting no abnormalities or incidental findings unrelated to the indications for EEG. Two of the cases with concomitant frontal intermittent

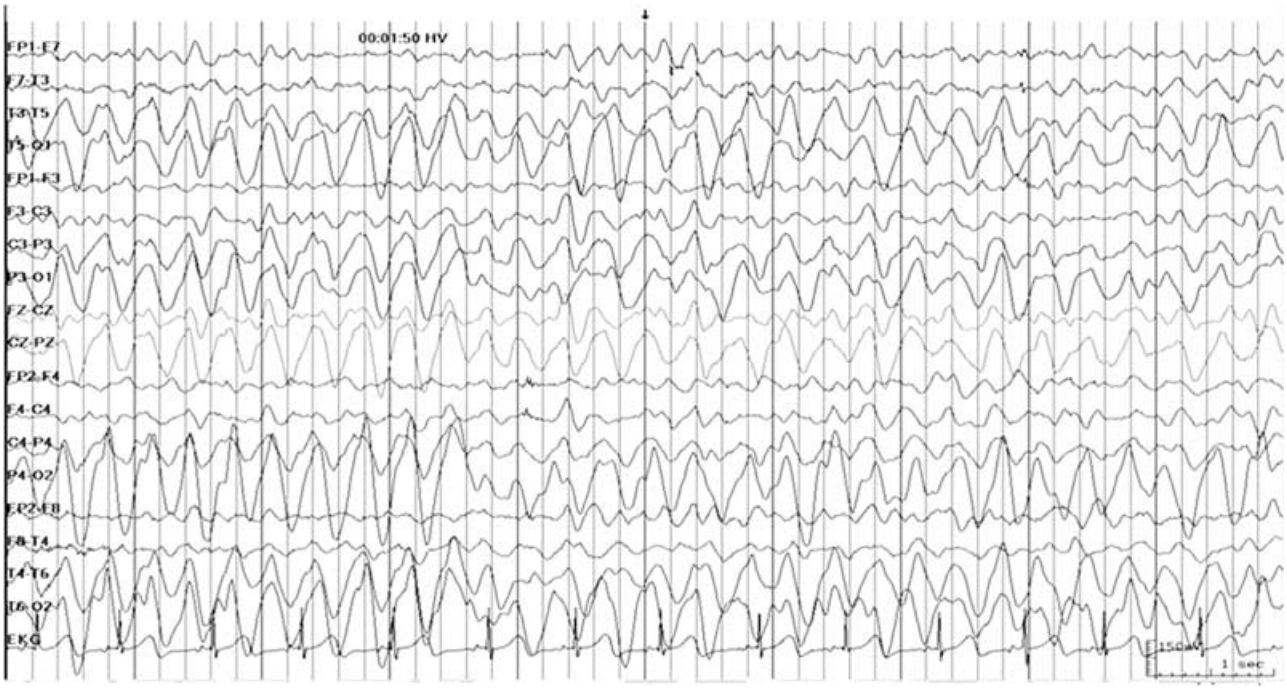

FIG. 1. EEG sample of a five-year-old boy with childhood absence epilepsy. During hyperventilation, long runs of 3 Hz OIRDA appear.

rhythmic delta activity had structural brain findings probably unrelated to the occipital rhythmic delta: in one case, the MRI showed a prolonged T_2 signal in the right frontal white matter; in the second study, right sylvian fissure enlargement in a child with absence seizures was noted.

DISCUSSION

Initially, the significance of occipital intermittent rhythmic delta activity was thought to be similar to that of frontal intermittent rhythmic delta activity. However, early publications focused mostly on the frontally located rhythm (FIRDA), whereas the occipital equivalent was rarely studied. Frontal intermittent rhythmic delta activity was considered for years to arise from posterior fossa and midline tumors (Cobb, 1945). As modern neuroimaging techniques such as CT and MRI were developed, it became obvious that this assumption was erroneous (Scollo-Lavizzari and Matthis, 1981; Fariello et al., 1982; Watemberg et al., 2002). Indeed, one study introduced the notion of "zeta" waves. These sharply contoured biphasic delta waves appear on different locations and seem to be highly predictive of structural brain lesions. However, the zeta waves are easily distinguished from the sinusoidal or sawtooth pattern seen in intermittent rhythmic delta activity (Dunne and Silber, 1991). Frontal intermittent rhythmic delta activity is probably related to metabolic derangement, particularly in the presence of previous ischemic brain insults (Watemberg et al., 2002) in adults. In children, the significance of the frontal rhythm is still unclear, although it is not associated with acute encephalopathy and does not appear to be epileptic in nature (Watemberg et al., 2003).

TABLE 2. *EEG localization of independent epileptiform discharges on studies depicting occipital intermittent rhythmic delta activity*

Epileptiform activity	Number of cases
Frontal	3
Frontal and occipital	2
Centrotemporal	2
Occipital	2
Temporal	2
Generalized	1
Total	12

The difference in the EEG location of both frontal and occipital intermittent rhythmic delta activity was previously considered to be age-related expressions of the same pathophysiologic process, with adult patients showing frontal predominance, whereas the occipital area was more commonly involved in children (Cobb, 1945; Sharbrough, 1987). However, recent studies have shown that OIRDA is probably an epileptiform pattern. Riviello and Foley, studying intermittent rhythmic delta activity in pediatric patients, found that the location of the rhythm was occipital in 87% of cases, and that 81% of patients had active epilepsy, mostly idiopathic generalized epilepsy. Moreover, epileptiform discharges were present in 70% of the records (Riviello and Foley, 1992).

Another study compared 77 EEG records depicting OIRDA and the medical records of the patients with those of matched controls. All but one case were children younger than 18 years; the only adult patient in this series was also young (22 years). Seizures were significantly more frequent among OIRDA patients, their EEG tracings were more likely to depict spike-and-wave complexes, and generalized tonic–clonic and absence seizures were the most prominent seizure types among patients with OIRDA. Mean frequency of the occipital rhythm was 2.89 Hz. Of note, the pattern was unilateral in 21% and enhanced by hyperventilation in more than one half of the cases. Moreover, it intermixed with epileptiform discharges in 30% of the epilepsy patients, compared with 12.5% of patients without seizures (Gullapalli and Fountain, 2003).

Our findings support the notion that OIRDA is a pediatric EEG pattern associated with epilepsy. However, we found some differences in the clinical characteristics of the patients as well as on the EEG findings: Whereas the majority of our cases underwent an EEG study for recent convulsions, most patients had localization-related epilepsy. In these cases, OIRDA did not intermix with epileptiform activity, and it was not time related to seizure occurrence. Conversely, most of the records (five of seven) of the four patients with absence seizures depicted epileptiform discharges ("occult spikes") intermixed with OIRDA, and the frequency of the rhythmic delta activity was faster than that seen in cases with localization-related epilepsy. This finding suggests that, although OIRDA is certainly associated with epilepsy, the electrographic characteristics of OIRDA may differ between focal and primary generalized epilepsies. Paroxysmal posterior rhythmic delta activity was detected specifically in patients with atypical absence seizures, who also had a higher incidence of focal absence seizure onset (Holmes et al., 1987). Only one of the four patients with absence seizures in our series had atypical absences. Although the number of patients is small, our findings suggest that OIRDA also occurs in association with typical absence seizures.

Contrary to Gullapalli's work (Gullapali and Fountain, 2003), we did not detect any case in which the occipital delta rhythm evolved into an absence seizure. This difference may be related to the small number of patients in our series. However, the authors did not elaborate on the features of the absence epilepsy cases and the relative frequency of absence seizures in their series. Hence, whether OIRDA preceding the seizure may be a characteristic of typical absence epilepsy or rather occur in association with atypical absences is not clear.

Although the association between OIRDA and epilepsy seems well established, the occurrence of this pattern in encephalopathic patients is still occasionally reported. This electrographic pattern was the initial finding suggesting CNS involvement in a patient with CNS salmonella infection (Buoni et al., 2005). It also was detected in a 3.5-year-old girl with juvenile Huntington disease and seizures. In this case, the EEG also depicted occipital spikes (Ulrich et al., 2004). One patient in our series had progressive encephalopathy when the EEG demonstrated OIRDA. This 11-year-old boy had subacute sclerosing panencephalitis and eventually died of the disease. The occipital delta rhythm was noticed in one of four EEG studies performed during the 4-month period preceding his death, when the pace of neurologic deterioration increased. The study depicting OIRDA was obtained at the beginning of this 4-month period.

In summary, OIRDA is found almost exclusively in children. This finding is probably epileptiform in nature, as suggested by our series and most published studies. However, this pattern does not appear to be pathognomonic of epilepsy, and it may be occasionally encountered in encephalopathic patients. Previous studies have found that most children whose EEGs depicted OIRDA had primary generalized epilepsy. However, we detected a higher proportion of cases with localization-related epilepsy. Furthermore, the frequency of the OIRDA discharge appears to be higher when it occurs in association with absence epilepsy, and these cases are more likely to depict epileptiform activity intermixed with the rhythmic delta pattern than are cases of focal epilepsy.

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