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Benign Childhood Epilepsy Syndromes

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Abstract

Epileptic syndromes usually carry a devastating outcome and poor prognosis; patients usually present with epileptic encephalopathy, delayed milestones, multiple comorbidities and refractory seizures, although not all epileptic syndromes behave the same, among the big group of children with epilepsy who do not fit within any of the currently recognized syndromes, epilepsy in many children follows a benign course.

Keywords: benign, epilepsy, syndromes, Rolandic, Panayiotopoulos, Gastaut

1. Introduction

International League Against Epilepsy (ILAE)'s definition of a benign epilepsy syndrome is "a syndrome characterized by epileptic seizures that are easily treated, or require no treatment and remit without sequelae". Many children who have epilepsy syndromes that are not usually considered benign, nevertheless, have a good outcome. Many children with congenital hemiplegia who develop focal seizures have only a few seizures, respond well to medication, and eventually successfully discontinue treatment [1].

Benign epilepsy syndromes have been classified according to age of onset into three types: benign neonatal epilepsy syndromes with onset during early neonatal period; benign infantile epilepsy syndromes with onset outside the neonatal period and before first birthday; and benign childhood epilepsy syndromes with its subclassification; these usually occur in children older than 1 year [2].

1.1. Benign neonatal epilepsy syndromes

Most of early-onset neonatal convulsions are characterized by their poor prognosis, except for fifth-day seizures, which carry an excellent outcome. Exclusion of toxic, metabolic, hypoxic-ischemic insult and structural abnormalities is a must prior to diagnosis of benign familial neonatal convulsions. Benign epilepsies in the neonatal period are generally characterized by good prognosis and long-term favorable outcome.

1.2. Benign infantile epilepsy syndromes

Benign infantile epilepsies, with presumed genetic origin, with an increased risk of epileptic seizure disorders are present in children with positive family history of benign focal epilepsies [3].

1.3. Benign childhood epilepsy syndromes

Benign childhood epilepsies are present in about one fourth of children with afebrile seizures. They are also called benign childhood focal seizures and related idiopathic epilepsies, which are subclassified into three types: epilepsy with centrotemporal spikes (CTSs) or “Rolandic” epilepsy, Panayiotopoulos syndrome with its two forms, namely early-onset benign childhood occipital epilepsy and late-onset idiopathic childhood occipital epilepsy (Gastaut type), and idiopathic photosensitive occipital lobe epilepsy of childhood [2].

2. Epilepsy with centrotemporal spikes “Rolandic epilepsy” versus “benign childhood epilepsy with centrotemporal spikes (BECTS)”

2.1. Definition

In 1958, Beaussart was the first to recognize benign focal epilepsies. Lombroso established its recognition in the United States later on. It is now considered the single most common epilepsy syndrome after febrile seizures [4].

The term “Rolandic Epilepsy” is widely used by pediatricians; BECTS was so labeled as centrotemporal spikes (CTSs), which are more predominantly seen in the central (rolandic) fissure; nevertheless, infrequent or even rare spikes may be located in the temporal electrodes bilaterally [5].

The presence of CTS remained a prerequisite for diagnosis of BECTS, though many studies showed that BECTS may take place without CTS and on the contrary some children may show CTS with no history of seizures and it may be encountered in children with other benign focal epilepsies. The word “temporal” may be mistakenly linked to manifestations similar to those taking place in temporal lobe seizures, though this has not been suggested as a characteristic feature of BECTS [5].

2.2. Epidemiology

Age of onset ranges between 1 and 15 years. Up to 75% of cases have onset of seizure between 7 and 10 years. There is marked male predominance with boys being more often affected than girls, with a ratio of 3:2.

2.3. Clinical manifestations

Rolandic epilepsy, being idiopathic, is expected to occur in children who are otherwise normal, though recent studies showed that the presence of excessive spike and wave epileptiform discharges has been linked to learning difficulties and lower school performance.

They are characterized by hemifacial sensorimotor symptoms, oro-pharyngo-laryngeal manifestations, speech arrest and hypersalivation. Secondary generalization is frequent. From all these multiple seizure types, the most recognizable form remains facial symptoms in the form of oro-pharyngo-laryngeal symptoms in addition to hemifacial motor, sensory or more frequently combination of sensorimotor symptoms which is encountered in more than 30% of patients usually in the form of unilateral funny feeling with paresthesia and numbness associated with hemifacial twitches involving tongue, inner cheeks, teeth, gums, pharynx and larynx.

Sensory manifestations of the hemifacial seizures are usually described as numbness or tingling sensations in the corner of the mouth. Speech arrest is very common in Rolandic seizures. The child with speech arrest will often try to communicate using other means, such as gestures. Aphemia (motor aphasia) and aphonia (inability to produce sound as a consequence of laryngeal dysfunction) have been reported. Hypersalivation is reported in around one-third of cases. Ictal syncope has also been recently reported as a rare feature of Rolandic seizures. Panayiotopoulos has argued that it is an anarthria or dysarthria (implying an articulatory disturbance). In support of this, it appears to occur equally in left and right-sided seizures.

Motor manifestations start with abrupt, sustained or episodes of focal clonic contractions involving the lower limb, with deviation of the ipsilateral angle of the mouth, and less frequently, widespread motor components to involve the ipsilateral upper limb. These motor manifestations are responsible for gurgling, grunting and guttural noises, sometimes producing a so-called death rattle.

In more than one-half of Rolandic seizures, consciousness is retained throughout the seizure, such that the child can often give a vivid description of his or her experiences after the seizure. Spread and secondary generalization (generalized tonic clonic seizures [GTCSs]) is much more likely with seizures occurring in sleep; Rolandic epilepsy is one of the commonest causes of nocturnal GTCS in children, exceptionally associated with GTCS when awake.

Seizures are usually brief, mostly lasting 1–2 minutes and may be longer if they progress to GTCS. Focal motor and hemiconvulsive status epilepticus (SE) are well described. Hemiconvulsive SE is more common in younger children, may be followed by Todd's paresis, usually

sparing the face. Opercular status is encountered as part of the atypical evolution sometimes encountered in the syndrome. Convulsive SE is exceptional.

2.4. Diagnostic evaluation and differential diagnosis

There is controversy regarding the need of investigations for cases with BECTS. Most investigations, other than EEG, are expected to be normal. Neuroimaging is not required if the clinical presentation, taking into account age, the absence of any comorbidities, seizure semiology and EEG features, is typical.

Neuroimaging, preferably MRI is considered a necessity in the presence of atypical features, abnormal neurological exam, developmental regression or abnormal disease course. MRI brain may show structural brain lesions involving the rolandic regions, coincidental finding of other abnormalities, especially white matter changes and hippocampal abnormalities have been reported, though their significance remains an area for further studies.

2.4.1. EEG

Interictal EEG shows high amplitude sharp and slow wave foci, which are usually localized to the central or centrottemporal electrodes. Three-quarters of seizures occur in non-rapid-eye movement (non-REM) sleep, usually shortly after sleep commences or before wakening [6].

The EEG trait characteristic of Rolandic epilepsy is considered to be inherited in an autosomal dominant manner. The interictal EEG in children with Rolandic epilepsy usually shows a normal background but with the hallmark centrottemporal/Rolandic spikes, complexes of high amplitude epileptiform sharp and slow waves are mainly seen in the central electrodes (C3/C4) or in between central and temporal electrodes (C5/C6). EEG findings can be seen unilateral or bilateral with or without secondary generalization; synchronous as well as asynchronous reports have been delineated [7].

Various studies of CTS utilizing EEG single- or multiple-dipole modeling computerized techniques showed that modeling of the negative spike component can be done by a single and stable tangential dipole source along the central (rolandic) region, the negative pole is maximally delineated in the centrottemporal region while the positive pole is maximally delineated in the frontal regions.

CTSs are highly activated by sleep with preservation of normal sleep architecture. The interictal EEG be normal in rare settings. Trivial findings in the form of subtle background slowing most likely considered as a postictal effect, antiepileptic medication effect, or may be seen if CTSs are particularly abundant. Extracentral sharp and slow wave complexes such as occipital, parietal, frontal and midline regions may occur concurrently with CTS. They are of similar morphology to CTS. Rarely, generalized discharges occur. An unusual feature of the EEG apparent in some children with rolandic epilepsy is the provocation of extreme somatosensory evoked spikes (giant somatosensory evoked potentials) in the contralateral hemisphere by tapping of the fingers or toes [6].

CTS maximally seen at the age of 8–9 years, incidence of 2–3% in normal children and even in children with neurological deficits without history of seizures or epilepsy. There is no link between CTS' frequency, location, persistence and clinical scenario, manifestation, progression of seizures or long-term prognosis. Before the onset of the ictal discharge, CTSs become sparse. There are very few ictal EEG recordings of Rolandic seizures. The ictal discharge consists of unilateral slow waves intermixed with fast rhythms and spikes located in the central regions [6].

Marked EEG abnormalities over many years may not correlate with the clinical condition; some patients will be entirely normal with markedly abnormal EEG abnormalities without any linguistic or neuropsychological deficits and good school achievement.

2.4.2. Magnetoencephalography (MEG)

MEG shows that the dipoles of the prominent negative sharp waves of rolandic discharges appear as tangential dipoles in the central (rolandic) region, with positive poles being situated anteriorly. In rolandic epilepsy, equivalent current dipoles of spikes are located and concentrated in the rolandic regions and have regular directions [6].

2.5. Treatment

Prognosis is not influenced by regular AED treatment. Frequent or diurnal seizures are considered a reasonable indication to start regular antiepileptic medications. Sudden unexplained death in epilepsy (SUDEP) will remain a worry for many parents who will consider regular medications.

Seizure control is easily reached with monotherapy in most of the cases, though recent studies had raised the possibility that it is the secondary generalized tonic clonic seizure (GTCS) that is controlled not the focal ones as previously assumed. There is a lack of strong evidence-based protocol for choosing a first or second line antiepileptic, though the most commonly used medication is still carbamazepine, with oxcarbazepine, valproate and levetiracetam considered as second line choices [8].

2.6. Course and prognosis

Most cases with rolandic epilepsy will have less than ten seizures, single seizures are common. Up to 20% will have frequent seizures, especially at the start of the disorder. The prognosis, however, remains excellent, with remission within 1–2 years of onset and certainly before late adolescence. Incidence of occurrence of GTCS is more or less comparable to normal population (<2%). Patients with rolandic epilepsy are still at higher risk of developing absence seizures later on. Despite being labeled as benign seizures, rolandic epilepsy has been associated with multiple neurodevelopmental effects, cognitive impairment and learning difficulties mainly demonstrated as a consequence of CTS; this has been an area of research and studies focusing on the difference between ictal and interictal EEG abnormalities and their overall effect on the patient even in the absence of clinical seizures. Some patients with BECTS will suffer from

multiple speech, reading, behavioral and sleep problems, it is controversial if regular antiepileptic medications should be used in such scenarios and their usefulness to control or even reverse these conditions keeping in mind the side effects of AEDs themselves should be taken in consideration when deciding to start regular antiepileptic medications.

Less than 1% of children with rolandic epilepsy have the so-called atypical evolutions. These include the development of severe linguistic, cognitive or behavioral problems. If such problems develop in a child with rolandic epilepsy, a sleep EEG should be obtained, because continuous spike-and-wave during slow-wave sleep (CSWS) may be present. Landau-Kleffner syndrome is sometimes said to develop from rolandic epilepsy. Atypical focal epilepsy of childhood in which other seizure types, including tonic and atypical absence seizures, may also develop in children with otherwise typical rolandic epilepsy. CSWS may also be seen in children with opercular status characterized by continuous positive or negative myoclonias around the mouth or elsewhere in the face and pseudobulbar problems [9, 10].

3. “Panayiotopoulos syndrome” or “early-onset benign childhood occipital epilepsy (Panayiotopoulos type)”

3.1. Introduction and definition

In 1973, this syndrome was first described by Panayiotopoulos in a 30-year prospective study. After rolandic epilepsy, it is the most common of the benign focal epilepsy syndromes of childhood, which is manifested with autonomic seizures (epileptic seizures characterized by altered autonomic function occurring at the onset of the seizure or as the sole manifestation of the seizure). It is likely to be misdiagnosed as a nonepileptic disorder [11].

Panayiotopoulos syndrome seizures manifest with emesis-like symptoms with retching, nausea, with or without other autonomic symptoms. Episodes of syncope-like behavior have been frequently reported. Seizures are usually long lasting. Interictal EEG usually showed focal high amplitude sharp and slow epileptiform discharges with shifting predominance throughout various cortical areas.

3.2. Epidemiology

Panayiotopoulos syndrome cases vary between late infancy where cases present in the late infancy period and the age of early adolescence at the age of 14 years. However, most cases lie between preschool age (3–6 years) with peak onset age (4–5 years). Boys and girls are equally affected. Prevalence is 6% (1–15 years) and 13% in those (3–6 years). Therefore, a clinician might expect to see at least one case of Panayiotopoulos syndrome for every three cases of rolandic epilepsy [12].

3.3. Clinical manifestations

Panayiotopoulos syndrome is an idiopathic epilepsy syndrome; it occurs in children who are otherwise normal. No specific precipitants can be identified.

Two-thirds of seizures occur during sleep, including daytime naps, and they usually begin with emetic symptoms (nausea, retching with or without vomiting). Some mothers will find their children retching or even vomiting in bed, if seizures start during night sleep, though most of the kids will wake up complaining of nausea and tendency to vomit.

During wakefulness, seizures usually commence with feeling of nausea, mostly associated with behavioral changes, irritability and agitation. Repetitive vomiting is less frequently encountered throughout the seizure onset, usually over many hours. In rare occasions, seizures in Panayiotopoulos syndrome can happen without any feature of the classic “emetic triad”. Pallor as well as various autonomic manifestations can concomitantly take place; pupillary abnormalities (mydriasis usually more frequently encountered than miosis), cyanosis and flushing are less common. Urinary and occasionally fecal incontinence may occur [11].

Hyperthermia may be encountered during or immediately after a seizure and may represent a true ictal symptom, rather than being a precipitant of the seizure. Tachycardia is certainly a feature of seizures recorded on EEG with simultaneous ECG recording. Cardiorespiratory arrest during a typical seizure of Panayiotopoulos syndrome has been reported in few case reports. Rarer ictal symptoms that have been reported include headache and other “cephalic sensations,” hypersalivation and coughing. Breathing changes are sometimes reported, particularly before convulsions. Episodes of ictal syncopal-like attacks infrequently occur, which have been assumed to result from brief cortical hypoperfusion.

Seizures usually commence during full wakefulness, they start as simple partial seizure, shortly after that, consciousness will be impaired with emergence of a complex partial seizure with some preservation of ability to respond to external stimuli. Behavioral changes in the form of restlessness, agitation and terror, being apparently reported in full consciousness. The occurrence of autonomic manifestations like pallor, while the kid is atonic and unresponsive, raises the possibility of syncope. Seizures often end in hemiconvulsions (in about 20% seizures) or GTCS (also in about 20% seizures). 44% seizures that lasted for 30 minutes or more (maximum is 7 hours) represent a form of nonconvulsive status epilepticus and might reasonably be classified as autonomic status epilepticus, may end spontaneously or as short hemiconvulsions/GTCS and returned to normal within a few hours of such episodes. Convulsive status epilepticus is exceptional. The mean duration is around 9 minutes.

Initial reports of Panayiotopoulos syndrome described lateralizing manifestations such as aversive movement of the head and, as an initial manifestation, eyes to one side; although this feature is not consistent over subsequent reports, data re-evaluation confirmed that is a common feature that occurs when consciousness is impaired, which usually takes place sometime after seizure starts.

3.4. Diagnostic evaluation and differential diagnosis

Panayiotopoulos syndrome is frequently mistaken for paroxysmal nonepileptic disorders and occasionally for other types of epilepsy; this is mainly due to its peculiar ictal clinical features and the presence of abnormal interictal EEG changes associated with Panayiotopoulos syndrome.

Seizures in Panayiotopoulos syndrome are usually prolonged and it is not uncommon that many patients will present to emergency room during a clear ictal state; it is sometimes confusing for the attendant physician especially if the presentation is with impaired level of consciousness and vomiting to consider epileptic seizure in the differential diagnosis. Top differential diagnoses will be encephalitis, meningitis and herpetic meningoencephalitis; this will lead the patient to the intensive care unit with broad spectrum antibiotics and antiviral management.

Long-lasting seizures may also be confused with acute confusional migraine and, if vomiting is particularly prominent, with cyclical vomiting syndrome or gastroenteritis. Some seizures may simply be dismissed as travel sickness.

Children with Panayiotopoulos syndrome commonly present to emergency departments while still seizing or in the immediate postictal period.

Panayiotopoulos syndrome should be considered in the differential diagnosis of all previously well young children, (3–6) years, who have rapid onset of emetic symptoms followed by impaired (often fluctuating) consciousness. Eye or head deviation may be a useful finding. However, it may still be appropriate to manage the child for a suspected encephalopathy.

If Panayiotopoulos syndrome is suspected from the history, the most useful investigation is likely to be the EEG (including sleep study if necessary). Symptomatic epilepsies may mimic Panayiotopoulos syndrome, so even if the history is typical, most authorities recommend neuroimaging.

3.4.1. EEG

Interictal EEG usually shows normal background with high-amplitude sharp, sharp and slow-wave foci (sometimes labeled as functional spikes), which is similar, in morphology; but not in location, to those seen in rolandic epilepsy. The EEG abnormalities in Panayiotopoulos syndrome are accentuated in sleep. Cloned-like, repetitive and multifocal spike wave complexes in which repetitive spike or sharp and slow wave complexes appear concurrently in different brain locations of one or both hemispheres.

Previously the occipital location of these changes was emphasized, along with their occurrence in long trains (occipital paroxysms) and abolition by central fixation (fixation-off sensitivity). Although fixation-off sensitivity is common, photosensitivity is exceptional. It now appears that these features were overemphasized, and in Panayiotopoulos syndrome the characteristic functional spikes can occur in multiple locations, albeit with a posterior predominance. Focal or diffuse slowing may be seen postictal. However, spikes' frequency, distribution and persistence are not time-locked to clinical manifestations, progression, seizures' frequency or intensity and are not consistent with prognosis of Panayiotopoulos syndrome.

Various ictal EEG changes have been reported; trains of focal rhythmic delta or theta activity associated with the presence of epileptic spikes. These changes used to be delineated mainly in the posterior areas, but may be anterior. Rarer EEG findings: small, spikes, slow waves

intermixed with small spikes and brief generalized discharges. Occasionally, repeated EEGs can all be normal.

In some cases, EEG findings of patients with Panayiotopoulos syndrome may be misleading and may lead to wrong diagnosis. EEG findings may look similar or even identical to those of other benign epilepsies such as BECTS or Gastaut idiopathic childhood occipital epilepsy, which is highly possible is higher if proper clinical history is not obtained. Multifocal spike discharges and bursts of repetitive multifocal spike wave complexes within the delta range may look similar to EEG findings associated with some malignant epilepsies such as the Lennox-Gastaut syndrome, though they run a totally different clinical course.

3.4.2. MEG

MEG study for patients with Panayiotopoulos syndrome usually shows equivalent current dipoles of spikes concentrated in the rolandic regions and occipital areas. In MEG combined with MRI, equivalent current dipoles cluster preferentially in cortical locations along the parietal-occipital, the calcarine or the central fissure. The equivalent current dipole clustering may be unilateral or bilateral, monofocal or multifocal. These findings are in keeping with the condition being a multifocal rather than a purely occipital epilepsy. The directions of each equivalent current dipole in each area are quite regular as if three small round toothbrushes are placed in each of the three areas.

3.4.3. MRI

MRI is considered a better modality than CT scan. However, if MRI will require sedation or general anesthesia, CT may be appropriate.

3.5. Treatment

Regular prophylactic AED medication is usually reserved for patients with frequent seizures, which distressed them and their families or even interfere with their daily living activities and regular life style. There is a lack of well-structured study to suggest the first line AED to be used. Carbamazepine and sodium valproate are considered equally efficacious, though carbamazepine was found to exaggerate seizures in a minority of children with PS [13].

Panayiotopoulos syndrome is considered a benign syndrome, so it is important to try to avoid side effects of various AEDs. It is considered a good practice to try to wean AED after 2 years if the child remained seizure-free throughout this period; it is still controversial whether or not to have a normal EEG prior to that as clinical improvement may be reached prior to electrographical one. I think that the emergence of association between Panayiotopoulos syndrome and cardiorespiratory arrest will be a valid indication to continue AED for longer duration; this area still needs further studies.

3.6. Course and prognosis

Seizures in Panayiotopoulos syndrome are considered infrequent. 30% of cases with Panayiotopoulos syndrome will encounter only one seizure and only 5–10% will have more frequent seizures reaching more than 10, though sometimes seizures are very frequent. One-fifth will have one or more seizures typical of one of the other benign focal epilepsies of childhood, especially rolandic epilepsy [14].

The duration of active seizures is short; remission usually occurring within 1–2 years from onset. Seizures in adult life are no greater than in the general population [14, 15].

There are few case reports of atypical evolutions in Panayiotopoulos syndrome, including the development of absences and drop attacks [1, 14].

4. Idiopathic childhood occipital epilepsy (late-onset childhood occipital epilepsy, “Gastaut type”, and idiopathic photosensitive occipital lobe epilepsy)

4.1. Introduction and definition

“Idiopathic childhood occipital epilepsy” includes both late-onset childhood occipital epilepsy –Gastaut type and idiopathic photosensitive occipital lobe epilepsy. This is because both conditions share many common features and it is not clear that they merit separation into two distinct syndromes [1, 16].

Idiopathic childhood occipital epilepsy with and without photosensitivity was first established as an epileptic syndrome by Gastaut. Recently, it has been classified separately by the ILAE Task Force as late-onset childhood occipital epilepsy (Gastaut type) and idiopathic photosensitive occipital lobe epilepsy. Idiopathic childhood occipital epilepsy can be defined as an idiopathic focal seizure disorder of childhood manifested mainly by elementary visual seizures and ictal blindness, which are often frequent and usually occur without impairment of consciousness [1, 16].

Idiopathic photosensitive occipital epilepsy is an idiopathic focal seizure disorder mainly of childhood manifested mainly by elementary visual seizures provoked by various forms of environmental light stimulation [1, 16].

The likelihood of remission in these syndromes is considerably less than rolandic epilepsy and Panayiotopoulos syndrome. Some children with these conditions remit completely [1, 16].

4.2. Epidemiology

Idiopathic childhood occipital epilepsy usually starts at toddler age group in children as young as 3 years and extends to involve adolescents as old as 15 years of age. Peak age of onset is around 8 years. Boys and girls are equally affected. Furthermore, idiopathic photosensitive occipital epilepsy may start as early as the second year of life or as late as young adult life.

However, it peaks at around 12 years of age. There is probably a slight female preponderance, but nowhere near as great as for photosensitivity alone [16].

Panayiotopoulos estimated that idiopathic childhood occipital epilepsy accounted for about 2–7% of all benign focal epilepsies of childhood.

4.3. Clinical manifestations

Idiopathic childhood occipital epilepsy hallmark is visual hallucinations which the patient described as small multicolored patterns that run in circles, most commonly occurring unilaterally in the peripheral visual field; they increase in size and number with seizure progression, sometimes moving horizontally across the visual field, with or without other more complex movements. Normal vision may be hindered by these hallucinations, but in others it may not. Visual illusions as shape and distance distortions have been reported; even more complex visual hallucinations as formed shapes and faces may be encountered, though they are less frequent [12, 16].

Ictal blindness is considered the second most common complaint reported by patients with idiopathic childhood occipital epilepsy after visual hallucinations. Ictal blindness is usually bilateral, but may be unilateral or even partial involving less than one half of a visual field. It usually occurs in the form of sudden black outs, though others will describe that everything goes white. Ictal blindness is usually an initial seizure manifestation but sometimes follows visual hallucinations. Other ictal ocular symptoms are relatively common, e.g., ocular pain and ictal eye deviation, often with simultaneous head deviation in about 70% of cases. Forced eye closure and eyelid blinking are other reported phenomena [12, 16].

Seizures are usually brief, lasting for few seconds, though prolonged seizures lasting for few minutes have been described, mainly those seizures with ictal blindness which may last for even hours; this has been described as “status amauroticus” [12, 16].

Headache has been described as a common manifestation in idiopathic childhood occipital epileptic seizures (post ictal > ictal). It often has a migrainous character. Autonomic manifestations have been reported in idiopathic photosensitive occipital lobe epilepsy, e.g., vomiting which characterizes Panayiotopoulos syndrome [12, 16, 17].

Patients with idiopathic childhood occipital epilepsy usually maintain their consciousness during most seizures, though occasionally impaired level of consciousness will be reported in cases of secondarily generalization with occurrence of generalized tonic clonic seizures. Occurrence of temporal lobe symptoms has been reported and is considered to result from local spreading of epileptiform discharges.

Seizures usually occur in day time with high frequency; multiple seizures take place on a daily or sometimes weekly basis. Nocturnal seizures with hemiconvulsions or even generalized tonic clonic seizures are not uncommon [12, 16].

Photosensitivity in idiopathic photosensitive occipital lobe epilepsy is significant, as seizures may be provoked by variable light sources, e.g., video-games, watching television. Photosensitivity ranges between different subjects; some will have high photosensitivity resulting in

high seizure frequency while others with less photosensitivity will have few seizures. Nevertheless, spontaneous seizures without photosensitivity have also been reported. Some cases with idiopathic childhood occipital epilepsy have developed absences and myoclonic jerks provoked by photic stimulation [12, 16].

4.4. Diagnostic evaluation and differential diagnosis

These syndromes, like all occipital epilepsies, are very prone to misdiagnosis as migraine. However, the elementary visual hallucinations are unlike those of migraine. In the latter they tend to be black and white, rather than colored, and have jagged or sharp contours rather than being predominantly rounded. They may mimic symptomatic occipital lobe epilepsies, and neuroimaging, preferably MRI, is indicated [12, 16].

4.4.1. EEG

In late-onset childhood occipital epilepsy ‘Gastaut type’, interictal background is normal, occipital paroxysms are characteristic and isolated occipital spikes may be seen. Extraoccipital paroxysmal abnormalities may occur, but are much less common than in Panayiotopoulos syndrome [12, 16].

The ictal EEG is expected to show attenuation of occipital paroxysms followed by appearance of an occipital discharge of fast rhythms, fast spikes or both.

In some subjects, EEG abnormalities may only be seen in sleep; occasionally both awake and sleep EEGs may be consistently normal [12, 16].

In idiopathic photosensitive occipital lobe epilepsy, interictal EEG is expected to have a normal background. No spontaneous epileptiform abnormalities or else there may be occipital spikes or paroxysms. Extraoccipital epileptiform abnormalities may also be seen. Intermittent photic stimulation will, in all subjects, show occipital or generalized photoparoxysmal responses [12, 16].

4.5. Treatment

Given the frequency of seizures in idiopathic childhood occipital epilepsy, including the likelihood of occasional GTCS, regular AED treatment is considered necessary in most if not all subjects. There are no controlled studies comparing alternatives, although carbamazepine appears to be most often used in subjects who are not photosensitive. Broad spectrum agents such as sodium valproate and levetiracetam, active against focal and generalized seizures and photosensitivity, would appear to be reasonable choices. However, it appears that carbamazepine, not usually considered a useful drug for photosensitivity, may sometimes be effective [12, 16].

Attempt withdrawal after two seizure-free years is associated with significant risk of relapse. Some subjects with idiopathic photosensitive occipital lobe epilepsy who are only mildly photosensitive and who do not have spontaneous seizures can remain seizure-free by avoiding precipitants. Others will require AED treatment.

4.6. Course and prognosis

Prognosis for both idiopathic childhood occipital epilepsy and idiopathic photosensitive occipital lobe epilepsy is variable. About 50–60% of Gastaut type have remission of seizures within 2–4 years of them starting. However, in a significant minority, seizures will continue into adulthood [18].

In children with idiopathic photosensitive occipital lobe epilepsy who are only mildly photosensitive and can control their exposure to relevant provoking factors, freedom from seizures may be easily achieved. For others, particularly those who are highly photosensitive, the likelihood of seizures continuing into adult life is high [18].

Atypical evolutions in idiopathic childhood occipital epilepsy with cognitive deterioration and CSWS. Carbamazepine is sometimes implicated in precipitating such atypical evolutions [18].

5. Other described benign focal epilepsies of childhood

The syndromes discussed previously are the only benign focal epilepsies of childhood currently recognized by the ILAE. However, others have been proposed and are more or less well characterized: benign childhood seizures with affective symptoms, benign childhood focal seizures associated with frontal or midline spikes, benign focal epilepsy in infants with central and vertex spikes and waves during sleep and Benign childhood seizures with affective symptoms are associated by and frequent extreme somatosensory-evoked spikes [19, 20].

5.1. Benign childhood seizures with affective symptoms

The onset is between 2 and 9 years of age. They are characterized by multiple, usually short, daytime and nighttime seizures in which the predominant symptom appears to be fear or terror, accompanied by autonomic disturbances (pallor, sweating, abdominal pain and salivation), arrest of speech and mild impairment of consciousness with automatisms [21].

Interictal EEG shows sharp and slow wave complexes similar to those in rolandic epilepsy but located in the frontotemporal and parietotemporal electrodes [21].

This is likely to be an intermediate phenotype between Panayiotopoulos syndrome and rolandic epilepsy. Benign childhood seizures with affective symptoms are associated by and frequent giant somatosensory evoked potentials. Remission in 1–2 years from onset is expected [21].

This putative disorder is mainly defined by its interictal EEG features reflected in its name. These features are, however, said to often be associated with a phenotype characterized by mainly daytime versive seizures, which are infrequent and have an excellent prognosis [21].

5.2. Benign childhood focal seizures associated with frontal or midline spikes

This putative disorder is mainly defined by its interictal EEG features. These EEG features can be seen in children with febrile seizures, rolandic epilepsy, Panayiotopoulos syndrome and idiopathic childhood occipital epilepsy.

5.3. Benign focal epilepsy in infants with central and vertex spikes and waves during sleep

Benign focal epilepsy in infants with central and vertex spikes and waves during sleep has been recently described as a new benign syndrome [22].

It lies in the transition zone between benign infantile seizures and Panayiotopoulos syndrome with age of onset usually in the first two years of life. Males and females are equally affected. Clinical examination, including developmental assessment, should be normal; investigations and imaging are always normal [3].

Benign focal epilepsy in infants with central and vertex spikes and waves during sleep usually starts with staring, arrest of activity, autonomic features with facial cyanosis, loss of consciousness and tonic contractions of both upper limbs; rarely clonic movements and automatisms may be encountered. Seizures usually last for less than 5 minutes, most of the time they occur during daytime, though nocturnal seizures have been described. They are usually infrequent with less than four seizures annually, but they may occur in clusters [3, 22].

Interictal EEG changes mainly encountered in non-REM sleep stages, consist of central spike and wave epileptiform discharges of low amplitude, localized to the vertex [3, 22].

Benign focal epilepsy in infants with central and vertex spikes and waves during sleep carries a favorable prognosis in families with strong history of benign epilepsies. Prognosis is excellent with more or less complete seizures remission, normal neurodevelopment and even EEG normalization before the fourth year of life [3, 22].

6. Genetics

Benign focal epilepsies of childhood do not follow simple Mendelian inheritance, which has strong concordance for idiopathic generalized epilepsies in monozygotic twins, but not for rolandic epilepsy. Autosomal dominant genetic linkage has been reported to 15q14 for BECTS [23].

Researchers found that EEG traits characterizing these disorders may surprisingly show Mendelian inheritance even if the seizure phenotype did not; this is still an area of debate and needs further studies [24].

Vadlamudi et al. found strong concordance for idiopathic generalized epilepsies in 26 monozygotic twins, but no concordance for Rolandic epilepsy in six monozygotic twins [25].

Mendelian inheritance in individual families with forms of benign focal epilepsy has been established. EEG trait characterizing these disorders may show Mendelian inheritance, even if the seizure phenotype does not [23].

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