Chapter 9  Benign Childhood Focal Seizures and Related Epileptic Syndromes

Benign Childhood Seizure Susceptibility Syndrome

Clinical note  Benign childhood focal seizures and related epileptic syndromes are the commonest and probably the most fascinating and rewarding topic in paediatric epileptology. They affect 25% of children with non-febrile seizures and form a significant part of the everyday practice of paediatricians, neurologists and clinical neurophysiologists who care for children with seizures. Rolandic seizures (RS) are widely recognised and are associated with an excellent prognosis thanks to appropriate research and publications. Paediatricians have been receptive to and have made excellent use of this knowledge. Panayiotopoulos syndrome (PS), a common disorder with dramatic clinical and EEG manifestations, eluded us until recently. PS has now been formally recognised in the new ILAE diagnostic scheme and is becoming more readily diagnosed by physicians. Less common phenotypes, such as the Gastaut type-idiopathic childhood occipital epilepsy (G-ICOE) and idiopathic photosensitive occipital lobe epilepsy have also been recognised and defined. Furthermore, there are also children who manifest with seizures of predominantly affective symptoms, and there are claims of other benign childhood seizures associated with certain interictal EEG foci, such as frontal, midline or parietal, with or without extreme somatosensory evoked spikes.

Considerations on Nomenclature

These are detailed in the individual description of each of these benign childhood focal syndromes. Overall, benign childhood focal syndromes and their main representatives, BCECTS and PS, do not fulfit the diagnostic criteria of ‘epilepsy’ defined as “chronic neurological condition characterised by recurrent epileptic seizures”. BCECTS and PS are age-limited (not “chronic”) and at least one-third of patients have a single (not “recurrent”) seizure. They should be classified among “Conditions with epileptic seizures that do not require a diagnosis of epilepsy”, which is a new concept in the ILAE diagnostic scheme to incorporate “febrile, benign neonatal, single seizures or isolated clusters of seizures and rarely repeated seizures (oligoepilepsy)” (Table 1.7).

It should also be emphasised that functional spikes of whatever location occur in 2–4% of children with or without seizures including apparently normal children without seizures and even more often children with non-epileptic neurological or medical disorders. (Figure 9.1).

All these conditions may be linked together in a broad age-related and age-limited benign childhood seizure susceptibility syndrome (BCSSS), which may also constitute a biological continuum with febrile seizures and benign infantile and neonatal seizures. It is my thesis that the clinical, EEG, pathophysiological and management aspects of BCSSS should be properly re-examined and redefined. The 1989 ILAE classification recognised three “age-related and localization-related (focal, local, partial) epilepsies and syndromes” (Table 1.5):

- Benign childhood epilepsy with centrotemporal spikes (BCECTS)
- Childhood epilepsy with occipital paroxysms (Gastaut type)
- Primary reading epilepsy

The new diagnostic scheme rightly reclassified “reading epilepsy” as a reflex epileptic syndrome (Table 1.7) and recognised three syndromes of “idiopathic childhood focal epilepsy”:

- Benign childhood epilepsy with centrotemporal spikes
- Early onset benign childhood occipital epilepsy (Panayiotopoulos type)
Late onset childhood occipital epilepsy (Gastaut type)

Benign Childhood Epilepsy with Centrottemporal Spikes (Rolandic Seizures)

Benign childhood epilepsy with centrotemporal spikes or Rolandic seizures/epilepsy is the commonest manifestation of a childhood seizure susceptibility syndrome that is age related and genetically determined.

This chapter is based on an exhaustive review of the literature regarding all aspects of Rolandic seizures and their EEG manifestations. The history of the ‘discovery of benign Rolandic epilepsy’ has been vividly described by the main French protagonists Marc Beaussart and Pierre Loiseau.

Demographic Data

Onset of RS is between 1 and 14 years; in 75% of patients, onset is between 7 and 10 years, and there is a peak at 8–9 years. There is a 1.5 male predominance. Prevalence is around 15% in children with seizures aged 1–15 years. Incidence is 10–20/100,000 children aged 0–15 years.

Clinical Manifestations

The cardinal features of RS are infrequent, often single, focal seizures consisting of unilateral facial sensorimotor symptoms, oro-pharyngo-laryngeal (OPL) manifestations, speech arrest and hypersalivation.

Hemifacial sensorimotor seizures occur in approximately one-third of patients. These are mainly motor seizures, which may be entirely localised in the lower lip manifesting with sudden, continuous or bursts of clonic contractions, usually lasting from a few seconds to 1 minute. Ipsilateral tonic deviation of the mouth is also common. More rarely, hemifacial convulsions may appear nearly simultaneously or spread to the ipsilateral upper extremity. Involvement of the leg is rare.

Hemifacial sensory seizures are less common and consist of numbness in the corner of the mouth. Consciousness is usually preserved.

Hemifacial sensorimotor symptoms may be the only ictal manifestations, but are often associated with an inability to speak and hypersalivation.

Oro-pharyngo-laryngeal ictal manifestations, which occur in more than half of seizures (53%), are the most characteristic of all other ictal symptoms of RS. They consist of unilateral sensory and motor manifestations inside the mouth, tongue, inner cheek, gums, teeth and pharyngo-laryngeal regions. Sensory symptoms manifest with unilateral numbness and more commonly paraesthesias (tingling, pricking, freezing and their variations), and are usually diffuse on one side or, exceptionally, may be highly localised to even one tooth. Motor OPL symptoms produce strange sounds, such as death rattle, gargling, grunting, guttural sounds and their combinations.

Arrest of speech is another common ictal symptom that occurs in more than 40% of RS. The child is inarticulate and attempts to communicate with gestures. A few mainly laryngeal sounds, not words, may be uttered, particularly at the beginning.
Benign childhood epilepsy with centrotemporal spikes (best known among paediatricians as Rolandic epilepsy) is defined by the ILAE as follows:  

“Benign childhood epilepsy with centrotemporal spikes is a syndrome of brief, simple, partial, hemifacial motor seizures, frequently having associated somatosensory symptoms which have a tendency to evolve into generalised tonic clonic seizures. Both seizure types are often related to sleep. Onset occurs between the ages of 3 and 13 years (peak 9–10 years) and recovery occurs before the age of 15–16 years. Genetic predisposition is common, and there is a male predominance. The EEG has blunt high-voltage centrotemporal spikes, often followed by slow waves that are activated by sleep and tend to shift or spread from side to side.”

The following should be considered in future revisions of the classification of BCECTS:
(a). Most of centrotemporal spikes are, in fact, Rolandic spikes; they are rarely located in the temporal electrodes
(b). The word ‘temporal’ is misleading because these children do not have symptoms from the temporal lobes
(c). BCECTS may occur without centrotemporal spikes and conversely centrotemporal spikes may occur in children without seizures or other clinical phenotypes of benign childhood seizure susceptibility syndrome
(d). Similar clinical features may appear in patients with spikes in other than centrotemporal locations
(e). Children with centrotemporal spikes may manifest with symptoms typical of PS.

Rolandic seizures/epilepsy may be the most appropriate nomenclature. Rolandic epilepsy is very well established and well identified by neurologists, neurophysiologists and paediatricians with this form of benign childhood focal seizures. They all understand it as a benign seizure syndrome of children with ictal symptoms, originating from a well-known anatomical region of the brain, the inferior part of the pre-and post-central gyrus.

Rolandic fissure (or central fissure) is a well-established anatomical name that can not change, though the central (or Rolandic) sulcus was probably first described by the French anatomist Vicq d’Azy and not by the Italian anatomist Luigi Rolando. Also, epileptic symptoms do not come from the Rolandic (central) fissure, but from the pre- and post-central gyrus.

In this book I use BCECTS, Rolandic seizures or Rolandic epilepsy synonymously though I would prefer the term Rolandic seizures.

There is no impairment of the cortical language mechanisms. The child is perfectly able to understand what is being said, but unable to utter a single intelligible word. Some authors call this aphemia or aphiaphonia. However, aphemia means motor aphasia or pure word mutism and does not appear to be correct, and aphiaphonia is an inability to produce sounds by laryngeal mechanisms, which also does not appear to be the case in RS. The arrest of speech in RS is more of an anarthria, that is loss of the power and coordination for the articulation of words, which also explains why this is equally common in left or right sided RS.

Patient note My right hand was numb and stiff. My mouth opened and I could not speak. I wanted to say I cannot speak. At the same time, it was as if somebody was strangling me.

She was trying to speak but only noises came out of her mouth as if her tongue was tied up in her mouth.

Some RS patients are dysarthric rather than anarthric that is they were able to pronounce some words but with difficulty as:

Patient note if there are stones in my mouth.

Hypersalivation is one of the most characteristic ictal symptoms of RS and probably occurs in as many as one-third of
cases. It is often associated with OPL symptoms, but is also associated with pure hemifacial seizures and may be the most pronounced ictal manifestation. Hypersalivation is not just frothing:

Patient note  Suddenly my mouth is full of saliva, it runs out like a river and I can not speak.

Ictal syncope may occur probably as a concurrent symptom of PS (page 237).

Patient note  She lies there, unconscious with no movements, no convulsions, like a wax work, no life.

Consciousness is fully retained in more than half (58%) of RS and the patient is able to describe the events after the end of the fits well.

Patient note  I felt that air was forced into my mouth, I could not speak and I could not close my mouth. I could understand well everything said to me. Other times I feel that there is food in my mouth and there is also a lot of salivation. I can not speak.

Secondarily GTCS are reported in between one- and two-thirds of children with RS. Primarily GTCS are not part of the syndrome of RS.

Duration of RS is usually brief, lasting for 1–2 min, but may become longer if seizures progress to convulsions.

Circadian distribution. Three-quarters of seizures occur during non-REM sleep, mainly at sleep onset or just before awakening. Seizures during sleep are usually longer and may progress to GTCS, which rarely occurs during wakefulness.

Status Epilepticus

Generalised convulsive status epilepticus is exceptional. Though rare, focal motor status epilepticus and hemiconvulsive status epilepticus are more likely to occur.

Patient note  While skating, he felt that the left side of his tongue was numb and that he could not see well. This was followed within seconds by repetitive and continuous left-sided clonic hemifacial spasms, involving the mouth and eye that ended 40 min later with left hemiconvulsions. There was postictal Todd’s paralysis.

Hemiconvulsive status epilepticus may be more common in children aged 2–5 years; this is often associated with postictal Todd’s paralysis, which generally does not include the face.

Opercular status epilepticus usually occurs in atypical evolutions of BCECTS or exceptionally it may be carbamazepine-induced. These are often associated with EEG continuous spikes and waves during slow-wave sleep (page 183). The status may last for hours to months and consists of continuous unilateral or bilateral contractions of the mouth, tongue or eyelids, positive or negative subtle perioral or other myoclonia, dysarthria, anarthria or speech arrest, difficulties in swallowing, buccofacial apraxia and hypersalivation.

Aetiology

Benign childhood epilepsy with centrotemporal spikes is genetically determined and there is evidence of linkage with chromosome 15q14. The mode of inheritance is unknown. Autosomal dominant inheritance with age-dependent penetrance refers to the EEG centrotemporal spikes, and not to the clinical syndrome of BCECTS (see review in ref1). A multifactorial pathogenesis with hereditary impairment of brain maturation has been proposed by Doose and associates. However, according to a recent study conventional genetic influences may be less important than other mechanisms, which need to be explored. This study compared the concordance of twins with Rolandic epilepsy with the concordance of a twin sample of IGEs. All eight twins (six monozygous and two dizygous) with RS were discordant. Monozygous pairwise concordance was 0 (95% confidence interval, 0–0.4) for Rolandic epilepsy compared with 0.7 (95% confidence interval, 0.5–0.9) for 26 IGE monozygous pairs.
Important note

Siblings or parents of patients with BCECTS may rarely have the same type of seizures or other phenotypes of BCSSS, such as PS (page 240). Febrile seizures are common (10–20%) prior to RS.

My view that RS are part of a BCSSS is detailed on page 262. All these benign childhood conditions are linked together through a common, genetically determined, mild and reversible, functional derangement of the brain cortical maturational process.

Pathophysiology

The ictal manifestations of RS agree well with the symptoms elicited by electrical stimulation of the lower part of the precentral and postcentral gyrus in man. Hypersalivation, like other autonomic manifestations in childhood focal seizures, is difficult to explain; it is extremely rare in adults.

Diagnostic Procedures

Apart from the EEG, all tests are normal.

Brain imaging is not needed when the diagnosis of RS is certain though 15% of patients with RS may have abnormal findings because of static or other brain diseases unrelated to the pathophysiology of RS. Further, hippocampal abnormalities have been detected in some children with RS on MRI and proton magnetic resonance spectroscopy which may be incompatible with such an age-related and benign seizure disorder.

The presence of brain lesions has no influence on the prognosis of RS.

Interictal EEG

Centrotemporal spikes are the hallmark of the syndrome of BCECTS (Figures 9.2, 9.3, 9.4 and 9.5). They are characterised by their morphology, amplitude and duration, location and field distribution, frequency and pattern of occurrence, reactivity to external stimuli and the sleep-wake cycle, as well as age-dependence and evolution.

Although called centrotemporal spikes, these are mainly high amplitude sharp and slow wave complexes localised in the C3/C4 (central) or C5/C6 (midway between central and temporal) electrodes (Figure 9.3). The main spike (sharp wave) component is diphasic with a maximum surface, negative, rounded peak that is followed by a smaller positive peak (Figure 9.2). This is followed by a negative or negative-positive slow wave. A relatively minute positive spike often precedes this spike–slow wave complex (Figure 9.2). The amplitude of the main spike (or sharp wave) component often exceeds 200 µV, though it may be much smaller or much higher. The negative phase is larger than the positive phase of the spike, as well as the preceding or following components of the spike–slow wave complex. CTS may be unilateral, but are more often bilateral, independently right or left. They are abundant (4–20/min) and usually occur in clusters.

CTS increase during stages I–IV of sleep by a factor of 2–5 times without disturbing the sleep organisation (Figure 9.2).

Rarely, children with RS may have a normal EEG, the spikes may be very small or CTS appear only during sleep stages (3–35%). In serial EEG, CTS may appear right or left, infrequent or abundant, small or giant, alone or with functional spikes in other locations.

In this book, I comply with the nomenclature “centrotemporal spikes”, though they are rarely temporal; “Rolandic spikes” or simply “central spikes” would be a more accurate name (see Figure 9.3).

Dipoles of Centrotemporal Spikes

The main negative spike component of CTS can usually be modelled by a single and stable tangential dipole source along the Rolandic region, with the negative pole maximum in the centrotemporal region and the positive pole maximum in the frontal regions (Figure 9.3). The tangential dipole and the location of CTS have been confirmed...
Concurrent Spikes in Locations Other than the Centrocortical Region

CTS may occur simultaneously in the same EEG with morphologically similar sharp and slow waves in other locations, such as the midline, parietal, frontal and occipital regions. These multifocal sharp waves are more frequently seen in serial EEGs. Occipital spikes are usually the first to appear.

**Important note** The frequency, location and persistence of CTS do not determine the clinical manifestations, severity and frequency of seizures or the prognosis.

Centrotemporal Spikes in Normal and Children without Rolandic Seizures

CTS occur in 2–3% of normal school-age children, of whom less than 10% develop RS. CTS are age-dependent, appearing at a peak age of 7–10 years, often persisting despite clinical remission, and usually disappearing before the age of 16 years. They are common among relatives of children with RS. Age-dependent CTS frequently occur in a variety of organic brain diseases with or without seizures, such as cerebral tumours, Rett syndrome, fragile X syndrome and focal cortical dysplasia. Furthermore, CTS may incidentally be found in non-epileptic children with various symptoms, such as headache (Figure 9.4), speech, behavioural and learning difficulties.

**Definitions**

*Sharp wave:* A transient, clearly distinguished from background activity, with pointed peak at conventional paper speeds and duration of 70–200 ms i.e. over 1/14–1/5 s approximately. The main component is generally negative relative to other areas. Amplitude is variable.

*Spike:* A transient, clearly distinguished from background activity, with pointed peak at conventional paper speeds and duration of 20 to under 70 ms i.e. over 1/50–1/14 s approximately. The main component is generally negative relative to other areas. Amplitude is variable.

According to the above definition what we call centrotemporal spikes are centrotemporal sharp waves because their duration is usually more than 70 ms.

Niedermayer explained: “EEG spikes should be differentiated from sharp waves (i.e. transients having similar characteristics but longer duration). However, it is well to keep in mind that this distinction is largely arbitrary and serves primarily descriptive purposes. It is certainly not incorrect to use the term 'spike' and 'sharp wave' synonymously when a local paroxysmal event is discussed, although purists of nomenclature would regard this as a breach of etiquette.”

Extreme Somatosensory Evoked Potentials/Spikes

After sleep, the most common form of activation of CTS (10%–20%) is somatosensory stimulation mainly of the fingers or toes (Figures 9.2 and 9.4). These are called extreme somatosensory evoked spikes (ESES), extreme somatosensory evoked potentials or giant somatosensory evoked spikes. Like normal somatosensory evoked potentials, their location depends on the site and side of stimulation (Figures 9.2 and 9.4), but their size and morphology is identical to that of CTS. ESES correspond to mid- or long-latency somatosensory evoked potentials with peaks at 35–80 ms depending on the height of the individual and the site of the stimulation (Figure 9.2). ESES persist during sleep. ESES, like spontaneous CTS, occur in children with or without seizures and disappear with age. They may be detected in EEGs with or without spontaneous CTS or other functional spikes of childhood.

**Techniques to Elicit Extreme Somatosensory Evoked Spikes**

Any type of mechanical or electrical stimulus can elicit ESES in susceptible children providing that it is properly applied. It must be abrupt and strong enough (without being uncomfortable), and delivered to the appropriate sensitive...
body region. Percussion of the distal parts of the legs (toes and heels) or arms (palms and mainly tips of fingers) with a reflex hammer or with the plantar tips of the examiner’s fingers is very effective in eliciting ESES (Figure 9.2). A hammer can also be connected to a channel of the EEG in order to mark the exact timing of the stimulus and measure the latency of the evoked spike. Electrical stimulation with digital electrodes as in orthodromic sensory nerve testing, or electrical stimulation of the nerve as in antidromic sensory nerve testing, is equally effective for eliciting ESES, but this is used only for research purposes (Figure 9.2). It may not be necessary for routine EEG, because it is no more efficient than the mechanical stimulation described above, which is a more child-friendly method.

In clinical EEG practice, asking the child to tap together the palmar surface of the tips of his/her fingers of both hands is an easy method of testing for ESES (Figure 9.4). The child should be instructed to strike them with sufficient strength and at random intervals of varying frequency. This may elicit either bilateral or unilateral ESES (Figure 9.4).

Important note

Generalised Discharges

The reported prevalence of generalised discharges in RS varies from as low as 0% to as high as 54%. In my studies, generalised discharges occurred in about 4% of patients with RS and consisted of brief 1–3 s generalised bursts of 3–5 Hz slow waves intermixed with small spikes. These brief generalised discharges are identical to those seen in PS (Figure 9.6, page 242).

Important note

The combination of a normal child with infrequent seizures and an EEG showing disproportionately severe focal epileptogenic activity is highly suggestive of benign childhood seizure susceptibility syndrome.

Ictal EEG

There are very few reports of ictal EEG of RS. One example captured with video EEG is shown in Figure 9.5. There is an initial paucity of spontaneous CTS prior to the onset of the ictal discharge, which appears in the ipsilateral Rolandic regions and consists of slow waves intermixed with fast rhythms and spikes.

Evolution and Prognosis

Remission occurs within 2–4 years of onset and before the age 16 years. The total number of seizures is low. The majority of patients have less than 10 seizures; 10–20% have a single seizure only. Around 10–20% may have frequent seizures, but these also remit with age.

Children with RS may develop reversible linguistic abnormalities during the active phase of their disease. Hospital-based studies emphasise learning or behavioural problems that require intervention. A few patients (<1%) may progress to atypical evolutions of more severe syndromes of linguistic, behavioural and neuropsychological deficits, such as Landau-Kleffner syndrome, atypical focal epilepsy of childhood, or epilepsy with continuous spikes and waves during slow wave sleep.

The prognosis of RS is invariably excellent, with a less than 2% risk of developing infrequent generalised seizures in adult life; absence seizures may be more common than GTCS.

Development, social adaptation and occupation of adults with a previous history of RS is normal.

Patient note

The only problem was with five patients who had difficulties in obtaining their driving licences and one patient who despite a 15-year seizure-free period was still on phenytoin because of concerns of her physician regarding her driving licence.

For unknown reasons social levels of patients with Rolandoic epilepsy seem to be even higher than for non-epileptic controls.

Management


Children with RS may not need antiepileptic medication, particularly if the seizures are infrequent, mild or nocturnal, or the onset is close to the age of natural remission of this age-limited disorder. Patients with frequent seizures and secondarily GTCS or with comorbid conditions (tics, attention-deficit hyperactivity disorder, learning disability) may need medication. In a recent study, AEDs significantly reduced GTCS, but did not reduce focal seizures. On an empirical basis, carbamazepine is the preferred AED. However, some children might experience particular learning difficulties and exaggeration and new types of seizures while receiving carbamazepine.

Within days after re-introduction of carbamazepine, she suffered nearly continuous, brief atonic attacks of head and arm drop and also absences (case 17.3 in ref 1)

Lamotrigine may be contraindicated in RS, because of case reports with exacerbation of the condition and new types of seizures.

See details in “Management of benign childhood focal seizures” (page 257).

**Panayiotopoulos Syndrome**

Panayiotopoulos syndrome (PS) is a childhood-related idiopathic benign susceptibility to focal, mainly autonomic, seizures and autonomic status epilepticus. Affected children have normal physical and neuropsychological development. Autonomic manifestations are the cardinal seizure symptoms in PS, and have immense pathophysiological, clinical and treatment implications. All functions of the autonomic system may be affected during the ictus. Nearly half of these seizures last between 30 min and 7 hours, and constitute autonomic status epilepticus.

**Demographic Data**

Age at onset is 1–14 years with a peak at 4–5 years; in 76% of cases, onset occurs at 3–6 years of age. Boys and girls are equally affected. Children of all races are vulnerable. Prevalence is around 13% in children 3–6 years old with one or more non-febrile seizures and 6% in the 1–15-year age group. In the general population, 2–3/1000 children may be affected. These figures may be higher if cases that are currently considered to have atypical features are included.

**Clinical Manifestations**

Seizures comprise an unusual constellation of autonomic, mainly emetic, symptoms, behavioural changes, unilateral deviation of the eyes and other more conventional ictal manifestations. Consciousness and speech, as a rule, are preserved at seizure onset. The seizure commonly starts with autonomic manifestations (81%), which are mainly emetic (72%). In a typical presentation, the child is fully conscious, able to speak and understand, complains “I feel sick”, looks pale and vomits.

He complained of nausea and he looked pale. Five min later he vomited while still standing… He gradually became disorientated, but was still able to walk. However, 10 min from onset his eyes turned to the right and he became unresponsive.

**Ictus Emeticus**

The full emetic triad (nausea, retching, vomiting) culminates in vomiting in 74% of seizures; in others, only nausea or retching occur and, in a few cases, emesis may not be apparent. Emesis is usually the first apparent ictal symptom, but may also occur long after the onset of other manifestations.

**Considerations on Classification and Nomenclature of Panayiotopoulos Syndrome**

Panayiotopoulos syndrome has been recognised in the new diagnostic scheme as: “Early onset benign childhood occipital epilepsy (Panayiotopoulos type)”. However, PS is not “occipital” epilepsy.
(a) Onset is with autonomic manifestations, which are unlikely to be of occipital origin. Of all the other seizure symptoms, only eye deviation, which is often not the first ictal symptom, may originate from the occipital lobes.

(b) Interictal occipital spikes may never occur.

(c) Even ictal EEG has documented anterior or posterior origin.

Currently, most authors prefer the eponymic nomenclature “Panayiotopoulos syndrome” to include all patients with this syndrome irrespective of EEG spikes or topographic terminology\(^4\,14;\,76;\,77;\,79–82;\,84\) as in the original study of Panayiotopoulos.\(^70\)

In that study of 900 seizure-patients of all ages, only 24 had ictal vomiting and all of these were children.\(^70\) Of these 24 children with ictal vomiting:

(a) 21 had normal development with EEG occipital spikes (12 patients), extra-occipital spikes (5 patients), brief generalised discharges (1 patient) or normal EEG (3 patients)

(b) 3 had an abnormal neurological state (symptomatic childhood autonomic epilepsy).

Of the 21 idiopathic patients (which constitutes what is now known as Panayiotopoulos syndrome), attention was directed towards the predominant group of occipital spikes and occipital paroxysms (hence the name “occipital epilepsy” or “epilepsy with occipital paroxysms”).\(^85\) The fact that the other 9 patients without occipital spikes had the same disorder with those of occipital spikes was documented much later.\(^86\) The terms “early onset” and “late onset” have been used adjunctively with “childhood occipital epilepsy” or “epilepsy with occipital paroxysms” in order to discriminate between PS (with median age of onset in early childhood) and G-ICOE (with median age of onset in late childhood). It should be clarified that these terms (“early onset” and “late onset”) were first introduced at a time that G-ICOE was the only recognised syndrome associated with “occipital paroxysms”\(^85\) and prior to the documentation that PS manifests with multi-focal EEG spikes with one-third not having occipital spikes. For those few that they may disapprove certain eponyms (see for example reference\(^87\)) the most precise descriptive term I can propose for PS is “benign childhood autonomic seizures and autonomic status epilepticus” and not “early onset benign childhood epilepsy with occipital spikes”.

Typically, nausea is the first complaint of a child who suffers a seizure (while awake or who wakes from sleep) prior to vomiting and other ictal manifestations. This initial stage does not suggest an epileptic seizure; the child simply complains of feeling sick and being unwell, and looks pale. The patient may be quiet or agitated, vacant or restless but fully conscious and able to understand and answer questions. Ictus emeticus at this stage is no different from any other disease that causes emesis – just a child who feels sick or wants to be sick.

On returning home from school, she looked tired and had a nap. After half an hour, she woke up looking pale and complained of feeling sick. She ran to the toilet and vomited repeatedly. Then her eyes deviated to one side and she became unresponsive and flaccid for 10 min. Soon after, she started recovering, answering simple questions and 1 hour later she was playing again as if nothing had happened.

**Patient note**

<table>
<thead>
<tr>
<th>Timing of the Vomiting</th>
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<tr>
<td>When vomiting occurs, it commonly starts 1–5 min after the onset of nausea, while the child is still conscious and otherwise well. Less often, vomiting may occur later during other more conventional seizure symptoms.</td>
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**Intensity and Duration of Vomiting**

This varies considerably from mild to often severe and repetitive. Usually, the child vomits 3–5 times; however, some children may repeatedly vomit for hours leading to dehydration, while others may vomit only once.

**Other Autonomic Manifestations**
Autonomic manifestations other than ictus emeticus may occur concurrently or appear later in the course of the ictus. These include pallor and less often flushing or cyanosis, mydriasis and less often miosis, cardiorespiratory and thermoregulatory alterations, coughing, incontinence of urine and/or faeces, and modifications of intestinal motility. Hypersalivation (probably a concurrent Rolandic symptom) may occur. Headache and more often cephalic auras may occur particularly at onset.

Pallor is one of the commonest ictal manifestations. It mainly occurs at onset, usually with emetic symptoms. Exceptionally, pallor may be among the first symptoms without apparent emesis.

Cyanosis is less common than pallor. It principally occurs during the evolution of the seizures, often while the child is unresponsive.

Incontinence of urine and faeces occur when consciousness is impaired prior to, or without, convulsions: “became unresponsive and incontinent of urine” is a common association. These symptoms do not occur at onset.

**Definitions of Autonomic Seizures and Autonomic Status Epilepticus**

*Autonomic auras* consist of a subjective awareness of a change in the activity of autonomic nervous system function.\(^8^8\)

*Autonomic seizures* consist of episodic altered autonomic function of any type at onset or as the sole manifestation of an epileptic event. These may be objective, subjective or both. They must be distinguished from secondary (indirect effects) on the autonomic system by other seizure symptoms.\(^8^9\)

*Pure autonomic seizures* are those that consist solely of episodic altered autonomic function from onset to the end.

In the absence of a definition for autonomic status epilepticus, the terms I propose are:

*Autonomic status epilepticus*: is an autonomic seizure that lasts for more than 30 minutes.\(^4^;^9^0\)

*Pure autonomic status epilepticus* is a pure autonomic seizure that lasts for more than 30 minutes.\(^4^;^9^0\)

**Terminology of the Emetic Components**

*The emetic process* consists of the triad of nausea, retching and vomiting, which are three separate functional entities occurring independently of each other and in any combination.

*Vomiting* refers to the forcible oral expulsion of gastric contents and is usually preceded by symptoms (e.g. pallor, salivation, sweating) attributed to autonomic nervous system discharges, which are not, however, essential for the act of vomiting.

*Ictus emeticus*: emetic manifestations (nausea, retching, vomiting alone or in combination) caused by seizure discharges.

*Ictal vomiting*: forcible oral expulsion of gastric contents due to seizure discharges.

Mydriasis is sometimes so prominent that it may be reported spontaneously.

Her pupils were as big as her eyes.

Mydriasis occurs concurrently with other marked autonomic manifestations. Dilated pupils may not be reactive to light.

Miosis is rare and occurs with other severe autonomic manifestations while the child is unresponsive.
Hypersalivation is also rare in PS, in contrast to its common occurrence in RS. Combined speech arrest and hypersalivation, as in RS, is even rarer.

Cephalic auras, though rare, are of interest, because they are considered to be autonomic manifestations and because of the diagnostic confusion they may cause with migraine if not properly evaluated. Cephalic auras commonly occur with other autonomic symptoms, mainly nausea, at seizure onset. Occasionally, the child may also complain of “headache” but whether the complaint of “headache” is a true perception of pain, discomfort or some odd sensation in the head is uncertain.

**Patient note**

“funny feeling in my head”, “warm sensation”, “pressure”, “headache”

Coughing may occur as an initial ictal symptom either with or without ictus emeticus. It is described as “strange coughing” or “cough as if about to vomit”.

**Thermoregulatory Changes**

Raised temperature may be subjectively or objectively documented during the seizure or immediately post-ictally. Whether this is a coincidental finding, a precipitating factor or an ictal abnormality is uncertain. It could be any of these. However, pyrexia recorded immediately after seizure onset is probably an ictal autonomic manifestation.

**Abnormalities of Intestinal Motility**

Diarrhoea (3%) is occasionally reported during the progression of seizures.

**Breathing and cardiac irregularities** are rarely reported, but may be much more common in a mild form. Breathing changes prior to convulsions include descriptions of “heavy, irregular, abnormal breathing” or “brief cessation of breathing for a few seconds”. Tachycardia is a consistent finding, sometimes at the onset, of ictal EEG (Figures 9.8 and 9.9).

Cardiorespiratory arrest is exceptional, but is potentially fatal without immediate medical intervention (case 37 in ref4).

**Patient note**

At the age of 3 years, while dozing in his mother’s car, his head went back, eyes were “rolling”, colour clay-like grey, pupils dilated, unresponsive and incontinent of urine. Arms were initially rigid, but then he became floppy. At this stage, 5–10 min from onset, a Swiss paediatrician who happened to be present diagnosed cardiopulmonary arrest and resuscitated him: “Lips blue and then white. No respiration. No heart beat and wide pupils without reaction to light. External heart massage and mouth-to-nose resuscitation, less than 1 minute. Total time of asystole reckoned to be 2 min. Unconsciousness went on for 20 min then he started to cry and he recognised his mother”. He was well after a few hours’ sleep.4

I have been made aware of three other children with PS and cardiorespiratory arrest. Sadly, one of them died. Though tragic and exceptional, this should be expected to happen in view of the frequent occurrence of autonomic status epilepticus in children (page 239).

**Ictal Syncope**

Ictal syncope is an intriguing and important ictal feature of PS.4 It is a common and dramatic occurrence. In at least one-fifth of seizures, the child becomes ‘completely unresponsive and flaccid like a rag doll’ before and often without convulsions or in isolation.

**Terminology of the Cephalic Auras**

Cephalic auras are ictal symptoms of non-specific sensory perceptions involving or limited to the head.91
While talking to her teacher, suddenly and without warning, she fell on the floor pale, flaccid and unresponsive for 2 min. She had a complete recovery, but 10 min later she complained of feeling sick, vomited repeatedly and again became unresponsive and flaccid with pupils widely dilated for 1 hour. She had an unremarkable recovery and was normal after a few hours’ sleep.

She complained of ‘dizziness’ and then her eyes deviated to the left, she fell on the floor and she became totally flaccid and unresponsive for 5 min.

I proposed the descriptive term ‘ictal syncope’ to describe this state, because ‘unresponsiveness with loss of postural tone’ are the defining clinical symptoms of syncope. Other authors prefer ‘syncope-like symptoms’.

Ictal Behavioural Changes

Ictal behavioural changes usually consist of restlessness, agitation, terror or quietness, which appear at the onset of seizures, often concurrently with emetic or other autonomic manifestations. These symptoms are often similar to those occurring in ‘benign childhood epilepsy with affective symptoms’ (page 254).

Patient note He was happily playing and asking questions when he started complaining that he was feeling sick, and became very pale and quiet. He did not want to drink or eat. Gradually, he was getting paler and paler, and kept complaining that he felt sick. He then became restless and frightened. Ten min from the onset, his head and eyes slowly turned to the left. The eyes were opened, but fixed to the left upper corner. We called his name, but he was unresponsive. He had completely gone. We tried to move his head, but this was fixed to the left. There were no convulsions. This lasted for another 15 min when his head and eyes returned to normal and he looked better, although he was droopy and really not there. At this stage he vomited once.

At age 9 years, on return from school, he looked tired and pale. He said that his head was killing him “something that would cause me to be sick”. In 10 min, he started screaming and banging his head on the wall. Within the next 20 min, he gradually became disorientated and floppy ‘like a rag doll’. He was staring.

Conventional Seizure-Symptoms

In PS, pure autonomic seizures and pure autonomic status epilepticus occur in 10% of patients. They start and end solely with autonomic symptoms. In all others, autonomic manifestations are followed by conventional seizure-symptoms and these in order of prevalence are:

- Impairment of consciousness (94%)
- Deviation of the eyes (60–80%)
- Hemiconvulsions (26%)
- Generalised convulsions (20%)
- Speech arrest (8%)
- Visual hallucinations (6%)
- Other manifestations occur less than 3% each.

Impairment of Consciousness

Though initially fully conscious, the child gradually or suddenly becomes confused and unresponsive. Impairment of consciousness may be mild or moderate, with the child retaining some ability to respond to verbal commands, but often talking out of context. Complete unresponsiveness is probably exceptional at the beginning of the seizure. In diurnal seizures observed from onset, clouding of consciousness usually starts after the appearance of autonomic and behavioural symptoms, becoming progressively worse until complete unresponsiveness is reached. Good awareness may be preserved throughout the ictus in around 6% of seizures.
Definition of Ictal Syncope

‘Ictal syncope’ denotes transient loss of consciousness and postural tone that occurs in a seizure before or without convulsions. Transient loss of consciousness and postural tone are the defining symptoms of syncope irrespective of underlying cause. Ictal syncope is purely a name for a cluster of seizure symptoms that, until now, have lacked a descriptive term. Ictal syncope does not occur or may be very rare in other types of seizures, except in postictal states after GTCS.

Deviation of the Eyes

Unilateral deviation of the eyes with, or rarely without, ictal vomiting is a common ictal manifestation, which seldom occurs at onset. The eyes shift to the extreme of one side and the head may also turn ipsilaterally. This pursuit-like deviation of the eyes may be brief (for min) or prolonged (for hours). It may be continuous or less often intermittent, with eyes returning to the midline and shifting again towards the same side. The eyelids remain open, but may be half open or open wide and, at this stage, consciousness is often, but not invariably, impaired.

Deviation of the eyes may occur without vomiting in 10–20% of patients and, in some children, the eyes may be open wide and remain in the midline before other convulsions occur.

Other ictal symptoms in order of prevalence are speech arrest (8%), hemifacial spasms (6%), visual hallucinations (6%), OPL movements (3%), unilateral drooping of the mouth (3%), eyelid jerks (1%), myoclonic jerks (1%), ictal nystagmus and automatisms (1%). These probably reflect the primary area of seizure discharge generation. The seizures may end with hemiconvulsions often with Jacksonian march (19%) or generalised convulsions (21%).

Ictal visual symptoms, such as elementary visual hallucinations, illusions or blindness, occur after more typical seizure symptoms of PS.

Hemiconvulsive (2%) or generalised convulsive status (2%) is exceptional.

The same child may have seizures with marked autonomic manifestations and seizures in which autonomic manifestations may be inconspicuous or absent. Seizures without autonomic manifestations are rare (7%).

The clinical seizure manifestations are roughly the same irrespective of EEG localisations though there may be slightly less autonomic and slightly more focal motor features at onset in children without occipital spikes.

Seizures without Autonomic Manifestations

Such seizures are rare (7%) and occur in patients who may also have additional autonomic seizures.

Case 3 of ref had three seizures with ictus emeticus. An additional lengthy diurnal seizure manifested only with deviation of the eyes and mild impairment of consciousness prior to generalised convulsions.

Duration of Seizures and Autonomic Status Epilepticus

Nearly half (44%) of the seizures last for more than 30 min and can persist for up to 7 hours (mean about 2 hours), constituting autonomic status epilepticus. The rest of the seizures (54%) last from 1–30 min with a mean of 9 min. Lengthy seizures are equally common in sleep and wakefulness. Even after the most severe seizures and status, the patient is normal after a few hours’ sleep. There is no record of residual neurological or mental abnormalities. The same child may have brief and lengthy seizures. Hemi-convulsive or convulsive status epilepticus is exceptional (4%).

Despite the high incidence of autonomic status epilepticus, convulsive status epilepticus is exceptional in PS.
Circadian Distribution

Two-thirds of seizures start in sleep; the child may wake up with similar complaints while still conscious or else may be found vomiting, conscious, confused or unresponsive.

Clinically, while asleep, “he suddenly got up with both eyes open, vomited several times and then showed a prolonged atonic state with cyanosis and irregular respiration for 3 min” (from ictal EEG documentation by Oguni and associates).72

The same child may suffer seizures while asleep or awake.

A 10-year-old boy of normal development suffered an episode of autonomic status epileptics at 5 years of age. Half an hour after he had gone to sleep, he woke up looking pale and then complained that he felt sick before vomiting repeatedly. Within min his eyes deviated and fixed laterally, and soon after he became flaccid ‘like a rag doll’, unresponsive, and incontinent of urine and faeces. His breathing was short and shallow, and stopped for a few seconds before he started vomiting again. Two hours after onset, he had brief hemiconvulsions with Jacksonian marching for 5 min. On arrival at hospital, he was recovering and his temperature was mildly raised to 37.4°C. A CT brain scan, CSF examination and relevant blood tests were normal. Triple treatment for encephalitis was started, though he was entirely normal and apyrexial after a few hours of sleep. EEG the next day showed a few scattered occipital and central spikes. Treatment was stopped 4 days later.

A similar autonomic status epilepticus occurred 6 months later on a ferryboat while on holiday. He said that he felt sick and looked pale. He vomited a couple of times, and then his eyes turned to one side and he talked out of context and vomited again. “Then I knew that he was having another fit” his mother said. “He was as if drifting in and out of sleep. He did not become unconscious, but he was continuously vomiting for several hours.” On arrival at hospital 3 hours later, he was improving and able to talk and walk. He was diagnosed and treated for dehydration. He was normal the next morning. A new EEG showed repetitive multifocal spike-wave complexes.

No further seizures occurred in the next 5 years, and his development and EEG were normal.

Precipitating Factors

There are no apparent precipitating factors other than sleep. Fixation off sensitivity is an EEG phenomenon, which may not be clinically important.

Many seizures have been witnessed while a child is travelling in a car, boat or aeroplane. There are two explanations for this: (a) the seizures are more likely to be witnessed during travelling; or (b), children are more vulnerable because travelling also precipitates motion sickness which is particularly common in children.

Aetiology

PS, like RS, is probably genetically determined. Usually, no family history of similar seizures exists, though siblings with PS or PS and Rolandic epilepsy have been reported.14,71,74,79 There is a high prevalence of febrile seizures (about 17%). Also, there may be a high incidence of abnormal birth deliveries, but these all need re-evaluation.4

PS, Rolandic epilepsy and all other benign childhood focal seizures are probably linked together by a common, genetically determined, mild and reversible functional derangement of the brain cortical maturational process that I proposed to call ‘benign childhood seizure susceptibility syndrome’ (see Figure 9.1 and pages 223).

Considerations on the Classification of Autonomic Status Epilepticus

Childhood autonomic status epilepticus, though common and specific in childhood, is ignored in all classifications even now, long after its first description by Panayiotopoulos.4,70,90 The new ILAE diagnostic scheme2 recognises four forms of focal status epilepticus (Table 1.4).2,95
(a). Epilepsia partialis continua of Kozhevnikov
(b). Aura continua
(c). Limbic status epilepticus (psychomotor status)
(d). Hemiconvulsive status with hemi-paresis

From a clinical point of view aura continua is classified into:

1. somatosensory, i.e. dysaesthesia phenomena that involve the trunk, head and extremities,
2. aura continua that involve the special senses (visual, auditory, vertiginous, gustatory and olfactory).
3. aura continua with predominantly autonomic symptoms, and with psychic symptoms.

It is anticipated that future revisions of the ILAE classifications will recognise this type of age-related autonomic status epilepticus. This is mandated by its high prevalence and its high rate of misdiagnosis and mismanagement. Four-fifths of childhood autonomic status epilepticus occur in PS and the remaining one-fifth occurs in symptomatic childhood epileptic disorders. Ignoring these facts, as indeed happens even now, results in avoidable morbidity and probably mortality.

**Pathophysiology**

Autonomic symptoms of any type are often encountered in seizures, focal or generalised, in adults or children and they are implicated in occurrences of sudden death. However, autonomic seizures and autonomic status with ictus emeticus and ictal syncope, with the symptomatology and the sequence as detailed in this chapter, are specific in childhood. This clinical picture does not occur in adults: only about 30 cases of ictal vomiting have been reported, but not in the same sequence as in children – adult patients usually have amnesia about the vomiting, which often occurs after the seizure has started with other symptoms (see page 373). An explanation for this is that children are vulnerable to emetic disturbances as exemplified by the ‘cyclic vomiting syndrome’, a non-seizure disorder of unknown aetiology that is also specific to childhood. Ictal syncope is even more difficult to explain.

Symptoms at the onset of seizures are important, because they indicate the possible location of the epileptic focus. However, autonomic and emetic disturbances are of uncertain value with regard to localisation in PS and may occur in seizures starting from the anterior or posterior regions. The localisation of ictal vomiting in adults does not appear to apply in children (page 373).

Clinical and EEG findings indicate that, in PS, there is a diffuse cortical hyperexcitability, which is maturation-related. This diffuse epileptogenicity may be unequally distributed, predominating in one area, which is often posterior. The preferential involvement of emetic and other autonomic manifestations may be attributed to epileptic discharges triggering low-threshold emetic centres and hypothalamus of vulnerable children. In other words, it is likely that in vulnerable children a “weak” epileptic electrical discharge (irrespective of localisation) activates at its onset susceptible autonomic centres to autonomic seizures and autonomic status epilepticus. This is prior to the generation of clinical manifestations from brain regions that are topographically related to the ictal electrical discharge (occipital, frontal, central, parietal and less often temporal) with seizure thresholds higher than those of the autonomic centers.

**Diagnostic Procedures**

By definition, in an idiopathic syndrome, neurological and mental states and high resolution MRI are normal. The most useful laboratory test is the EEG (Figure 9.6). The most important determinant of the neurodiagnostic procedures is the state of the child at the time of first medical attendance.

- The child has a typical brief or lengthy seizure of PS, but has fully recovered before arriving at the accident and emergency department or being seen by a physician. A child with the distinctive clinical features of PS, particularly ictus emeticus and lengthy seizures, may not need any investigations other than EEG. However,
because approximately 10–20% of children with similar seizures may have brain pathology, an MRI may be indicated.

- The child with a typical lengthy seizure of PS has partially recovered, though is still in a postictal stage, tired, mildly confused and drowsy on arrival at the accident and emergency department or when seen by a physician. The child should be kept under medical supervision until full recovery, which is the rule after a few hours of sleep. Then guidelines are the same as above

- The child is brought to the accident and emergency department or is seen by a physician while ictal symptoms continue. This is the most difficult and challenging situation. Symptoms may dramatically accumulate in succession, and demand rigorous and experienced evaluation. A history of a previous similar seizure is reassuring and may help to avoid unnecessary investigation.

**Electroencephalography**

In about 90% of cases, the EEG reveals functional, mainly multi-focal, high amplitude sharp-slow wave complexes (Figure 9.6). Spikes may appear anywhere. They are often independent, and occur at various posterior locations and, less often anterior locations, in the same or the contralateral hemisphere, and may appear as cloned-like, repetitive, multifocal spike-wave complexes. In order of prevalence, the complexes most commonly occur in occipital, frontal and centrotemporal regions; right and left regions are equally involved. Midline spikes occur in 17% of cases.

**Important note** The EEG shows great variability in functional focal spikes at various electrode locations. All brain regions are involved, though posterior areas predominate (Figures 9.6 and 9.7).

Two-thirds of patients (68%) have at least one EEG with occipital paroxysms or, more commonly, occipital spikes, which are often (64%) concurrent with extra-occipital spikes in at least one EEG. The other third (32%) never show occipital spikes (Figures 9.4 and 9.6). Instead, they have extra-occipital spikes (21%) only, a consistently normal EEG (9%) or brief generalised discharges only (2%). An EEG with multifocal spikes in more than two, and often many, brain locations occurs in one-third of patients; single spike foci are rare (9%). Cloned-like, repetitive, multifocal spike-wave complexes may be characteristic features when they occur (19%). They have never been studied or reported before in idiopathic epilepsies. On the contrary, multifocal repetitive spikes are considered to be suggestive of a bad prognosis and indicative of symptomatic epilepsies (page 169). Cloned-like, repetitive, multifocal spike-wave complexes do not determine the prognosis, because they occur equally in children with single or multiple seizures.

Spikes are usually of high amplitude and morphologically similar to CTS. They often show stable dipoles in the occipital regions.

Small and even inconspicuous spikes may appear in the same or a previous EEG of children with giant spikes. Though rare, positive spikes or other unusual EEG spike configurations may occur.

Brief generalised discharges of slow waves intermixed with small spikes may occur either alone (4%) or more often with focal spikes (15%).

The EEG spikes may be stimulus sensitive; occipital paroxysms are commonly (47%) activated by the elimination of central vision and fixation, while CTS may be elicited by somatosensory stimuli (Figure 9.4). EEG occipital photosensitivity is an exceptional finding.

Functional spikes in whatever location are accentuated by sleep. If a routine EEG is normal, a sleep EEG should be performed. There is no particular relationship between the likelihood of an abnormal EEG and the interval since the last seizure. EEGs recorded a short or long time after a seizure are equally likely to manifest with functional spikes, which may occur only once in serial routine and sleep EEGs.

The background EEG is usually normal, but diffuse or localised slow wave abnormalities may also occur in at least one EEG in 20% of cases, particularly postictally.
Definition of Cloned-Like, Repetitive, Multifocal Spike-Wave Complexes

See reference 4

Cloned-like repetitive multifocal spike wave complexes are repetitive spike or sharp and slow wave complexes that appear concurrently in different brain locations of one or both hemispheres. There may be just two discrete foci unilaterally or contralaterally, but the complexes are usually multifocal and often give the impression of generalised discharges or secondary bilateral synchrony. They are stereotypically and identically repetitive in the same and often subsequent EEGs from the same patient, which is the reason that I coined the name ‘cloned-like’. On the surface, in routine EEG recordings, they appear synchronous but usually one spike focus leads the others by a few milliseconds. The leading focus commonly occurs alone without spikes in other locations. Cloned-like repetitive multifocal spike-wave complexes can occur without the primary spike and can be so abundant that they dominate the EEG and obscure its background, which is otherwise normal. On other occasions, they are scarce and appear in a well-organised EEG with normal background.

**Important note** EEG abnormalities, particularly functional spikes, may persist for many years after clinical remission; they disappear when the patient reaches the mid-teens. Conversely, spikes may appear only once in one of a series of EEGs.

Frequency, location and persistence of functional spikes do not determine clinical manifestations, duration, severity and frequency of seizures, or prognosis.

Ictal EEG

Typical autonomic seizures and autonomic status epilepticus of PS have been documented by ictal EEG (Figures 9.8 and 9.9). The seizure discharge consists mainly of rhythmic theta or delta activity, usually intermixed with small spikes. Onset is unilateral, often posterior, but may also be anterior and not strictly localised to one electrode. Figure 9.8 shows autonomic status epilepticus captured from the onset on video EEG. Ictal clinical manifestations may start minutes after the onset of the electrical seizure discharge (Figure 9.8).

Differential Diagnosis

PS is easy to diagnose, because of the characteristic clustering of clinical seizure semiology, which is often supported by interictal EEG findings. However, despite sound clinical EEG manifestations, PS escaped recognition for many years for a number of reasons. Ictus emeticus is rarely considered a seizure event. When it is associated with a deteriorating level of consciousness, followed by convulsions, encephalitis or other acute cerebral insults are the prevailing diagnoses in the acute stage. If the child is seen after complete recovery, atypical migraine, gastroenteritis, motion sickness or a first seizure are likely diagnoses. Similarly, ictal syncope has only recently been recognised as an important clinical manifestation of PS; ictal syncope may be misdiagnosed as cardiogenic syncope, pseudoseizure or a more severe encephalopathic state.4

**Important note** The main problem is to recognise emetic and other autonomic manifestations as seizure events, and not to dismiss them or erroneously consider them as unrelated to the ictus and as a feature of encephalitis, migraine, syncope or gastroenteritis.

Similarly, 10–20% of autonomic seizures and autonomic status epilepticus with a similar presentation to PS are due to heterogeneous cerebral pathology, such as focal or diffuse brain lesions of diverse aetiology. These autonomic seizures are also restricted to childhood. In these symptomatic cases, there is often abnormal neurological or mental symptomatology, abnormal brain imaging, and background EEG abnormalities (Figure 9.10). Also, patients commonly have additional types of seizures without autonomic symptoms that continue in adult life. Management and treatment is similar to any other form of lesional epilepsy.

- **Gastaut-type idiopathic childhood occipital epilepsy** has entirely different clinical manifestations, despite
common interictal EEG features when occipital paroxysms occur in PS. Visual seizures are predominant in G-ICOE. Visual symptoms in PS, when present, occur with other typical clinical, mainly autonomic, vomiting and behavioural manifestations. They are not the predominant seizure symptom. They are never the sole symptom of a seizure and do not occur alone. Usually, they develop after the seizure has started. Their presence signifies spreading of the discharge to the occipital regions of the brain. Exceptionally (in 1% of cases), visual symptoms occur at onset suggesting an occipital origin for the epileptic discharge in these patients.

- Rolandic seizures have different clinical manifestations. Emesis, when it occurs, is a concomitant symptom of PS. Conversely, in some cases of PS, there are concurrent Rolandic symptoms. This should not be a problem as the management and the prognosis of both syndromes is the same.

- Cases of PS with seizures occurring when the child is febrile may be diagnosed asfebrile seizures, but this again is of no prognostic significance.

**Important note**
An EEG demonstrating multifocal spikes may be indispensable in the diagnosis of patients with PS if clinical information is inadequate or emetic manifestations are inconspicuous.

**Prognosis**

PS is a remarkably benign condition despite the high incidence of autonomic status epilepticus. One-third of patients (27%) have only a single seizure, half (47%) have 2–5 seizures and only 5% have more than 10 seizures, which can sometimes be very frequent, but again the outcome is favourable. Furthermore, the active seizure period is very brief and remission commonly occurs within 1–2 years from onset. The risk of developing epilepsy in adult life is probably no more than that in the general population. However, one-fifth of patients (21%) may develop another type of infrequent, usually Rolandic (13%), seizures during their childhood and early teens. These seizures are also age-related and remit before the age of 16 years. Atypical evolutions with absences and drop attacks, such as those occurring in Rolandic epilepsy, are exceptional.

However, despite its benign course, some seizures of PS may occasionally manifest with potentially fatal autonomic symptoms, such as cardiorespiratory arrest.

**Management**

Current guidelines for febrile seizures, if appropriately modified, may provide the basis for similar guidelines for PS. Based on the risks and benefits of the effective therapies, continuous anticonvulsant therapy is not recommended for children with one or brief seizures. Most clinicians treat recurrent seizures with carbamazepine. Lengthy seizures are a medical emergency and rectal diazepam is prescribed for home administration.

See details in “Management of benign childhood focal seizures” (page 257).

**Clinical note**
**Diagnostic tips**
Paediatricians should be alerted by lengthy autonomic seizures and electroencephalographers by frequent multifocal spikes in a normal child with one or a few seizures.

In terms of the EEG, it is important to remember that frequent epileptogenic foci in a normal child with infrequent seizures should raise the possibility of the benign childhood focal seizures.

**Gastaut-Type Idiopathic Childhood Occipital Epilepsy (Idiopathic Childhood Occipital Epilepsy)**

Gastaut type-idiopathic childhood occipital epilepsy (G-ICOE) or idiopathic childhood occipital epilepsy of late onset is a rare manifestation of a childhood seizure susceptibility syndrome that has an age-related onset, is often age limited and may be genetically determined. It is a pure form of idiopathic occipital epilepsy.

**Demographic Data**

Age at onset is 3–15 years with a mean around 8 years of age. Girls and boys are equally affected.
accounts for about 2–7% of benign childhood focal seizures.

**Clinical Manifestations**

Seizures are purely occipital and primarily manifest with elementary visual hallucinations, blindness or both (see also detailed symptomatology of occipital lobe epilepsy, page 417). They are usually frequent and diurnal. They develop rapidly within seconds and are brief, lasting from a few seconds to 1–3 min, rarely longer. Elementary visual hallucinations are the commonest and most characteristic ictal symptom, and are most likely to be the first and often the only clinical manifestation. Ictal elementary visual hallucinations mainly consist of small multicoloured circular patterns that often appear in the periphery of a visual field, becoming larger and multiplying during the course of the seizure, frequently moving horizontally towards the other side (Figure 9.11).

**Patient note** I see millions of small, very bright, mainly blue and green coloured, circular spots of light, which appear on the left side and sometimes move to the right

Other occipital symptoms, such as sensory illusions of ocular movements and ocular pain, tonic deviation of the eyes, eyelid fluttering or repetitive eye closures, may occur at the onset of the seizures or appear after the elementary visual hallucinations.

Deviation of the eyes is the most common (in around 70% of cases) non-visual symptom. It is often associated with ipsilateral turning of the head and usually starts after visual hallucinations, but may also occur while the hallucinations still persist. It may be mild, but more often it is severe and progresses to hemiconvulsions and GTCS. It has been well established that children with benign occipital seizures may have motor focal seizures of eye deviation ab initio without visual hallucinations. It is likely that these cases have a better prognosis and shorter seizure life span than those with G-ICOE, but this has not yet been properly investigated. ¹⁰⁹

**ILAE Definition**

The 1989 ILAE Commission named this syndrome “childhood epilepsy with occipital paroxysms” and defined it as follows: “The syndrome of childhood epilepsy with occipital paroxysms is, in general respects, similar to that of benign childhood epilepsy with centrotemporal spikes. The seizures start with visual symptoms (amaurosis, phosphenes, illusions, or hallucinations) and are often followed by a hemiclonic seizure or automatisms. In 25% of cases, the seizures are immediately followed by migrainous headache. The EEG has paroxysms of high-amplitude spike waves or sharp waves recurring rhythmically on the occipital and posterior temporal areas of one or both hemispheres, but only when the eyes are closed. During seizures the occipital discharge may spread to the central or temporal region. At present, no definite statement on prognosis is possible.”

This purely ‘idiopathic’ and purely ‘occipital’ epilepsy has been recognised in the new diagnostic scheme² as: “Late onset childhood occipital epilepsy (Gastaut type)”. ²

Uncertainty in regard to prognosis is reflected by the fact that the term ‘benign’ (used in all other benign childhood focal seizures) is not included in the ILAE descriptive terminology of this syndrome.²;¹³

Forced eyelid closure and eyelid blinking are interesting ictal clinical symptoms of occipital seizures that occur in approximately 10% of patients, usually at a stage in which consciousness is impaired. They signal impending secondary GTCS.

Ictal blindness, appearing ab initio or less commonly after other manifestations of occipital seizures, usually lasts for 3–5 min. It can occur alone and can be the only ictal event in patients who may, at other times, have visual hallucinations without blindness.

**Patient note** Everything went suddenly black, I could not see and I had to ask other swimmers to show me the direction to the beach.
Complex visual hallucinations, without the emotional and complicated character of temporal lobe seizures, visual illusions and other symptoms resulting from more anterior ictal spreading, may rarely occur ab initio or as a result of seizure progress that may terminate in hemiconvulsions or generalised convulsions.

Ictal headache, or mainly orbital pain, may occur and often precedes the visual or other ictal occipital symptoms in a small number of patients. Consciousness in not impaired during the elementary or complex visual hallucinations, blindness and other occipital seizure symptoms (simple focal seizures), but may be disturbed or lost in the course of the seizure, usually prior to eye deviation or convulsions.

Ictal syncope is rare.4

Patient note A 10-year-old child had frequent visual seizures of elementary hallucinations and blindness. He also had 4 episodes of ictal syncope in its most pure and challenging form: brief (1–2 minutes), transient loss of consciousness and postural tone with pallor of sudden onset and sudden recovery. They occurred whilst sitting or standing. He falls down and becomes unresponsive. One episode witnessed by a physician was described “clumsiness, vacant, unresponsive for a minute or so. No convulsions”. Another one was witnessed by his parents: “he was next to us in a shop. We heard a bang and saw him on the ground. Colour white not blue. He was out for a few seconds.” He never had anything like this in the next 9 years of follow-up. This is case 26 in ref.1

Occipital seizures of Gastaut-type childhood occipital epilepsy may rarely progress to extra-occipital manifestations, such as hemiparaesthesia. Spreading to produce symptoms of temporal lobe involvement is exceptional and may indicate a symptomatic cause.

Postictal headache, mainly diffuse, but also severe, unilateral and pulsating, or indistinguishable from migraine headache occurs in half of the patients and, in 10% of these cases, it may be associated with nausea and vomiting.1,110

Patient note I then have a left-sided severe throbbing headache for an hour or so.

Circadian Distribution

Visual seizures are predominantly diurnal and occur at any time of the day. Longer seizures, with or without secondary hemi- or generalised convulsions, tend to occur either during sleep, causing the patient to wake, or after awakening. Thus, some children may have numerous diurnal visual seizures and only a few secondarily GTCS that are exclusively nocturnal or that occur on awakening.

Frequency of Seizures

Patients experience brief visual seizures frequently (often several each day or weekly), if untreated. However, propagation to other seizure manifestations, such as focal or generalised convulsions, is much less frequent (monthly, yearly or exceptionally).

Aetiology

There may be an increased family history of epilepsies (37% of cases) or migraine (16% of cases),107 but a family history of similar seizures is exeptional.111 G-ICOE is considered to be a late onset phenotype of BCSSS (see page 262).

Pathophysiology

The seizures are purely of cortical occipital origin.109

The mechanisms of postictal headache, which is common even after minor idiopathic or symptomatic visual seizures, with or without a predisposition to migraine, are unknown. It is likely that the occipital seizure discharge triggers a genuine migraine headache through trigeminovascular or brain-stem mechanisms.110;112
The occipital paroxysms are bilateral and synchronous when they occur, because they are activated in the bi-occipital regions by the elimination of fixation and central vision (Figures 9.4;9.12). They are not due to a thalamocortical mechanism, driven by a thalamic pacemaker, as proposed by Gastaut and Zifkin.

**Diagnostic Procedures**

By definition, all tests other than the EEG are normal. However, high resolution MRI is probably mandatory, because of the high incidence of symptomatic occipital epilepsies with the same clinical EEG manifestations.

**Electroencephalography**

The interictal EEG shows occipital paroxysms often demonstrating fixation-off sensitivity (Figure 9.12). However, some patients may have only random occipital spikes, while others may have occipital spikes in only the sleep EEG and a few may have a consistently normal EEG. Centrotemporal, frontal and giant somatosensory spikes may occur, but less often than in PS. Whether occipital photosensitivity is part of this syndrome or not is debated (page 469).

Ictal EEG, preceded by regression of occipital paroxysms, is characterised by the sudden appearance of an occipital discharge that consists of fast rhythms, fast spikes or both (Figures 9.9 and 9.13). This is of much lower amplitude than the occipital paroxysms. Elementary visual hallucinations are related to the fast spike activity that may spread to the other hemisphere. Complex visual hallucinations may occur when the discharge is slower. In oculoclonic seizures, spikes and spike and waves are slower and a localised ictal fast spike rhythm may occur prior to the deviation of the eyes. Ictal EEG during blindness is characterised by pseudoperiodic slow waves and spikes, which differs from that seen in ictal visual hallucinations.

There are usually no postictal abnormalities.

**Differential Diagnosis**

The differential diagnosis of G-ICOE is mainly from probably symptomatic (cryptogenic) or symptomatic occipital epilepsy, coeliac disease, migraine with aura, and basilar or acephalgic migraine where misdiagnosis is high.

The differential diagnosis from migraine should be easy if all clinical elements are properly assessed and synthesised as described in Table 12.3, page 427.

**Important note** Basilar migraine with occipital spikes does not exist; the relevant reports described cases with genuine G-ICOE (see page 427) imitating basilar migraine.

**Patient note** In a typical case of G-ICOE, an 8-year-old child started complaining of elementary visual hallucinations that consisted of unilateral multicoloured small circles lasting for a few seconds. Their frequency and duration increased over the next few months and these symptoms were followed by diffuse headache. A diagnosis of migraine with aura was made and relevant treatment was initiated without success. On the contrary, the child had two seizures in which the same visual hallucinations became so intense as to obscure his vision, followed by deviation of the eyes and head. An EEG showed bilateral occipital paroxysms with fixation-off sensitivity. MRI was normal. The child had two episodes of complete blindness without convulsions. The diagnosis of basilar migraine was suspected, but visual hallucinations continued, one of them ending with GTCS. No more seizures of any type occurred after treatment with carbamazepine, which continued for 3 years. By 20 years of age, the patient was attending university, was well, free of seizures with no treatment and had a normal EEG.

Symptomatic occipital epilepsy often imitates G-ICOE; neuro-ophthalmological examination and brain imaging may be normal, and high resolution MRI may be required to detect subtle lesions. Occipital seizures of mitochondrial disorders, Lafora disease and coeliac disease should be considered (see details on pages 422–3).
Occipital paroxysms are by far more common in G-ICOE than PS (Figures 9.7 and 9.12). They are defined as ‘long runs of repetitive, high amplitude sharp and slow wave complexes in the occipital regions, which are morphologically similar to the CTS’. The word ‘paroxysm’ in EEG terminology is a ‘Group of waves that appears and disappears abruptly, which is clearly distinguishable from background activity by its different frequency, morphology or amplitude’. The individual complexes of the occipital paroxysms show a diphasic spike component with a main surface negative peak in the occipital electrodes and an amplitude often exceeding 200 µV. This is followed by a smaller positive peak and a high amplitude, negative slow wave. Occipital paroxysms are often bilateral and synchronous, but may also be unilateral and predominantly right sided. Occipital paroxysms appear and persist only when the eyes are closed and may not be recorded in children who keep their eyes open during the EEG. This is because occipital paroxysms are activated by the elimination of fixation and central vision (fixation-off sensitivity, Figure 9.12).

Occipital paroxysms may persist long after remission of clinical seizures, but also may not be detected interictally during the active seizure phase.

Random occipital spikes that do not have the repetitive character of occipital paroxysms are more common than occipital paroxysms.

Reminder about Occipital Spikes

Occipital spikes are not pathognomonic of a particular syndrome, because they also occur in a variety of organic brain diseases with or without seizures, in children with congenital or early onset visual and ocular deficits, and even in 0.5–1.2% of normal preschool-age children. They are common in young children with a peak age at first discovery of 4–5 years and “tend to disappear in adult life, and the subsidence of the EEG abnormality is usually accompanied by a cessation of seizures”.

Prognosis

The prognosis is unclear, though available data indicate that remission occurs in 50–60% of patients within 2–4 years from onset. Seizures show a dramatically good response to carbamazepine in more than 90% of patients. However, 40–50% of patients may continue to have visual seizures and infrequent secondary GTCS, particularly if they have not been appropriately treated with carbamazepine. Rarely, atypical evolutions to epilepsy with continuous spike waves during slow wave sleep with cognitive deterioration have been reported.

Although no significant differences were found in basic neurophysiological functions between patients with G-ICOE and control groups, patients’ performance scores for attention, memory and intellectual functioning were lower.

Management

In contrast to other phenotypes of the BCSSS, patients with Gastaut-type childhood occipital epilepsy often suffer from frequent seizures and therefore medical treatment, mainly with carbamazepine, is probably mandatory. Secondary GTCS are probably unavoidable without medication.

A slow reduction in the dose of medication 2–3 years after the last visual or other minor or major seizure may be advised, but if visual seizures reappear, treatment should be restored.

Other Phenotypes of Benign Childhood Seizure Susceptibility Syndrome

There are well-documented reports of children suffering from benign childhood focal seizures with clinical EEG manifestations that can not be classified as typical cases of Rolandic epilepsy, PS or Gastaut-type childhood occipital epilepsy. Their existence verifies the unified concept of BCSSS. They may represent atypical presentations of the recognised syndromes within the BCSSS.
Benign childhood seizures with affective symptoms are a rare clinical phenotype of the BCSSS. This disorder has features in common with both PS (behavioural and autonomic symptoms) and Rolandic epilepsy (arrest of speech and hypersalivation). Dalla Bernandina et al. consider ‘benign partial epilepsy with affective symptoms’ a relatively rare variant of RS.

**Demographic Data**

The onset of afebrile seizures occurs at 2–9 years of age and both sexes are equally affected. One-fifth of patients have febrile seizures and family history of epilepsy is common (36%).

**Clinical Manifestations**

Seizures are brief, but multiple, diurnal and nocturnal, and manifest with terror and screaming, autonomic disturbances (pallor, sweating, abdominal pain, salivation), chewing and other automatisms, arrest of speech and mild impairment of consciousness.

The predominant seizure symptom is sudden fear or terror:

> “This terror was expressed by the child starting to scream, to yell or to call his mother; he clung to her or to anyone nearby or went to a corner of the room hiding his face in his hands. His terrorised expression was sometimes associated with either chewing or swallowing movements, distressed laughter, arrest of speech with glottal noises, moans and salivation, or some kind of autonomic manifestation, such as pallor, sweating or abdominal pain, that the child expressed by bringing his hands onto his abdomen and saying ‘It hurts me, it hurts me’. These phenomena were associated with changes in awareness (loss of contact) that did not amount to complete unconsciousness.”

The seizures are brief, lasting between 1–2 min and a maximum of 10 min.

Half of the children have frequent (several times a day) seizures from the onset, which may occur with the same semiology whether awake or asleep. Some children may have brief and infrequent nocturnal RS at the same time as the affective attacks. Generalised seizures do not occur.

**Diagnostic Procedures**

All tests, apart from the EEG, are normal.

The interictal EEG shows high amplitude sharp and slow wave complexes, which are morphologically similar to Rolandic spikes and are located around the fronto-temporal and parieto-temporal electrodes. In common with the other benign childhood focal seizures, EEG abnormalities are exaggerated by sleep and may be associated with generalised discharges.

Ictal EEG discharges are localized to the fronto-temporal, centrotemporal or parietal areas or may be diffuse. They are stereotypical in each individual patient.

**Prognosis**

The response to treatment is excellent and remission is reported to occur within 1–2 years. At the active stage of the disease, behavioural problems may be prominent, but subside with the seizures.

**Treatment**

In the active phase of the disease and because of frequent seizures antiepileptic medication, mainly with carbamazepine, may be needed.

Benign Childhood Epilepsy with Parietal Spikes and Frequent Giant Somatosensory Evoked Potentials

Benign childhood epilepsy with parietal spikes and frequent giant somatosensory evoked potentials may be another
phenotype of BCSSS. The defining features are EEG spikes in the parietal regions, which are often elicited by tactile stimulation. However, somatosensory evoked giant potentials/spikes (page 229) are not specific for any syndrome as they also occur in 10–20% of children with RS, in a few patients with PS (Figure 9.4) and in children without seizures (Figure 9.4).

Clinically, patients suffer from versive seizures of the head and body, often without impairment of consciousness. These are mainly diurnal and infrequent. Multiple daily seizures and focal status epilepticus are exceptional.

**Benign Childhood Focal Seizures Associated with Frontal Spikes or Midlines Spikes**

Benign childhood focal seizures associated with frontal spikes or midline spikes have been described and long follow-up reports have confirmed a benign course, but no systematic studies have been published. However, it should be remembered that EEG spike foci of various locations are also seen in Rolandic epilepsy and more commonly in PS. Midline spikes are more common in children than in adults and they are not specific for any type of epilepsy. Of six children with at least one EEG having midline spikes only, five had normal development with febrile seizures (one case), Rolandic epilepsy (one case), PS (one case), a single occipital complex partial seizure (one case) and brief seizures with loss of consciousness only (one case). The only symptomatic case had generalised convulsions.

**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 BCSSS. In terms of age, this is on the borderline between benign infantile seizures (page 124) and BCSSS. Age at onset is in the first 2 years of life with both sexes equally affected. Infants are normal and all tests other than EEG are normal. Seizures consist mainly of staring, motion arrest, facial cyanosis, loss of consciousness and stiffening of the arms. Clonic convulsions and automatisms are rare. Duration is from 1–5 min. Seizures are mainly diurnal (but may also occur during sleep) and may occur in clusters, but are generally infrequent (1–3 seizures/year).

Interictal EEG abnormalities are seen only in non-REM sleep and consist of small, mostly singular, spikes and waves localised in the vertex and central electrodes.

There is a strong family history of epilepsy with benign epilepsies prevailing.

The prognosis is excellent with remission of seizures, normal development and normalisation of the EEG before the age of 4 years.

**Management of Benign Childhood Focal Seizures**

Short- and long-term treatment strategies of benign childhood focal seizures are empirical and there is no consensus regarding their management. However, current practice parameter guidelines for febrile seizures, if appropriately modified, may be the basis for similar guidelines in benign childhood focal seizures and, particularly, Rolandic epilepsy and PS. Based on the risks and benefits of the effective therapies, continuous antiepileptic medication is not recommended for children who have had only one or brief seizures. Most clinicians treat recurrent seizures with carbamazepine, but in exceptional cases this may worsen seizures. Lengthy seizures are a medical emergency; rectal diazepam is prescribed for home administration. Recurrent and lengthy seizures create anxiety in parents and patients, and, as such, appropriate education and emotional support should be provided.

**Acute Management of a Child with Prolonged Seizures**

Control of the seizure is paramount. On the rare occasions that the child is febrile, treatment of possible fever and the underlying illness is also important.

Long-lasting seizures (> 10 min) or status epilepticus (> 30 min–hours) is a feature in two-thirds of children (70%) with PS. This is a genuine paediatric emergency that demands appropriate and vigorous treatment as for status epilepticus. Early, usually parental administration of appropriate AEDs is more effective than late emergency
The treatment of prolonged febrile seizures and febrile status epilepticus is described on pages 123.

**Prophylactic Management of Benign Childhood Focal Seizures**

Continuous anticonvulsant treatment is usually not recommended. Although there are effective therapies that could prevent the occurrence of additional seizures, the potential adverse effects of such therapy are not commensurate with the benefit. The great majority of children with benign focal seizures do not need anticonvulsant treatment even if they suffer lengthy seizures or have more than two recurrences. The risks are small and the potential side effects of drugs appear to outweigh the benefits.

In patients with recurrent seizures and/or when parental anxiety associated with seizures is severe, small doses of antiepileptic medication may be effective in preventing recurrence. There is no convincing evidence, however, that any therapy will alleviate the possibility of recurrences. In deciding management for a child with benign childhood focal seizures the following should be considered:

- Most children have an excellent prognosis: 10–30% of patients may have only a single seizure and seizures may be infrequent (usually 2–10) in 60–70%. However, 10–20% of patients may have frequent seizures, which are sometimes resistant to treatment.
- Remission of benign childhood focal seizures is expected in all patients by 15–16 years of age at the latest.
- The possibility of future epilepsy is a most unlikely event and probably not higher than that in the general population.
- There is no evidence that the long-term prognosis is worse in untreated children, though they may not be protected against seizure recurrences.
- Some children become frightened even by simple focal seizures and some parents are unable to cope with the possibility of another fit despite firm reassurances.
- Persistence and frequency of EEG functional spikes are not predictive of clinical severity, frequency and degree of liability to seizures.

Continuous prophylaxis consists of daily monotherapy using any antiepileptic drug with proven efficacy in focal seizures, usually carbamazepine. However, carbamazepine may exaggerate seizures in a minority of children with RS. Recently, sulthiame has been revived as an excellent drug for the treatment of benign childhood epilepsy with centrotemporal spikes. Sulthiame may be the drug of choice in children with BCSSS and cognitive impairment, because it often normalises the EEG. Of the newer drugs, levetiracetam has been reported as therapeutic in the few children in whom it has been tried. Lamotrigine may be associated with significant seizure and cognitive deterioration.

**Stopping Medication**

Methods of withdrawing medication differ between experts, though all agree that there is no need to continue medication 1–3 years after the last seizure and certainly not after 14 years of age when most benign childhood focal seizures remit or 16 years of age when they are practically non-existent. My practice is to start gradual withdrawal of medication 2 years after the last seizure, making sure that the child does not have any minor seizures. However, I do not adhere to fixed rules and may continue medication until the age of 13–15 years depending on the severity, frequency and age at onset of seizures. Thus, in a child that had frequent, severe and difficult to control fits in early childhood, I would not stop medication if the child had a seizure-free period of 2–3 years by the age 7 years. Conversely, for a child who had three or four nocturnal seizures at age 11 and 12 years, I would certainly slowly discontinue medication after a 2-year seizure-free period. I advise very slow withdrawal, reducing the dose in monthly steps so that the drug is completely discontinued approximately 6 months later. The reason for this is that I expect that any possible seizure recurrence during the process of very slow drug withdrawal would manifest with mild, brief and simple focal without secondary generalised convulsions. In the case of barbiturates and diazepines, slow withdrawal of
medication is mandatory in order to avoid risking a possible withdrawal seizure.

Parental Attitude, Reaction and Education in Benign Childhood Focal Seizures

By Thalia Valeta

The traditional goal of care for children with epilepsies has been optimal seizure control. This has now been widely broadened to include optimal health-related quality of life outcomes. These outcomes assess the physical, mental and psychological functioning of the epileptic child within his/her family, educational and social environment, and reflect the impact of seizures and their treatments on patients and their parents. Parents are integral to the functioning and quality of life of their children and, therefore, the health-related quality of life of a child largely depends on the parents’ attitudes, reactions, education and adjustment. Thus, it is crucial that parents are given sufficient time and opportunity to discuss their concerns with the specialists, who should provide the following:

- an accurate diagnosis of the child’s condition including precise cause, risk of recurrence, prognosis, and type and length of management, as well as possible hereditary factors and adverse reactions of treatments
- an assessment of the immediate and future effects on the physical, mental, behavioural, educational, family, social and vocational aspects of their life
- information about the parental role in preventing (leisure and other activities) or early detection of an epileptic attack including means of terminating a seizure, particularly if this is lengthy and life threatening.

The adjustment and other problems experienced by children with a diagnosis of epilepsy and their parents have always been a concern to clinicians and health-care providers. It is understandable that this has been primarily focused on parents of children with severe and intractable epilepsy, who face profound challenges in dealing with their child’s frequent and severe seizures, the additional physical, social and psychological problems, and ongoing quest of seizure control through a variety of drugs, diet and surgery. However, the significant burden of anxieties placed on the parents of children with benign epileptic conditions has been less emphasised because, comparatively, they are more fortunate with regard to prognosis, management, current and future prospects, and responsibilities. The only exception to this is in relation to febrile seizures where parental reactions and concerns have been well documented and properly addressed. These reactions are often severe and contrast with the physician’s perception of febrile seizures as a uncomplicated and benign condition. In one such study, 52 parents of a child with febrile seizures were interviewed about their immediate and long-term reactions. Most parents knew little about febrile convulsions before the fit and most of them thought the child was dying (77%), was suffocating or had meningitis (15%). Afterwards, parental behaviour altered: 60% experienced restless sleep, 29% had dyspepsia, and 6% watched over their child at night and 8% when feverish. Parents with previous knowledge of febrile seizures took more appropriate measures, but only 21% positioned the child correctly during the seizure.

It is now part of good clinical practice to provide the parents of children with febrile seizures specific information about fever and febrile seizures, and comprehensive instructions about what to do if a seizure happens again. The parents of children with recurrent febrile seizures should be advised on what to do if a seizure recurs since early treatment provided by parents is more effective than late treatment in an emergency facility. The guidelines also provide room for starting anti-epileptic drug treatment in “the unlikely event of parental insistence” or “situations in which parental anxiety associated with seizures is severe”.

Talking extensively to parents of children with benign childhood focal seizures I realised that, despite the fact that these seizures are of excellent prognosis, they are a dramatic experience for patients and their parents. Consequently, I have initiated an on-going study aiming to define and analyse the psychosocial effects of benign childhood focal seizures on parents and children. I have designed a questionnaire in order to identify the main concerns of parents about the seizure itself and its impact on the child’s development and future. The questionnaire includes questions on beliefs and attitudes about epilepsy, as well as concerns about the prognosis, necessity of evaluation and information on daily care. I hope that the results of my study will assist the patient and parents, inform the physician and, consequently, help to improve the treatment outcome.
Questionnaire on Parental Attitude and Reactions to Benign Childhood Focal Seizures

I. Parental reaction during and after the seizure prior to medical attention
   a. What did you think was happening to your child?
   b. What was your reaction and how did you help your child?
   c. How did you feel?

II. Parental reaction after the child had recovered
   a. How did you feel?
   b. What do you think about the future?
   c. Did you think that this was a one-off event?
   d. Did you take any further actions?

III. Parental reaction after consultation with the attending physician
   a. Was this useful?
   b. How did you feel? (1) re-assured, (2) uncertain or (3) anxious.

IV. Short- and long-term effects of the event on you as parents, other members of the family and the child
   a. Did you discuss the event with the child and what did you say?
   b. Did your attitude change towards the child?
   c. If yes, in what way did it change?
   d. How did the event affect you over the following weeks/months/years (fears, sleep, appetite, work, relation with the child and other members of your family)?
   e. Did the event change the child’s behaviour and attitude?
   f. Did the event affect the family?

V. When the diagnosis of your child was established and explained to you did this affect your state of mind and your reactions?

VI. Any questions and comments that you may still have?

The commonest fears and expressions were:

- I thought he/she was dying, choking, asphyxiated, electrocuted never to come around again
- I thought he had a stroke
- I was terrified, petrified

The doctor told me that because the seizure was longer than half an hour this may affect the brain and that time will tell.

We sleep with our daughter in between us on a large bed, and we keep an eye on her as she enters and exits sleep.

The most dominant points emerging from this study are:

- Uncertainty of what this event was.
The majority of parents felt uncertain about what this event was and that they were not given sufficient information or reassurance, and some were told that the child had not had a seizure. Initially, some children were diagnosed as having encephalitis, atypical migraine, fainting, gastroenteritis or motion sickness.

- Anxiety of what the cause of the event was.

This was often associated with a feeling of guilt, either of parental acts directly associated with the event (child relatively unattended, preceding parental arguments, child involved in leisure activity that may have caused the attack), heredity or with previous events in the child’s development (birth, trauma, illness, family history of illness).

- The effect of the seizure on the child’s development. “Is this going to affect his/her brain?” Most parents were reassured that one brief seizure would not affect the child’s development. However, some parents of children with lengthy seizures were left with the impression that, because the seizure was prolonged, it may have had some adverse effect on the child and only “time will tell”.

- No specific advice was provided about the possibility of relapses and what the parents should do if such a seizure recurred.

These results indicate that there is a need for supportive family management, education and specific instructions about emergency procedures in possible subsequent seizures. Demonstrations of first aid practices for seizures are necessary. Parents of young children should be given general information about benign childhood focal seizures and, in particular, Panayiotopoulos syndrome in which seizures may last for many hours, compounded by physicians’ uncertainty regarding diagnoses, management and prognosis. Parents who have watched their child during a fit need specific information and psychological support to overcome anxiety and panic. Anxiety may result in overprotection, which interferes with parent-child separation and independence. Educating parents about the epilepsies and different types of seizure through seminars, courses and lectures will help to alleviate the social stigma surrounding these conditions, which parents often pass on to their children who are the patients. Parents must be offered training to remain calm and confident about their children’s condition in order to improve the quality of life of both the child and the family. Medicine can now profit from a holistic approach to care through counselling and helping people with epilepsies and their families. Practice parameter guidelines regarding parental management and education of children with benign seizures should be updated.

**Unified Concept for the Benign Childhood Focal Seizures (Benign Childhood Seizure Susceptibility Syndrome)**

Benign childhood focal seizures with focal EEG sharp slow wave complexes are a group of syndromes of probably one disease, which, in my opinion, share common clinical and EEG characteristics. Seizures are infrequent, usually nocturnal and remit within 1–3 years from onset. Brief or prolonged seizures, even status epilepticus, may be the only clinical event of the patient’s lifetime. Ictal autonomic manifestations, such as hypersalivation, vomiting, headache, pallor or sweating, and ictal syncope, which is unusual in other epileptic syndromes, are frequent and may occasionally appear in isolation. The clinical and EEG characteristics of one syndrome may evolve into another or a child may simultaneously develop features of another form of benign childhood focal seizures. Febrile seizures are common. Neurological examination and intellect are normal, but some children may experience mild and reversible neuropsychological problems during the active stage of the disorder. Brain imaging is normal. There are usually severe EEG abnormalities, which are disproportionate to the frequency of seizures. Epileptogenic foci, irrespective of their location, manifest as abundant, high amplitude, sharp, slow wave complexes that occur mainly in clusters. They are often bilateral, independent or synchronous, frequently combined with foci from other cortical areas or brief generalised discharges, and are exaggerated in sleep stages I–IV. A normal EEG is rare and should provoke a sleep EEG study. Similar EEG features resolving with age are frequently found in normal school-age children (2–4%) and children having an EEG for reasons other than seizures.

There is no reason to suggest that all these syndromes differ from each other merely because an ‘epileptogenic’ focus is a little anterior or posterior, lateral or medial to the centroparietal regions. A unified concept of benign childhood focal seizures is also suggested by the frequency of more than one type of benign childhood focal seizures in an affected child, siblings or both.
It is likely and I have proposed that all these conditions are linked together by a common, genetically determined, mild and reversible, functional derangement of the brain cortical maturational process. This is often clinically silent and manifests in more than 90% with EEG sharp and slow waves with an age-related localisation (Figure 9.1). The remaining minority have infrequent focal seizures with symptoms that are also localisation- and age-related and dependent. It is possible that a few of these children, with or without seizures, also have usually minor and fully reversible neuropsychological symptoms that are rarely clinically overt and that can only be detected by formal neuropsychological testing. Finally, there may be a very small number of patients (<1%) in whom this derangement of the brain maturation process may be further derailed in a more aggressive condition with seizures, neuropsychological manifestations and EEG abnormalities of various combinations and various degrees of severity, such as in atypical benign focal epilepsy of childhood, Landau-Kleffner syndrome, and epilepsy with continuous spikes and slow waves during sleep.

My overall impression is that benign childhood focal seizures, their clinical and EEG manifestations and evolutions, need appropriate prospective studies, such as those performed for febrile seizures.

**Febrile Seizures and Benign Childhood Seizure Susceptibility Syndrome**

One of the most interesting aspects of benign childhood seizures is their striking age-related sequence. Common benign seizure disorders are specific to children and do not occur in adults. The fact that children are particularly susceptible to seizures is well documented.

There are three main periods of age-related childhood susceptibility to seizures. Febrile mainly generalised convulsions first appear in early childhood at a peak age of 18–22 months. Rolandic focal seizures occur in late childhood at a peak age of 7–10 years. Panayiotopoulos syndrome covers the intermediate period, occurring at a peak of 4–5 years, and manifests with mainly autonomic seizures (Figure 9.14). Let me analyse this further as this is likely to be highly significant in our understanding of the disordered age-related maturational processes.

In the first, early period (febrile seizures), the brain is vulnerable to seizures that are triggered by fever and mainly manifest with convulsions that are commonly generalised. The second intermediate period (Panayiotopoulos syndrome) consists of spontaneous seizures that are often prolonged for hours and manifest principally with autonomic and mainly emetic symptoms. The third late period (Rolandic syndrome) consists of spontaneous focal, motor or sensorimotor seizures.

These three periods of clinical seizure susceptibility also have peculiar EEG accompaniments. The EEG is practically normal in the first period of febrile seizures, shows mainly posterior and multifocal spikes in the intermediate period of Panayiotopoulos syndrome and Rolandic spikes in the late period of the Rolandic syndrome.

All these indicate that the brain in early childhood has a low threshold to generalised convulsions provoked by fever with a relatively silent EEG spike capacity. Subsequently, the autonomic system and particularly the emetic centres become vulnerable, the seizure discharges may be self-sustained and the cortex exhibits a diffuse epileptogenicity, which is unequally distributed and mainly affects the posterior regions. Finally, in the third period of late childhood, brain epileptogenicity shrinks to around the Rolandic regions to produce the distinctive clinical and EEG manifestations of the Rolandic syndrome.

These are incontrovertible facts that tell us something very important about the developing brain that we have not yet explored. We should also consider the neonatal and the early infantile periods, because they also have their own peculiarities as indicated by the benign familial and non-familial neonatal seizures that occur around the first few days of life, and the benign infantile focal seizures of the Watanabe-Vigevano syndrome. This point is exemplified by reports of children with neonatal seizures who later developed Rolandic or PS. Maihara et al. described a family with benign familial neonatal seizures in which two siblings later developed RS and EEG centrotemporal spikes. Lada et al. reported an otherwise normal boy who first had benign neonatal seizures, then two febrile seizures at the age of 18 months and 3 years. This was followed by a brief autonomic seizure of PS with ictal vomiting and deviation of the eyes occurred during sleep at 6 years of age; the EEG showed occipital paroxysms. I have also described a boy who, at 8 weeks old, had three focal seizures of right-sided convulsions involving the face and upper limbs (benign infantile non-familial seizures of Watanabe-Vigevano syndrome). Subsequent EEGs were normal and
treatment stopped at age 10 months. He was well until the age of 7 years when he started having RS seizures and later developed epilepsy with continuous spikes and waves during slow wave sleep (see Figures 6.1 and 7.12). The brain MRI was normal (case 17.2 of Panayiotopoulos1).

**Benign (Isolated) Focal Seizures of Adolescence**

Benign (isolated) focal seizures of adolescence156–163 constitute an idiopathic, short-lived and transient period of seizure susceptibility during the second decade of life.

**Demographic Data**

This is a seizure susceptibility of the second decade of life with a peak at 13–15 years of age. There is a 71.2% male preponderance. According to Loiseau and Louiset (1992)157 a quarter of focal seizures with an onset between 12 and 18 years of age have a benign course (i.e. they are single or occur in a cluster of up to five seizures during 36 hours, never to occur again). The disorder may account for between 7.5%158 and 22%160 of patients having simple focal seizures in the second decade of life. Around 200 cases have been described.157;160–162;164;165

**Clinical Manifestations**

The syndrome manifests with a single or a cluster of 2–5 focal, mainly motor and sensory, seizures, which progress to secondary generalized tonic-clonic convulsions in 50% of cases. There are no epileptic events preceding or following this limited seizure period, which lasts for no more than 36 hours. The physical and mental states of the patients are normal.

The seizures are, by definition, focal, but the temporal lobes are rarely involved.

**Consideration on Classification**

Benign (isolated) focal seizures of adolescence are not a recognised syndrome in the new diagnostic scheme.2 Loiseau and Jallon (2002)162 consider that this is a syndrome of “isolated focal seizures of adolescence”, which is in accordance with my proposition to classify them among “Conditions with epileptic seizures that do not require a diagnosis of epilepsy”.2

Most of the seizures are diurnal (87%). The commonest ictal clinical manifestations are motor, usually without Jacksonian marching, and somatosensory. Visual, vertiginous and autonomic symptoms are reported in one-fifth of cases. Experiential phenomena, such as those seen in temporal lobe seizures, practically never occur. The teenager is fully aware and can give a reliable account of the onset of the clinical manifestations (simple focal seizures) in the majority of episodes (88%). However, consciousness rarely remains intact throughout the whole event; the seizures usually evolve to impaired cognition, and/or to secondary GTCS, which occur in 50% of cases.

**Diagnostic Procedures**

Laboratory tests and brain imaging are normal. The EEG may show some minor, non-specific, abnormalities without spikes or focal slowing. In a recent report, 9 of 37 cases had functional spikes,161 which is incompatible with this syndrome; these patients probably suffered from benign childhood focal seizures as described earlier in this chapter.

**Differential Diagnosis**

These patients are difficult to diagnose, as there are no specific features at onset to differentiate them from others with similar clinical manifestations, but of different aetiology, such as symptomatic or cryptogenic focal epilepsies. My practice is to investigate all adolescents with onset of focal seizures with MRI and EEG, which, if normal, would make the diagnosis of benign focal seizures of adolescence more likely. A definitive diagnosis cannot be made until the patient has been free of seizures for 1–5 years.157;160

**Prognosis**
The prognosis is excellent; in 80% of patients, there is a single, isolated seizure event and, in the remaining 20%, a cluster of 2–5 seizures all occurring within 36 hours.

**Management**

No drug treatment is needed because only one or a cluster of 2–5 focal seizures (which can not be predicted) occurs within 36 hours.

**References**


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**Footnotes**

* **Author’s note:** Searching through appropriate sources of information, I found no work or reference to the reactions and needs of parents with children of benign focal seizures. The on-going study of Thalia Valeta is the first of this kind.
Figure 9.1

Schematic presentation of benign childhood seizure susceptibility syndrome.*

Top: Over 90% of functional spikes are clinically silent.

Middle: Prevalence of functional spikes by location. Centropetalal spikes predominate followed by occipital spikes.

Bottom: Rolandic seizures (64%) are 2.5 times more common than PS (25%), but this figure may change with increasing awareness of PS and the inclusion of less typical cases.

* The percentages are approximate estimations from available relevant clinical and EEG data on the prevalence of clinical phenotypes of benign childhood focal seizures and functional spikes in childhood.\textsuperscript{1,4}
Figure 9.2

Video EEG of an 11-year-old girl with Rolandic seizures who has been in remission since the age of 8 years.

Top: High amplitude centrotemporal spikes (in fact these are central spikes) occur independently on the right or left, and are markedly exaggerated during natural sleep.

Top extreme right: Typical morphology and polarity of CTS in Laplacian montage.

Bottom: ESES, which are evoked by tapping fingers or toes. Note that their location corresponds to the location of the activating stimulus.

Bottom extreme right: ESES from another patient, which were evoked by electrical stimulation of the right thumb (onset at arrow). Peak latency of the somatosensory spike is 58 ms.
Figure 9.3

Centrotemporal spikes are mainly Rolandic not Centrotemporal.

Top, middle and bottom: The same EEG sample is shown in 3 different montages.

This is from an 8-year-old boy referred for an EEG because of “recent GTCS and a 2-year history of unilateral facial spasms. Previously, the EEG and CT brain scan were normal. No medication. Focal seizures with secondarily generalised convulsions?”

The EEG showed frequent clusters of repetitive centrotemporal spikes on the left. Because the spikes appeared to be of higher amplitude in the temporal electrode (T3) (black arrows), the technologist rightly applied additional

1:35
electrodes at C5 and C6 (Rolandic localisation). This showed that the spike is of higher amplitude in the left Rolandic region (C5) (open arrows). Another EEG 16 months later, showed a few small spikes in the right frontal and central midline electrodes.
Activation of functional centrotemporal and occipital spikes. Facing page

Top: Video EEG of a 6-year-old girl with headaches and abdominal pains of recent onset (case 16.2 in ref¹). Neurological examination and MRI were normal. Symptoms improved over the following year. She never had
seizures and her development was normal. EEG showed normal background with the following abnormalities:

(a). Spontaneously central spikes, occurring independently on the right or left.

(b). High amplitude central spikes elicited by somatosensory stimulation of the contralateral side. Simultaneous stimulation of the fingers of the hands by the patient herself elicited simultaneous bilateral central spikes.

(c). Brief, mainly anterior, bursts of polyspikes.

(d). Brief and high amplitude generalised discharges of 3–5 Hz slow waves interspersed with small spikes or small polyspikes.

Note that the ESES are bilateral and synchronous when the stimulus is also bilateral and synchronous (tapping together and simultaneously the palmar tips of her fingers). Unilateral tapping evoked contralateral ESES.

Middle: ESES of a patient with PS (case 17 in ref4). At the age of 8 years, this boy had a single nocturnal seizure, which started with repetitive vomiting and “he was lost”. He then clenched his teeth and became rigid, but there were no clonic convulsions. The last follow-up, at the age of 14 years, disclosed no further seizures and normal development, though school performance was moderate. Three EEGs showed right CTS that were also evoked by somatosensory stimuli. Occipital spikes were never observed.

Bottom: Sample EEG from a patient with PS and occipital paroxysms (case 6 in ref4). Occipital paroxysms are bilateral and synchronous, because they are activated in the hyperexcitable occipital cortices by the elimination of central vision and fixation.
Interictal EEG

Fp2 - F4
F4 - C4
C4 - P4
P4 - O2

Fp1 - F3
F3 - C3
C3 - P3
P3 - O1

Fp2 - F8
F8 - T4
T4 - T6
T6 - O2

Fp1 - F7
F7 - T3
T3 - T5
T5 - O1

Ictal EEG

\[ 150 \mu V \quad 1 \text{ sec} \]

↑ Onset of the discharge

09:59

\[ 150 \mu V \quad 1 \text{ sec} \]

△ Onset of clinical manifestations

15.47

End of seizure

16.32

200 \mu V \quad 1 \text{ sec}
Figure 9.5

Video EEG of a 10-year-old girl with Rolandic seizures (case 5.1 in ref^1).

*Top:* High amplitude right-sided centrotemporal spikes (C5 and C6 electrodes were not applied).

*Bottom:* Onset of ictal discharge in the right centrotemporal regions during sleep. Arrow shows onset of clinical manifestations that started with contractions of the left facial muscles (note muscle artefacts on the left),
Figure 9.8

EEG of a 4-year-old boy with autonomic status epilepticus recorded from onset to termination.

*Top:* High amplitude spikes and slow waves are recorded from the bifrontal regions prior to the onset of the electrical discharge, which is also purely bifrontal (arrow).

*Bottom:* First clinical symptoms with three or four coughs and marked tachycardia appeared 13 min after the onset of the electrical discharge, when this had become bilaterally diffuse. Subsequent clinical symptoms were tachycardia, ictus emeticus (without vomiting) and impairment of consciousness. No other ictal manifestations occurred until termination of the seizure with diazepines 70 min after onset.

Another lengthy autonomic seizure was recorded on video EEG 1 year later. The onset of symptoms was different with mainly tachycardia and agitation.
From Panayiotopoulos (2004) with the permission of the Editor of *Epilepsy and Behaviour*. Figure courtesy of Dr Michael Koutroumanidis, MD from the Department of Clinical Neurophysiology and Epilepsies, Guy’s & St Thomas’ NHS Trust, UK.
Figure 9.9

Ictal EEGs in Panayiotopoulos syndrome (top) and Gastaut-type childhood occipital epilepsy (bottom).

Top: Samples of continuous EEG recording from the onset to the end of a 9-minute seizure during sleep stage II in an 8-year-old girl. Clinically, the seizure manifested with awakening, eyes opening, frequent vomiting efforts and complaints of frontal headache. The ictal EEG started with remission of the interictal occipital paroxysms and the appearance of occipital sharp rhythms progressing to monomorphic rhythmic theta activity in the bi-occipital regions, but mainly involving the right hemisphere in a wider posterior distribution. The slow activity slowed down with the progress of the seizure and ended without postictal abnormalities. The ECG showed significant tachycardia during the ictus.

Bottom: Ictal EEG during a visual seizure in a boy with Gastaut-type childhood occipital epilepsy. The seizure starts in the left occipital region with fast rhythms associated with visual symptoms. This spreads, 4 s later, to the parietal regions and the child sees a bundle of coloured balloons swinging in his right hemifield. This lasted for 40 s and was followed by slow waves that progressively became slower and diffuse over the whole brain. At this stage, he complained of clouded vision. This boy was normal physically and intellectually, and also had a normal CT brain scan. At the age of 3 years, he had a nocturnal, left hemiconvulsion. His first EEG showed occipital paroxysms with fixation-off sensitivity. Since the age of 4 years, he had started having frequent, brief visual seizures (simple, coloured, visual hallucinations) provoked by sudden darkness.

From Beaumanoir (1993) and reproduced with the kind permission of the author and the publisher John Libbey.
Figure 9.6

EEG variability in Panayiotopoulos syndrome in 6 children with autonomic seizures.

Samples from EEGs of six children with typical clinical manifestations of PS. Spikes may occur in all electrode locations, and they are usually of high amplitude and frequent or repetitive (cloned-like repetitive multifocal spike wave complexes), but may also be small and sparse. Brief generalised discharges of small spikes and slow waves may be present.
Occipital paroxysms in their classical form with fixation-off sensitivity from the initial study of Panayiotopoulos (1981).

**Top:** EEGs from two patients with PS and G-ICOE. In routine EEG, repetitive, high amplitude, occipital sharp and slow wave complexes (occipital paroxysms) occur immediately after the eyes are closed and persist for as long as the eyes remain closed. The EEG normalises immediately after the eyes are opened and for as long as the eyes remain open. The activation of the occipital paroxysms is due to the elimination of central vision and fixation (left of the vertical bar, symbol of eyes with glasses) and inhibition by fixation (right of the vertical bar, symbol of eyes without glasses).

**Bottom:** Effect of darkness on occipital paroxysms:
(a) Complete darkness activates the occipital paroxysms even when eyes are open.
(b) The occipital paroxysms become continuous in darkness irrespective of whether the eyes are open or closed.

Modified from Panayiotopoulos (1981) and reproduced with the permission of the Editor of *Neurology*. 

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**Figure 9.7**

Case 1 Panayiotopoulos Syndrome | Case 2 Gastaut type idiopathic childhood occipital epilepsy

EEGs from two patients with PS and G-ICOE. In routine EEG, repetitive, high amplitude, occipital sharp and slow wave complexes (occipital paroxysms) occur immediately after the eyes are closed and persist for as long as the eyes remain closed. The EEG normalises immediately after the eyes are opened and for as long as the eyes remain open. The activation of the occipital paroxysms is due to the elimination of central vision and fixation (left of the vertical bar, symbol of eyes with glasses) and inhibition by fixation (right of the vertical bar, symbol of eyes without glasses).

Effect of darkness on occipital paroxysms:
(a) Complete darkness activates the occipital paroxysms even when eyes are open.
(b) The occipital paroxysms become continuous in darkness irrespective of whether the eyes are open or closed.

Modified from Panayiotopoulos (1981) and reproduced with the permission of the Editor of *Neurology*. 

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EEG of a 10-year-old girl with autonomic seizures and autonomic status epilepticus identical to those of PS (case 60 in ref^4). The neurological findings and development were normal. However, the EEG had features that were markedly different to the idiopathic cases (rectangle). This prompted an MRI examination, which showed an extensive dysembryoblastic neuroepithelial tumour in the right temporo-parieto-occipital regions, corresponding to severe EEG abnormalities in the same areas.

Since 7 years of age, she had experienced about 10 brief nocturnal seizures lasting 5–10 min each. During these episodes, she jumps out of bed, calls her parents, and complains of feeling sick and of headache. She screams and vomits, her eyes stare and roll, her eyelids blink, her pupils are dilated and she sweats profusely.

A dramatic diurnal autonomic status epilepticus occurred at school. She complained of frontal headache and she had gone out for fresh air when she started feeling “funny”. She screamed that she was hot, and she was sweating and vomiting. Subsequently, she became vacant, her speech slurred slightly, her eyes twitched and she dribbled a lot. She asked for water, but did not drink. She talked “gobbledygook” and gradually got worse. Her head deviated to the left and she became unresponsive. This was followed by a series of left hemiconvulsions lasting 5–10 min each. Convulsions were stopped with intravenous diazepam. She gradually recovered. The neurological examination was normal. She was amnesic of the events the next morning and she looked well, complaining only of photophobia.

The CT brain scan was normal. CSF and other relevant investigations were normal. She received triple therapy for encephalitis for 4 days after which she was discharged home.
Figure 9.11

Elementary visual hallucinations as perceived and drawn by patients with visual seizures.

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Occipital paroxysms in their classical form with fixation-off sensitivity in a 10-year-old boy with Gastaut-type childhood occipital epilepsy (case 26 in ref 1).

Occipital paroxysms occur as long as fixation and central vision are eliminated by any means (eyes closed, darkness, +10 spherical lenses, Ganzfeld stimulation). Under these conditions, eye opening is not capable of inhibiting the spikes. Symbols of the eyes open or closed without glasses denote that the recording was made with the lights on and whenever fixation was possible.

Symbols of the eyes open or closed with glasses denote that the recording was made when fixation and central vision were eliminated by any of the above means.

From Panayiotopoulos 1999 and reproduced with the permission of the publisher, John Libbey.
Ictal video EEG telemetry of an 11-year-old girl with frequent daily visual seizures since 7 years of age. The seizures were intractable to any appropriate medication. The child had learning difficulties, but all appropriate tests, including high resolution brain MRI, were normal. This may be a case of probable symptomatic occipital epilepsy rather than G-ICOE.

Note that the first ictal symptom is blurring of vision. The ictal EEG starts with small bi-occipital spikes followed by symmetrical, episodic, fast activity in the posterior regions.
Figure 9.14

Diagramatic age-related presentation of febrile, Rolandic and Panayiotopoulos syndromes.