Benign childhood focal seizures and related epileptic syndromes

Benign childhood focal seizures and related epileptic syndromes are the most common 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. Rolanic seizures are widely recognised. Panayiotopoulos syndrome (PS), a previously unrecognised common disorder with dramatic clinical and EEG manifestations, has now been formally recognised by the ILAE. It has further been highlighted by editorials and reviews in medical journals, examined in an expert consensus, featured as the main theme of a recent issue of Epilepsia and is becoming more readily diagnosed by physicians. Less common phenotypes, such as the idiopathic childhood occipital epilepsy of Gastaut (ICOE-G) and idiopathic photosensitive occipital lobe epilepsy, have also been recognised and defined. Furthermore, there are also children who present with seizures of predominantly affective symptoms, and claims have been made for other benign childhood seizures associated with certain inter-ictal functional EEG foci, such as frontal, midline or parietal, with or without extreme somatosensory evoked spikes (ESEs). 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 neonatal and benign infantile seizures. BCSSS should be properly re-examined and redefined.

The term ‘functional spikes’ refers to transient focal EEG abnormalities of sharp waves that occur in children with or without epileptic seizures and which disappear in the late teens. Functional spikes of childhood are of low epileptogenic potential and they occur in 2% to 3% of normal children (Table 12.1).

Considerations on classification

The ILAE Task Force recognises three syndromes of idiopathic childhood focal epilepsy (Table 5.2):1
1. benign childhood epilepsy with centrotemporal spikes (BCECTS) (rating score 3)
2. early onset benign childhood occipital epilepsy (Panayiotopoulos type) (3)
3. late-onset childhood occipital epilepsy (Gastaut type) (2).

The rating score in parenthesis reflects on the certainty with which the ILAE Core Group believed that each syndrome represents a unique diagnostic entity on a range of 1–3 (with 3 being the most clearly and reproducibly defined).3

The 1989 ILAE classification recognised three ‘age-related and localisation-related (focal, local, partial) epilepsies and syndromes’ (Table 5.1):
1. BCECTS
2. childhood epilepsy with occipital paroxysms (which is now called ‘late-onset childhood occipital epilepsy [Gastaut type]’)
3. primary reading epilepsy.
‘Reading epilepsy’ is now rightly classified as a reflex epileptic syndrome (Table 5.2).

Considerations on classification and nomenclature are detailed in the individual description of each syndrome. Overall, benign childhood focal syndromes and their main representatives, BCECTS and PS, do not fulfil the diagnostic criteria of ‘epilepsy’ defined as ‘chronic neurological condition characterised by recurrent epileptic seizures’.\(^{14}\) BCECTS and PS are age limited (not ‘chronic’) and at least a third of patients have a single (not a ‘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 that incorporates ‘febrile, benign neonatal, single seizures or isolated clusters of seizures and rarely repeated seizures (oligoepilepsy)’ (Table 5.2).\(^{2,3}\)

### Benign childhood epilepsy with centrotemporal spikes

**Synonyms:** BCECTS, rolandic seizures, rolandic epilepsy.

BCECTS\(^{1,15–22}\) is the most common manifestation of benign childhood seizure susceptibility syndrome (BCSSS).

**Considerations on nomenclature**

I use the terms ‘BCECTS’, ‘rolandic seizures’ and ‘rolandic epilepsy’ synonymously, although I prefer the term ‘rolandic seizures’ for the following reasons:

- the term ‘rolandic seizures’ has long been established and is better known than BCECTS among paediatricians
- most ‘centrotemporal spikes’ (CTSs) are rolandic spikes; they are rarely located in the temporal electrodes
- the word ‘temporal’ is misleading because children with this form of epilepsy do not have symptoms from the temporal lobes
- BCECTS may occur without CTSs and conversely CTSs may occur in children without seizures or other clinical phenotypes of BCSSS\(^{23}\)
- similar clinical features may appear in patients with spikes in locations other than at centrotemporal sites.

**Demographic data**

Onset is from age 1 to 14 years; 75% start between 7 and 10 years (peak 8 or 9 years).\(^{1,17}\) There is a 1.5 male predominance. Prevalence is around 15% in children aged 1–15 years with seizures. Incidence is 10–20 per 100,000 children aged 0–15 years.

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**EEG functional spikes in normal children (% median and range)**

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Centrotemporal spikes</th>
<th>Occipital spikes</th>
<th>Frontal spikes</th>
<th>Generalised discharges</th>
</tr>
</thead>
<tbody>
<tr>
<td>5–12</td>
<td>2.25 (0.7–3.5)</td>
<td>0.15 (0.0–0.4)</td>
<td>0.10 (0.1–0.6)</td>
<td>1.00 (0.1–1.1)</td>
</tr>
<tr>
<td>1–5</td>
<td>0.40 (0.3–0.4)</td>
<td>0.90 (0.8–1)</td>
<td>0.05 (0.0–0.1)</td>
<td>0.20 (0.1–0.3)</td>
</tr>
</tbody>
</table>

Table 12.1 Modified with permission from Panayiotopoulos (1999).\(^1\)