Benign Rolandic and Occipital Epilepsies of Childhood

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KEYWORDS

• Rolandic epilepsy • Occipital epilepsy
• Panayiotopoulos syndrome • Sleep

Benign childhood focal seizures represent the most common epileptic manifestations in childhood and affect approximately 22% of children. Three identifiable electro clinical syndromes are coded by the International League against Epilepsy (ILAE)[1]: benign epilepsy with centrotemporal spikes (BECTS), Panayiotopoulos syndrome (PS), and the idiopathic childhood occipital epilepsy of Gastaut (ICOE-G).

The term "*benign*" refers to the positive prognosis of these disorders in regard to the EEG pattern and the seizures. However, a significant number of children with BECTS present various cognitive deficits affecting language and memory functions the severity of which associated with the intensity and the duration of interictal epileptic discharges (IED) and resolve with EEG normalization.[2–6]

Since the publication of the ILAE classification of epileptic syndromes,[7] the group of focal idiopathic epilepsies has been enlarged to a subgroup of epileptic encephalopathies (EE) with continuous spike-and-waves during slow-wave sleep (CSWS) in which the appearance and persistence of IED are associated with cognitive regression. BECTS and EE with CSWS represents opposite ends of a spectrum, behavioral and cognitive deficits are often milder in BECTS and severe in the CSWS epileptic encephalopathy.[8–14]

BENIGN CHILDHOOD EPILEPSY WITH CENTRO-TEMPORAL SPIKES

BECTS, also known as *Rolandic Epilepsy*, is the most common among the benign focal epilepsies of childhood, occurring in 15–25% of pediatric epilepsy patients.[15–18] The age at onset is between 2 and 14 years, with a peak of incidence (80% of the cases) at 5 to 10 years. Absence seizures develop in approximately 2% of cases.[19,20]

Loiseau and Duché[21] specified 5 criteria for the diagnosis of BECTS: (1) age at onset between 2 and 13 years; (2) lack of neurologic/intellectual deficit at the time of the onset; (3) partial seizures with motor signs, often associated with somatosensory symptoms or precipitated by sleep; (4) a spike focus located in the centrotemporal area with normal background activity on the interictal

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EEG; and (5) spontaneous remission (generally during adolescence).

Seizure Manifestations

Seizures are usually brief, lasting for 1 to 3 minutes, and their clinical manifestations include (see Video 1):

- Unilateral facial motor symptoms (30% of cases), in the form of clonic contractions, mostly localized in the lower lip that may extend to the ipsilateral hand. Symptoms can include numbness in the corner of the mouth.[19,22]
- Oro-pharyngo-laryngeal symptoms (53%), consisting of unilateral sensorimotor symptoms or paresthesias inside the mouth, associated with vocalizations.[19,22]
- Speech arrest in 40% of patients, who are unable to utter intelligible words and can communicate only with gestures.[19,22]
- Hypersalivation (30%), often associated with hemifacial seizures.[19,22]

Generalized convulsive status epilepticus is rare. Opercular status epilepticus occurs in atypical evolutions of BECTS or, exceptionally, it may be induced by carbamazepine.[23] This 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, buccofacial apraxia and hypersalivation.[24]

EEG Features

The interictal EEG is distinctive in BECTS, showing centrotemporal spike-and-wave discharges which have a tangential dipole that is negative in the centrotemporal area and positive frontally. Centrotemporal spikes (CTS) are the hallmark of the BECTS syndrome and are mainly localized in the left central (C3) and right central (C4) or the supra-sylvian electrodes (C5, C6 using the International 10-10 system of electrode placement) and not in the temporal ones.[25] CTS are markedly activated by drowsiness, occur independently over both hemispheres at frequencies of 4 to 20 discharges per minute, and usually occur in clusters (**Fig. 1**). Recent studies show that the main negative spike component is modeled by a stable tangential dipole source, with the positive pole was maximum in the frontal region, while the negative pole was maximum in the central region.[26] The main spike (sharp wave) component is diphasic with a maximum surface, negative, rounded peak that is followed by a smaller positive peak. The amplitude of the main spike 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. About 4% of patients with rolandic epilepsy also show brief bursts of 3-5 Hz slow waves with internalized small spikes lasting 1-3 seconds, without overt clinical symptoms.[27] The frequency, location and persistence of CTS are not specific for BECTS.[27,28] In fact,

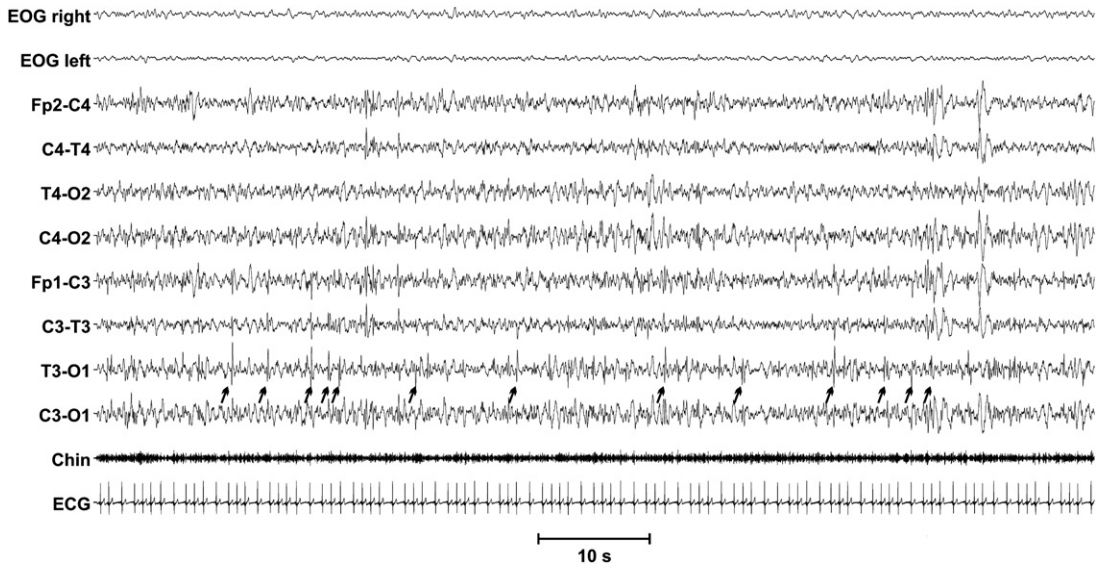

Fig. 1. Example of EEG tracing of a patient with rolandic epilepsy with typical spikes in the left fronto-centrotemporal area with spreading in the right corresponding area (*arrows*).