Benign focal epilepsies represent almost one-fourth of all childhood epilepsies and are a frequent occurrence in clinical practice. Because the prognosis is always excellent in patients with benign focal epilepsies, it is necessary to consider the risks and benefits of chronic anti-epileptic drug (AED) administration. By contrast, benign focal epilepsies, especially benign childhood epilepsy with centrotemporal spikes (BCECTS), occasionally presents severe aggravation of epileptic manifestations and marked cognitive, behavioral and language impairments. Fejerman [1] designated these forms of epilepsy as “atypical evolution” of benign focal epilepsies. Is BCECTS always benign? When and how should BCECTS be treated? Management of treatment may be required to remit seizures as soon as possible to achieve optimal prognosis in BCECTS with atypical features such as increased number of seizures, onset of inhibitory seizures, electroencephalography characteristics such as electrical status epilepticus during slow wave sleep and cognitive or behavioral involvement.

AED treatment is usually not recommended for patients with a first attack. Continuous treatment should be considered only for patients with frequent seizures (second or third attack) and when the ictal events are disruptive to the patient or family. The results of a descriptive study indicate that BCECTS may not always be benign in terms of neurodevelopmental outcomes, even if seizure outcomes are favorable. Several difficulties have been reported, such as problems maintaining attention, language and otorrheic disturbances, learning deficits for verbal material and subtle motor and visuomotor disabilities. These disturbances may relate to the topography and frequency of paroxysms. In previous 3-dimensional MRI volumetric studies, frontal and prefrontal lobe volumes and the prefrontal-to-frontal lobe volume ratio in particular showed growth disturbances during the seizure period in the two patients presenting atypical evolution [2]. This suggests that children with atypical evolution of BCECTS may experience frontal lobe dysfunctions. Moreover, during active seizure periods frequent spike-waves coupled with the occurrence of frequent seizures may be associated with prefrontal lobe growth retardation, which relates to neuropsychological problems and ultimately to neuropsychological outcome. Current research suggests that damage to the frontal regions during childhood may interrupt normal maturational processes and organization, resulting in impairments to neurobehavioral development. Integrative executive functions may thus rely on the health of frontal lobe tissue and connectivity with the rest of the cortex. Children with BCECTS may have cognitive and behavioral impairments related to frontal lobe dysfunctions.

In considering outcomes for children with BCECTS, control of seizures must be weighed against the incidence of neurological impairments, either transitory or persistent. The best goal for children with BCECTS is to make them as free from restrictions as possible so that they can lead a normal life irrespective of AED treatment [3]. In general, BCECTS is associated with very good prognosis, with electrical normalization in puberty. However, in rare cases (approximately 2%), atypical evolutions lead to significant neuropsychological impairments. Therefore, determined how to treat BCECTS should be in an individual manner based on active seizure period and the attitudes of the patients’ parents toward seizure recurrences and possible adverse effects of AED. Although cases with atypical evolutions leading to significant neuropsychological impairments are rare, these events need to be recognized as early as possible. Management of treatment may be required to remit seizures as soon as possible to achieve optimal prognosis in BCECTS with atypical features.

References

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