Benign Partial Epilepsy withRolandicSpikes

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Abstract

Benign epilepsy of childhood with rolandic (centro temporal) spikes (BECRS) is the most frequent of all childhood epileptic syndromes. It is characterized by partial epileptic seizures, mainly during sleep or just before awakening, with involvement of the mouth, face and speech organs or otherwise generalized. The EEG is typical and appears to be the cornerstone of the diagnosis. It is diphasic consisting of high-voltage, spikes or sharp waves followed by slow waves. This pattern used to be called “centro temporal” or “rolandic spikes”. They always disappear by adolescence. It is an extremely rare disease in which antiepileptic drugs (AED) are hardly needed. BECRS represents 13% to 23% of all epileptic patients in childhood. Incidence is 21:100,000 children under 15. A Mendelian dominant pattern of transmission with low penetrance and expressivity limited by age was postulated, the gene is now known to be located on chromosome 15q14.

Keywords
Benign epilepsy of childhood with rolandic spikes (BECRS), childhood epileptic syndromes, partial epileptic seizures

Disease name and synonyms
- Benign epilepsy of childhood with rolandic spikes (BECRS)
- Benign epilepsy of childhood with centro temporal spikes (BECTS)
- Silvian epilepsy

Definition
BECRS is the most common epileptic syndrome during childhood. It usually consists of partial seizures, which affect the mouth, face and speech organs and mainly occur during sleep and tends to remit and even disappear at adolescence for typical forms. The signs have been described by French neurologists since the 1950s and named by Beaussart in 1972 (1-3). BECRS has been recognized worldwide (4) and included by the International League Against Epilepsy (ILAE) as one of the epileptic syndromes.

Incidence and prevalence
BECRS represents 13% to 23% of all epileptic patients in childhood (5,6). The incidence is...
21/100,000 children under 15 years. The boy to girl ratio is 3:2. It always begins after 2 years of age and disappears before 16, regardless of the clinical manifestations developed during its course (7,8).

Clinical manifestations
Seizures (9) usually occur during sleep (80% of patients), with only 20% occurring in awake children and 15% in both states. When they appear during full consciousness, they are clinically characterized by simple partial seizures, which affect the mouth, face and speech organs, causing, usually with some kind of grunting noise, mouth and tongue paresthesia and drooling. Postictal confusion and amnesia are seldom observed after simple partial seizures of BECRS. Because of these clinical characteristics, the attacks have often been called 'silvian seizures'. Because the crises are very uncommon, they were only exceptionally recorded by the EEG tracings. When they affect very young children, as is the case for 10% of patients, the number of crises is high, generalized or unilateral, prolonged in time and source of high concern for their relatives. Usually, they tend to subside at most after 3 years of their onset.

EEG manifestations
The EEG is typical and the cornerstone of the diagnosis. It is diphasic consisting of high-voltage, spikes or sharp waves followed by slow waves. This pattern used to be called 'centro temporal' or 'rolandic'spikes'. They tend to occur in clusters but can also be isolated. Marked activation of rolandic spikes in drowsiness and sleep is very usual and 30% of them appear only during that state (10). Peaks could not be found in around 1/3 of EEG done throughout entire evolution of BECRS. It may sometimes take several months to see these peaks, even if the EEG is taken during sleep. Over 60% of cases show unilateral rolandic spikes, but they tend to alternate from one side of the brain to the other, and follow-up recordings showed that they tended to shift to and away from the centro temporal area, becoming bilateral or appearing at other sites, for example frontal or occipital. The EEG background is normal although mild slowing could be seen in some cases. Rolandoic spikes can be seen in 1-2% of healthy children between 5 and 12 years old (11). Therefore it is not exceptional to find them in children who had never suffered from epileptic seizures and that had undergone an EEG for other reasons such as cranial trauma, learning difficulties or headache.

Genetics
Rolandoic spikes are detected in 34% of the patients' siblings, but only 15% of the latter had ever experienced a seizure during their lives (12). For that reason, a Mendelian dominant pattern of transmission, with low penetrance and expressivity limited by age was postulated (13,14). The fact that the focus vanishes during adolescence makes it almost impossible to follow beyond this age. It is now known that the gene is located on chromosome 15q14 (15). However, recent twin studies suggest that heredity is multifactorial (16). Incidentally, some patients with another 'rolandoic spikes' might coincidentally suffer from another cerebral pathology (17). In that case, the evolution could be the expression of a genetic factor and behave in a similar self-limited benign fashion. It should be emphasized that rolandic spikes are not pathognomonic of BECRS. For which the diagnosis is based on the characteristics of attack and the clinical manifestations. When partial complex seizures appear, a diagnosis of BECRS should always be considered. Furthermore, the diagnosis of BECRS probably cannot be seriously retained without at least one EEG tracing showing rolandic spikes.

Incidence of seizures
It varies widely among patients. A recent meta-analysis (18) showed that, among the affected population, 15% had only one attack throughout the entire evolution; 62% experienced between 2 and 5 and only 23% suffered more than 5. On the other hand, since most of attacks occur during sleep, the social stigma is greatly minimized. The invariable good evolution had led some authors to advise against treating these children on antiepileptic drugs (AED) (19), mainly because, even in the worst scenario, the activity span seldom lasts more than 3 years.

Atypical forms
In spite of their consistent benignity, during the disease course some patients show life-threatening features with, increasingly severe seizures and learning difficulties, and a decline of cognitive capabilities. These patients were described as having atypical partial benign epilepsy of childhood (20). In such cases, the rate and intensity of seizures rises and new manifestations appear, mainly drop attacks and atypical absences, and the clinical picture could be mistaken for Lennox-Gastaut syndrome. As a general rule, these seizures can be associated with neuropsychological impairments, affecting mainly language in Landau-Kleffner syndrome (21), opercular epileptic syndrome (22), all of which have increased EEG paroxysms, mainly
during sleep, which could generate continuous spike and wave during slow sleep (23). The main problem is that all of these patients could be helped by standard AED, but also that almost all of them develop this exacerbation some time after having started AED therapy (24) and they would get better if drugs were discontinued (25).

**Treatment of side effects**

There is strong evidence that some AED cause BECRS to worsen in some patients (26). Much information on the potential harmful effects of AED on these patients has been collected. In particular carbamazepine has been blamed, but valproate (27), phenobarbital, phenitoin (28) and new AED (29-31) could also be responsible. Although it has sometimes been thought that treatment of EEG paroxysms could provide some benefit in terms of the cognitive capabilities of these children, evidence shows that the opposite was true when AED were given (32). For that reason, when it is decided to prescribe AED for BECRS, the patient must be monitored very closely, looking for any intellectual deficiency that could signal a cognitive decline, which can usually be reversed only by AED discontinuation.

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