Is rolandic epilepsy really benign?
Epilepsia rolândica é realmente benigna?

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Epilepsies can be considered as benign as long as they do not compromise the long-term quality of life of the person. Benign epilepsy with centrotemporal spikes (BECTS) or rolandic epilepsy (RE) is usually considered as a good example of this situation. RE has a number of clinical and electroencephalography (EEG) features that indicate a favourable outcome.

Classically, RE is described as partial epilepsy of childhood characterized by absence of neurological deficits, motor partial seizures, peculiar EEG centrotemporal spikes and spontaneous recovery. There are also children with peculiar EEG centrotemporal spikes with no clinical seizure, called EEG traits for RE.

Although RE is considered a benign syndrome and, in theory, with no neurological impairment, children with RE may have neuropsychological problems more often than the general population. Even if neuropsychological involvements are considered as mild, the impact of RE on cognitive functions is far from being negligible.

Delayed speech and learning disabilities have been reported, but their incidence and the long-term impact remains undetermined. The basic mechanism of cognitive compromise, in this syndrome, also remains unclear. It is difficult to determine, whether the deficits are due to the basic brain dysfunction responsible for the epilepsy, or to other factors such as, the effects of ongoing epileptic activity on developing cognitive functions.

Neuropsychological studies of children with RE show that they have normal intelligence but often have limited weaknesses in various domains such as language, visuospatial abilities or isolated attendant problems.

Some studies tried to demonstrate that cognitive disorders, which can occur in RE, could be temporary and more evident in the active phase of the disease. These studies seem to correlate cognitive disorders with an increase in epileptic activity during this period. Attempts to correlate these deficits with the frequency of clinical seizures and with the side of discharges, for example, left or right side, unilateral or bilateral foci, have given conflicting results.

On the other hand, when EEGs of children with dyslexia were analyzed, 10% showed EEG traits of RE. Ten per cent means 2-5 times higher than the expected rate that is estimated to be about 2-4% in the child population. The results raise a number of questions about the relationship between dyslexia and a specific EEG trait and RE and language or any other neuropsychological impairment.

Other interesting point is that language disabilities and academic impairments found in RE are also common in relatives of RE probands. The incidence of language, reading disabilities and academic impairments in relatives of RE probands are 5.4 times that in the general population.

The strong proband and familial associations with reading disabilities and academic impairments and the similarity in neurocognitive profiles between probands and siblings, suggest that these neurodevelopmental traits in RE should be also genetically influenced.

The role of genetic influences has been sought for a long time. The hypothesis of shared genetic influences, recently has been demonstrated as a familial pattern of risk for reading disability, speech sound disorder, presence of EEG traits of RE, and migraine in families with RE.

These results strongly suggest that susceptibility to comorbidities in RE is inherited and not the direct result of recurrent seizures or discharges. Clinically, relevance of these studies is that early evaluation and intervention may benefit these children academically.
Long-term neuropsychological evaluation showed that neuropsychological impairments remain even after seizure remission, suggesting a persistent course that does not correlate with the time course of either seizures or EEG abnormality in RE, although the seizures and EEG abnormality may well complicate neuropsychological performance, as suggested by the association of attendant impairment and centrotemporal spikes.

Besides genetic evaluation, scans with functional MRI using word-generation and reading tasks in children with RE has been done comparing age-matched controls. Data has shown that functional connectivity with the sensorimotor network was reduced in patients with RE compared to controls. Reduced functional connectivity was demonstrated between the sensorimotor network and the left inferior frontal gyrus, Broca’s area, in children with RE. These findings might link epileptiform activity originating from the sensorimotor cortex to language impairment.

Structural scans using T1-weighted and diffusion-weighted MRI at 3T and corresponding research has shown that in RE, structural connectivity is reduced for several connections involving rolandic regions, from which the epileptiform activity originates. Most of these aberrant tracts involve the left hemisphere, especially the pars opercularis of the inferior frontal gyrus (Broca’s area) and the supramarginal gyrus.

Although the underlying mechanism may be multiple, the accurate diagnosis of comorbid conditions is of great importance for research, pathophysiology comprehension and for clinical prognosis and a better approach.

In this fascinating scenario that Oliveira EP et al. authors of rolandic epilepsy and dyslexia, a paper published in this number of Arquivos de Neuro-Psiquiatria, studied the prevalence of dyslexia and other language impairments in a group of 31 patients with clinical and electrographically defined diagnosis of RE. What stands out in this study is the detailed clinical and neuropsychological assessment that allowed the authors to diagnose dyslexia. The choice of control group is of fundamental importance. If there is a probability of relatives being affected by epileptiform discharges at centrotemporal regions or by comorbidities, including language impairments, relatives would not be the appropriate control group for later comparison of data. In this paper, the control group was recruited from regular schools.

Oliveira et al. findings show significant evidence for occurrence of dyslexia in patients with RE. Out of 31 patients, six (19.4%) had a diagnosis of dyslexia. Dyslexia diagnosis is not so simple to assume and in this paper, the authors showed that a detailed evaluation was necessary for a precise diagnosis. The main finding of their study contributes significantly to understanding school performance of children with RE. The findings are extremely important for patient rehabilitation.

Beyond the rehabilitation of the patient, their results allow and encourage future studies in several aspects, including clinical and neuropsychological long-term evolution, which are scarce in the literature.


