Original article

Epileptic Disord 2007; 9 (4): 432-6

Are dyslexia and dyscalculia associated with Rolandic epilepsy? A short report on ten Italian patients

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Received February 23, 2007; Accepted October 13, 2007

ABSTRACT – Rolandic epilepsy (RE) is the most common childhood epilepsy syndrome with a good, long-term outcome. Nevertheless, some studies indicate that children with RE have more scholastic and neuropsychological problems than controls. The purpose of this study was to describe neuropsychological findings in a small group of Italian children with RE, focusing on dyslexia and dyscalculia. Possible correlations between these findings and the age-at-onset of seizures, duration of active epilepsy, frequency, type and localization of epileptic discharges were examined. Children affected by RE, aged nine to eleven years were selected from patients admitted to the outpatient service of our Clinic. They underwent cognitive evaluation, specific evaluation for dyslexia and dyscalculia, and awake and sleep EEG recordings. We found two patients out of the ten with dyscalculia, one of whom also had characteristics of dyslexia. This small study suggests that dyscalculia and dyslexia might be more frequent than expected in children with RE. No significant correlations between this finding and EEG, seizure-frequency or age-at-onset of epilepsy were found in our patients.

Key words: dyslexia, dyscalculia, rolandic epilepsy, language, neuropsychology

Rolandic epilepsy (RE) is the most common childhood epilepsy syndrome with a good outcome (Holmes 1993). Nevertheless some authors have suggested that children with RE may have more neuropsychological deficits and scholastic underachievement than controls (Deonna 2000, Croona *et al.* 1999, Weglage *et al.* 1997, Saint-Martin *et al.* 2001, Metz-Lutz *et al.* 2006, Pinton *et al.* 2006, Deltour *et al.* 2007).

The pathophysiology of cognitive dysfunction in RE is unknown. Two hypotheses, which both remain controversial, have been considered: the first that EEG discharges may interfere with cognitive processing, the second that EEG traits and cognitive impairments are independent symptoms of a common, underlying pathology (Carlsson *et al.* 2000).

There are studies indicating that numerical processing could be localized

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doi: 10.1684/epd.2007.0138

to the parietal lobes (Landerl *et al.* 2004), and that a difference in the functional organization of parieto-occipital areas could be present in dyslexia (Leisman 2002).
Few studies focusing on dyslexia and dyscalculia in RE have been published. Staden *et al.* (1998) demonstrated language deficits, including dysfunction in written language, in children with RE, related to the distribution of epileptiform discharges mainly over the centrotemporal region.
Carlsson *et al.* (2000) reported worse reading skills, with a greater number of reading errors, in a group of 15 dyslexics with Rolandic EEG traits as compared to 15 dyslexics with a normal EEG.
Papavasiliou *et al.* (2005) reported a group of children with RE that scored significantly lower than their peers on written language skills such as spelling, reading aloud, reading comprehension and dyslexia testing.
A recent study (Wolff *et al.* 2005), showed a correlation between spike location and selective cognitive deficits in children with benign partial epilepsy; in particular, children with left perisylvian spikes performed significantly less well in language tests.
Possible factors in the learning dysfunction in childhood epilepsy include the seizures themselves, the effect of subclinical EEG discharges and adverse effects of AED: early onset of seizures, higher lifetime total seizures, presence of multiple seizure types and multiple AEDs are factors that have correlated significantly with academic achievement outcomes in childhood epilepsy (Bailet, 2000). However, Monjauze (2005) and Papavasiliou (2005) reported a dissociation between the resolution of seizures and persistence of learning problems, suggesting long-term consequences.
The purpose of this study was to describe the neuropsychological findings, focusing on reading and mathematical skills, in a group of Italian children with RE. Possible correlations between these findings and age-at-onset of seizures, duration of active epilepsy, frequency, type and localization of epileptic discharges were examined.

Methods

Children were selected from patients admitted to the outpatient service of our Clinic over a period of six months (January-June 2006) according to the following inclusion criteria:
– diagnosis of RE according to the ILAE diagnostic criteria, without any atypical features,
– normal prenatal and perinatal history, psychomotor development and neuroradiological investigations,
– absence of psychiatric/behavioural disturbance,
– negative family history for learning disabilities,
– age between nine and eleven years, since the Italian dyscalculia battery used is standardized for this age range.

They were submitted to a neuropsychological protocol including:
– cognitive evaluation (WISC-R and Raven's coloured progressive matrices test),
– specific evaluation for dyslexia:
• MT standardized reading tests (Cornoldi - evaluating accuracy, speed and text comprehension),
• standardized reading battery (Sartori-Job - evaluating accuracy and speed of words and non-word reading),
– specific evaluation for writing skills: standardized writing test (Tressoldi-Cornoldi - evaluating writing skills and orthographic accuracy),
– specific evaluation for dyscalculia: standardized dyscalculia battery (BDE) (Biancardi-Nicoletti) (the battery consists of different subtests: counting, reading, writing, repetition of numbers, semantic codification, multiplication tables, multiplications, sum and subtraction with numbers up to ten, sum and subtractions with numbers above ten, and four written operations. The results are expressed with a global quotient "QNC").
The diagnostic criteria for dyslexia and dyscalculia that we used were:
– IQ ≥ 85,
– reading/mathematical skills more than 2SD lower than expected for age (for dyscalculia battery, a global quotient "QNC" below 70),
– no neurological/sensory deficits.

Awake and sleep EEG recordings were performed for all patients and a specific "awake score" and "sleep score" were given: score 1: ≤ 20 spikes in two minutes of recording, score 2 : > 20 and ≤ 70 spikes in two minutes, score 3: > 70 spikes in two minutes.
Both neuropsychological evaluation and EEG recordings were performed within one week.

Results

Patients

We included ten patients (five females and five males) with a mean age of 10,5 years (SD: 0,70, Median: 11, Range 9-11). Seizure onset was at a mean age of 7,3 years (SD: 1,76, Median: 8, Range 4-9). Mean duration of epilepsy was 3,2 years (SD: 1,61, Median: 3, Range 1-6).
Seizure-frequency varied from no recurrence after an initial seizure to more than three seizures/year; case 7 had only one episode but had a short duration of follow-up (only one year).
Six patients were on valproate therapy, three had never had therapy and one patient was off-therapy (VPA discontinued two years after epilepsy onset) *(table 1)*.

EEG findings

EEG abnormalities were bilateral in two (centro-temporal), right-sided in six (parieto-temporal in one, centro-temporal

Table 1. Cases included in the study, with main clinical, EEG and neuropsychological features.

Patients /gender	Age at onset of seizures (years)	Seizures frequency	AEDs	EEG Awake Score*	EEG Sleep Score*	Localization of epileptic focus	Age at Neuropsych. Evaluation (years)	WISC-R: Total IQ (VQ;PQ)	Raven (percentiles)	Reading tests	Writing test	Dyscalculia quotient (QNC)
Case 1 F	6	2-3/year	VPA	1	2	Right C-T	11	106 (103; 109)	95°	Severe problems	Appropriate	56
Case 2 M	8	2 at onset	VPA	1	2	Left C-T	10	107 (107; 106)	50°	Appropriate	Appropriate	< 50
Case 3 M	8	> 3/year	VPA	3	3	Bilateral C-T	11	118 (125; 104)	75°	Appropriate	Appropriate	92
Case 4 F	9	1 at onset	None	1	2	Bilateral C-T	11	93	50°	Appropriate	Appropriate	80
Case 5 F	5	1 at onset	Off- VPA	1	1	Left C-P	11	93 (100; 88)	50°	Appropriate	Appropriate	98
Case 6 F	9	2 at onset	VPA	3	3	Right P-T	11	125 (127; 117)	95°	Appropriate	Appropriate	118
Case 7 M	9	1 /year	None	1	3	Right C-T	10	121 (122; 115)	50°	Appropriate	Appropriate	105
Case 8 M	4	2 at onset	VPA	1	1	Right C-T	9	134 (137; 120)	95°	Appropriate	Appropriate	119
Case 9 M	7	2 at onset	VPA	2	2	Right C	10	125 (136; 108)	75°	Appropriate	Appropriate	103
Case 10 F	8	1 at onset	None	1	3	Right C-T	11	112 (118; 102)	75°	Appropriate	Appropriate	87

* EEG score 1: ≤ 20 spikes in two minutes, score 2: > 20 and ≤ 70 spikes in two minutes, score 3: > 70 spikes in two minutes. AEDs: antiepileptic drugs; C: central; T: temporal; P: parietal.

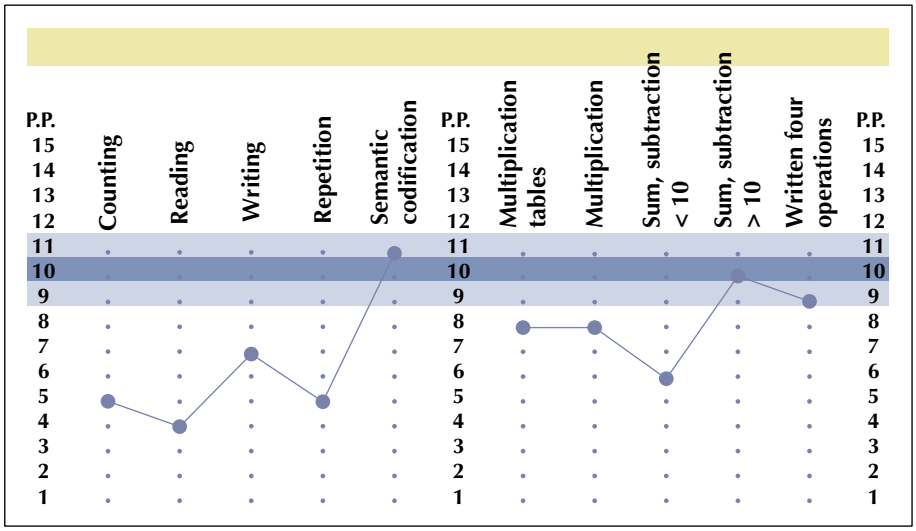

Figure 1. Dyscalculia battery result of case 2. P.P. stands for "age-corrected scaled score". The scores are below the normal range (9-11) for all subtests except for semantic codification, sum and subtraction with numbers above ten and four written operations. The worst results are in the counting, reading and repetition subtests.

in five) and left-sided in two patients (fronto-central in one and centro-parietal in one).
The EEG awake and sleep scores are shown in *table 1*:
– cases 5 and 8 had the least active EEG with awake and sleep scores of 1,
– cases 1, 2 and 4 had an awake score of 1 and a sleep score of 2,
– cases 10 and 7 had a mildly active awake EEG (score 1) and very active EEG during sleep (score 3),
– case 9 had moderately active awake and sleep EEGs, both with a score of 2,
– cases 3 and 6 had very active awake and sleep EEGs with a score of 3 in both.

Neuropsychological findings

Cognitive evaluation revealed an IQ within a normal range in all children. Raven's coloured progressive matrices revealed grades between the 50th and 95th percentiles in all cases.
In the dyscalculia battery, two children (cases 1 and 2) achieved a score markedly below the normal range *(table 1)*.
In the MT reading tests, all cases had a normal score for speed, accuracy and comprehension, except for case 1 who failed in all three parameters.
In the writing tests, all patients achieved scores within the average range.

Discussion

According to the literature, the prevalence of dyslexia is about 6%, although the percentages found are extremely variable, while that of dyscalculia varies from 5.4% to 3.6%. (Shalev *et al.* 2000, Lewis *et al.* 1994) Dyslexia seems to be more frequent in males, while for dyscalculia there is no gender difference.

In our series, we found two cases with dyscalculia (one male, case 2 and one female, case 1) and one with dyslexia (male case 2).
Our findings were consistent with the literature (Lewis *et al.* 1994) in that our dyscalculic patients showed good skills in semantic codification while the worst performances were recorded in reading and writing complex or long numbers, repeating multiplication tables and in tasks related to automatization procedures of counting (e.g. counting backwards) *(figure 1)*.

With regard to the question of whether the seizures themselves and the sub-clinical EEG discharges could represent possible factors of learning dysfunction in childhood epilepsy, we did not find any correlation between higher EEG scores and poor cognitive performance as both our patients with learning disabilities had mild to moderately active EEG (awake score 1 and sleep score 2). In *case 1*, the seizure-onset was at 6 years and in *case 2* at 8 years, within the mean age-at-onset of the whole series (7.3 ± 1.76 years). While *case 1* still has active epilepsy with two-three seizures per year, *case 2* had only two isolated seizures at onset, and has been followed up for two years, suggesting a dissociation between the resolution of seizures and persistence of learning problems, as reported elsewhere in literature (Monjauze 2005, Papavasiliou 2005).

With regard to the localization of the epileptiform discharges, most of our patients had a unilateral centro-temporal focus, including both patients with dyscalculia, who presented with a left- and right-sided focus respec-

tively; the latter patient was found to be dyslexic. Our results are in contrast with some data in the literature that suggest that the parietal lobe is implicated both in dyslexia and in dyscalculia (Landerl 2004, Leisman 2002), or a left perisylvian involvement in language deficits (Wolff 2005). However, they agree with other findings for learning dysfunction and language problems in patients with left-sided, right-sided or bilateral centro-temporal spikes (Papavisiliou 2005, Monjauze 2005, Carlsson 2000, Staden 1998).

Although no definitive conclusions can be drawn from such a small series RE, often considered to be an epilepsy syndrome with a good outcome, seems to represent a possible risk factor for dyslexia and dyscalculia. Our findings do not provide any insight into the underlying mechanism for the cognitive dysfunction in RE. However, they do support the recommendation that neuropsychological testing should be performed in patients with RE so that the need for appropriate therapeutic intervention can be recognised and provided promptly. □

Acknowledgement. The authors are grateful to Professor Frank MC Besag for his assistance in the preparation of this paper.

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