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

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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...
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 lan-
guage, 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 dys-
lexics with Rolandic EEG traits as compared to 15 dys-
lexics 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, chil-
dren 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, pre-
sence 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 neuropsy-
chological findings, focusing on reading and mathemati-
cal 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 out-
patient 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 de-
  velopment 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 writ-
  ing test (Tressoldi-Cornoldi - evaluating writing skills and
  orthographic accuracy),
– specific evaluation for dyscalculia: standardized dyscal-
  culia battery (BDE) (Biancardi-Nicoletti) (the battery con-
  sists of different subtests: counting, reading, writing, rep-
  etition 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 record-
ing, 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 ini-
tial 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 discon-
tinued 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.

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

* 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.
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 (Monjaune 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 (Papavasiliou 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.

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References


