The variants of reading epilepsy
A clinical and video-EEG study of 17 patients with reading-induced seizures

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Summary
We present the clinical and electrographic data of 17 patients with reading-induced seizures documented with ictal video-EEG studies during provocation with language related tasks. The median age at onset was 15 years (range 11–22 years) and the male : female ratio was 2.4. Fourteen patients had no spontaneous seizures of any type while the remaining three had infrequent generalized tonic–clonic seizures during nocturnal sleep. Two distinct electroclinical ictal patterns were confirmed on video-EEG analysis. (i) Fifteen patients had reading-induced jerks which invariably involved the region of the jaw but also included the upper limbs in five of them. Ictal EEG discharges were noted in 12 patients; these were brief but varied in terms of morphology and spatial distribution, with a clear tendency for left-sided predominance. All but one of these patients had similar myoclonic seizures induced by linguistic activities other than reading, the phenomenon probably justifying the term ‘language-induced epilepsy’. Some patients had evidence of transient cognitive impairment associated with the reading-induced jaw or limb jerks. Three patients had a sibling with reading epilepsy but there was no other family history of epileptic seizures. (ii) Two patients had reading-provoked paroxysmal alexia without motor symptoms, associated with prolonged focal ictal EEG abnormalities. Reading provoked a subclinical, continuous and reproducible EEG activation over the left posterior temporal area. We propose that ictogenesis in reading or language-induced epilepsy is based on the reflex activation of a hyperexcitable network that subserves the function of speech and extends over multiple cerebral areas on both hemispheres. The parts of this network responding to the stimulus may, secondarily, drive the relative motor areas producing the typical regional myoclonus. This network hyperexcitability can be genetically determined and its clinical expression is age-related.

Keywords: EEG; reading epilepsy; myoclonus; paroxysmal dyslexia; network hyperexcitability

Abbreviations: GTCS = generalized tonic–clonic seizure; ILAE = International League Against Epilepsy

Introduction
Reading epilepsy is a distinct type of reflex epilepsy in which seizures are precipitated by reading. The condition was first described by Bickford et al. (1956), who distinguished two types, ‘primary’ and ‘secondary’ reading epilepsy. In primary reading epilepsy, stereotyped and similar ictal symptoms consisted of a clicking sensation or movement in the jaw, occurring only in relation to reading, and these would progress to a generalized convulsion if reading continued. Reading-provoked jaw-clicking symptoms were in each instance associated with 3–6-Hz bilateral and synchronous discharges maximal in the occipital and parietal areas. Patients with secondary reading epilepsy had seizures under other conditions too (Bickford et al., 1956). Subsequent reports on similar cases have essentially validated the original clinical features of primary reading epilepsy which is presently recognized as an idiopathic localization-related epilepsy with the following characteristics: ‘all or almost all seizures are precipitated by reading (especially aloud) and are independent...
of the content of the text. They are simple partial motor seizures, involving masticatory muscles, or visual. If the stimulus is not interrupted, secondary generalized tonic–clonic seizures (GTCSs) may occur. The syndrome may be inherited. Onset is typically in late puberty and the course is benign with little tendency to spontaneous seizures. Physical examination and imaging studies are normal, but EEG shows spikes or spike-waves in the dominant parietal-temporal region. Generalized spike and wave may occur” [Commission on Classification and Terminology of the International League Against Epilepsy (ILAE), 1989].

However, review of the literature suggests that reading epilepsy is less homogeneous than is currently appreciated. Reading epilepsy is not always idiopathic, and a few well-documented cases with symptomatic jaw myoclonus have been described (Lee et al., 1980; Ritaccio et al., 1992; Radhakrishnan et al., 1995, their patient 2). Furthermore, reading is not always the sole triggering stimulus. In some patients jaw myoclonus has been elicited by additional triggers, both simple such as photic stimulation, pattern and television, and complex such as talking, writing, arithmetic and decision-making processes. Ictal EEG features are also inconsistent as some patients have focal and others have generalized discharges (Wolf, 1992). Radhakrishnan et al. (1995) concluded that there was not sufficient electroclinical evidence to justify the inclusion of reading epilepsy amongst the localization-related idiopathic epilepsies, and they proposed an alternative classification among the idiopathic generalized epilepsies with seizures precipitated by specific modes of activation. While the description of a patient with reading-induced absence seizures (Singh et al., 1995) provided further support for the classification as a generalized epilepsy, there are earlier reports describing reading-precipitated prolonged partial seizures without ictal motor elements, manifested as pure dyslexia (Gastaut and Tassinari, 1966).

The purpose of this study was to delineate the clinical and electroencephalographic spectrum of reading epilepsy by analysing the video-EEG of 17 patients with seizures provoked by reading. Fifteen of these patients had the typical orofacial/jaw myoclonus, whereas the remaining two had prolonged partial seizures manifested with alexia. Particular emphasis was given to the partial variant of reading epilepsy which, since its early description (Gastaut and Tassinari, 1966), has received little attention, being uncommon and probably underdiagnosed.

**Methods**

**Patients**

We studied 17 patients with reading-induced seizures, seen at the National Hospital for Neurology and Neurosurgery, Queen Square (Patients 2–7), at the department of Clinical Neurophysiology and Epilepsies, St. Thomas’ Hospital, London (Patients 8–10, 16 and 17), at the Schweizerische Epilepsieklinik Zürich, Switzerland (Patients 1, 14 and 15) and at the Neurology Division, IWK Children’s Hospital, Halifax, Canada (Patients 11–13). The diagnosis was based on the patients’ own account that reading was the single or the main seizure-triggering stimulus, and it was confirmed by reproducing the habitual clinical seizures through reading provocation under continuous video-EEG monitoring.

**Electroencephalography**

All patients had routine scalp EEG with photic stimulation according to the international 10–20 system of electrode placement. Reading during continuous video-EEG monitoring provoked clinical seizures in all patients. In all four centres, the effect of other language-related triggering stimuli such as talking and writing was investigated by direct questioning, and it was appropriately tested during video-EEG monitoring. The effect of non-linguistic higher cognitive activities, such as calculating, decision making and spatial tasks, was specifically sought by history taking in all patients and formally tested when this was indicated. All video tapes and EEG tracings were reviewed by two of the investigators (M.K. and M.J.K.).

**MRI**

MRIs were obtained for Patients 1–7, 11, 16 and 17 on a 1.5-T Siemens SP63 Magnetom scanner (Siemens, Erlangen, FRG) using a 3D MPRAGE (magnetization prepared rapid gradient echo) sequence 10/4/200/1 (TR/TE/TI/NEX), flip angle 12°, matrix size 256 × 256 and 128 sagittal partitions in the third dimension as described in detail previously (Van Paesschen et al., 1997). Patients 8–10 and 11–15 were scanned on a 1.0-T Siemens Magnetom scanner (Siemens, Erlangen, Germany) with turbo spin echo axial T2-weighted (TR 4500 TE 90) and coronal spin echo T1 (TR 660 TE 14) images, and angled coronal STIR (short tau inversion recovery) of the temporal lobes (TR = 400, TE = 30 and TI = 120).

**Results**

The clinical data of the 17 patients with reading epilepsy are presented in Table 1, and the EEG and brain MRI findings are shown in Table 2.

**Demographic data**

Between 1991 and 1996, 12 male and five female right-handed patients with reading epilepsy were seen; the male : female ratio was 2:4. The mean age at onset was 15 years (range 11–22 years). The mean age at diagnosis was 24 years (range 12–30 years), and the median time from the first reading-induced seizure until the diagnosis of reading epilepsy was made was 7 years (range 0–15 years).
Previous medical history and neurological examination
Patient 2 had haemophilus meningitis at the age of 2 years. No risk factors for epileptic seizures such as prolonged early childhood convulsions or head trauma were found in the remaining 16 patients. Neurological examination and intellect were normal in all subjects.

Family history of epilepsy
The 17 patients belonged to 16 families. Patients 11 and 12 were brothers. Their other brother, aged 23 years, had difficulties with learning to read, but myoclonic seizures have never been noticed. Another sister was completely symptom-free. They had two half-brothers who were younger than the age of symptom onset. The brother of Patient 7 gave a similar description of reading-induced jaw jerks and GTCS. The sister of Patient 8 gave a clear description of talking-induced, but not reading-induced, jaw jerks. Two brothers of Patient 3 had a history of febrile convulsions but did not develop epileptic seizures thereafter. There were no other family members with seizures in 11 families, and the family history of Patient 4 (adopted) is not known.

MRI
Brain MRI was normal in 15 but showed a left temporal, probably perinatal, infarct in Patient 3 and an arachnoid cyst at the left temporal pole in Patient 16 (Fig. 1).

The two variants of reading epilepsy
Two distinct patterns of reading-induced seizures were recognized: 15 patients (1–15) had the typical form of reading-induced seizures with orofacial/jaw myoclonus which is henceforth referred to as myoclonic reading epilepsy. The remaining two patients (16 and 17) had prolonged partial seizures manifested with dyslexia and strictly focal EEG abnormalities, henceforth referred to as partial reading epilepsy. The clinical seizure characteristics and the interictal and ictal EEG findings of the two forms are presented separately.

Myoclonic reading epilepsy
Ictal manifestations. Video-analysis and direct observation confirmed jaw jerks in all patients. These were described as ‘jerking’, ‘twitching or clicking of the jaw’, ‘jaw tic’ and ‘blocked jaw’. In addition, Patients 1, 2 and 13 reported ‘tightening of the throat’, Patient 6 described contractions of her diaphragm that moved her jaw, while Patients 2 and 7 also experienced numbness around the mouth and stiffness of the right arm. Six patients (3, 4, 9, 10, 12 and 15) reported ‘loss of track of the reading text’, five patients (1, 4, 6, 7 and 11) described themselves as ‘sticking on the word’, Patient 3 described a frequent ‘light bulb on/off’ sensation, and Patient 15 a ‘thinking block’. Other associated ictal symptoms included a ‘shock-like body sensation’ (Patients 1, 3, 6 and 15), feeling of fear (Patients 3 and 4), lightheadedness (Patient 3) and ‘difficulty in getting the words out’ (Patients 4, 11, 12 and 15). Ictal stammering was noted in Patients 1, 6, 9, 11, 12 and 15, eyelid blinking in Patient 4, and staring in Patient 11. Reading-induced axial myoclonus or jerks of the upper extremities were reported or captured on video-EEG in five patients (6, 10, 11, 12 and 14). The severity of the jerks and their distribution varied, even in individual patients. Patient 10 had jaw and axial jerks on different occasions, while Patient 11 sometimes had coarse, clinically evident jaw jerks, but at other times they were inapparent on detailed video analysis.

Other seizure types. Jaw myoclonus had evolved into GTCS if reading was continued on a single occasion in five patients, and more than once in nine patients. None of the patients gave a history of other seizure types including spontaneous myoclonic jerks, except Patients 2 and 6, who reported occasional spontaneous nocturnal GTCS before the onset of the typical reading-provoked myoclonic jerks.

Precipitating factors other than reading. Talking, especially when argumentative, was an effective triggering stimulus in 14 patients, though it was less potent than reading. Interestingly, the sole case with jaw jerks induced by reading alone (Patient 8) had a sister with identical symptoms elicited exclusively by talking fast. The sister was never examined or given any medication, but gave the following written account of her symptoms: ‘I had jaw jerking but I can’t remember having one recently. It used to happen quite frequently when I was about 14, at school, and always when I was talking. I was talking quite rapidly at the time and it was like a very quick uncontrollable spasm. It didn’t last long enough for anyone else to notice, but my flow of speech was disrupted’. Seizures were also triggered by writing in six patients (1, 4, 6, 11, 14 and 15). Two patients (12 and 13) gave a history of thinking-induced jaw jerks. None of the patients had a history of seizures being induced by photic stimulation.

Interictal EEG findings. Standard EEG during the awake resting state showed normal background activity in all but one case (Patient 2) who had mild, non-specific bitemporal slowing. Spontaneous epileptiform discharges were noted in four patients. These were focal in Patients 6, 9 and 11, and both focal and generalized in Patient 12 (Table 2). None of the patients showed photoparoxysmal responses on photic stimulation.

Ictal EEG findings. Brief spike/sharp-wave discharges, or sharp theta wave discharges associated with reading-induced regional jerking were observed in 12 patients. Five of these patients (4, 6, 9, 11 and 15) had a number of video
Table 1 Clinical data of 17 patients with reading epilepsy

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at onset (years)</th>
<th>Age at diagnosis (years)</th>
<th>Triggers other than reading (history)</th>
<th>Effective triggers (tested)</th>
<th>Site of onset (spread)</th>
<th>Associated symptoms/signs</th>
<th>Evidence of cognitive impairment</th>
<th>Type of seizures</th>
<th>Unprovoked seizures</th>
<th>Family history</th>
<th>Anti-epilepsy drug</th>
<th>Treatment response</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>34/F</td>
<td>19</td>
<td>Writing, talking, shorthand</td>
<td>Reading, writing</td>
<td>Jaw, lips, tongue</td>
<td>‘Shock-like feeling, tightening of throat’, stammering</td>
<td>‘Stuck on the word’</td>
<td>MS, a single GTCS</td>
<td>No</td>
<td>No</td>
<td>CBZ initially, then CZP and VPA</td>
<td>SF</td>
</tr>
<tr>
<td>2</td>
<td>25/M</td>
<td>11</td>
<td>Stressful conversation</td>
<td>Reading</td>
<td>Jaw</td>
<td>‘Tightening of throat, numbness around the mouth (ictal) and R arm (postictal)’</td>
<td>No</td>
<td>MS, a single GTCS</td>
<td>No</td>
<td>No</td>
<td>CBZ, VPA</td>
<td>Still MS, no further GTCS</td>
</tr>
<tr>
<td>3</td>
<td>37/M</td>
<td>14</td>
<td>Stressful conversation</td>
<td>Reading</td>
<td>Jaw</td>
<td>‘Shock-like feeling fearful, lightheaded’</td>
<td>‘Light bulb off/on’, loss of track</td>
<td>MS, GTCS</td>
<td>No</td>
<td>Two brothers had FC</td>
<td>Still MS, no further GTCS</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>35/M</td>
<td>15</td>
<td>Writing, stressful conversation</td>
<td>Reading, talking</td>
<td>Jaw</td>
<td>‘Fearful’, ‘difficulty in getting the words out’, eye-blinking</td>
<td>‘Stack on the word’, loss of track</td>
<td>MS, GTCS</td>
<td>No</td>
<td>Unknown</td>
<td>PHT for a while</td>
<td>Still MS, last GTC at 32 years</td>
</tr>
<tr>
<td>5</td>
<td>21/M</td>
<td>20</td>
<td>Talking, stressful conversation</td>
<td>Reading</td>
<td>Sudden brief mouth twitching R &gt; L</td>
<td>‘Shock-like feeling’, stammering</td>
<td>No</td>
<td>MS, a single GTCS</td>
<td>No</td>
<td>No</td>
<td>None</td>
<td>Still MS, further GTCS</td>
</tr>
<tr>
<td>6</td>
<td>57/F</td>
<td>13</td>
<td>Talking, shorthand</td>
<td>Reading</td>
<td>Jaw, diaphragm, abdominal and chest muscles</td>
<td>Stiffness of R arm</td>
<td>‘Stuck on the word’</td>
<td>No</td>
<td>MS, GTCS</td>
<td>No</td>
<td>Brother with RE</td>
<td>CZP</td>
</tr>
<tr>
<td>7</td>
<td>24/M</td>
<td>15</td>
<td>Talking</td>
<td>Reading</td>
<td>Jaw</td>
<td>Stiffness of R arm</td>
<td>‘Stuck on the word’</td>
<td>No</td>
<td>MS, a single GTCS</td>
<td>No</td>
<td>No</td>
<td>CZP</td>
</tr>
<tr>
<td>8</td>
<td>34/F</td>
<td>15</td>
<td>None</td>
<td>Reading</td>
<td>Jaw</td>
<td>None</td>
<td>None</td>
<td>Loss of track</td>
<td>MS</td>
<td>No</td>
<td>No</td>
<td>CZP</td>
</tr>
<tr>
<td>9</td>
<td>31/M</td>
<td>22</td>
<td>Talking</td>
<td>Reading, talking</td>
<td>Jaw</td>
<td>Stammering</td>
<td>Loss of track</td>
<td>MS</td>
<td>No</td>
<td>No</td>
<td>CZP</td>
<td>SF</td>
</tr>
<tr>
<td>10</td>
<td>27/F</td>
<td>18</td>
<td>Talking</td>
<td>Reading</td>
<td>Jaw (upper limbs, trunk)</td>
<td>None</td>
<td>Loss of track</td>
<td>MS</td>
<td>No</td>
<td>No</td>
<td>CZP</td>
<td>SF</td>
</tr>
</tbody>
</table>

**Myoclonic reading epilepsy**

- **1**: Writing, talking, shorthand; reading, writing; jaw, lips, tightening of throat; stammering; ‘Shock-like feeling, tightening of throat’, stammering; ‘Stuck on the word’; MS, a single GTCS; No; No; CBZ initially, then CZP and VPA; SF

- **2**: Stressful conversation; reading; jaw; ‘Tightening of throat, numbness around the mouth (ictal) and R arm (postictal)’; ‘Shock-like feeling fearful, lightheaded’; ‘Light bulb off/on’, loss of track; MS, GTCS; No; No; CBZ, VPA, Still MS, no further GTCS

- **3**: Stressful conversation; reading; jaw; ‘Shock-like feeling fearful, lightheaded’; ‘Light bulb off/on’, loss of track; MS, GTCS; No; Two brothers had FC; Unknown; Still MS, no further GTCS

- **4**: Writing, stressful conversation; reading, talking; jaw; ‘Fearful’, ‘difficulty in getting the words out’, eye-blinking; ‘Stack on the word’, loss of track; MS, GTCS; No; No; PHT for a while; Still MS, last GTC at 32 years

- **5**: Talking, stressful conversation; reading; sudden brief mouth twitching R > L; jaw, diaphragm, abdominal and chest muscles; stiffness of R arm; ‘Shock-like feeling’, stammering; ‘Stuck on the word’; MS, GTCS; No; No; PHB; SF

- **6**: Talking, shorthand; reading; jaw, diaphragm, abdominal and chest muscles; stiffness of R arm; ‘Shock-like feeling’, stammering; ‘Stuck on the word’; MS, GTCS; No; No; DPH; SF

- **7**: Talking; reading; jaw; stammering; ‘Stuck on the word’; MS, GTCS; No; No; Brother with RE; None; Still MS

- **8**: Talking, talking; reading; jaw; stammering; ‘Stuck on the word’; MS, GTCS; No; No; Sister talking-induced jaw jerks; None; Still MS

- **9**: Talking; reading; jaw (upper limbs, trunk); None; Loss of track; MS, GTCS; No; No; CZP; SF

- **10**: Talking; reading; jaw (upper limbs, trunk); None; Loss of track; MS, GTCS; No; No; CZP; SF
### Table 1

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at onset (years)</th>
<th>Sex</th>
<th>Age at diagnosis (years)</th>
<th>Triggers</th>
<th>Effective treatment</th>
<th>Other signs/symptoms</th>
<th>Associated evidence of unprovoked seizures</th>
<th>Family history of epilepsy</th>
<th>Response to treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>11</td>
<td>14</td>
<td>M</td>
<td>24</td>
<td>Writing, Reading</td>
<td>Jaw, tongue</td>
<td>'Difficulty in talking (body). Getting the word out', sometimes staring, dysphasic/stammering</td>
<td>Dyslexic</td>
<td>None</td>
<td>Poor</td>
</tr>
<tr>
<td>12</td>
<td>12</td>
<td>M</td>
<td>18</td>
<td>Talking, Reading</td>
<td>Jaw, 'Difficulty in thinking (fingers in both hands) out', Stammering</td>
<td>Loss of track</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>13</td>
<td>15</td>
<td>F</td>
<td>21</td>
<td>Talking, Reading, Jaw, face</td>
<td>'Tightening of naming throat'</td>
<td>Loss of track</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>14</td>
<td>20</td>
<td>M</td>
<td>21</td>
<td>Writing, Reading, Jaw</td>
<td>None</td>
<td>Loss of track in getting words out, Writing (upper limbs) a single talking</td>
<td>GTCS</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>15</td>
<td>16</td>
<td>M</td>
<td>26</td>
<td>Writing, Reading, Jaw</td>
<td>'Shock-like feeling', 'Think in getting words out', Stammering, 'Difficulty of thinking'</td>
<td>Loss of track</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>16</td>
<td>16</td>
<td>M</td>
<td>23</td>
<td>None</td>
<td>Reading</td>
<td>No</td>
<td>No</td>
<td>Motor or Sensory Dysphasia, GTCS No</td>
<td>No</td>
</tr>
<tr>
<td>17</td>
<td>22</td>
<td>M</td>
<td>24</td>
<td>None</td>
<td>Reading</td>
<td>No</td>
<td>No</td>
<td>Motor or Sensory Dysphasia, GTCS No</td>
<td>No</td>
</tr>
</tbody>
</table>

**Legend:**
- M = Male; F = Female; MS = Myoclonic seizure; GTCS = Generalized tonic–clonic seizure; PS = Partial seizure; CZP = Clonazepam; SF = Sodium Valproate; VPA = Sodium valproate; DPH = Diphenylhydantoin; PHB = Phenobarbitone; CZP = Clonazepam; CLB = Clobazam; FC = Febrile convulsions; Followed by: postictal depression (in minutes) (Patient 2).
Table 2 MRI and EEG findings of 17 patients with reading epilepsy

<table>
<thead>
<tr>
<th>Patient</th>
<th>MRI</th>
<th>Intercital (standard record)</th>
<th>Ictal (reading activation)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Left frontal-central and temporal spike-wave/sharp-wave discharges, spreading rapidly to the right.</td>
</tr>
<tr>
<td>2</td>
<td>Normal</td>
<td>Non-specific bitemporal slowing</td>
<td>Jaw jerks were not associated with clear EEG changes.</td>
</tr>
<tr>
<td>3</td>
<td>Left temporal infarct (perinatal)</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Bilateral synchronous sharp-and-wave complex maximal over the vertex. Bilateral sharp waves on central regions. Jaw jerks were not always associated with EEG changes.</td>
</tr>
<tr>
<td>4</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Jaw jerks were not always associated with EEG changes.</td>
</tr>
<tr>
<td>5</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Bisynchronous frontotemporal small spikes, right more than left</td>
</tr>
<tr>
<td>6</td>
<td>Normal</td>
<td>Normal background, occasional small spikes over the left centrotemporal region.</td>
<td>Bilateral synchronous single sharp wave discharges over the parasagittal areas, maximal in the left central-parietal region. Some generalized discharges also showed left sided predominance. EEG changes did not always accompany clinical manifestations nor were associated consistently with EMG potentials.</td>
</tr>
<tr>
<td>7</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Jaw jerks were not associated with clear EEG changes.</td>
</tr>
<tr>
<td>8</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Single sharp wave left temporal</td>
</tr>
<tr>
<td>9</td>
<td>Normal</td>
<td>Normal background, sharp theta wave left mid-temporal.</td>
<td>Single left temporal sharp theta wave. Some jaw jerks were not associated with any EEG changes.</td>
</tr>
<tr>
<td>10</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Jaw jerks were not associated with clear EEG changes.</td>
</tr>
<tr>
<td>11</td>
<td>Normal</td>
<td>Normal background, small spikes over the left central and anterior to mid temporal areas, slightly activated by hyperventilation</td>
<td>Bisynchronous single spikes or multiple spike-and-wave complexes, maximal over the central or centrotemporal regions (left more than right). Jaw jerks were not associated with EEG changes on other recording.</td>
</tr>
<tr>
<td>12</td>
<td>Normal</td>
<td>Normal background, left anterior to mid temporal and occasionally central spike-and-wave discharges. Also, bisynchronous symmetrical spike–wave discharges with frontal maximum.</td>
<td>Repetitive spikes in the left central and temporal area, phase-reversing at C3 and T3, or showing equipotentiality at C3–T3.</td>
</tr>
<tr>
<td>13</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity</td>
<td>Bisynchronous frontocentral and generalized spike or spike-and-wave discharges. Naming a sequential series of pictures provoked a diffuse rhythmic activity at 8–9 Hz with generalized onset followed 3 s later by a GTCS (medication withdrawn)</td>
</tr>
<tr>
<td>14</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Generalized 3-Hz polyspike or spike-and-wave discharges</td>
</tr>
<tr>
<td>15</td>
<td>Normal</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Bisynchronous symmetrical sharp theta discharges with frontal maximum. Some jaw jerks were not associated with EEG changes RE with prolonged partial seizures manifested with alexia Left temporal rhythmic 0.8–1-Hz delta activity with equipotentiality at T3–T5 (subclinical). Partial seizure with left temporal onset of slow activity transformed into rhythmic sharp theta activity, restricted to the left.</td>
</tr>
<tr>
<td>16</td>
<td>Arachnoid cyst at the left temporal pole</td>
<td>Normal background, no paroxysmal activity.</td>
<td>Subclinical low voltage spike–wave discharges alternating with runs of regular low voltage slow activity over the left temporoparietal area.</td>
</tr>
<tr>
<td>17</td>
<td>Normal</td>
<td>Normal background without paroxysmal abnormalities. Brief low voltage spike-and-wave discharges during sleep 8–9 Hz sharpened rhythmic activity restricted to the left.</td>
<td>Partial seizure with left temporal onset of low voltage</td>
</tr>
</tbody>
</table>

Recorded, or directly observed, jaw jerks without noticeable electrographic changes. None of the jaw jerks in Patients 2, 7, and 10 were associated with EEG changes. In total, eight out of 15 patients had myoclonic jerks without EEG changes on at least one occasion. Regarding topography, clearly unilateral, ictal focal discharges were noted in four patients.
The variants of reading epilepsy

(1, 8, 9 and 12); they were all lateralized to the left side (Fig. 2). In one of them (Patient 8), a previous EEG during reading was reported as showing brief generalized spike-wave discharges associated with jaw jerks. Bilateral synchronous discharges were observed in the remaining eight patients. These were generalized in four patients (3, 11, 14 and 15), of frontotemporocentral distribution in two patients (4 and 5), and both bisynchronous frontocentral and generalized in two patients (6 and 13). Bilateral synchronous EEG findings were symmetrical in five patients (3, 4, 13, 14 and 15), lateralized to the left side in two patients (6 and 11) (Fig. 3) and to the right side in one (Patient 5). Ictal EEG changes were not invariably associated with muscle potentials in surface electromyography from the jaw and submental region (Fig. 3).

Anti-epileptic medication and outcome. Twelve patients were on anti-epileptic drug therapy (Table 1). Six of these were well controlled with clonazepam, given either alone (four patients) or in combination with sodium valproate (one patient) or clobazam (one patient), two with sodium valproate alone and one with phenobarbitone and diphenylhydantoin. The three remaining patients were partially controlled with sodium valproate, either alone (one patient) or in combination with carbamazepine and diphenylhydantoin, with the reading-provoked myoclonic jerks being less frequent and not evolving into GTCS. Patient 4 still experienced jaw jerks whilst reading. He was temporarily given diphenylhydantoin after his first convulsive seizures, long before the diagnosis of reading epilepsy was made, but received no anti-epileptic drugs thereafter. Patients 5 and 7 were also on no medication.

Reading epilepsy with prolonged focal seizures manifested with alexia

Ictal manifestations. Patients 16 and 17 had prolonged reading-induced partial seizures manifesting mainly with alexia. Mild dysphasia was evident on video tape analysis in Patient 16 and was suggested by history in Patient 17. Both described an associated dizziness and an ill-defined feeling of discomfort. No other ictal or postictal symptoms or clinical changes were elicited by history or observed on analysis of video tapes.

Other seizure types. In Patient 16 partial seizures evolved into GTCS on several occasions. In Patient 17 paroxysmal alexia never evolved into GTCS. He never had spontaneous daytime seizures but suffered infrequent nocturnal convulsions coinciding with the onset of the reading-provoked seizures.

Precipitating factors other than reading. There were no precipitating factors other than reading.
EEG findings. The routine EEG was normal in both patients (16 and 17). Reading characteristically provoked a subclinical focal EEG activation over the left posterior temporoparieto-occipital area. The effect was almost immediate, appearing whenever reading started and ceasing whenever it stopped. Ictal EEG changes were prolonged and clearly focal over the left posterior temporoparieto-occipital region.

Anti-epileptic medication and outcome. Both patients were started on anti-epileptic drug therapy after their first GTCS (Table 1). Treatment with phenobarbitone, diphenylhydantoin and clonazepam over a period of 7 years was ineffective in Patient 16, while Patient 17 received sodium valproate for 2 years, also without apparent benefit. Introduction of carbamazepine resulted in considerable improvement in both patients, with less frequent reading-induced partial seizures and rare GTCS associated with poor compliance.

Case reports

Patient 16. A 24-year-old right-handed student was referred at the age of 23 years with a history of infrequent GTCS over the last 7 years. He suffered a single febrile convolution at the age of 20 months lasting 3–5 min for which no medication was given. The first GTCS occurred at the age of 16 years whilst studying for school exams, the second in the following month whilst reading in the classroom, and a third a month later under similar conditions. Serial EEGs were normal and a brain MRI scan showed an arachnoid cyst in the left temporal pole (Fig. 1). Phenytoin and phenobarbital were given without substantial therapeutic effect. On referral, he was on phenytoin 400 mg and clobazam 30 mg/day. He had suffered 12 GTCS in total. Close questioning on possible precipitating factors revealed that all but one of the GTCSs occurred during reading and were invariably preceded by a stereotyped symptomatology. The patient described himself as 'sticking to a word, unable to understand its meaning. It is impossible to pass over it, I repeat the word several times. If I keep trying to read I may have a fit'. He was able to prevent GTCS by stopping reading as soon as he became dyslexic. He never experienced any other symptoms, but occasionally had an associated, rather ill-defined, dizzy feeling. His awareness always remained intact during these episodes. He never had symptoms while talking, writing or solving mathematical problems. There was no evidence of clinical or EEG photosensitivity at any time.

Prolonged video-EEG studies showed an entirely normal background without paroxysmal abnormalities during the resting state, hyperventilation and photic stimulation. Reading aloud an English text induced almost immediately bursts of slow activity at 1–1.5 Hz over the left temporal area, phase reversing over the posterior and, less often, the mid-temporal regions. Paroxysmal abnormalities ceased immediately when the patient stopped reading (Fig. 4, upper trace). A prolonged partial seizure was recorded in the last reading session (Fig.
The variants of reading epilepsy

1) when the patient started reading aloud a text on medical statistics, in his native language (Greek). The patient remained silent throughout the entire event. He was apparently able to understand and to give non-verbal responses to simple questions as well as to carry out simple commands. No motor manifestations or other behavioural changes were noted on video-EEG. The patient spoke for the first time 20 s after the end of the electrical discharge, denying in a slightly paraphasic manner to read further ‘I cannot read (instead of ‘read’) any more’. Introduction of carbamazepine 1000 mg daily resulted in considerable improvement.

**Patient 17.** This 29-year-old right-handed man was referred at the age of 24 years with a 2-year history of infrequent nocturnal convulsive seizures for which he was treated with sodium valproate, without apparent therapeutic effect. He had also had episodes of alexia since the age of 22 years. These usually occurred after prolonged reading and invariably consisted of gradual loss of ability to recognize, first, the infrequently encountered letters, then the commonest ones like the letter ‘A’, and finally the numbers. These episodes would last for 1–2 min and the ability to understand the reading material would resume in the reverse order. During the seizure he could understand other people talking to him, but he was only able to respond basically. He described an associated slight dizziness and a feeling of discomfort, but he never experienced these symptoms or any other symptom, suggesting epileptic seizure activity when he was not reading. Episodes of paroxysmal alexia occurred almost daily and were worse when he was tired. None of them ever evolved into a GTCS. Reading figures, talking, writing, playing cards or chess and solving mathematical problems never provoked any symptoms.

The patient had prolonged video-EEG studies during the alert resting state, multiple reading sessions and all night natural sleep. The EEG during the resting state, hyperventilation and photic stimulation was normal. Reading consistently activated the EEG, resulting in frequent asymptomatic brief small spike-and-wave discharges over the left temporoparietal area (Fig. 4, lower trace), alternating with runs of low voltage regular slow activity at 3–4 Hz, lasting up to 4 s. One of the patient’s habitual seizures was recorded on video-EEG (Fig. 6) and lasted for 75 s. During the subsequent all night natural sleep, spike-and-wave discharges were apparent over the left temporal area. Replacement of sodium valproate with carbamazepine 600 mg/day led to a considerable improvement, with only occasional episodes of reading-induced alexia over a follow-up period of 5 years. A single nocturnal GTCS occurred due to poor compliance. Follow-up video-EEGs showed only occasional bursts of left-sided slow waves evoked by reading. Talking, writing and solving mathematical problems never exerted any influence on the EEG, and generalized discharges were never recorded.

**Discussion**

We report 17 patients with reading epilepsy who were uniformly investigated with state of the art video-EEG and

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**Fig. 4** Partial variant of reading epilepsy. Reading-provoked subclinical focal EEG activation over the left posterior temporoparietal areas in Patients 16 and 17. The upper recording (Patient 16) shows irregular bursts of low voltage monomorphic slow activity during reading aloud. Note that paroxysmal abnormalities cease as soon as the patient stops reading (arrow) when instructed to do so. The lower recording (Patient 17, same montage) shows small spike-wave discharges during silent reading with clear equipotentiality over T3–T5. The resting EEG was normal in both patients.
Reading-induced partial seizure manifested with alexia in Patient 16. Soon after the patient started reading aloud, rhythmical monomorphic delta activity at 0.8–1 Hz appeared over the left temporal area showing a relative equipotentiality over T3–T5 channel and extending to the ipsilateral parietal–occipital region (first arrow). The patient was still able to read but 9 s later (20 s after behavioural onset, second arrow) he stuck on a word, hesitated and repeated it for a couple of times as though he tried to understand it, and pressed the event button. The EEG normalized for the next 20 s and then an irregular low voltage slow activity at 3.5–5 Hz appeared on the left (third arrow), followed by a sharpened rhythmic theta activity at 5 Hz (fourth arrow). The electrical seizure ceased 94 s after the onset of the first behavioural changes, followed by a short period of left postictal suppression. For other behavioural changes, see text. The numbers on the left side of the arrows indicate the time in seconds after the first behavioural changes. The seizure is depicted in a discontinuous manner to include only the periods during which distinct behavioural and electrical changes occurred.

structural neuroimaging studies. On the basis of ictal video-EEG studies we were able to identify two seizure types with distinct semiological and electrographic features: (i) myoclonic jerks of the jaw that may spread to the upper limbs, associated usually, but not invariably, with brief discharges of variable morphology and spatial distribution; and (ii) paroxysmal alexia with prolonged, and strictly focal, ictal EEG discharges.

In all but one patient with the myoclonic form, typical seizures indistinguishable from those induced by reading could also be provoked by other language-related higher cognitive functions. In these patients myoclonic jerks were frequently associated with some form of cognitive impairment. Three of our patients had a sibling with reading epilepsy, suggesting a specific genetic background for the syndrome. The family history of epileptic seizures was otherwise unremarkable in this series.

**Clinical features**

The age at onset of the reading-induced seizures appears to be one of the most constant features of reading epilepsy. The median age of onset of 15 years (range 11–22 years) in our study is similar to the figures given by Radhakrishnan et al. (1995) (median 17.5 years; range 10–46 years) and Wolf (1992) (mean age at onset of 17.7 years with only three cases <12 years and five >25 years), indicating that reading epilepsy is clinically manifested after the acquisition of reading skills has been completed. The male : female ratio is also similar in these major series [2.4 in the present study, 1.8 in the report by Wolf (1992) and 1.9 in the report by Radhakrishnan et al. (1995)], confirming that reading epilepsy is a syndrome with male preponderance.

**Myoclonic reading epilepsy**

The typical reading-induced jaw or orofacial myoclonus was by far the commonest and most constant ictal manifestation. The severity and frequency of the myoclonic seizures varied even in individual patients. In five of them reading-provoked jerks occasionally spread to the upper limbs, as previously reported (case 3 of Stevens, 1957; five cases of Radhakrishnan et al. (1995)). None of our patients gave a history of spontaneous myoclonic jerks. This is in contrast to the five patients (2, 9, 10, 14 and 17) of Radhakrishnan et al. (1995) who had spontaneous myoclonic jerks in addition to the reading-provoked ones, but also calculation-induced seizures (2, 9, 14 and 17), suggesting a more widespread cortical hyperexcitability than in pure reading or language-induced epilepsy. An overlap of reading epilepsy with juvenile myoclonic epilepsy was suggested on the basis of some clinical findings such as the age at onset, bilateral myoclonic
The variants of reading epilepsy

Fig. 6 Prolonged reading-induced partial seizure manifested with alexia in Patient 17. After a series of small spike-wave discharges induced by silent reading (first arrow), the EEG normalized for 4.5 s. The patient then pressed the event button signaling his inability to understand the text, and this coincided with a single small spike-wave complex (second arrow). Three seconds later, a sharpened, low amplitude, rhythmical activity at 8–9 Hz appeared synchronously on the left hemisphere being more evident on the posterior temporal areas and lasted for 20 s. The EEG became entirely normal again (third arrow) but the patient clearly stated that he could read again perfectly 50 s later. The patient remained calm and silent throughout the seizure and no motor or other clinical manifestations were observed by careful video analysis.

jerks, strong influence of heredity, progression to GTCS, response to sodium valproate and persistence through life (Radhakrishnan et al., 1995). In reading epilepsy, however, myoclonic jerks are reading-induced and involve mostly the jaw, whereas, in patients with juvenile myoclonic epilepsy, jerks affect the extremities, and precipitation with reading has not been described (Janz, 1989; Canevini et al., 1992; Panayiotopoulos, 1996). Furthermore, absences are present in up to one-third of patients with juvenile myoclonic epilepsy (Panayiotopoulos et al., 1989; Janz and Waltz, 1995), but are rare in reading epilepsy.

Although ictal manifestations in reading epilepsy are said to occur without impairment of consciousness, one of our patients reported a ‘light bulb off/on sensation’, and a second reported a ‘think block’ when jaw jerks occurred. Similar clinical evidence of brief impairment of consciousness can be found in patient 2 of Bingel (1957), in patient 1 of Saenz-Lope et al. (1985), and in patients 4, 15 and 19 of Radhakrishnan et al. (1995). Many of our patients described themselves as ‘sticking to the word’ or, more frequently, reported ‘loss of track of the reading text’ when a jaw jerk occurred, which made them restart reading from the beginning of the sentence. The phenomenon seemed to correlate with the severity of the motor manifestations, and indeed was present in all four patients with widespread reading-induced myoclonus. While the ictal stammering can be explained on the basis of the brief ictal jaw/or perioral clonic or tonic activity which impedes the flow of speech, the semiological implication of the ‘losing track’ symptom may be more complex. Forster et al. (1969) reported a patient with ‘loss of place in the text’, while Brooks and Jirauch (1971) described a patient with limited comprehension of the reading material on attempting to read, in spite of the jerking. These symptoms may suggest a selective cognitive impairment associated with the brief ictal spike-wave discharge as it has been shown to occur with subclinical epileptiform discharges. Transient cognitive impairment has been associated mainly with generalized (Mirsky and Van Buren, 1965), but also focal discharges (Kooi and Hovey, 1957), so that left-sided discharges tend to interfere more with verbal tasks relative to spatial tasks, and the converse holds for right-sided discharges (Aarts et al., 1984). As the ictal electroclinical phenomena of the myoclonic form of reading epilepsy are very brief, and the involvement of the cortical neurons is not massive, an overt ‘impairment of consciousness’ is unlikely to occur. Nevertheless, as the act of reading is, at the same time, the provoking stimulus and evidently the most specific test for detecting a transient impairment of language functions, symptoms such as ‘loss of track of the text’ or ‘sticking to a word’ may imply that reading-induced myoclonic seizures are not always as simple as currently appreciated (Commission on Classification and Terminology of the ILAE, 1989).
Partial reading epilepsy

Prolonged alexia, strikingly different from the transitory ‘loss of track in the text’ of the myoclonic form, was the prevailing ictal manifestation in two patients. In addition, the postictal speech of Patient 16 was slightly paraphasic, and mild expressive dysphasia was suggested by history in Patient 17. Both patients reported an associated feeling of discomfort and dizziness. Ictal semiology suggested a selective involvement of the dominant angular region without propagation to neighbouring areas. Motor manifestations were never reported or observed.

Precipitating factors

Reading was the principal triggering factor in all patients. In addition, the habitual regional myoclonus could be elicited in 14 patients by talking, and in six by writing, albeit less frequently than by reading. The sister of Patient 8, who had jaw jerks only when reading, reported jaw jerks elicited exclusively by talking fast, the phenomenon suggesting that different language modalities may trigger identical symptoms in subjects belonging to the same pedigree. Talking and writing were not effective seizure triggers in the two patients with the reading-induced partial seizures manifested with alexia.

In his detailed review Wolf (1992) found that among 91 patients, 29 (32%) also had talking- or writing-induced regional jerking which, on the latter occasions, would also involve the arm used for writing. Radhakrishnan et al. (1995) also reported that talking, writing and even listening to conversation were effective seizure triggers in six of their 20 patients. Cirignotta et al. (1986) reported two patients with myoclonic jerks precipitated by writing and only occasionally by reading. Therefore, accumulated clinical evidence indicates that in reading epilepsy all three facets of language function may be specific seizure-provoking stimuli, perhaps to the extent that the term ‘language-induced epilepsy’, coined by Geschwind and Sherwin (1967), may be justified.

The role of non-linguistic higher cognitive processes in precipitating seizures in reading epilepsy is less clear. Radhakrishnan et al. (1995) reported that six of their patients had seizures when engaged in cognitive tasks such as calculating, concentrating or playing chess. Two of our patients with the myoclonic form gave a history of thinking-induced seizures. Similar evidence is found in a few case reports (case 3 of Bingel, 1957; Hosokawa et al., 1965; case 9 of Forster, 1977; case 2 of Wolf, 1978) but in the last case the seizures when playing chess were different from the habitual reading-induced seizures (Wolf, 1978). Despite some overlap regarding the seizure triggers, there are significant differences between reading epilepsy and the other higher cognitive reflex epilepsies. In thinking epilepsies, myoclonic jerks involve mainly the upper limbs, clinical absences are present in about 60% of patients, and photosensitivity is elicitable in one-third of patients (Goossens et al., 1990).

Conversely, in most cases of reading epilepsy, myoclonic jerks are characteristically restricted to the jaw or perioral musculature, reading-induced absences have so far been described in only three patients (Alajouanine et al., 1959; Matricardi et al., 1991; Singh et al., 1995), and photosensitivity is rare (Wolf, 1992). Furthermore, reading epilepsy has a strong genetic component while a family history of epileptic seizures in thinking epilepsy does not appear to be different from that in other patients with idiopathic generalized epilepsies (Goossens et al., 1990).

Genetics

Reading epilepsy has a distinct genetic background. Reading-induced jaw jerking has been described in paired cases of parent and child, suggesting dominant inheritance (Matthews and Wright, 1967; case 9 of Radhakrishnan et al., 1995), as well as between monozygotic twin pairs (Forster and Daly, 1973). Furthermore, a clear inheritance of spike-and-wave reflex activation during reading has been well documented in asymptomatic family members of patients with reading epilepsy (Daly and Forster, 1975). In the recent meta-analysis by Wolf (1992), 18 of 69 index patients (with reading epilepsy and available information on family history) had first-degree relatives with epilepsy or epileptic seizures of known type; 11 of them had reading epilepsy, three had idiopathic generalized epilepsies, two had febrile seizures, one had GTCS since the age of 3 years, and one had symptomatic localization-related epilepsy. Radhakrishnan et al. (1995) found that four of the 18 patients with a known family history had first-degree relatives with epileptic seizures and one had reading epilepsy, while the information in the remaining three is not clear. We found three families with siblings affected with reading epilepsy, and none of our patients had a family history of any other form of epilepsy. The high incidence of reading epilepsy among the first-degree relatives [15 subjects: 11 in the review by Wolf (1992); patient 9 of Radhakrishnan et al. (1995); and three in the present report] of the index cases with known family history reported so far [103 subjects: 69 in the review by Wolf (1992); 18 in the report of Radhakrishnan et al. (1995); and 16 patients in the present report] reflects a specific genetic background for the syndrome, but the mode of inheritance is still unclear. On the other hand, the few reported patients with reading epilepsy and relatives with idiopathic generalized epilepsy do not suggest any clear genetic link. More substantiated hypotheses regarding the question raised recently on the possible relationship between reading epilepsy and juvenile myoclonic epilepsy need a larger number of patients with reading-induced seizures and electroclinical features suggesting juvenile myoclonic epilepsy, while families with members affected by both conditions will allow molecular genetic studies. It should be noted, however, that none of the patients reported so far had a family member with juvenile myoclonic epilepsy. Similarly, idiopathic localization-related epilepsies, such as benign epilepsy with centrotemporal or occipital
spikes, have not been reported among family members of patients with reading epilepsy.

**EEG features**

**Myoclonic reading epilepsy**

**Interictal findings.** We found left-sided focal interictal spontaneous epileptiform abnormalities in four (27%) of our patients; one of them had in addition symmetric generalized spike-and-wave discharges. Radhakrishnan et al. (1995) reported seven patients (35%) with spontaneous bilateral spike-and-wave discharges and none with focal abnormalities. Of 104 patients with reading epilepsy reported earlier, Wolf (1992) found 11 (11%) with bilateral spike-and-wave discharges, and five (5%) with paroxysmal temporal abnormalities. Nine per cent of patients were photosensitive in this review (Wolf, 1992), but none in the Mayo Clinic abnormalities. Of 32% showed bilateral symmetrical discharges, 38% (1992), 32% showed bilateral symmetrical discharges, 38% bilateral lateralized and 30% unilateral. Lateralization to the dominant hemisphere was noticed in 78% and to the non-dominant side in 10%, and it switched between sides in 12% of patients. Unilateral or bilateral regional discharges were noted in fairly equal proportions. Bilateral asymmetric and focal discharges were most often lateralized to the left (six patients) than the right hemisphere (one patient) (Table 2). Ictal changes were not always associated with EMG potentials from the jaw muscles.

Ictal EEG findings have been found to vary widely in spite of the rather uniform clinical correlates (jaw jerk). Of 73 available ictal traces in the literature reviewed by Wolf (1992), 32% showed bilateral symmetrical discharges, 38% bilateral lateralized and 30% unilateral. Lateralization to the dominant hemisphere was noticed in 78% and to the non-dominant side in 10%, and it switched between sides in 12% of patients. Unilateral or bilateral regional discharges were more often temporal-parietal (80%) than frontocentral (20%). Radhakrishnan et al. (1995) reported a significantly higher proportion of patients with generalized symmetrical ictal discharges (75%). Discharges were strictly lateralized to the dominant side in only 10%, and were generalized with dominant side preponderance in 15% of patients. Despite this obvious variability, ictal EEG phenomena have been used as a major criterion as to whether reading epilepsy is a localization related or a generalized syndrome (Commission on Classification and Terminology of the ILAE, 1989; Wolf, 1989, 1992; Radhakrishnan et al., 1995).

**Electroclinical correlation.** Patients with reading-induced myoclonic seizures involving the upper limbs did not show a specific discharge pattern. Five out of 10 patients with some clinical evidence of selective cognitive impairment showed ictal EEG changes lateralized towards the left hemisphere; Patients 1, 9 and 12 had left focal abnormalities, and Patients 6 and 11 had bilateral discharges with left-sided preponderance. Two patients (7 and 10) showed no ictal EEG changes while generalized symmetrical discharges were noted in three patients (3, 4 and 15), of whom Patient 3 reported an associated ‘light bulb off/on’ sensation, and Patient 15 a ‘thinking block’. Similar left-sided predominance was noted in two previous case reports with clear evidence of selective cognitive impairment; the patient of Forster (1969) with ‘loss of place in the text’ associated with bitemporal spiking, sharper on the left, and with some generalization, and the patient of Brooks and Jirach (1971) with ‘only limited comprehension of the reading material’, associated with synchronous bilateral discharges, accentuated often, and occasionally only on the left.

**Partial reading epilepsy**

In both patients with partial reading epilepsy, reading provoked a continuous, subclinical activation of the EEG which was localized over the language-dominant posterior temporal area. This was specific, reproducible and clearly distinguishable from the typical brief and frequently bilateral synchronous reading-induced discharges of the myoclonic form which were intermittent and indeed ictal (Table 3). This focal activation finally resulted in prolonged partial seizures associated also with unilateral and strictly focal EEG abnormalities clinically expressed as prolonged alexia with varying degrees of dysphasia.

Similar electroclinical features have been rarely reported. Chavany et al. (1956) reported a case with reading-provoked left occipital sharp transients and ictal symptoms consisting of the letters changing place vertically and horizontally and becoming entirely incomprehensible. This was associated with a rhyththmic theta activity of left occipital predominance which was replaced 20 s later by delta activity at 1.5–2 Hz, the whole electrical event lasting for ~2 min. Gastaut and Tassinari (1966) recorded a partial seizure which consisted of a fast rhythmic discharge lasting for 50 s and remained localized at the left parieto-occipital region. According to their description ‘the last word was seen by the patient in a foggy way, then it became fixed even though the patient had the eyes closed, the letters changed place and the word became distorted.’ A second seizure in the same subject was preceded by a laryngeal sensation which coincided with a brief generalized burst of spike-and-wave activity of right predominance. Their patient tried to read the last word many times, and the EEG showed a rapid rhythmic activity arising from the right parietal region, followed by multiple spikes and sharp waves which became generalized. The patient suffered a left versive seizure with subsequent generalization.
Table 3 EEG findings in 17 patients with reading epilepsy

<table>
<thead>
<tr>
<th>Routine EEG</th>
<th>Myoclonic form (n = 15)</th>
<th>Partial form (n = 2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. and (%) of patients</td>
<td>No. and (%) of patients</td>
<td></td>
</tr>
<tr>
<td>Background</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>14 (93)</td>
<td>2 (100)</td>
</tr>
<tr>
<td>Bitemporal slow waves</td>
<td>1 (7)</td>
<td>–</td>
</tr>
<tr>
<td>Spontaneous epileptiform discharges</td>
<td>4 (27)</td>
<td>–</td>
</tr>
<tr>
<td>Unilateral, focal (left temporal-central)</td>
<td>4</td>
<td>–</td>
</tr>
<tr>
<td>Generalized with frontal maximum</td>
<td>1*</td>
<td>–</td>
</tr>
<tr>
<td>Nil</td>
<td>11 (73)</td>
<td>2 (100)</td>
</tr>
<tr>
<td>Paroxysmal abnormalities on reading provocation</td>
<td>Brief, intermittent, associated with jaw jerks</td>
<td>Almost continuous, subclinical, focal (left posterior temporal-parietal)</td>
</tr>
<tr>
<td>Ictal discharges</td>
<td>12 (80)</td>
<td>2 (100)</td>
</tr>
<tr>
<td>Bilateral, symmetric</td>
<td>5 (33)</td>
<td>–</td>
</tr>
<tr>
<td>Bilateral, asymmetric</td>
<td>3* (20)</td>
<td>–</td>
</tr>
<tr>
<td>Brief, focal (left central–temporal-frontal)</td>
<td>4 (27)</td>
<td>–</td>
</tr>
<tr>
<td>Prolonged focal (posterior temporal–parietal–occipital)</td>
<td>–</td>
<td>2 (100)</td>
</tr>
<tr>
<td>Nil</td>
<td>3 (20)</td>
<td>–</td>
</tr>
</tbody>
</table>

*This patient had also focal discharges. †Two with left- and one with right-sided predominance.

Patient 12 of Radhakrishnan et al. (1995) gave a history of inability to comprehend the meaning of the words without any jaw myoclonus before three GTCSs induced by reading. This patient had a left posterior temporoparietal seizure during pentylentetrazol activation while reading. In addition to the partial seizures, and independent of these, he had reading-induced jaw jerks associated with generalized spike-wave discharges (personal communication).

Reading-induced partial seizures manifested with alexia may be apparently idiopathic (Gastaut and Tassinari, 1966; Radhakrishnan et al., 1995) or of symptomatic origin (Chavany et al., 1956). The normal neurological status, resting EEG activity and brain MRI of Patient 17 are consistent with idiopathic epilepsy. In Patient 16, the relevance of the arachnoid cyst is uncertain. The partial form of reading epilepsy appears to be rare, as only five cases have been reported during the last 43 years. It is possible that the lack of the typical jaw jerk may have led to the misdiagnosis of some cases, or to the misplacement of others in the group of the poorly defined ‘secondary’ reading epilepsy which has received little attention in recent years. Occasionally, partial and myoclonic seizures may coexist in the same subject (patient 12 of Radhakrishnan et al., 1995).

Mechanisms of epileptogenesis

Several pathophysiological mechanisms have been proposed to explain seizure precipitation in reading epilepsy, but pathogenesis still remains obscure. These mechanisms include proprioceptive input from the peripheral muscles (jaw, larynx and eyes), abnormalities of the visual system, heredity, and central language and cognitive mechanisms (Ramani, 1998), but they do not operate uniformly in every patient. For example, proprioceptive impulses from the jaw muscles may be effective in some patients (Bickford et al., 1956; Brooks and Jirauch, 1971) but not in others (Geschwind and Sherwin, 1967; Ramani, 1983). Similarly, the complexity (Christie et al., 1988) and comprehension (Kartsounis, 1988) of the reading material have been shown to be potent and even critical triggers, but unintelligible text may also be particularly provocative (Stevens, 1957; Wolf, 1978), and even nonsense reading may be as effective as logical reading (Bickford et al., 1956). In most of the reported cases, no single factor can be exclusively implicated as the critical precipitating stimulus, and a cumulative effect of different factors has been postulated (Christie et al., 1988).

Anti-epileptic treatment and outcome

Of the 14 patients who received anti-epileptic drugs, nine are seizure-free but continue on medication. The remaining five patients continue to have a reduced number of reading-induced seizures, and no further GTCS. The response to the anti-epileptic medication appeared to depend on the seizure type. Clonazepam was particularly effective in myoclonic seizures, as noted by others (Login and Kolakovich, 1978; Hall and Marshall, 1980; Murphy and Yamada, 1981). Sodium valproate is considered effective in patients with the myoclonic form (Vanderzant et al., 1982), but failed to eliminate the reading-induced seizures in three out of six patients to whom it was given. In patients with prolonged partial seizures, treatment should probably be considered on an individual basis, but drugs for partial seizures seem to be the first choice (see Patients 16 and 17 here and case 12 of Radhakrishnan et al., 1995).
factors implicated so far should be regarded as having facilitatory action on an altered neuroanatomical substrate rather than indicating different pathogenetic mechanisms. Radhakrishnan et al. (1995) proposed that the interaction between hyperexcitable focal cortical areas and cortico-reticular systems, under provocation by linguistic or other higher cognitive activities, may give rise to primary generalized myoclonus and explain the EEG variability. Ramani (1998) suggested that reading may elicit an abnormal response from a vulnerable link in the underlying neural systems, and that the location of the defect at the input (visual registration), processing (meaning extraction) or output (motor expression) stage may determine the diverse clinical subtypes accordingly.

Concept of network hyperexcitability
The classical model of centrencephalic epilepsy has been replaced by the concept of the generalized corticoreticular epilepsies (Gloor, 1968, 1979) according to which diffuse cortical hyperexcitability plays a major role, responding with spike-and-wave activity to normal afferent volleys. It can be reasonably hypothesized that in epilepsies with seizures precipitated by specific modes of activation, a localized cortical hyperexcitability of either genetic or acquired origin may give rise to an epileptic process after the stimulus specific for that area is applied. In photosensitive epilepsies, intermittent photic stimulation can produce occipital spikes before the onset of generalized discharges (Panayiotopoulos et al., 1970; Jeavons and Harding, 1975), the occipital cortex is not uniformly hyperexcitable (Wilkins et al., 1981), and epileptogenesis is thought to be associated with the synchronization of the individual neurons within the visual cortex by the stimulus (Binnie et al., 1985). Such a hypersensitive area is not easily conceivable in reading epilepsy. Reading is a complex cognitive process including visual analysis, memory functions, then either semantic processing or a more mechanical grapheme-to-phoneme conversion, followed by articulation and acoustic monitoring. These subsystems of the reading process are integrated into a network that expands over different brain areas. In PET studies investigating brain activity during normal reading, the left middle temporal, left frontal and left medial extra-striate cortices, as well as right sided homologous regions, have been activated (Petersen et al., 1990; Wise et al., 1991). Right hemisphere activity during reading suggests that, even with normal right-handed subjects, processing of written words is not restricted to the left hemisphere (Price et al., 1994). The left posterior temporal and left inferior parietal cortex have recently been shown to be associated with word processing (Price et al., 1996). In patients with reading-induced seizures, endogenous opioids were released only during or after reading-induced epileptic seizures in both temporal and frontal lobes. These hyperexcitable cortical areas are part of the normal reading network (Koepp et al., 1997, 1998). Ictal SPECT (single photon emission computed tomography) also showed focal hyperperfusion of the frontal lobes bilaterally and the left temporal area in a right-handed subject with reading-induced jaw jerks (Miyamoto et al., 1995).

Therefore, in reading epilepsy this hyperexcitability should be conceptualized as a network or regional, involving multiple, non-contiguous anatomically cortical areas in both hemispheres rather than being localized. This concept can explain the fact that precipitating stimuli include all aspects of language function (talking, writing and even listening to conversation).

Network hyperexcitability and the type of clinical seizure
Reading-induced seizures are usually brief myoclonic, but in some patients may be prolonged and clearly focal with paroxysmal alexia or, exceptionally, absences. The underlying mechanisms accounting for these different clinical manifestations are not clear. Although ictal PET studies in Patients 16 and 17 were not performed, it can be hypothesized on the basis of the existing electroclinical evidence that in the clearly partial variety, the hyperexcitable zone is highly localized at, and restricted to, posterior temporal areas implicating the dominant angular gyrus and its connections with the associative visual cortex. Conversely, ictal functional neuroimaging studies in patients with the myoclonic variant clearly showed bitemporal and bifrontal areas of hyperexcitability (Miyamoto et al., 1995; Koepp et al., 1998), consistent with the observations in patients with symptomatic reading-provoked myoclonic seizures. The patient of Lee et al. (1980) had language-induced seizures after a stroke in the dominant frontal cortex, the patient of Ritaccio et al. (1992) had reading-induced seizures after removal of a left frontal arteriovenous malformation, and patient 2 of Radhakrishnan et al. (1995) had language- and calculating-induced seizures after the acute onset of right hemiparesis, presumably due to a stroke.

Clinical ictal manifestations in myoclonic reading epilepsy have striking similarities to cortical reflex myoclonus, which is related to the hyperexcitability of a small area of the sensorimotor cortex, and typically involves a few adjacent muscles (Hallett, 1985). However, the relationship between the stimulus and the response in reading epilepsy is not immediately evident. Although the proprioceptive bombardment from the jaw or laryngeal muscles in consequence of reading has been shown in some patients to facilitate the occurrence of the myoclonic jerk (Bickford et al., 1956; Brooks and Jirauh, 1971; Meyer and Wolf, 1973), the common precipitating mechanism in all patients with reading epilepsy is the formal act of reading, that is the transformation of the displayed linguistic material into speech (Wolf, 1992). Furthermore, in patients with reading-induced seizures, the sensorimotor cortex is not hyperexcitble per se, as spontaneous jaw jerks never occur. To explain the reading-
induced regional myoclonus, one has to embark on the hypothesis that the hyperexcitable neuronal network that subserves speech may drive the relative motor cortex effectively through a direct transcortical pathway, as has been thought to occur in reflex myoclonus elicited by highly specific somatosensory stimuli (Hallett et al., 1979; Marsden et al., 1983). According to this model of epileptogenesis, the bilaterally hyperexcitable speech network under reading provocation may eventually give rise to bilateral and synchronous myoclonic jerks which may not be necessarily symmetric. The phenomenon of the bilateral involvement of the masticatory and lower facial muscles, in spite of their predominantly contralateral innervation, can also be explained on this basis. Neuronal excitation in the motor strip may spread and, if the process is not interrupted, secondary generalization may occur, sometimes through a ‘cortical march’ (Saenz-Lope et al., 1985). Depending on the level of the network hyperexcitability, the cumulative effect of more factors may be needed for the generation of the response, as for example reading aloud (implying additive auditory and possibly proprioceptive input) as opposed to silent reading, and comprehension of a complex text versus mechanical reading. Also, prolonged effort with sustained concentration and the resulting fatigue (Stevens, 1957; Gastaut and Tassinari, 1966; Ramani, 1998), and other non-specific factors, such as sleep deprivation, alcohol intake and menstruation (Ramani, 1998), may lower the epileptic threshold of the speech network.

Network hyperexcitability and pattern of the ictal EEG phenomena
Non-uniform cortical hyperexcitability may lead to generalized discharges with variable degrees of symmetry, for example, in patients with reflex seizures induced by calculation, card or board games, and spatial tasks (Goossens et al., 1990), and in photosensitive epileptic patients (Binnie et al., 1981). In reading epilepsy, the concept of the variable bilateral cortical hyperexcitability at multiple levels clearly allows any asymmetry or regional predominance of a generalized ictal EEG discharge, and even regional or focal discharges.

Such an interpretation is admittedly highly speculative and certainly, the issue of epileptogenesis in reading epilepsy remains open to alternative hypotheses. Nevertheless, this theory can explain the clinical and EEG phenomena in reading epilepsy and their heterogeneity, and the effectiveness of the various linguistic processes as seizure triggers, and it is consistent with the recent functional neuroimaging findings.

On the classification of reading epilepsy
Clinical classification of reading epilepsy
Radhakrishnan et al. (1995) proposed the classification of reading epilepsy into (i) pure or overlap syndromes, according to whether reading is the single specific trigger or there are additional closely related triggers and (ii) primary or lesional syndromes, according to the aetiology. In addition to the above, reading epilepsy can be differentiated according to the type of the clinical seizure into the most common myoclonic form, the less frequent form with prolonged paroxysmal alexia, and occasionally into a form with reading-induced absences. These distinctions are worthwhile and useful in terms of diagnosis, treatment and possibly prognosis. However, more complex classifications should not be encouraged, as reading or language epilepsy can only be intrinsic (Gastaut and Tassinari, 1966) and specific (Bickford, 1973).

Position of reading epilepsy in the current classification system of the ILAE
The present classification of reading epilepsy among the localization-related idiopathic epilepsies with age-related onset (Commission on Classification and Terminology of the ILAE, 1989) is based on the assumption that the regional myoclonus, as always developing in clear consciousness, is the clinical expression of a simple motor partial seizure. Further arguments include the relationship to a specific cortical area (the angular region of the language-dominant hemisphere), the age-related onset and the relatively benign course, in the sense that seizures remain strictly bound to the precipitating factor. Generalized spike-and-wave discharges, when present, are attributed to a genetic trait (Wolf, 1989, 1992). Conversely, Radhakrishnan et al. (1995) expressed the view that the age at onset, the anti-epileptic drugs to which it is more responsive, the natural history and the EEG findings are distinct features from those of benign epilepsy with centrotemporal or occipital spikes. These authors emphasized the overlap between reading epilepsy and epilepsies with seizures precipitated by language-related activities other than reading and high cognitive processes; they stressed the electroclinical and therapeutic similarities and proposed the classification of reading epilepsy amongst the idiopathic generalized epilepsies with seizures precipitated by specific modes of activation.

In his critical review, Ramani (1998) emphasized the inadequacy of the distinction between primary and secondary types as a classification tool, and pointed out that the clinical heterogeneity of reading epilepsy defies its position in any single category of the present classification schema of the ILAE. This author proposed that the different subtypes (of the myoclonic form) can be categorized separately into the groups of idiopathic generalized (age-dependent), idiopathic localization-related and symptomatic localization-related epilepsies, depending on clinical and EEG features, while Bickford’s secondary reading epilepsy can be included into the group of symptomatic generalized epilepsies (Ramani, 1998).

Our present findings confirm that, apart from the myoclonic...
reading- or language-induced epilepsy which represents a valid entity, given its relatively uniform clinical semiology (Ramanı, 1998), a second variant also exists with clearly partial seizures manifested by prolonged episodes of dyslexia. These observations, along with the recent description of a patient with reading-induced absence seizures (Singh et al., 1995), indicate that the electroclinical spectrum of reading epilepsy is much wider than currently believed. Gastaut and Tassinari (1966) were the first to question the consistency of the electroclinical criteria of reading epilepsy that were originally delineated by Bickford (1956) and proposed the term ‘intrinsic’ or ‘perceptive’ instead of ‘primary’ as more emphatic of the cause–effect link between reading and seizures, focusing on the specificity of the reflex mechanism rather than the type of the seizures or their ‘idiopathic’ or ‘symptomatic’ origin. We concur that there is no reason for splitting reading epilepsy into separate syndromes that share the same precipitating factors and pathophysiology, and in this sense reading epilepsy cannot fit into any given category of the present classification system of the ILAE with the dual dichotomy between ‘generalized’ and ‘focal’ or ‘partial’ for seizures, and ‘generalized’ and ‘localization-related’ for syndromes. Even if a separate categorization was pursued, it would present no difficulty regarding the clearly partial and absence subtypes, but would result in obvious inconsistencies regarding the myoclonic variant. Although the demonstration, with functional neuroimaging, of the bilateral hyperexcitable parts of the neuronal network which integrate speech is a reasonably sufficient argument for positioning myoclonic reading epilepsy among the localization-related syndromes (Commission on Classification and Terminology of the ILAE: proposal for revised classification of epilepsies and epileptic syndromes, 1989), the fact that it is manifested in at least two out of three of the patients with generalized seizures [in which the first clinical changes indicate initial involvement of both hemispheres and the ictal encephalographic patterns are initially bilateral (Commission on Classification and Terminology of the ILAE: proposal for revised clinical and encephalographic classification of epileptic seizures, 1981)] makes such a classification more confusing than informative. From the clinician’s point of view, reading- or language-induced myoclonic seizures are neither partial nor generalized, but bilateral and synchronous focal motor resulting from the simultaneous activation of parts of the speech network that expands over both hemispheres, with a potential for rapid spread and secondary generalization if exposure to the stimulus is not interrupted. Further electrophysiological and functional neuroimaging studies in epilepsies with seizures precipitated by specific modes of activation are expected to facilitate understanding of epileptogenesis, and any future classification system should encompass ongoing developments. Until then, an effective definition of reading- or language-induced epilepsy should be based largely on clinical semiology, and any attempt to position the syndrome in the current classification system of the ILAE would be premature.

Proposed definition

Reading epilepsy is a distinct form of reflex epilepsy in which all, or almost all, seizures are precipitated by the act of reading. In many patients, clinically identical seizures can also be provoked by the other linguistic activities to the extent that the term language-induced epilepsy should be justified. The clinical spectrum of reading- or language-induced epilepsy is wide. Regional myoclonic jerks that most often involve the masticatory muscles and the tongue, but may also extend to the upper extremities, is the most frequent and remarkably uniform seizure pattern. Some patients have prolonged, clearly partial, seizures manifested with alexia and possibly dysphasia, while occasionally absences may occur. Seizures usually evolve into GTCS if reading persists. Ictal EEG changes show considerable heterogeneity in terms of discharge morphology and scalp topography. In the myoclonic variant, discharges are brief, bilateral synchronous or focal, while in the partial variant they are prolonged and focal. There is a clear tendency for left hemisphere predominance. Reading or language epilepsy is most often idiopathic and rarely symptomatic. Ictal functional neuroimaging studies show multiple cortical hyperexcitable areas that are part of the neuronal network which subserves the function of speech. Secondary excitation of the relative motor areas may produce the typical regional myoclonus and the underlying physiological mechanism may relate to cortical reflex myoclonus. This network hyperexcitability can be genetically determined, and its clinical expression is age-related. Reading or language epilepsy is non-progressive and optimal treatment relates to the electroclinical variant; patients with the myoclonic form are usually best treated with clonazepam or sodium valproate, whereas those with the partial variant respond to anti-epileptic drugs that are effective for partial seizures.

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The variants of reading epilepsy


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