Acquired epileptic dysgraphia: a longitudinal study

MAYOR, Claire, et al.

Abstract

A male presenting with benign partial epilepsy with rolandic spikes from the age of 7 years was evaluated at age 11 years for worsening of his epilepsy associated with a specific regression of graphomotor skills. A longitudinal study over nearly 2 years showed an improvement in handwriting to an almost normal level under modified antiepileptic therapy. A detailed analysis with a computer-monitored graphics table showed at first a rapid improvement of skills followed by protracted slower progress. We argue that the initial rapid recovery of skills was directly linked to the improvement of his epilepsy. The slower late acquisition of motor programmes that had never been fully established was due to longstanding interference by his epilepsy. The specificity of the deficit within the graphomotor system and its possible neurobiological basis are also discussed. The analytical method and approach used in a single patient might provide an example for other patients in whom epilepsy can interfere in the acquisition, progress, and maintenance of new skills and can be responsible for selective deficits.

Reference


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Acquired epileptic dysgraphia: a longitudinal study

Claire Mayor Dubois MA, Neuropaediatric Unit, Department of Paediatrics, Centre Hospitalier Universitaire Vaudois, Lausanne; Pascal Zesiger PhD, Faculty of Psychological and Educational Sciences, University of Geneva, Geneva 4; Eliane Roulet Perez MD, Neuropaediatric Unit, Department of Paediatrics; Malin Maeder Ingvar MD, Clinical Physiology Unit; Thierry Deonna* MD, Neuropaediatric Unit, Department of Paediatrics, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland.

*Correspondence to last author at Neuropaediatric Unit, Department of Paediatrics, CHUV, 1011 Lausanne, Switzerland. E-mail: thierry.deonna@chuv.hospvd.ch

A male presenting with benign partial epilepsy with rolandic spikes from the age of 7 years was evaluated at age 11 years for worsening of his epilepsy associated with a specific regression of graphomotor skills. A longitudinal study over nearly 2 years showed an improvement in handwriting to an almost normal level under modified antiepileptic therapy. A detailed analysis with a computer-monitored graphics table showed at first a rapid improvement of skills followed by protracted slower progress. We argue that the initial rapid recovery of skills was directly linked to the improvement of his epilepsy. The slower late acquisition of motor programmes that had never been fully established was due to longstanding interference by his epilepsy. The specificity of the deficit within the graphomotor system and its possible neurobiological basis are also discussed. The analytical method and approach used in a single patient might provide an example for other patients in whom epilepsy can interfere in the acquisition, progress, and maintenance of new skills and can be responsible for selective deficits.

Acquired cognitive deficits of epileptic origin are now increasingly recognized in children, irrespective of the presence of clinical seizures and particularly within the spectrum of idiopathic, partial, or rolandic epilepsies (Deonna et al. 2000). These epilepsies can manifest themselves as a loss or disruption of an already mastered cognitive function, as delayed acquisition, or both. Except for major syndromes such as acquired epileptic aphasia, these disorders are not easily documented, especially when the deficit is moderate and insidious in its development.

We had the opportunity to study prospectively a male whose handwriting was affected selectively by a focal cortical epileptic dysfunction and to document the extent and nature of this deficit at repeated intervals for nearly 2 years, using a computer-monitored graphics table and a comparative set of data from aged-matched, typically developing children.

We studied the specificity and the nature of the patient’s graphomotor disorder by assessing several spatial, temporal, and kinematic parameters of a writing sample, and correlated the evolution of the deficit and its dynamics of recovery with electroencephalogram (EEG) abnormalities and antiepileptic treatment. We discuss how partial epilepsy starting in the early learning phase of a specific skill, in this case handwriting, can interfere with the development and consolidation of these skills.

The patient is a French-speaking, right-handed male who presented with right-sided clonic motor seizures affecting the hand at the age of 7 years. An EEG showed typical focal sharp waves in the rolandic areas, left more than right; a diagnosis of benign partial epilepsy with rolandic spikes (BPERS) was made.

In subsequent years the child had several seizure recurrences and the epileptic discharges on the EEG remained very frequent, leading to some changes in antiepileptic medication.

His history revealed normal development, including oral and written language. His parents reported no difficulty in learning to use and handle a pencil nor in the acquisition of letter tracing in the first years of school. But they observed progressive handwriting difficulties as soon as the epilepsy started at the age of 7 years (Fig. 1).

The child was referred to our hospital at the age of 11 years when his epilepsy worsened. He and his parents mentioned occasional uncontrolled sudden jerks of the right hand, writing difficulties (slow and painful), and difficulties in keeping balance. At that time he was treated with carbamazepine and sodium valproate. The neurological examination was normal. There was no asymmetry of deep tendon reflexes, no clonus, no dystonia, no difficulty with rapid arm and finger movements and no writer’s cramp nor sensory abnormality. Specifically, there was no loss of tone while keeping arms outstretched (negative myoclonus).

EEG showed very frequent epileptic asynchronous biphasic discharges in the left and right hemispheres, more pronounced than in previous tracings, with intermittent generalized spike waves during sleep. Cerebral magnetic resonance imaging was normal.

The neuropsychological assessment at that time (11 years; Table I) revealed normal oral language skills (Verbal IQ=94, clinical observation of speech and language), with a lower non-verbal quotient (Performance IQ=75). This lower result in the performance scale was due to speed constraints. There was no visuo-attentional deficit; there were good visuo-motor
and visuo-spatial skills. However, there were severe handwriting deficits (computer-monitored graphics table) associated with spelling errors.

Carbamazepine was stopped and the child was maintained on sodium valproate. The occasional jerking of the hand disappeared rapidly. The child was generally much more alert, and his handwriting improved within 3 months. A rapid recovery of adequate sound-to-print conversions was noticed in spelling. A few months later, sodium valproate was replaced by sulthiame because of persistent measured abnormalities in handwriting and epileptic EEG discharges. Repeated handwriting examinations over the following 10 months showed a marked improvement but performances on several measures remained much below that of control children of the same age. Subsequently, normalization of handwriting was nearly complete about 2 years after the regression.

Method
To evaluate precisely the nature and the severity of the writing deficit and not only the final product, we used a model on a computer-monitored graphics table that has been standardized using typically developing children (Zesiger 1995).

Graphomotor study

Task, apparatus, and procedure
The graphomotor task consisted of writing the same word (‘dame’, i.e. lady) with variations of size and speed under six conditions: normal (spontaneous handwriting), large (twice

Table I: Results of first neuropsychological assessment at age 11 years (T1), time of maximal handwriting deficit

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Assessment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Behaviour</td>
<td>Slow, passive, easily tired</td>
</tr>
<tr>
<td>WISC III</td>
<td>FSIQ 83 (mean 100), VIQ 96, PIQ 75(^*)</td>
</tr>
<tr>
<td>Written language</td>
<td></td>
</tr>
<tr>
<td>Reading comprehension test (LOBROT L3)</td>
<td>Slow but correct</td>
</tr>
<tr>
<td>Spelling test (LOBROT O3)</td>
<td>Many sound-to-print conversion errors (4th quartile)</td>
</tr>
<tr>
<td>Visual gnosis skills (Test of Visual Perceptual Skills)</td>
<td>Perceptual quotient 125 (mean 100)</td>
</tr>
<tr>
<td>Constructive praxic abilities</td>
<td></td>
</tr>
<tr>
<td>Kohs cubes (WISC III)</td>
<td>Standard score 10 (mean 10)</td>
</tr>
<tr>
<td>Rey Complex Figure Test (copy)</td>
<td>50th centile</td>
</tr>
<tr>
<td>Memory</td>
<td></td>
</tr>
<tr>
<td>Digit Span</td>
<td>Span 4, standard score 6 (mean 10)</td>
</tr>
<tr>
<td>Rey Complex Figure Test (immediate recall)</td>
<td>90th centile</td>
</tr>
<tr>
<td>Attention (Conners’ Continuous Performance test)</td>
<td>No sign of attention difficulties (global index 0), despite slow reaction times (centile 1.4)</td>
</tr>
</tbody>
</table>

WISC III, Wechsler Intelligence Scale for Children, French adaptation, (Wechsler 1996); FSIQ, Full-scale IQ; VIQ, Verbal IQ; PIQ, Performance IQ; Rey Complex Figure Test (Rey 1959); Test of Visual–Perceptual Skills (Gardner 1982); Conners’ Continuous Performance Test (Connors 1995); LOBROT, Batterie d’Épreuves Pour Mesurer la Lecture et l’Orthographe (Lobrot 1967); Digit Span, from WISC III.

\(^*\)PIQ 111 when controlled 10 months later (acceleration of processing speed, allowing in particular for normalization of subtest ‘coding’).

Figure 1: Evolution of epilepsy and handwriting skills. Broken line showing onset and course of handwriting problems is based on retrospective anamnestic data. Solid line starting with prospective quantitative study at successive periods (T1 to T5) indicates dynamics of improvement.
the usual size), as small as possible, normal, as fast as possible, and slow (half usual speed). This procedure is frequently used to evaluate a participant’s ability to control handwriting size and tempo (Van Galen 1991, Teulings and Schomaker 1993). Six trials per condition were elicited, with a total of 36 attempts. The child wrote on a normal sheet of white paper lying on the graphics table.

The writing tests were recorded by means of a Wacom UD (Wacom, Saitama, Japan) series graphics table (sampling rate 200Hz) monitored by a PC running MS-DOS. The writing device was a wireless, special ballpen (Wacom UP-401) with a built-in pressure-sensing device.

The child was allowed to practice a few times before being tested. He was instructed to start writing on hearing an auditory signal that indicated the beginning of data acquisition.

The handwriting skills were assessed five times over nearly 2 years: at 11 years (T1), 11 years 3 months (T2), 11 years 8 months (T3), 11 years 10 months (T4), and 12 years 10 months (T5) guided by parental reports of the evolution of the condition, changes of medication, and EEG controls (Fig. 1).

**Data analysis**

A computer program was used to display on a monitor the trajectory and absolute velocity pattern of each attempt. Segment boundaries were determined by searching for the minima in the velocity pattern of the recorded writing movement that coincided with consecutive up and down strokes. The first and last segments were excluded because of the high variability known to characterize these strokes. Therefore, 15 strokes (out of 15 for the word ‘dame’) were used for analysis.

Several temporal, spatial, and kinematic measures were computed on each stroke. They included: (1) dysfluency (discontinuity of movement measured by the number of velocity extremes per stroke); (2) duration (seconds), trajectory length (centimetres); and (3) mean and maximal velocities (centimetres per second).

Results were compared with those for a control group of typically developing 11- and 12-year-old children (12 per age group; Zesiger 1995) and expressed as z-scores (mean=0, a standard deviation [SD] of 1 was considered normal).

**EEG STUDY**

The EEG (recorded with 21 electrodes) included waking and sleep (nap) records. Four EEGs were performed at the hospital at the same time as the assessments of handwriting skills (SD 1 month), i.e. at T1, T2, T3, and T5. The five previous tracings (waking only) made by the referring physician from the age of 8 years were also reviewed. The location and density of discharges and possible clinical EEG correlates were noted. A score ranging from 0 to 4 was given to indicate the frequency of the focal and generalized epileptiform activities (0, no discharge; 1, occasional discharges; 2, intermittent discharges; 3, very frequent discharges; 4, subcontinuous discharges).

**Results**

**FIRST ASSESSMENT OF HANDWRITING SKILLS (T1)**

At 11 years the child showed severe alterations of handwriting skills: calligraphy was very poor with large, irregular, and dented letters (Fig. 2).

During dictation he wrote very slowly (4 words per minute). Sound-to-print conversion rules were not always applied (e.g. casserole→caslole; escargot→scagot).

Analysis of the handwriting samples showed deficits for nearly all measures: severe dysfluency, increased duration of production (pauses), and increased trajectory length (Table II). Although the mean velocity seemed to be within the normal range, the presence of high-velocity peaks (maximal velocity) indicated that strokes were produced with sudden, jerky movements. All these phenomena were observed throughout the six conditions.

**EVOLUTION OF GRAPHIC SKILLS**

Three months after the first assessment (T2), the child’s graphic skills showed a marked improvement: trajectory length and maximal velocity had normalized. However, despite the marked decrease in dysfluency and duration, graphic skills remained clearly pathological (Table II). Sound-to-print conversion rules were correctly applied.

The pattern of evolution during the following year (11 years 3 months to 11 years 10 months) showed specific, persistent difficulties for fluency and duration that normalized at 12 years 10 months (scores within 1SD of the mean for 12-year-old children; Fig. 3). The dynamics of evolution of these two measures, which are related to the degree of mastery of handwriting, showed a rapid initial improvement followed by much slower progress in the next year (Fig. 3).

Within this 1-year follow-up, the child passed from performances inferior to the mean for 8-year-olds to normal scores for 11- to 12-year-old children. The improvement was particularly marked in the spontaneous writing condition. A slower rhythm of improvement was noted in the conditions ‘small/large’ and ‘fast’ writing, in which the child obtained results comparable to those of, respectively, 10- and 9-year-old control children at the age of 12 years 10 months.

**Figure 2:** Samples of patient’s handwriting (dictation) at T1 (11 years 0 months old) and T5 (12 years 10 months old).
Paroxysmal EEG discharges were frequent for at least 2 to 3 years before the study, with bilateral asymmetric focal sharp waves predominant in the left centro-temporal region, but with variations from tracing to tracing. During the worst period, discharges still increased with marked diffusion, especially during sleep, but there were no continuous spike waves during sleep. After the withdrawal of carbamazepine the epileptic activity decreased significantly and decreased even more while the child was under sulthiame (Table III).

Discussion
The acquired handwriting deficit in this child was limited to that function and could not be explained neither by praxic, visuo-spatial and attentional problems nor by an associated hemiparesis, a sensory deficit, or a dystonia (writer’s cramp; Guerrini et al. 1999).

Evolution of the deficit was closely correlated with the epileptic activity. The physiopathology is probably the same as that seen in other types of prolonged neurological deficit occasionally seen in BPERS (such as oromotor function), which are somehow due to focal inhibition (as observed in the phenomenon of negative myoclonus; De Saint-Martin 1999). The first graphomotor difficulties were reported at the onset of seizures. A rapid deterioration in handwriting skills occurred 3 years later at a time of worsening of the epilepsy, probably precipitated by the use of carbamazepine. We think that epileptic activity interfered with the normal course of handwriting acquisition and was responsible first for a slowing of learning processes, and then for the regression of graphomotor skills. Recovery was initially fast (between 11 years and 11 years 3 months) but incomplete. Continuous progress at a slower pace was observed during the 1 year 6 months follow-up, with improvement in the epilepsy. This double-step evolution suggests that when the temporary aggravation of his epilepsy was stopped, the child initially recovered the handwriting skills that had previously been acquired, and then more slowly pursued further mastery of this function. The motor programming abilities were probably not normally developed at the time of the epileptic deterioration. This is consistent with the parents’ report that handwriting difficulties appeared with the onset of epilepsy at 7 years of age. From that time onwards the epilepsy probably interfered with further acquisition of handwriting skills and, more specifically, prevented or impeded the development of motor programmes. The slow progress observed over 1 year, when epileptic activity on the EEG had markedly decreased, might be explained by an acquisition of new skills (but delayed in time) that had never been mastered rather than a slow recovery of previously acquired functions.

The reason why apparent bilateral and/or relatively widespread distribution of the epileptic disturbance provokes such a specific fine-motor programming deficit is difficult to understand. We think it possible that a very discrete cortical dysfunction involving networks involved in the handwriting function occurred in this patient with otherwise typical BPERS. Owing to the variability of the main location(s) of epileptic foci in BPERS (Pan and Lüders 2000), it is clear that different neural

Table III: EEG results and antiepileptic drugs

<table>
<thead>
<tr>
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<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Spike focus (waking)(^a)</td>
<td>R&gt;L</td>
<td>R&gt;L</td>
<td>R&gt;L</td>
<td>L&gt;R</td>
<td>L&gt;R</td>
<td>L&gt;R</td>
<td>L&gt;Rb</td>
<td>ND</td>
<td></td>
<td>ND</td>
</tr>
<tr>
<td>Right</td>
<td>4</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td></td>
<td>0</td>
</tr>
<tr>
<td>Left</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Generalized</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td></td>
<td>0</td>
</tr>
<tr>
<td>Sleep EEG</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>↑ F &amp; G spikes.</td>
<td>F &amp; G</td>
<td>F=1, G=1</td>
<td>ND</td>
</tr>
<tr>
<td>Antiepileptic drug (mg)</td>
<td>No</td>
<td>CBZ</td>
<td>VPA</td>
<td>VPA CBZ (400)</td>
<td>VPA CBZ (400)</td>
<td>VPA</td>
<td>Sulthiame</td>
<td>Sulthiame</td>
<td>Sulthiame</td>
<td></td>
</tr>
</tbody>
</table>

R, right; L, left; ND, not done; F, focal; G, generalized; CSWS, continuous spike waves during sleep; CBZ, carbamazepine; VPA, valproate.

\(^a\)Focal sharp waves in centro-temporal and/or fronto-central location with variable extension. Scoring: 4, subcontinuous spikes; 3, very frequent spikes; 2, frequent spikes; 1, rare residual spikes; 0, no spike. \(^b\)With arms outstretched, no negative myoclonus was observed.

Table II: Patient’s results in graphomotor study at T1 to T5 assessments

<table>
<thead>
<tr>
<th>Parameter</th>
<th>T1 (11:0)</th>
<th>T2 (11:3)</th>
<th>T3 (11:8)</th>
<th>T4 (11:10)</th>
<th>T5 (12:10)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dysfluency(^a)</td>
<td>8.4</td>
<td>4.7</td>
<td>3.7</td>
<td>3.1</td>
<td>1.1</td>
</tr>
<tr>
<td>Duration (s)</td>
<td>5.9</td>
<td>3.5</td>
<td>3.2</td>
<td>2.4</td>
<td>1.2</td>
</tr>
<tr>
<td>Length (cm)</td>
<td>3.8</td>
<td>1.3</td>
<td>1.0</td>
<td>1.3</td>
<td>1.0</td>
</tr>
<tr>
<td>Mean velocity (cm/s)</td>
<td>-0.9</td>
<td>-0.8</td>
<td>-0.9</td>
<td>-0.9</td>
<td>-1.5</td>
</tr>
<tr>
<td>Maximal velocity (cm/s)</td>
<td>1.8</td>
<td>0.8</td>
<td>0.6</td>
<td>0.8</td>
<td>0.9</td>
</tr>
</tbody>
</table>

\(^a\)Dysfluency is measured by number of velocity extremes per stroke. Values are z scores, indicating standard deviation from mean.
networks can be altered leading to various selective functional deficits. For instance, Morrell (1995) showed in an electrophysiological study of Landau–Kleffner syndrome (viewed by many as a severe variant of BPERS) that the source of the epileptogenic activity was unilateral and extremely localized in the auditory cortex, despite frequent bilateral discharges (continuous spike-waves on the EEG during sleep).

During the worst period, in addition to the massive deterioration in the child’s handwriting skills, there was a slight impairment of the orthographic programmes, as shown by the presence of errors that violated sound-to-print conversion rules (Ellis 1982). Very quickly, however, the deficits apparently became restricted to motor programming (a highly increased duration of production and severe dysfluency) and parameter setting levels (parameterization=control of size and tempo; Van Galen 1991), coinciding with improvement in the EEG. Motor programming remained altered to a greater extent than the other parameters of handwriting skills. Thus the nature of the handwriting deficit seemed to correspond to an atypical pattern of handwriting skills rather than normal but immature development (Meulenbroek and Van Galen 1989, Mojet 1991). All deficits resolved at nearly 13 years of age. Interestingly, however, the analysis of fluency in one condition of speed variation (fast writing) and in one of size variation (small writing) did not show a completely normal pattern. It seems that either the parameter setting level remained the most difficult process for the child to master, or the late acquisition of motor programmes was not yet firmly established and required additional (sensory) control when constraints were introduced.

In this case epilepsy probably affected the constitution of an internal representation of movement (Rijntjes et al. 1999). The very localized epileptic neuronal dysfunction in the rolandic area interfered with the normal consolidation of the high-level motor memory (Gandolfo et al. 2000) or disturbed the normal shift from acquisition in the prefrontal region to established motor representation in the posterior part of the parietal cortex (Shadmer and Holcomb 1997). After an initial rapid but only partial recovery, the child showed a much more gradual improvement. This was because of inadequate or insufficient learning of a skill whose consolidation depended on a network that had long been involved in the epileptic disorder.

The computerized measures on a graphics table enabled us to evaluate different levels of the writing act, and its disruption and recovery, which is rarely possible in these circumstances. Other ‘cognitive’ deficits, now increasingly described in children with BPERS, can be thought about in the same terms (Deonna et al. 2000). Depending on the type of function involved in the epileptic process and the degree of mastery of the function when epilepsy becomes active (i.e. the age at onset and severity of epilepsy in relation to the normal developmental tempo of this function), a very different impact, degree of recovery, and rapidity of change can be expected.

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References

**Figure 3:** Evolution of dysfluency and duration in child’s handwriting, as a function of age.


Rey A. (1959) *Rey Complex Figure Test*. Paris: Éditions du Centre de Psychologie Appliquée.


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**BPNA**

British Paediatric Neurology Association XXX Annual Conference

The 30th Annual Conference of the British Paediatric Neurology Association will be held in the city of Sheffield from Friday, 23 January until Sunday, 25 January 2004. The venue for presentations is the excellently equipped Sheffield Hallam University, located in the centre of the city. The Gala Dinner will be held in the beautiful ballroom of the Royal Victoria Holiday Inn.

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*The genetic basis of neuronal migration: lissencephaly, cryptogenic infantile spasms and mental retardation with epilepsy*

Keynote guest lectures will cover a wide range of subjects:

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Professor Paul Griffiths, Academic Professor of Neuroradiology, University of Sheffield

*The Zebrafish: A new model organism for human disease*

Professor Phil Ingham, Professor of Developmental Genetics, University of Sheffield

*What radiosurgery (gamma knife) can offer children*

Mr Andras Kemeny, Consultant Neurosurgeon, Sheffield

*Can we diagnose that a baby has been shaken?*

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