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Benign partial epilepsy of childhood: a longitudinal neuropsychological and EEG study of cognitive function

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The study combined prospective neuropsychological and EEG results of 22 children presenting with typical benign partial epilepsy with rolandic spikes (*n=19*) and occipital spikes (*n=3*). The aims were to assess the types of cognitive problems which may be encountered in this population, to evaluate the course of cognitive and learning capacities during the active phase of epilepsy, and to see if there was a correlation with paroxysmal activity on the EEG. Average age at entry in the study was 8.4 years and each child was seen two to four times over a period of 1 to 3 years. EEGs showed persistent spike foci in most cases that worsened in three cases, but there were no continuous spike–waves during sleep. No child had persistent stagnation, marked fluctuations, or a regression in cognitive abilities. Of 22 children, 21 had average IQ (>80). Eight children had school difficulties requiring special adjustment. No single cognitive profile was identified. Four children had delayed language development and eight children had transient weak scores in one isolated domain (verbal ,visuospatial, memory) which improved or normalized during the course of the study with concomitant EEG improvement or normalization. In two of the three children with aggravation of the paroxysmal EEG activity, clinical changes were documented. A proportion of children with typical benign partial epilepsy with rolandic spikes showed mild, varied, and transient cognitive difficulties during the course of their epilepsy, and in most cases this probably had a direct relation with the paroxysmal EEG activity.

In recent years it has been increasingly realized that a significant proportion of children with benign partial epilepsy with rolandic spikes (BPERS) and variants (benign partial epilepsy with occipital spikes: BPEOS) often have associated learning problems which cannot be simply attributed to a psychological reaction to the disease, as originally believed (Beaussart 1972).

There have been a few neuropsychological studies on children with benign partial epilepsy, and those that have been published show very different areas of study and experimental methodologies (see Table I). There is some consensus that these children as a group do not do as well as children without the condition, but the type and severity of reported cognitive and behaviour problems do not appear uniform with either minor transient problems or more severe specific disabilities. The rare detailed studies on this latter aspect have given contradictory results; one study emphasized specific language problems while the other found normal language but problems with visual perception (Heijbel and Bohman1975; Piccirilli et al. 1988, 1994; d'Alessandro et al. 1990; Weglage et al. 1997; Staden et al. 1998).

Independent of the nature of the problems, the major question now is whether cognitive impairment is due to the basic brain dysfunction which caused the epilepsy or if these problems are related to the focal or multifocal paroxysmal activity, which is characteristically prolonged over the years, and often quite intense, even when not accompanied by clinically recognized seizures. The idea that focal epileptic discharges can be of importance independently of clinical seizures stems from two lines of observation. First, transient cognitive dysfunction during focal EEG discharges, as opposed to periods free of discharges, can be documented in some cases during continuous performance tests (Binnie et al. 1992, Pressler 1997). Second, and perhaps more importantly, several cases of BPERS with transient oromotor, speech, language, or cognitive deficits occurring during a very active stage of the syndrome have been reported (sometimes with so-called continuous spike–waves during sleep), suggesting a strong connection between cognitive function and epilepsy (Deonna et al. 1993, Prats et al. 1998).

The question arises whether the cognitive and behavioural problems often encountered in BPERS and BPEOS are in effect minor manifestations of the epileptic dysfunction and if so, whether treatment is justified and efficient. The finding that children with BPERS as a group do not do as well cognitively, behaviourally, and scholastically as a control group of typically developing individuals does not prove the direct role of epilepsy, because the differences are often subtle and can have several explanations. Only prospective studies, in which the child provides their own control data, could perhaps show if a weak domain of function, a stagnation or regression of function, or a learning failure correlates with the activity of the epilepsy as documented by clinical evaluation and EEG follow-up (Weglage et al. 1997).

The problem is all the more important because a consensus is now emerging that most cases of BPERS (the most frequent form of idiopathic epilepsy of childhood, do not require antiepileptic medication for the seizure disorder itself because of it is relatively benign and has a good prognosis.

This study aimed to investigate the range and type of the

cognitive difficulties in an unselected population presenting with BPERS.

No first assumptions were made as to any specific deficit and each child was studied with an extensive battery of common tests (see Appendix). A more focussed evaluation was made in cases where a specific deficit emerged in the course of the investigation or was identified in the child's history. It was postulated that, although BPERS is characterized by an epileptic focus (or independent foci) predominantly in the sylvian region, there would be variations in its exact localization, degree of local extension, and contralateral spread which might lead to varying levels of cognitive dysfunction, with involvement of different cognitive domains.

This study also aimed to investigate the course of cognitive development and learning capacities during the course of the epileptic syndrome. This part of the study aimed to see whether some children stagnate, regress, or fluctuate during the first years of the condition; to observe the impact of epilepsy on school performance; and to see whether there is any relation between school performance and activity of the epilepsy as judged by the paroxysmal spike–wave activity on the electroencephalogram.

Method

PARTICIPANTS

From 1992 to 1997, 22 children with typical clinical and EEG features of BPERS (Aicardi 1994) or BPEOS (Panayotopoulos 1989) were enrolled in a prospective study including clinical evaluation, a detailed neuropsychological evaluation (see Appendix), and waking and sleep EEGs. The basis for their recruitment was the epilepsy syndrome diagnosis. Participants were not included if they had been seen first because of cognitive or behavioural problems. The Pediatric Neurology Clinic and EEG Department at the Centre Hospitalier Universitaire Vaudois is the referral center for most childhood epilepsy cases in the Canton de Vaud, Switzerland so there was no special selection bias. In some children, epilepsy had started some time before inclusion in the study; this is indicated in Table II.

Developmental history was taken with special emphasis on acquired cognitive and behaviour problems and on alternative explanations for observed problems other than the epilepsy.

One child had iris coloboma and had experienced intrauterine growth retardation. She had otherwise typical BPERS and so was included.

In cases where antiepileptic treatment was given, this was according to clinical judgment, either because of frequent seizures, or behavioural or cognitive problems suspected to be related to the discharges as observed on the EEG. We elected to use sulthiame which has been found to be beneficial in open studies of benign partial epilepsy, and also in suppressing EEG discharges (Gross-Selbeck 1995). Nine children were already on treatment when enrolled in the study and this was not changed initially.

Table I: Published neuropsychological studies on BPERS[a]

Author (y)	*Areas studied*	*Nr of children (age)*	*Control data*	*Longitudinal study*	*Tests used*	*Conclusions*
Heijbel & Bohman(1975)	Intelligence, behaviour, school adjustment	16 (7–12y)	Yes	No	IQ, Bender behaviour scale	Patients worse than controls for visuomotor coordination
Piccirilli et al. (1988)	Language lateralization	22 (9–13y)[b]	Comparison left- and right-side EEG focus	No	Verbomanual concurrence task	Atypical cerebral organization
D'Alessandro et al. (1990)	Neuropsychological abilities in relation to side/bilateral of EEG focus	44 (9–13y)[b]	Yes	Partial (11/14)	IQ, attentional tasks, language, visuomotor	No IQ difference from controls, slight problems in some subtests, no specific deficits, no difference from controls 4 years later (11/44)
Piccirilli et al. (1994)	Attention problems in relation to focus side	43 (9–13y)[b]	Yes	No	Figure cancellation task	Patients with right and bilateral EEG foci worse on attention (left focus was same as controls)
Weglage et al. (1997)	Neuopsychological and behaviour characteristics in children with and without seizures (EEG focus only)	40 (6–12y)	Yes	No	IQ, psycholinguistic tests, motor performance, behaviour check-list	Children impaired in IQ, visuomotor, short-term memory positive correlation with frequency of spikes, not with side or presence of seizures
Staden et al. (1998)	Search for specific language dysfunction	20 (6–13y)	No	No	IQ, 12 standardized language tests (oral)	13/20 children with two or more low performances

[a] There have been many studies on this topic which have been reported in congress abstracts but to our knowledge not subsequently published.
[b] Probably same children are included in these studies.

PROCEDURE

Initial neuropsychological, developmental and school evaluation

Initially, each child was administered a complete neuropsychological examination that included: (1) an age-adequate intelligence scale, (2) tests assessing various aspects of oral and written language function, (3) visuospatial processing tests, (4) working memory and long-term memory tests, (5) attention, and (6) executive function assessments (see Appendix for specific tests used).

This extensive initial evaluation also served as a basis for comparison in case of deterioration over the course of the

Table II: Clinical data on 22 children

Participant Nr	*Age at diagnosis of epilepsy (y)*	*Age at study entry (and follow-up)*	*AED at 1st neuropsychological and follow-ups*	*Major neuropsychological results VIQ and PIQ*	*Course school/help*	*Comment*	*See table*
A1	4.7	6.2 (7.2, 8)	CBZ/VPA/S	94(89, 101)	Normal	–	
A2	8.3	10.6	CBZ/CBZ	114(114, 112)	Normal superior	Refused repeat tests	
A3	6.7	6.9	No/no	132 (123, 132)	Normal	No formal repeat tests (1y)	
A4	8.2	8.8 (10.1)	No/no	117(110, 120)	Normal	Immature graphomotor skills	
A5	8.4	8.7 (9.9)	CBZ/no	96(89, 104)	Normal	–	
A6	7.2	7.3 (10.5)	No/no	107(104, 109)	Mainstream school, repeated grade	3 years between 1st and 2nd testing	
A7	6.7	7.8 (9.2)	No/no	Slow speech 122(126, 110)	Speech slowing excellent student	[a]	
A8	6.2	7.1 (7.6, 8.2, 9.2)	No/S	89(93, 85)	Mainstream school, slow learning to read	[a]	
A9	4.4	4.7 (5.4, 5.9)	No/S	73(69, 79)[b]	Special class	[a]	
B10	9.4	11.6 (12.4)	CBZ/CBZ	Some weak areas 95(93, 98)	Normal	–	III
B11	9.5	11.2 (12.2)	CBZ/CBZ	Weak lexicon 101(93, 109)	Normal	–	III
B12	4.5	6.7 (7.3, 8)	CBZ/S	Weak visuospatial 84(87, 94)	Mainstream school psychotherapy	–	III
B13	9.2	9.5 (10.5)	No/S	92(89, 97)	Repeats grade special help	–	III
B14	9.8	9.9 (10.9)	No/no	110(111, 107)	Normal	Transient attention/ memory problems	III
B15	5	5.2 (6)	No/no	Weak visuoconstr. 90(105, 78)	Normal	–	III
B16	6	9.7 (10.7)	No/S	95(100, 91)	Mainstream school repeats grade	–	III
B17	9.3	10.7 (11.3, 11.9, 13)	No/S	Dysorthography 119(115, 120)	Recover spelling	See case report	III
C18	2	5.7 (6.8, 7.9)	VPA/CLB+S	DLD; PIQ 88	Mainstream school speech therapy	Early onset R hemifacial seizures	IV
C19	4.7	8.8 (9.7, 10.7)	VPA/S	DLD severe 79(73, 91)	Special school global learning problem	–	IV
C20	4.7	6.6 (7.7, 8)	CBZ/S	DLD 97(83, 114)	Mainstream school speech therapy	–	IV
C21	4.8	6.5 (7.3, 7.9)	No/no	DLD 86(88, 87)	Mainstream school speech therapy	–	IV
D22	11	11.5 (12, 13)	No/S	Borderline normal 71(74, 73)	Special school significant progress	Iris coloboma IUGR	

A Normal neuropsychological findings: participants 1–19 (participants 7–9; transient worsening of EEG).
B Transient weak results: participants 10–17.
C Developmental dysphasia: participants 18–21.
D In the borderline range: participant 22.
[a] Worsened EEG, see discussion results.
[b] Difficult cooperation. Does not reflect potential.
AED, anitepileptic drugs; CBZ, carbamazepine; CLB, clobazam; DLD, delayed language development; S, sulthiame; VPA, sodium valproate.

study. In addition, we pursued a more detailed assessment of deficient domains which we thought might fluctuate in the natural course of the syndrome or in relation to treatment.

Developmental history was taken with special attention to possible cognitive stagnation or regression or special behaviour problems which had occurred at any time before the diagnosis of epilepsy. Information about the child's school curriculum and possible special aids (speech and language therapy, psychotherapy, school support, etc.) was supplied during or after school. Whenever the child presented difficulties in school or was receiving special care, further information was collected from the special needs teacher or from the therapist.

Although no behavioural questionnaire or in-depth psychological interview was administered, we paid close attention to possible emotional or behavioural problems present at initial encounter or during clinical follow-ups and pursued these issues clinically if we suspected a significant psychological problem.

Examination at follow-up

Each child was regularly reexamined (every 6 or 12 months, sometimes more frequently when justified by the child's state) for a duration of 1 to 3 years in order to evaluate their cognitive and language development. A parental interview was again carried out to document the child's progress. Special attention was given to main school achievements that were supposed to be mastered during the study period (reading, spelling, and arithmetics) and contact was made with the teachers and therapists when needed.

Neuropsychological examination performed on these occasions varied from child to child; unless parents (teachers, therapists) mentioned the emergence of new concerns or difficulties, the domains that were weak or deficient at the initial assessment were focussed on.

ELECTROENCEPHALOGRAPHIC STUDY

Prospective EEG records were standardized (registration with 21 electrodes using the 10–to–20 international system) with both waking and sleep EEGs (after sleep deprivation or

Table III: EEG data on 22 children

Participant Nr	*Nr of EEGs (sleep EEGs)*	*Localized initial focus*	*Worsening of EEG*	*Persistence of focus (2 or more records)*	*Improvement*	*Normalization spontaneous or under treatment*
1[a]	6(6)	LO and RO	–	+ (4)	Disappearence of 2nd focus on S	–
2	3(2)	RCT	–	+ (2)	–	+on CBZ
3	1(0)	RCT	One tracing only			
4	3(3)	RCP	–	+ (3)	↓Waking state	–
5	2(2)	RCT and LCT	–	+ (2)	Disappeared 2nd focus	–
6[b]	2(2)	RCT	–	–	–	5y later spontaneous
7	2(2)	RCT	Appearance 2nd focus LCT	+ (2)	–	–
8	5(4)	RCT	↑Spikes 2nd–5th	+ (4)	–	–
9	3(3)	RC and LC	Appearance 3rd focus LTC	+ (3)	↓ on S	–
10	4(3)	RCT	–	+ (2)	Change of side of focus, never bilateral	Disappeared in wake state
11	3(3)	RCT	–	+ (2)	–	–
12[a]	6(6)	RO	–	+ (3)	Disappeared in 1/6 EEGs	–
13	3(3)	LCT	–	–	–	On S
14	2(2)	LCT	–	+ (2)	↓Spike frequency	–
15[a]	2(2)	LO and RO; LCT	–	+ (2)	Disappeared RO focus	–
16	3(3)	LCP and RP	–	+ (2)	_	On S
17	5(5)	LCT	–	+ (3)	↓ on S	–
18[c]	8(8)	LCT	–	+ (4)	–	On CLB and S
19	8(6)	LCP and RFC	–	+ (3)		On S
20	5(5)	RCT	–	+ (2)	↓ on CBZ	On S
21	3(3)	RCT and LCT; RO	–	+ (3)	Disappeared 3rd focus	–
22	4(1)	LCT	–	+ (2)	–	On S

[a] Benign partial epilepsy with occipital spikes (BPEOS).
[b] Almost continuous unilateral spike–waves during sleep.
[c] Electroclinical right hemifacial seizures occurred during left centrotemporal discharges in one of eight EEGs.
CBZ, carbamazepine; CLB, clobazam; S, sulthiame; R, right, L, left; CT, centrotemporal; CP, centroparietal, FC, frontocentral; O, occipital.
↑, increase in spike frequency (worsening); ↓, decrease in spike frequency (improvement); –, no; +, yes.

mild sedation), and synchronized video-recording. The EEGs were done at the beginning of the study at the time of the neuropsychological examination (±1 to 3 months) and repeated every 6 to 12 months at the same time as the repeated tests, for a duration from 1 to 3 years. Focal epileptic activity (single or multiple) with positive sharp waves was identified and the intensity of the focus was estimated visually in four grades (absent, rare, frequent, and continuous) both during waking and sleep. The reactivity of the epileptic activity was tested by sensory stimulation. Electroclinical manifestations were looked for. Change in location of the focus (foci) were noted. Some children had already had one or several EEGs before inclusion in the study and these results were also integrated in the prospective study.

The EEG was described as 'worsened' if a second or third EEG focus appeared or if the spike frequency increased.

Results

GENERAL DATA

Table II and Table III give an overview of the clinical, neuropsychological, and EEG findings of each individual participant.

Twenty-two children were studied (19 with BPERS and three with BPEOS). The average age at onset of epilepsy was 7 years (range 2 to 11 years) and average age at entry into the study was 8.4 years (range 4.5 to 11.6 years). Eight participants had no therapy throughout the study period, eight were on medication at entry into the study, and six were started on treatment at some point during the course of the study. The neuropsychological and EEG examinations were repeated from two to four or more times on each child depending on individual situation with two exceptions: one participant (participant 9) refused repeat testing and one (participant 22) was entered too late into the study. One child (participant 6) was seen twice, 5 years apart.

History of cognitive development, behaviour, and emotional problems

In two children, parents mentioned some problems in the first year after onset of the epilepsy. In the first child (participant 17) parents thought that the child had more difficulties expressing himself verbally the year after his first seizure. He was found to have unexpected difficulties with written language and had been studied in detail under therapy with sulthiame. In the second child (participant 16), a period of regression in behaviour and school performance was mentioned only after the onset of her epilepsy 3 years before inclusion in the study. Although no special questionnaire was used to evaluate behavioural and emotional problems, aspects of this dimension were noted. No child was found to have a major psychopathology. None of the children had a history and behaviour suggestive of typical attention-deficit–hyperactivity disorder or had been evaluated or diagnosed as such before or during the study. One participant received psychological support in relation to her parents' divorce. Some children were described as 'more nervous' before the time of diagnosis of epilepsy or during the observation period, but this was not significant.

NEUROPSYCHOLOGICAL RESULTS AND SCHOOLING

Of the 22 children, 21 had normal IQs. One had a borderline intellectual level (participant 22) and studied at a special school. She was born with intrauterine growth retardation (IUGR) and had iris coloboma. Four children had delayed language development (participants 18 to 21); all four needed school support (speech therapy). Eight children had isolated weak scores in one or several areas of neuropsychological testing which were clearly out of proportion to other results but which we did not consider a specific learning disability (participants 10 to 17, Table IV). These 'weak areas' consisted of a performance level below the 15th centile or an 18-month delay in a particular task. In two children it involved short-term visuospatial memory; in five, long-term memory (visuospatial and/or verbal); in two, visuospatial organization; and in five, some language difficulties (see Table IV and report on participant 17). Two of these eight children had to repeat a year in school. Of the nine children without any neuropsychological deficit, one had to repeat a year in school, and the other had to be put in a special class (participant 9).

COGNITIVE EVOLUTION

We did not encounter participants with regression in cognitive

Table IV: Course of eight children with transient weak results

Participant Nr	*Age at 1st testings (y)*	*Interval 1st repeated testing*	*Test domains initially affected*			*Course of paroxysmal activity*
			Memory	*Language*	*Visuospatial*	
10	11.6	10 mo	a	d		↓
11	11.2	12 mo	a	e		Persistent RCT focus
12	6.7	8 mo	b		h	Disappear (S)
13	9.5	12 mo	c	fg		Disappear (S)
14	9.9	13 mo	a			↓
15	5.2	9 mo			h	↓
16	9.7	8 mo	abc	f		Disappear (S)
17	10.7	27 mo		g		↓

a, long-term verbal memory; b, short-term visuospatial memory; c, long-term visuospatial memory; d, naming; e, lexicon; f, reading; g, spelling; h, visuospatial organization. (S) sulthiame.
↓, decrease in spike frequency (improved but not disappeared).

abilities during the study period. In all the eight children who had a definite weakness in one or several domains, there was always an improvement during follow-up period, sometimes after a period of stagnation (see Table IV).

Evolution of eight children with isolated weak results

Table IV shows only the domains which were weak at the initial testing and which were retested 8 to 27 months later in participants 10 to 17, meaning that all other results were normal in these children. The type of tasks involved in these cases allows us to rule out a test–retest effect. Most results improved or normalized during the follow-up period or some improved while others remained low in the same child. In none of these cases did the EEG actually worsen during this period.

It was evaluated whether an attentional deficit might be the only cause of poor performance for each child. In six of these eight children (participants 10, 11, 12, 13, 15, 17) we judged that attentional factors were not the cause of poor performance at the time of the weak results, as normal results on specific attention tests and good results on other tests required as much attention as tests which yeild poor performance. In two of the eight children (participants 14 and 16) attention could have been a contributing factor to poor performances; in participant 14, attention deficits may completely account for poor performance because an improvement was seen both in memory and attentional tasks. In participant 16, attention could have played a partial role because there was a slight deficit in the continuous performance test. However, visuospatial memory improved much more than verbal long-term memory suggesting that the problem was not solely due to attentional factors.

Children with delayed language development

Participants 18 to 21 had delayed language development (DLD); two with a family history of DLD and three came from a bilingual family environment but had major difficulties in both languages (see Table II). None had verbal auditory agnosia or a purely phonological disorder. They all had mixed phonological and/or lexicosyntactic problems. Progress in oral language was documented. The onset of written language acquisition which happened to occur at the time of their involvement in the study was also observed in detail. We tried to see whether a stagnation, regression, or conversely a catch-up in abilities would be observed in relation to the paroxysmal EEG activity. All children improved during the observation period but none reached normal levels. We could not demonstrate rapid improvement when the paroxysmal EEG activity disappeared (participants 19 and 20). We can neither conclude that there was a direct link between the children's language problem and epilepsy, nor rule out that epilepsy contributed to their slow and incomplete language recovery.

ELECTROENCEPHALOGRAPHIC RESULTS

EEG results can be seen in Table III. With the exception of two children (participant 3 had one waking and sleep record and participant 22 had four EEGs, only one with sleep) all had several waking and sleep records at the time of the neuropsychological evaluations (between two to eight tracings, mean 3.8 per patient). Thirteen children had a single unilateral focus throughout the study and six children showed two or more independent foci from the start of the study. No case had diffuse continuous spike–waves during sleep; one child had in a single tracing almost continuous unilateral discharges (participant 6) and on follow-up 4 years later had a normal EEG (untreated). In seven children, focal spikes were found in one or several tracings only during sleep. In 19 of 21 children, the paroxysmal EEG activity was found in at least two records. Three children had a worsening of the paroxysmal activity on successive tracings (participants 7 to 9). Only one of the children (participant 6 with 5 years between the two tracings) spontaneously normalized during the study period. In five children, the epileptic activity disappeared under treatment with sulthiame, while in three children it decreased but did not disappear.

Children with worsening paroxysmal EEG activity

Clinical EEG correlations for participants 7 to 9 are in Tables II and III. One child (participant 7) was very bright and an excellent student. Two years after his first evaluation, the EEG showed a second focus. Repeat testing showed normal performances; although he was slightly slower he progressed normally in school. His speech (which was also slow like his oromotor movements) seemed slower than before, although this could not be quantified. He could be considered as having an 'a minima' opercular syndrome (Colamaria et al. 1991). Treatment was not judged necessary.

Another child (participant 8) was found to have an increased frequency of the right focal spikes during his first 2 years of primary school. He was unexpectedly slow to learn to read, although still in the lower normal range, but this contrasted with his excellent oral language performances and metaphonological abilities. Follow-up over the next 18 months showed a total stagnation in this domain. In the third child (participant 9) who developed a third focus and had some poor results on the tests, it was difficult to make any conclusions due to factors which may have affected results such as poor cooperation, family situation, starting school, etc.

Correlation between lateralization of EEG focus and neuropsychological findings.

In the nine children without neuropsychological impairment, six had unilateral foci (all on the right side) and three had bilateral foci. In the 13 children with transient or persistent problems, nine had unilateral foci (five left and four right) and four had bilateral foci. In summary, no definite correlation could be made between type of problems and side/bilaterality of EEG foci.

PARTICIPANT 17

This participant was unusual for several reasons, so a special in-depth study of literacy skills (spelling), from 10.9 years to 12 years 11 months, was carried out. He was especially bright, had no history of language delay, and had an unexpected, isolated difficulty in spelling without other familial or personal evidence of dyslexia. These data suggest that this selective difficulty in spelling might be a manifestation of his epilepsy. This is why this child was studied in greater detail.

Participant 17 was a French-speaking, left-handed boy born in 1986. His early motor, cognitive, and language development were considered normal. No developmental abnormality was noted by the parents until he entered school, where he experienced difficulties from the first grade onwards

due to poor spelling. The parents recollect difficulty in verbal expression during that period. At age 9.3 years, he had a first nocturnal sylvian seizure. The EEG performed shortly afterwards showed a left centrotemporal focus that was increased during sleep.

The initial neuropsychological examination was performed when he came back after having a second identical seizure at age 10.7 years. He obtained high normal results of the WISC-R (FISQ 119, VIQ115, PIQ 120). His performance on Raven's matrices was excellent (>90th centile) and his visuospatial skills were good (normal high score on Rey's Complex Figure Test, see Appendix). All aspects of speech and oral-language processing were normal: speech production and discrimination, picture naming, verbal comprehension, and verbal fluency. The dichotic-listening test for words and digits revealed a performance that was almost at the ceiling level with a slight left-ear advantage for words only. This finding can be considered normal and cannot be taken to reliably indicate right-hemisphere dominance for language. The child also showed good results in phonological awareness tests from BELEC (see Appendix). Reading tests showed age-appropriate written comprehension (L3, see Appendix) and written word identification skills (MIM and REGUL from BELEC, see Appendix). Spelling was assessed by the test ORTHO3 (from BELEC) in which the subject is asked to write down, under dictation, 72 words containing different types of phonology-to-orthography mappings. Our participant's performance in this test was poor (Table V).

Given this particular profile of performance including age-adequate reading and clearly deficient spelling performance, we decided to conduct further investigations with the participant concerning the relations between reading and spelling. He was, therefore, asked to participate in an experiment which had been conducted with 144 normally developing French-speaking children aged 7 to 12 years (1st to 6th grades) and with several children presenting specific written language learning disorders. The tasks consisted of reading aloud and writing to dictation two sets of 96 matched pseudo-words and words varying in orthographic regularity. In this experiment, participant 17 again obtained normal results in reading compared to age-controlled subjects. By contrast, his results on spelling were clearly below that of the age-control children (see Table V). Participant 17 had relatively good skills in spelling regular words and pseudo-words, but had obvious difficulties in spelling irregular words. Most of his errors were phonologically plausible or regularization errors.

These results indicate that the child presented an isolated spelling deficit which was not accompanied by a reading deficit. This discrepancy between reading and spelling is at least partly compatible with the description of a surface dysgraphia, in which the alphabetic strategy (based on grapheme-to-phoneme and phoneme-to-grapheme correspondence rules) is mastered in both modalities, but the orthographic strategy (based on the memorization of specific orthographic patterns) is mastered for reading only (Frith 1985). According to Frith's model, this discrepancy is supposed to be observed in normal literacy acquisition. However, in the above mentioned experiment, large discrepancies between reading and spelling, such as the one noted in participant 17 have not been observed in normally developing children or in children with reading/spelling disabilities. Consequently, these results suggest that the specific spelling disorder experienced by participant 17 has a different origin compared to typical learning disabilities and can be viewed as an acquired deficit in which epilepsy might have played a decisive role.

Discussion

Important questions to ask are how representative our sample was of the general population of children with BPERS and BPEOS and whether the children were studied during the 'active' period of their syndrome. Our selection criteria and the results of the EEG suggest that they were indeed in the 'active' phase of their syndrome, because it was possible to record paroxysmal EEG activity in several of the tracings obtained repeatedly throughout the observation period.

We found a wide range of cognitive abilities from the superior to the borderline-normal. A significant proportion of children (nine of 22) had school difficulties requiring special adjustments, clearly more than would be expected in a population of normally developing children. We could not find a single pattern of dysfunction such as language disability, visuospatial problems, or executive dysfunction as reported in other studies (Weglage et al. 1997, Staden et al.1998). No child had a history suggestive of attention-deficit–hyperactivity disorder, although some children had poor results on some attention tasks which could have affected their school results.

Table V: Participant 17: Special study of written language (spelling) from age 10y 9mo to 12 y 11mo

	10y 9 mo (3/96)	*11y 4mo (10/96)*	*11y 10mo (4/97)*	*12y 11mo (6/98)*
ORTHO3				
Overall % of correct responses	66.3%	69.3%	66.3%	79.2%
Age equivalent	9 years	9 years	9 years	11 years
Experimental spelling test				
% of correct responses	61.5%	47.9%	64.5%	81.2%
Age equivalent	9 years	8 years	9 years	12 years
Words trained vs not trained in language therapy				
Trained				69.7%
Untrained				67.4%
EEG: left centrotemporal focus	+	+	+	0

Onset of medication (sulthiame) at age 10y 9mo, and onset of language therapy at 11y 10mo.
+, present; 0, absent.

We can exclude the role of antiepileptic therapy, because problems could be seen in children who were not on antiepileptic drugs (AED) and inversely some children on AED had normal functioning or even improved while on therapy.

In seven of the eight children who showed isolated weakness in particular areas of cognitive functioning (which normalized or improved at the 6 months to 2 years follow-up), there was a corresponding decrease or disappearance of the epileptic activity on the EEG; in no participants was an increase noted. These findings suggest that there was either a transient cognitive impairment related to focal EEG discharges at the moment of the initial tests (Binnie and Marston 1992, Pressler et al. 1997) or that another mechanism closely related to the epileptic activity was present. The case of participant 17, who compensated his severe written language problems (unanticipated in the early school years) is especially striking.

In the four children with delayed language development, we could not conclude that persistent focal discharges were affecting their progress in this domain or that their disappearance was associated with improvement, although we could not rule it out either. In the three participants where EEG actually worsened during the study, one had an increased slowness of speech, and the other was specifically slow in learning to read without the antecedant of oral-language retardation, familial reading problems, or personal early predictors of poor reading ability. These findings again suggest a direct role of the epilepsy. We did not encounter any child who had a major cognitive stagnation, regression in abilities, or marked short-term fluctuations in cognitive functioning during the follow-up period, possibly because of our selection criteria. Within the spectrum of children with 'focal sharp waves of genetic origin' (Doose 1989, p 210) those with BPERS and typical sylvian seizures may be those least at risk for cognitive impairment, because the epileptic focus affects mainly motor-cortical areas not involved in more complex cognitive processes or auditory recognition, which are so crucial for oral and written language development.

When the children first come to medical attention, it is never known for how long they may have had paroxysmal EEG discharges and if this could have interfered with their development possibly damaging, in a subtle but irreversible way, cortical networks in development. This could explain the deficits seen in some children and their persistently slow progress even after the epileptic activity has subsided. Doose and Baier's (1989) opinion that BPERS is a manifestation of a particular 'brain immaturity', which is itself also responsible for both the developmental problem and the epilepsy, can be challenged because a significant number of these children have normal to superior abilities and maintain them as seen in the current study.

Detailed longitudinal studies of individual children with rolandic epilepsy in very active stages of the disease and in whom a specific quantifiable aspect of cognitive function can be closely monitored, are still necessary to understand the role epileptic discharges play in cognitive development.

In children who are suspected of having a cognitive or learning problem with no clear explanations (especially if it has recently appeared), it seems reasonable to try antiepileptic therapy, and to repeat neuropsychological testing. Otherwise, children without cognitive problems would need treatment only when seizures are frequent or bothersome. They should, however, be followed up closely, remembering that most of them will probably outgrow the period of active epilepsy without consequences.

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Appendix I

List of tests used in the initial examination (*) and other regularly used tests throughout the study. Tests are listed by cognitive domain.

INTELLECTUAL ACHIEVEMENT

– *WISC: Wechsler Intelligence Scale for Children, French adaptation, Paris, Les Editions du Centre de Psychologie Appliquée, version–R, 1981, and version–III, 1996.
– *WPPSI-R: Wechsler Preschool and Primary Scale of Intelligence, French adaptation, Paris, Les Editions du Centre de Psychologie Appliquée, 1995.
– Raven's Progressive Matrices, French adaptation, Issy-les-Moulineaux, Editions Scientifiques et Psychologiques, 1981.

SPEECH AND LANGUAGE

– *EPD48: Epreuve de Discrimination Phonémique pour Enfants de 4 à 8 ans, Autesserre D Deltour JJ, Lacert P. Issy-les-Moulineaux, Etablissements d'Applications Psychotechniques, 1989.
– *TVAP: Test de Vocabulaire Passif et Actif pour Enfants (5–8 ans), Deltour JJ, Hupkens D. Braine-le-Château: L'Application des Techniques Modernes SPRL, 1983.
– *VOCIM: Test de Vocabulaire en Images, Légé Y, Dague P. Paris, Les Editions du Centre de Psychologie Appliquée, 1976.
– *SEVWF: Sevino's Word Fluency Test. Sevino O. Les fonctions exécutives chez l'enfant, Développement, structure et évaluation, unpublished doctoral thesis, University of Geneva, 1998.
– *EEL: Epreuves pour l'Examen du Langage, Chevrie-Muller C, Simon A-M, Decante P. Paris, Les Editions due Centre de Psychologie Appliquée, 1981.
– *BEP: Batterie d'Evaluation Psycholinguistique, Chevrie-Muller C, Simon AM, Le Normand MT, Fournier S. Paris, Les Editions du Centre de Psychologie Appliquée, 1988.
– Syllabe and word repetition: Rondal JA. L'évaluation de langage, Liège, Belgium, Mardaga, 1997.
– Epreuves testant les gnosies auditivo-phonétiques, Chevrie-Muller C, Ballan B, Simon AM, Oddos Y, Hourdin C, Housiin N. Issy-les-Moulineaux: Etablissements d'Applications Psychotechniques, 1979.
– ECOSSE: Epreuve de Compréhension Syntaxico-Sémantique, Lecocq P. Lille, France, Presses Universitaires du Septentrion, 1996.
– O-52: Epreuve d'Evaluation des Stratégies de Compréhension en Situation Orale. Khomsi A. Paris, Les Editions du Centre de Psychologie Appliquée, 1987.
– TLP: Test de Language pour Enfants de 5 à 10 ans. Caracosta H, Piterman Scoatarian S, Van Waeyenberghe M, Zivy J. Issy-les-Moulineaux: Editions Scientifiques et Psychotechniques, 1975.
– Dichotic listening: This material was developed within a collaborative research program coordinated by Claude Chevrie (INSERM Paris). The stimuli were tape-recorded at the Haskins Laboratories (National Institute of Child Health and Human Development Contract: N01 HD-5-2910, New Haven).

READING AND SPELLING

– *L3 and other subtests of the Batterie d'Epreuves pour Mesurer la Lecture et l'Orthographe. Lobrot M. Bureau d'Etudes et de Recherches de Beaumont-sur-Oise, 1967.

– BELEC: Batterie d'Evaluation du Langage Ecrit et de ses Troubles. Mousty P, Leybaert J, Alegria J, Content A, Morais J. Laboratoire de Psychologie Expérimentale, Université Libre de Bruxelles, 1994.

– Test de l'Efficience de la Lecture au Cours Préparatoire, Claire & Bruno. Giribone C, Hugon M. Paris, Les Editions du Centre de Psychologie Appliquée, 1987.

ARITHMETICS

–AME: Tests de Niveau Mathématique. Aufaure L. Issy-les-Moulineaux, Editions Scientifiques et Psychotechniques, 1972.

VISUAL AND VISUO-SPATIAL PROCESSING

– *RCFT: Rey Complex Figure Test. Rey A. Paris, Editions du Centre de Pschologie Appliquée, 1959.

– TVPS: Test of Visual-Perceptual Skills. Gardner MF. Burlingame, CA, Psychological and Educational Publications, 1982.

MEMORY

–*WPWDS: Word, Pseudo-Word and Digit Span, words from Van der Linden M, Closset A. Examen neuropsychologique de l'enfant, Liège (Belgium), undated; pseudo-word from BELEC (op.cit.) and Digit Span from the WISC-III (op.cit.).

– *CBTT: Corsi-Block Tapping Test, Adapted by Van der Linden & Closset (op.cit.).

– *RAVLT: Rey's Auditory Verbal Learning Test. Rey A. L'examen clinique en psychologie. Paris, Presses Universitaires de France, 1964.

– Rey's Visual Design Learning Test. Rey A. Epreuves mnésiques et d'apprentissage. Neuchâtel, Switzerland, Delachaux & Niestlé, 1968.

– Benton Visual Retention Test: French adaptation, Paris: Les Editions du Centre de Psychologie Appliquée, 1965.

– California Verbal Learning Test, French adaptation by Van der Linden M, Closset A. (op.cit.).

EXECUTIVE FUNCTIONS

– *HMK-ABC: Subtest 'Hand Movement' of the battery K-ABC, French adaptation. Kaufman AS, Kaufman NL. Les Editions du centre de Psychologie Appliquée, 1993.

– *TMT: Trail-Making-Test, adapted by Sevino O. (op.cit.). Further tests adapted by the same author have been used; Stroop Colour-Word Test, Verbal and Figural Fluency Tests.

ATTENTION

– *Z2B: Test des Deux Barrages. Zazzo R. Issy-les-Moulineaux: Editions Scientifiques et Psychologiques, 1992.

– *D2: Test d2 d'Attention Concentrée, French Adaptation. Bruxelles, Editest, 1966.

– *CPT: Conners' Continuous Performance Test. Toronto, Multi-Health Systems Inc. 1995.

MULTIPLE DOMAIN BATTERY

– FePsy: The Iron Psyche. Alphert WCJ, Aldenkamp AP. Heemstede, Instituut voor Epilepsiebestrijding, The Netherlands, 1995.

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