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# Cognitive functions in children with benign childhood epilepsy with centrotemporal spikes (BECTS)

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**ABSTRACT –** Benign childhood epilepsy with centrotemporal spikes (BECTS) is regarded as a benign form of epilepsy because of its usually favorable outcome, in terms of seizures. Eighteen children with BECTS were studied in terms of neuropsychological and learning abilities: intellectual quotient, oral language (phonological production, naming skills, verbal fluency and syntactic comprehension), drawing and visuo-spatial skills, visual and selective attention, verbal and visuo-spatial memory, reading, numeracy and spelling. The mean IQ of the population was within the normal range, but individual results were heterogeneous. Verbal functions and memory were normal. In contrast, drawing and visuo-spatial skills, attention and visuo-spatial memory were significantly weak compared to the normal range for age. Reading, numeracy and/or spelling ability were significantly delayed by one academic year or more in ten of the children. In conclusion, despite its benign outcome in terms of epilepsy, BECTS can be accompanied by specific cognitive disorders and low academic achievement.

**Key words:** benign childhood epilepsy with centrotemporal spikes, rolandic epilepsy, cognitive functions, learning skills, BECTS

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Benign childhood epilepsy with centrotemporal spikes (BECTS) is the most frequent form of epilepsy in children. According to the International Classi-

fication of Epilepsies and Epileptic syndromes (Commission on classification and terminology of the ILAE 1989), the sex ratio is approximately three boys to two girls. It occurs between three to 13 years in children with normal neurological and psychomotor development. Seizures are rare and most often occur during sleep. They are partial simple or complex motor seizures, involving the face and/or the arm, spreading sometimes to the ipsilateral leg and eventually secondary generalization. Oropharyngeal paresthesia is frequent. It is considered to be a "benign" epilepsy because it always disappears after adolescence (Lerman and Kivity 1975, Bouma *et al.* 1997). A familial history of epilepsy is found in 17 to 36% of cases, especially benign epilepsy with favorable outcome in adulthood (Dalla Bernardina *et al.* 2002). A genetic origin is suspected for this syndrome and linkage to a region on chromosome 15q14 has been demonstrated in some families (Neubauer *et al.* 1998). Other studies have shown a high incidence of centrotemporal spikes in non epileptic children with BECTS relatives (Heijbel *et al.* 1971, Doose and Neubauer 1997).

The absence of neuropsychological deficit has been considered a prerequisite for the diagnosis of BECTS. However, several authors have recently reported impairment of cognitive functions in children with this syndrome.

The aim of this study was to investigate neuropsychological and learning abilities in a series of children with BECTS, to determine whether or not neuropsychological deficits exist in children with BECTS, which cognitive functions are possibly affected, and their consequences on educational achievement. The study had methodological limits (18 patients only, no proper control population), as the first aim of the study was the validation of the BREV battery when compared to reference tests in epileptic children (Billard *et al.* 2002b). All types of epilepsy were then considered. After this first work, we then chose to analyse more specifically the cognitive functions in BECTS without the bias of aetiology or treatment.

## Methods

### Subjects

The study was a non-randomized, retrospective study. The subjects were selected from 202 children with epilepsy who had participated in the validation of the BREV battery and who had undergone evaluation of their neuropsychological and learning abilities (Billard *et al.* 2002b). Eighteen children with BECTS were selected from this population. The diagnosis of BECTS was made on the basis of electroclinical features before neuropsychological evaluation by neuropediatricians in different French neuropediatric centers. Diagnosis was confirmed retrospectively two years after the neuropsychological tests by the same neuropediatrician, at the time the present analysis was undertaken.

Sex, age-at-onset of epilepsy and at the time of the neuropsychological evaluation, duration of epilepsy, frequency of seizures, educational level and special educational needs were reviewed. We also looked for focus side on EEG (left, right or bilateral), whether or not a sleep EEG had been performed, if treatment had been administered, and if so, which one, and if neuroimaging had been performed.

### Methods

Verbal, performance and full intellectual quotients (VIQ, PIQ, FIQ) were evaluated using the Wechsler scale (WPPSI-R under six years and WISC-III over six years) according to French normative data (Wechsler 1995, 1996).

Supplementary verbal testing consisted of phonological production and naming skills, with two tests from the French EEL battery of Chevrie Müller (Chevrie-Müller *et al.* 1981), verbal fluency with the McCarthy scale subtest (McCarthy 1972) and syntactic comprehension with the French version of the North Syntax Screening Test (NSST) (Weil-Harpen *et al.* 1983).

Supplementary drawing and visuo-spatial abilities were evaluated by copying Rey's complex figure (Rey 1959).

Verbal memory was evaluated with the story recall subtest from the McCarthy scale (McCarthy 1972).

Visuo-spatial memory was evaluated by recall of the Rey's complex figure (Rey 1959).

Attention abilities (visual attention and selective attention) were evaluated by the BREV battery (Billard *et al.* 2002a) and by animal chequers and coding of the Wechsler scale (Wechsler 1995, 1996).

Reading and numeracy levels were evaluated with the deciphering and calculation subtests of the K-ABC battery (Kaufman and Kaufman 1993).

Spelling was evaluated with the BREV battery (Billard *et al.* 2002a).

### Statistical analysis

To evaluate intellectual ability, we calculated the mean VIQ, PIQ and FIQ for the population and compared them to the theoretical mean of 100. We also compared VIQ and PIQ according to the epileptic focus side, FIQ, VIQ and PIQ according to the presence or absence of antiepileptic drugs, and studied the relationship between sleep abnormalities on EEG and IQ. As the size of our population was too small to perform statistical tests involving the influence of focus side or sleep abnormalities seen on EEG, on IQ; only descriptive analyses were used for these items.

In order to obtain comparable results for all tests, specific verbal and non-verbal skills were evaluated according to individual scores normalized as follows: raw score/mean score for age x 10. We then calculated the mean of normalized values for the population for each specific

function and compared this score to the theoretical mean of 10.
Learning skills were expressed in terms of developmental age in months. The mean developmental age of the population for each skill was compared with the mean of chronological age. Reading and spelling were evaluated only in children aged over five years.
For spelling evaluated by the BREV battery, developmental age was determined in relation to educational age of the item that the child could spell and to the child's score for that item, each standard deviation corresponding to three months of developmental age. Thus a child who obtained a score within the mean for seven or eight years of age was allocated a developmental age of 7 years 6 months, and a child with a score -2SD was allocated a developmental age of seven years.
Learning retardation in reading, numeracy and spelling could thus be evaluated according to the difference between chronological age and developmental age. We investigated whether there was a correlation between retardation in reading, numeracy and spelling and levels of VIQ, PIQ and FIQ.
We used Student's t test for comparison between observed and theoretical means, and the paired t test to compare developmental and chronological ages for learning skills. The non-parametric Kruskal-Wallis test was used to compare mean IQs according to the use of an anti-epileptic treatment or not. Correlation between learning retardation and IQ was evaluated by the Spearman, non-parametric rank correlation test.

## Results

Clinical and electroclinical characteristics are summarized in *table 1*.
The study group comprised 18 children (11 boys, 7 girls) aged from 4 years 2 months to 8 years 10 months (mean 6 years 8 months). Age-at-onset of epileptic seizures was between three and eight years (mean 5 years 3 months). Epilepsy had been occurring for a duration of between three and 58 months at the time of the study (mean 20 months). Seizure frequency was low in all children (between zero and six seizures per annum, mean 1.94 seizures per child). Most children were being educated in classes appropriate for their age, except for three. Two children were in classes that were one year below their chronological age, and one in a class two years below. Five children were receiving speech therapy, and one was attending a multidisciplinary rehabilitation centre.
All children had had waking EEG examinations, and 10 a sleep EEG. Waking EEG demonstrated paroxysmal focal abnormalities in 16 of the 18 children tested. The other two children had previously had a focus, but it was no

**Table 1.** Clinical and paraclinical characteristics of the population

| | Age | Sex | Education level | Rehabilitation | Age at 1st seizure (years) | Duration of epilepsy (months) | Seizures/ year | EEG focus side | Sleep EEG *abnormal | Neuro imaging | Treatment |
|---|---|---|---|---|---|---|---|---|---|---|---|
| **1** | 4 y 2 m | M | KG 2 | - | 3 | 14 | 1 | None | Yes | MRI | Yes (VPA) |
| **2** | 4 y 7 m | F | KG 2 | - | 3 | 19 | 4 | None | Yes * | TDM | Yes (CBZ) |
| **3** | 4 y 10 m | M | KG 2 | - | 4 | 10 | 1 | Right | Yes * | MRI | No |
| **4** | 5 y 4 m | F | KG 2 | - | 4 | 16 | 3 | Right | Yes * | TDM | Yes (VPA) |
| **5** | 5 y 8 m | M | KG 3 | - | 4 | 20 | 4 | Bilateral | Yes * | TDM | Yes (VPA) |
| **6** | 5 y 10 m | M | KG 2 ° | MDC | 5 | 10 | 2 | Right | No | - | Yes (VPA) |
| **7** | 5 y 11 m | M | KG 3 | - | 5 | 11 | 0 | Left | No | - | No |
| **8** | 6 y 3 m | F | KG 3 | - | 3 | 39 | 1 | Right | Yes | TDM | No |
| **9** | 6 y 3 m | F | CP | Speech therapy | 6 | 3 | 0 | Bilateral | No | - | No |
| **10** | 6 y 4 m | F | KG 3 | Speech therapy | 5 | 16 | 3 | Right | Yes | TDM | Yes (VPA) |
| **11** | 6 y 9 m | F | CP | - | 5 | 21 | 1 | Right | Yes * | TDM | Yes (VPA) |
| **12** | 7 y 8 m | M | CE1 | Speech therapy | 6 | 20 | 0 | Right | Yes | TDM | Yes (VPA) |
| **13** | 7 y 11 m | M | CE1 | - | 7 | 11 | 4 | Left | Yes * | TDM | No |
| **14** | 8 y 2 m | M | CE2 | - | 4 | 50 | 3 | Right | No | - | No |
| **15** | 8 y 5 m | M | CE1 | - | 8 | 5 | 1 | Bilateral | No | - | No |
| **16** | 8 y 7 m | M | CE1 ° | - | 7 | 19 | 1 | Right | No | TDM | No |
| **17** | 8 y 9 m | F | CP # | Speech therapy | 7 | 21 | 6 | Left | No | - | Yes (VPA) |
| **18** | 8 y 10 m | M | CM1 | Speech therapy | 4 | 58 | 0 | Bilateral | No | TDM | No |

y = year (s) ; m= months (s) ; M = male, F = female ; KG 2 = year 2 kindergarten (4-5 years) ; KG 3 = year 3 kindergarten (5-6 years) ; CP = reception (6-7 years) ; CE1 and CE 2 = 1st and 2nd year primary (7-9 years) ; CM1 = 1st year junior (9-10 years) ; ° one year behind chronological age ; # two years behind chronological age ; MDC = multidisciplinary centre ; MRI = magnetic resonance imaging ; TDM = tomodensitometry ; VPA = sodium valproate ; CBZ = carbamazepine.

longer apparent at the time of the neuropsychological evaluation. In nine children the focus was right-sided, in three it was left-sided, and in four bilateral. Four of the 10 sleep EEG were normal, one demonstrated moderate activation of epileptic abnormalities, four had significant activation with generalized abnormalities in two cases, and one showed generalized spike-waves over more than 50% of sleep time (*table 2*). Nine children were receiving a single antiepileptic drug (eight sodium valproate and one carbamazepine). Neuroimaging was performed in 12 children, the results being normal in 11. One child had an arachnoid cyst of the posterior fossa and a mega cisterna magna.

The mean overall intelligence quotient of the population was normal (FIQ = 96), as were the mean VIQ and PIQ (98 and 95, respectively). The difference between this and the theoretical mean of 100 was not significant. However, the results were very heterogeneous between individuals (*table 2*). The profiles of three patients were homogeneous, with retardation and an FIQ of less than 70. The intellectual ability of five children was normal and homogeneous. Ten children had normal intellectual ability, but there was a significant verbal-performance discrepancy, with at least 15 points difference between scales: performance scores were lower for seven children and verbal scores for three children. We attempted to study the relationship between IQ profiles and the focus side as revealed by the EEG and/or the existence of abnormalities on sleep EEG. However, it was not possible to make statistical comparisons in view of the small study size. Nevertheless, of the seven children with lower performance scores, the focus was right-sided in four and left-sided in one. For the three children who had lower verbal scores, there was no predominant side. Three of the five children with homogeneous, normal intellectual ability had bilateral foci. *Table 3* summarizes the results of the mean IQ of the population according to the side of the focus. The mean PIQ was lower when the focus was right-sided.

Analysis of the abnormalities demonstrated on sleep EEG, showed that there did not appear to be any significant effect on intellectual ability. The children with the greatest abnormalities had mainly normal intellectual abilities, or even better than average. On the other hand, the patients with slight (patients eight and ten) or severe retardation (patient 12) had normal sleep EEGs. For patient 3, it could be wondered whether there was a relationship between the very active, right-sided epileptic focus and the discrep-

**Table 2.** Intellectual ability: individual results (VIQ, PIQ, FIQ) depending on IQ profile, side of focus on EEG, sleep EEG results

| Patient N° | VIQ | PIQ | FIQ | EEG focus | Sleep EEG |
|---|---|---|---|---|---|
| | ***FIQ < 70 and homogeneous*** | | | | |
| **6** | 68 | 66 | 64 | right | |
| **8** | 65 | 71 | 64 | right | normal |
| **10** | 69 | 69 | 66 | right | normal |
| | ***PIQ < VIQ*** | | | | |
| **9** | 82 | 65 | 69 | bilateral | |
| **16** | 110 | 77 | 93 | right | |
| **7** | 103 | 88 | 95 | left | |
| **3** | 108 | 89 | 99 | right | CSWS > 50% |
| **4** | 127 | 88 | 109 | right | gen. bursts |
| **1** | 132 | 100 | 117 | none | normal |
| **14** | 139 | 123 | 137 | right | |
| | ***VIQ < PIQ*** | | | | |
| **17** | 65 | 116 | 86 | left | |
| **2** | 76 | 119 | 96 | none | gen. bursts |
| **11** | 103 | 119 | 111 | right | activation + |
| | ***FIQ > 70 and homogeneous*** | | | | |
| **12** | 77 | 77 | 74 | right | normal |
| **15** | 102 | 91 | 96 | bilateral | |
| **5** | 114 | 107 | 107 | bilateral | activation +++ |
| **18** | 105 | 119 | 112 | bilateral | |
| **13** | 123 | 123 | 127 | left | activation +++ |
| **Mean** | 98 | 95 | 96 | | |
| **SD** | 24 | 21 | 22 | | |

(CSWS = continuous spike waves during sleep, gen bursts = generalised bursts, activation + = moderate activation, activation +++ = significant activation).

**Table 3.** Mean IQ results according to EEG focus side.

| | Mean IQ | Right EEG focus | Left EEG focus | Bilateral EEG focus |
|---|---|---|---|---|
| ***Patients (n=16)*** | | 9 | 3 | 4 |
| ***VIQ*** | 98.2 | 96.2 | 97.0 | 100.87 |
| ***PIQ*** | 94.8 | 86.6 | 109.0 | 95.5 |
| ***FIQ*** | 95.7 | 90.8 | 102.6 | 96.0 |

ancy of 19 points between the VIQ and PIQ, the latter being the weaker.

The administration of antiepileptic treatment (valproate or carbamazepine) did not influence the IQ results. There was no significant difference between the means for treated (VIQ = 92.3, PIQ = 95.6, FIQ 92.2) and untreated patients (VIQ = 104.1, PIQ = 94, FIQ = 99.1). The results for specific functions are summarized in *table 4*.

As regards verbal skills, there was no statistically significant difference from the theoretical mean of 10. Individual values were fairly heterogeneous for verbal fluency and naming skills, whereas it was more homogeneous for phonological production and syntactic comprehension.

Drawing and visuo-spatial skills scores were significantly lower than the mean, as were attention and visuo-spatial memory scores. In contrast, verbal memory was normal.

Non-verbal functions in the seven children with PIQ < VIQ were lower than those of the rest of the population (mean 3.37 *versus* 6.12), but the difference was not statistically different.

Scores were particularly low for drawing, for reasons we discuss below. Results and scores were particularly low for the youngest children, although patient 2 obtained a good score despite his young age.

Results for visuo-spatial memory were also low, with four children scoring zero. Attention scores were more homogeneous, particularly those for selective attention. Individual and mean scores for learning skills in reading, numeracy and spelling, and retardation in relation to chronological age are presented in *table 5*.

The mean developmental age for numeracy was lower than the mean chronological age for the population

**Table 4.** Normalised individual values for specific cognitive functions (reference value = 10) and individual IQ values.

| | Verbal functions | | | | Non-verbal functions | Memory | | Attention | | | IQ | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| **Patients** | **Phonological production** | **Naming skills** | **Verbal fluency** | **Syntactic comprehension** | **Drawing and VS abilities** | **Verbal memory** | **VS memory** | **Visual attention** | **Selective attention** | **Attention (AC or coding)** | **VIQ** | **PIQ** | **FIQ** |
| **1** | 12.0 | 12.5 | 24.7 | 10.9 | 1.3 | 10.0 | 0.0 | 1.9 | 6.7 | 10 | 132 | 100 | 117 |
| **2** | 10.5 | 10.2 | 13.2 | 8.1 | 10.3 | 0.0 | 2.4 | 11.5 | 11.0 | 13 | 76 | 119 | 96 |
| **3** | 9.9 | 12.2 | 13.2 | 10.5 | 0.0 | 13.5 | 0.0 | 8.0 | 7.7 | 8 | 108 | 89 | 99 |
| **4** | 10.8 | 11.6 | 16.4 | 12.1 | 1.8 | 7.5 | 1.0 | 12.3 | 10.6 | 9 | 127 | 88 | 109 |
| **5** | 10.6 | 10.7 | 9.9 | 11.3 | 1.2 | 12.5 | 2.0 | 6.9 | 9.4 | 7 | 114 | 107 | 107 |
| **6** | 10.3 | 4.4 | 4.6 | 7.3 | 0.0 | 0.0 | 0.0 | 5.7 | 9.4 | 5 | 68 | 66 | 64 |
| **7** | 10.3 | 12.7 | 13.8 | 8.3 | 4.7 | 12.5 | 4.9 | 8.2 | 9.9 | 7 | 103 | 88 | 95 |
| **8** | 10.3 | 6.5 | 11.2 | 8.3 | 0.5 | 8.9 | 0.8 | 10.2 | 4.6 | 9 | 65 | 71 | 64 |
| **9** | 10.8 | 6.5 | 10.5 | 10.8 | 0.5 | 10.7 | 0.0 | 6.3 | 7.6 | 2 | 82 | 65 | 69 |
| **10** | 9.8 | 4.6 | 7.2 | 8.3 | 11.2 | 10.7 | 3.0 | 4.5 | 3.0 | 3 | 69 | 69 | 66 |
| **11** | 10.4 | 7.9 | 15.1 | 11.0 | 9.3 | 11.7 | 4.5 | 9.2 | 10.3 | 10 | 103 | 119 | 111 |
| **12** | 10.1 | 11.6 | 12.8 | 10.5 | 4.9 | 2.7 | 2.8 | 6.9 | 10.1 | 3 | 77 | 77 | 74 |
| **13** | 10.1 | 8.5 | 7.7 | 9.6 | 4.9 | 11.0 | 8.3 | 8.3 | 10.1 | 12 | 123 | 123 | 127 |
| **14** | 10.1 | 11.1 | 8.7 | 11.4 | 9.1 | 8.2 | 5.3 | 11.5 | 9.1 | 11 | 139 | 123 | 137 |
| **15** | 10.1 | 13.2 | 9.7 | 10.2 | 9.4 | 12.3 | 2.1 | 11.5 | 9.6 | 7 | 102 | 91 | 96 |
| **16** | 10.1 | 13.5 | 9.4 | 8.8 | 6.3 | 8.3 | 6.9 | 5.7 | 9.5 | 3 | 110 | 77 | 93 |
| **17** | 7.9 | 3.7 | 7.2 | 9.6 | 6.6 | 7.1 | 7.4 | 10.9 | 10.1 | 13 | 65 | 116 | 86 |
| **18** | 10.1 | 13.0 | 10.6 | 11.7 | 9.1 | 10.7 | 9.5 | 10.5 | 10.1 | 9 | 105 | 119 | 112 |
| ***Mean*** | *10.24* | *9.68* | *11.43* | *9.93* | *5.04* | *8.80* | *3.38* | *8.33* | *8.81* | *7,83* | *98* | *95* | *96* |
| ***SD*** | *0.76* | *3.30* | *4.49* | *1.42* | *3.99* | *4.09* | *3.08* | *2.87* | *2.14* | *3,50* | *24* | *21* | *22* |
| **Related to 10** | NS | NS | NS | NS | p = 0.0001 | NS | p < 0.0001 | p = 0.024 | p = 0.03 | p = 0.02 | | | |

**Table 5.** Educational level for reading, numeracy and spelling (developmental age) and retardation compared to chronological age (negative values indicate retardation and positive values indicate advance compared to chronological age) (**in bold: 9 months retardation or more**). Results are expressed in months. Reading and spelling were not evaluated in children aged less than 60 months. Individual IQ values are repeated.

| Patients | Age | Reading | Numeracy | Spelling | Reading retardation | Numeracy retardation | Spelling retardation | VIQ | PIQ | FIQ |
|---|---|---|---|---|---|---|---|---|---|---|
| **1** | 50 | | 51 | | | 1 | | 132 | 100 | 117 |
| **2** | 55 | | 60 | | | 5 | | 76 | 119 | 96 |
| **3** | 58 | | 48 | | | **-10** | | 108 | 89 | 99 |
| **4** | 64 | 66 | 69 | 63 | 2 | 5 | -1 | 127 | 88 | 109 |
| **5** | 68 | 57 | 51 | 60 | **-11** | **-17** | -8 | 114 | 107 | 107 |
| **6** | 70 | 66 | 51 | 60 | -4 | **-19** | **-10** | 68 | 66 | 64 |
| **7** | 71 | 69 | 69 | 69 | -2 | -2 | -2 | 103 | 88 | 95 |
| **8** | 75 | 60 | 48 | 66 | **-15** | **-27** | **-9** | 65 | 71 | 64 |
| **9** | 75 | 63 | 57 | 63 | **-12** | **-18** | **-12** | 82 | 65 | 69 |
| **10** | 76 | 57 | 57 | 60 | **-19** | **-19** | **-16** | 69 | 69 | 66 |
| **11** | 81 | 90 | 81 | 84 | 9 | 0 | 3 | 103 | 119 | 111 |
| **12** | 92 | 81 | 87 | 87 | **-11** | -5 | -5 | 77 | 77 | 74 |
| **13** | 95 | 87 | 93 | 93 | -8 | -2 | -2 | 123 | 123 | 127 |
| **14** | 98 | 111 | 123 | 108 | 13 | 25 | 10 | 139 | 123 | 137 |
| **15** | 101 | 102 | 99 | 93 | 1 | -2 | -8 | 102 | 91 | 96 |
| **16** | 103 | 96 | 102 | 93 | -7 | -1 | **-10** | 110 | 77 | 93 |
| **17** | 105 | 72 | 84 | 72 | **-33** | **-21** | **-33** | 65 | 116 | 86 |
| **18** | 106 | 117 | 96 | 105 | 11 | **-10** | -1 | 105 | 119 | 112 |
| ***Mean*** | *80.2* | *79.6* | *73.7* | *78.4* | *-5.7* | *-6.5* | *-6.9* | *98* | *95* | *96* |
| ***SD*** | *18.2* | *19.9* | *22.7* | *17.1* | *12.2* | *12.5* | *9.7* | *24* | *21* | *22* |
| **Difference from chronological age** | | NS | p = 0.04 | p = 0.02 | | | | | | |

(80.2 months). Tests for reading and spelling conducted for 15 children with a mean age of 85.3 months showed that there was a significant difference between developmental age and chronological age for spelling but not for reading. Mean retardation was 5.7 months for reading, 6.5 months for numeracy and 6.9 months for spelling.

Individual scores for learning skills were particularly interesting. More advanced learning skills masked the extent of retardation in others and raised mean scores. However, in terms of educational level it was retardation that penalised children most. Of the 15 children aged less than 60 months, 10 children (67%) were retarded by nine months or more, *i.e.* the equivalent of at least one school year in one (4 children), two (2 children) or three (4 children) learning skills. Eight children had more than nine months' retardation in reading or spelling, and eight children in numeracy. If the three children with mental retardation were excluded (patients 6, 8 and 10), four children had more than nine months' retardation in reading, five in numeracy and three in spelling.

As shown in *table 6*, there was a correlation between the extent of retardation in learning skills and the level of intellectual ability. The correlation was particularly clear in the three children with mental retardation (patients 6, 8 and 10) and patient 9, whose profile was inconsistent but whose FIQ was also low (*table 5*). Nevertheless, the correlation was also very variable at an individual level. Five of the 18 children (27.7%) had normal intellectual ability,

**Table 6.** Spearman correlation scores (r) between reading, numeracy and spelling retardation and IQ levels.

| | VIQ | PIQ | FIQ |
|---|---|---|---|
| **Reading retardation** | r = 0.58<br>p = 0.01 | r = 0.52<br>p = 0.03 | r = 0.70<br>p = 0.001 |
| **Numeracy retardation** | r = 0.69<br>p = 0.001 | r = 0.51<br>p = 0.03 | r = 0.70<br>p = 0.001 |
| **Spelling retardation** | r = 0.65<br>p = 0.003 | r = 0.64<br>p = 0.004 | r = 0.81<br>p = 0.0001 |

or even higher, but had real learning difficulties (patients 3, 5, 16, 17 and 18).

## Discussion

BECTS has always been considered a benign epilepsy syndrome, in terms of seizures, which always disappear at puberty. However, in the initial description of BECTS in 1958, Beaussart has reported educational difficulties and behavior problems in his patients. He attributed them to the psychological consequences of the disease (Beaussart 1972). In view of the recognition of similar disorders in their patients, several authors have studied overall and specific cognitive functions in children with BECTS. Studies in various populations have generally been performed during the active phase of the disease and have focused on both overall intellectual ability and on specific functions such as verbal and non-verbal skills, memory, attention, behavior and learning skills (*table 7*). The neuropsychological studies which have been performed have used various methodologies, sometimes leading to heterogeneous results, but they appear to confirm the existence of cognitive disorders.

Intellectual ability has mainly been evaluated using Wechsler scales, but other tests have been used (*table 7*). Mean scores have been normal whatever the tests used. Some studies considered that an IQ lower than 80 was an exclusion factor, and this may have influenced results (D'Alessandro *et al.* 1990). When there was a control group, certain authors revealed a significant reduction in

**Table 7.** Published neuropsychological studies on BECTS.

| Authors | Year | Study type<br>Number of patients | Tests |
|---|---|---|---|
| Heijbel | 1975 | Retrospective 16 children<br>16 controls | Terman Merrill, Bender's Visual Motor Gestalt Test, parents' and teachers' interviews |
| D'Alessandro | 1990 | Retrospective longitudinal<br>44 children 9 controls | Wechsler scale, Trail Making Test, Stroop Color Word Test,<br>Tests with Barrage of Letters or Geometric Figures, Digit Span,<br>Corsi's Block Tapping Test, Rey's Auditory-Verbal Learning Test, Token test,<br>semantic fluency, Ferrari's naming test, Bender's Visual Motor Gestalt Test,<br>one-minute dotting test of Zazzo |
| Picirilli | 1994 | Retrospective 43 children<br>15 controls | Zazzo's cancellation task |
| Weglage | 1997 | Prospective 40 children<br>40 controls | Wechsler scale, Frostig's Developmental Test of Visual Perception,<br>German version of the Illinois test of Psycholinguistic Abilities,<br>Schoppe's motor performance task, Sonneville Visual Attention Task,<br>German version of the Child Behavior Check List of Achenbach |
| Staden | 1998 | Prospective 20 children | Wechsler scale, Children's Auditory Verbal Learning Test-2,<br>Test of Auditory Discrimination, Test for Reception of Grammar,<br>Clinical Evaluation of Language Fundamentals-R, Test of Word Knowledge,<br>Wechsler Objective Reading Dimension Test |
| Gündüz | 1999 | Prospective 20 children<br>15 controls | Cattell Culture Free Test and Wechsler scale, Griffith's language test,<br>non standardized test of articulation, Go-no-go Test,<br>Luria's alternating sequences of drawing, Raven's colored matrices |
| Metz-Lutz | 1999 | Prospective longitudinal<br>22 children | Wechsler scale, French version of LOLEA, Benton's Visual Form<br>Discrimination, Rey's Complex Figure, Forward digit repetition test,<br>Corsi's Block Tapping Test, Rey's Auditory-Verbal Learning Test,<br>Dichotic listening |
| Croona | 1999 | Prospective 17 children<br>17 controls | Raven's colored matrices, Digits forward, Block span of Milner,<br>Rey's Auditory-Verbal Learning Test, Story recall from Luria's modified<br>neuropsychological battery, Spatial learning test of Anderson,<br>verbal fluency, Trail-Making Test, Rey's Complex Figure, Tower of London,<br>neurobehavioral questionnaire |
| Deonna | 2000 | Prospective longitudinal<br>22 children | Wechsler scale, EDP48, TVAP, VOCIM, SEVWF, EEL, BEP,<br>Lobrot's battery, Rey's Complex Figure, Word, pseudo-word and digit span,<br>Corsi's Block Tapping Test, Rey's Auditory-Verbal Learning Test, HMK-ABC,<br>Trail-Making Test, Zazzo, D2, CPT and others |
| Baglietto | 2001 | Prospective longitudinal<br>9 children 9 controls | Wechsler scale, Trail-Making Test, Cancellation Task of Letters,<br>Numbers and Geometric Figures, Stroop Test, Digit Span,<br>Corsi's block Tapping Test, Token Test, Boston Naming Test,<br>semantic-phonological fluency, Bender's Visual Motor Gestalt Test,<br>Ghent-Poppelreuter Test, Street Gestalt Completion Test,<br>Judgement of Line Orientation |

IQ in children with epilepsy compared to controls (Weglage *et al.* 1997, Baglietto *et al.* 2001), but others did not (D'Alessandro *et al.* 1990, Gündüz *et al.* 1999, Croona *et al.* 1999). Weglage demonstrated a significant reduction in PIQ in children with epilepsy compared to controls, whereas VIQ was comparable (Weglage *et al.* 1997). Individual scores were not given in all studies, but heterogeneous IQs were demonstrated when they were (Staden *et al.* 1998, Metz-Lutz *et al.* 1999, Deonna *et al.* 2000, Baglietto *et al.* 2001). Some children had borderline intellectual ability (IQ < 85) or were retarded; others had verbal-performance discrepancy, the verbal scale being either higher or lower than the performance scale.

Specific cognitive functions have been studied by several teams, often using different methodologies (*table 7*).

In most cases, investigations into verbal skills using Wechsler scales have revealed no abnormalities. However, in view of the topology of paroxysmal abnormalities involving perisylvian language areas, some teams have focused on language studies and, by using more specific tools, have revealed slight changes in verbal function. These have included language deficit affecting verbal fluency, auditory discrimination, naming skills, morphosyntactic expression and prosody (D'Alessandro *et al.* 1990, Staden *et al.* 1998, Croona *et al.* 1999, Gündüz *et al.* 1999, Baglietto *et al.* 2001). In a recent article, Monjauze *et al.* (2005) reported language disorders in nine out of 16 children with BECTS. The areas most affected were morphosyntactic expression and written language.

Several studies have reported disorders of *visuo-spatial* and *visuo-motor skills* (D'Alessandro *et al.* 1990, Piccirilli *et al.* 1994, Weglage *et al.* 1997, Gündüz *et al.* 1999, Metz-Lutz *et al.* 1999, Baglietto *et al.* 2001), *attention* (D'Alessandro *et al.* 1990; Piccirilli *et al.* 1994; Metz-Lutz *et al.* 1999; Baglietto *et al.* 2001), *verbal* (Weglage *et al.* 1997, Croona *et al.* 1999, Hattori 2002) and *visuo-spatial memory* (Binnie *et al.* 1992, Metz-Lutz *et al.* 1999, Baglietto *et al.* 2001). Deonna also described disorders of visuo-spatial organization, attention, and verbal and visuo-spatial memory, but this study reported individual findings and was not a group study (Deonna *et al.* 2000).

Disorders related to frontal involvement such as disorders of planning, perseveration and fine motor and speed reduction have also been reported (Croona *et al.* 1999, Chevalier *et al.* 2000, Gündüz *et al.* 1999, Metz-Lutz *et al.* 1999).

Behavior disorders such as hyperactivity, aggression, opposition, difficulties with concentration and social relationships and even delinquency have also been reported with BECTS in studies based on parental questionnaires and interviews (Weglage *et al.* 1997, Croona *et al.* 1999, Yung *et al.* 2000, Chevalier *et al.* 2000). On the other hand, Heijbel and Bohman (1975) reported that they did not find any behavior disorders in the children that they studied.

Learning difficulties have rarely been reported in the literature concerning BECTS. Investigation of learning difficulties are not often described in detail and most of the evaluations have been undertaken by questionnaires to teachers or parents. Heijbel and Bohman, in a questionnaire-based study, found no reduction of educational performance in epileptic children (1975). This method of evaluation is not very sensitive, and mild cognitive function disorders may not be noticed by teachers. Some authors have reported learning difficulties but without detailing them (Staden *et al.* 1998, Gündüz *et al.* 1999, Deonna *et al.* 2000); others have cited disorders such as dyslexia (Metz-Lutz *et al.* 1999). Croona *et al.* found difficulties with reading comprehension as reported in questionnaires to teachers (1999). Yung *et al.* also reported specific learning difficulties involving reading, numeracy and spelling in 17% of 78 children (2000). Two recent studies reported difficulties with reading and spelling in 6 out of 16 patients (Monjauze *et al.* 2005) and in spelling, reading aloud and reading comprehension in 11 out of 32 BECTS patients (Papavasiliou *et al.* 2005). *According to all these studies, 17 to 41% of children with BECTS had learning difficulties*. These difficulties required therapy and even repeating school years or special education in 10% of children. In the study of Metz-Lutz *et al.* (1999), learning difficulties occurred particularly in children who had had more than three epileptic seizures and who were receiving antiepileptic treatment. In a recent prospective study involving 40 patients with BECTS, Massa *et al.* (2001) described the occurrence of behavior disorders and learning difficulties reported by parents or teachers in 28% of children, but 72% experienced no effects on behaviour or cognitive function.

Our results also confirmed the possibility of cognitive disorders although the mean intellectual ability in the study was not significantly different from the average for the general population. Our study revealed very clearly the heterogeneity of individual results, showing that group studies are not very reliable, and demonstrated the diversity of individual levels of disorder. We demonstrated disorders of attention and visuo-spatial memory and weaknesses in drawing and visuo-spatial skills. Learning difficulties were also found.

The characteristics of our population were comparable overall to those described in various publications, with onset of seizures between 3 and 13 years, low frequency of seizures and preponderance in boys, with a sex ratio of nearly 1:5. There was no particular pattern of previous history and the children had normal psychomotor development. Only patient 6 was attending a multidisciplinary rehabilitation centre before the onset of epilepsy, which might have made his inclusion debatable, but the retardation was not major and the clinical symptoms, EEG pattern and evolution corresponded to the diagnostic criteria of BECTS. Background activity on EEG was normal in all patients.

Neuroimaging did not reveal any anatomical lesions in the children examined. An arachnoid cyst of the posterior fossa and a mega cisterna magna were found in patient 3. However, because these abnormalities occur frequently and are not specific, we considered that they were not related to the symptoms.

Our study was retrospective and the initial aim of the neuropsychological evaluation was the validation of the BREV battery. Nevertheless, the findings, gathered in the context of a protocol comprising numerous complementary tests, performed by experienced researchers, and identical for all the children, were exploitable.

As reported in the literature, the overall mean IQ in our population, evaluated by Wechsler scales, was normal. At an individual level, intelligence quotients were variable from one child to another but were mostly normal. However, patients 6, 8 and 10 had mild mental retardation with homogeneous results, and patient 12 had an FIQ between 70 and 80. Lack of intellectual retardation on inclusion was defined by clinical criteria (psychomotor development, educational level...), as for all children with BECTS. Neuropsychological evaluations were not performed before diagnosis. Two of these children (patients 8 and 10) were respectively three and five years old at the time of diagnosis respectively, and were attending normal school. The learning retardation they presented was probably slight and difficult to appreciate at the time of diagnosis without neuropsychological evaluations because the children were very young and there was no effect on learning at kindergarten age. As discussed above, the inclusion of patient 6 is more debatable.

In the initial description of BECTS, the absence of intellectual retardation was considered to be a diagnostic criterion and a prerequisite (Beaussart 1972). Moreover, in certain studies, an FIQ lower than 80 was an exclusion criterion (D'Alessandro *et al.* 1990). Nevertheless, four studies of patients with BECTS included those with an FIQ lower than 80. All were prospective. Nine per cent of the 78 patients in the study by Deonna *et al.* (2000), and 5% of the 20 patients in the study by Staden *et al.* (1998, 2000) had an FIQ lower than 70. Two of the 22 patients in the study by Deonna *et al.* (2000), and four of the 22 patients in the study by Metz-Lutz *et al.* (1999) had FIQs between 70 and 80. In two of the last four patients, there was a verbal-performance discrepancy, with lower verbal functions. Evaluations were performed during the active phase of the epilepsy. The inclusion criteria were the presence of clinical and EEG characteristics of BECTS. The authors did not report age, psychomotor development or educational levels of the patients on inclusion. As in our study, the neuropsychological evaluations were undertaken after making the diagnosis of BECTS.

In terms of specific functions, attention deficit was found in our population as in other studies. Disorders of drawing and visuo-spatial performance were also observed in our group. The seven children who showed verbal performance discrepancies with lower PIQ had lower drawing and visuo-spatial abilities than the others. The difference was not statistically significant probably because of the small size of the group. Disorders of visuo-spatial and visuo-motor skills have been reported by several teams, but no precise information has been provided concerning drawing. Visuo-spatial memory disorder was revealed in our study. Particularly low scores for drawing and visuo-spatial memory were explained partly by the fact that we chose Rey's complex figure for all the children, including the youngest, whereas is not usually used before the age of 7-8 years in view of its complexity. Nevertheless, scores exist from the age of four years, thus justifying its use. Verbal memory was preserved, in contrast to reports by others (Weglage *et al.* 1997, Croona *et al.* 1999, Hattori 2002). As for verbal functions, no statistically significant disorder was revealed in our population. The results in the literature are also contradictory in this area, some authors describing abnormalities, particularly in verbal fluency, vocabulary and morphosyntactic expression and others reporting no language function disorders. However, we did not explore morphosyntactic expression. These findings, both personal and published in the literature, confirmed wide individual heterogeneity.

Patient 17 had difficulties suggestive of dyslexia, but this might have been fortuitous. Nevertheless, Deonna *et al.* (2000) reported the case of a child with unusual difficulties with spelling but not with reading, for whom a link with epilepsy was suspected. The possibility of learning disorders in three areas (reading, numeracy and spelling) appeared clear in our group of patients, with developmental ages lower than their mean chronological ages. Scores were significantly low in numeracy and spelling.

As in many other studies, there seemed to be a clear link between cognitive disorder and BECTS in our population. These findings lead to questions concerning the factors responsible for neuropsychological disorders in this context.

In general, evaluating the relationship between epilepsy and disorders of cognitive functions is complex in view of the different factors involved, such as age-at-onset, type, frequency and severity of seizures, the existence of underlying lesions, focus site, antiepileptic treatment, and the amplitude of the discharges. BECTS is a good model for such an evaluation as age-at-onset is fairly homogeneous, there are few seizures, treatment is not systematic and of short duration when given, and there are no underlying cerebral lesions. Our retrospective, non-randomized study did not reveal any particular factor.

Drug treatment is often cited as a potential bias in the interpretation of the existence of repercussions of epilepsy on cognitive function. In BECTS, the discovery of cognitive disorders in studies which did not include untreated patients (D'Alessandro *et al.* 1990, Piccirilli *et al.* 1994) and the absence of significant differences between treated and untreated groups (Deonna *et al.* 2000, Staden *et al.*

1998), as in our study, do not support a relationship between antiepileptic treatment and the occurrence of neuropsychological disorders, particularly as monotherapy is most often sufficient when treatment is necessary, and the treatment is usually of short duration in view of the spontaneous recovery in adulthood.

Disorders of cognitive function do not appear to be correlated with the frequency of epileptic seizures (Weglage *et al.* 1997, Carlsson *et al.* 2000).

The most frequently studied hypothesis in the literature involves the relationship between paroxysmal abnormalities on EEG and cognitive function. Centrotemporal spikes activated by sleep have been hypothesised as causal by several authors (D'Alessandro *et al.* 1990, Baglietto *et al.* 2001, Binnie 1993, Blom and Heijbel 1982, Metz-Lutz *et al.* 1999, Morikawa 2002, Pan and Lüders 2002, Papazian *et al.* 2003, Weglage *et al.* 1997). In fact, neuropsychological disorders are mainly related to the active phase of the disease and disappear when distant from the active period (D'Alessandro *et al.* 1990, Baglietto *et al.* 2001, Deonna *et al.* 2000, Weglage *et al.* 1997). Moreover, in one study undertaken with adults who had had BECTS in childhood but were in complete remission (absence of seizures without treatment, normal EEG), no disorders of cognitive function were found (Hommet *et al.* 2001). Massa *et al.* (2001) considered that the presence of five criteria of severity on EEG and their duration for at least six months, were predictive of accumulated risk of cognitive disorders, *i.e.* intermittent slow wave foci, multiple asynchronous foci, long rhythmic sequences of spikes, discharge of spike waves at 3 cycles/s, occurrence of symptoms occurring at precisely the same time as spike waves. We were unable to evaluate the correlation between cognitive or learning disorders with criteria of severity on EEG. As our study was retrospective and multicentered, the monitoring of epilepsy was not the same for all patients, and most children had only one EEG at the time of diagnosis, which is usual in this type of epilepsy.

One other approach consisted of investigating whether the presence of centrotemporal spike waves, with or without the typical pattern of seizures in BECTS, was accompanied by transitory cognitive retardation. Binnie (1993) evaluated eight children with centrotemporal spike waves on EEG. Six of them had all the electroclinical criteria of BECTS, one was asymptomatic and another had had an atypical seizure after the first EEG showing centrotemporal spike waves. Short-term memory was evaluated in each child using a video recorder and continuous EEG monitoring. The children had significant levels of errors in short-term recall tests, which were contemporaneous to the discharges, whether they were right- or left-sided. This study directly questioned the relationship between paroxysmal abnormalities and cognitive disorders, and suggested that the occurrence of transitory cognitive disorders might be related to the discharges.

Carlsson *et al.* (2000) also attempted to study the impact of paroxysmal spike waves on cognitive function. Thirty adolescents with dyslexia were divided into two groups according to the presence or absence of centrotemporal spike-waves on EEG and matched for sex and age. Two patients in the spike-wave group had clinical seizures. There was no difference between the two groups in terms of age, IQ or spelling ability. On the other hand, children with dyslexia and abnormalities on EEG made more mistakes in reading isolated words than those without abnormalities on EEG. Attention disorders in a cancellation task were also more frequent in the group with centrotemporal spikes. These findings suggest that paroxysmal abnormalities may have an effect on cognitive functions.

Other studies have attempted to establish a correlation between the topography of the focus and the type of neuropsychological deficit, but their results are rather contradictory. Such an evaluation is difficult because the focus in BECTS tends to shift from one side to another during evolution, or even becomes bilateral. Some studies have demonstrated predominant attention disorders, poor learning strategies and reduced visuo-spatial memory in children with right or bilateral focus (D'Alessandro *et al.* 1990, Piccirilli *et al.* 1994). In the study by Massa *et al.* (2001), the right-sided predominance of abnormalities on EEG in six children was consistent with the existence of visuo-spatial disorders. Similarly, in our population, qualitative analysis demonstrated that the results for children with right-sided focus were particularly low for non-verbal functions. These findings suggest that the existence of right-sided focus might be associated with cognitive disorders associated with the right hemisphere. Most authors found no obvious relationship between left-sided focus and oral or written language disorders using standard tests. One study aiming to investigate the precise relationship between the topography of the focus and disorders of cognitive function was performed by Piccirilli *et al.* (1988). Using a dual-task procedure (unilateral tapping associated with verbal activity or not), they studied hemispheric representation of language in 14 right-handed children with rolandic epilepsy and right- or left-sided focus matched with control children. Children with right-sided focus had similar results to controls (tapping with the right hand preferentially affected by verbal task) compared to children with left-sided focus, who presented a different pattern suggesting a bi-hemispheric representation of language. Piccirilli suggested disorder of representation of language in children with stable, left-sided focus. Hommet *et al.* (2001) obtained similar results when comparing hemispheric specialisation in 23 adults in complete remission from BECTS followed up from childhood to 33 non-epileptic adults and 10 adults in complete remission from childhood absence epilepsy. Hemispheric specialisation evaluated by a dichotic listening task and a dual-task procedure presented a reverse pattern for the dual-task procedure in adults with left-sided focus as determined by

the EEG in childhood. This study suggested persistence of a disorder of hemispheric language representation in adulthood and supported the hypothesis proposed by Piccirilli *et al.* (1988).

Although not related to an organic lesion, a paroxysmal focus might thus affect the organisation of cerebral function in the developing brain. Functional imaging studies in BECTS have not provided additional information. Using SPECT, Laub *et al.* (1992) studied the relationship between the topography of focus and cognitive function in nine children, but found no correlation between neuropsychological tests and resting cerebral blood flow.

Two main hypotheses have been advanced in an attempt to explain the relationship between spike discharges and neuropsychological disorders.

Some authors have suggested that paroxysmal abnormalities may alter cerebral mechanisms underlying cognitive activity and that the pattern of functional cerebral representation in patients with focal epilepsy depends on the focus side (Piccirilli *et al.* 1988, Hommet *et al.* 2001). These paroxysmal abnormalities might be responsible for an impairment of brain maturation even in the regions distant from the discharges. This would thus explain why functions depending on frontal regions, which are cerebral areas developing at the age-at-onset of this type of epilepsy, might often be affected (Metz-Lutz *et al.* 1999, Chevalier *et al.* 2000). The multiplicity of cognitive disorders observed in these children might thus be explained by the various localizations and areas of diffusion of epileptic foci in BECTS (Deonna 2000).

Doose proposed that epileptic activity and disorders of higher functions might be two independent elements, consequences of an underlying disorder of cerebral maturation, probably of genetic origin. He suggested a neurobiological approach by which genetic predisposition might lead to clinically variable phenotypes (BECTS, continuous spike-waves during sleep syndrome, Landau-Kleffner syndrome, benign occipital paroxysmal epilepsy, certain types of familial mental retardation and certain specific learning disorders) and/or EEG patterns (photosensitivity, spike waves, theta rhythms) that are associated in some families and that are related to hereditary disorders of cerebral maturation, which might explain the age-dependent nature and remission at puberty of these diseases (Doose *et al.* 2000).

These two hypotheses are not contradictory, and the two mechanisms are probably interrelated. The existence of a genetic determinant might result in a disorder of cerebral maturation and the occurrence of epilepsy is certainly plausible. In such a state of maturation and in the active phase of the epilepsy, it is also possible to envisage that accentuation of discharges on EEG might temporarily exacerbate cognitive disorders.

The probable relationship between interictal abnormalities and cognitive disorders raises the question if treatment indications should be limited to the frequency of seizures or should include presence of paroxysmal abnormalities (Weglage *et al.* 1997, Baglietto *et al.* 2001, Binnie 2003, Papazian *et al.* 2003). Care must also be taken not to aggravate abnormalities seen on EEG with certain drugs, as has been demonstrated in particular with carbamazepine (Prats *et al.* 1998). Controlled studies are lacking. Clobazam, ethosuximide and particularly corticosteroids have been described as very effective, but they do have side effects. Sulthiame has a specific effect on interictal paroxysms and on seizures, and has no side effects, but its effectiveness is reported transitory (Doose *et al.* 1988, Bast *et al.* 2003, Engler *et al.* 2003). There are no prospective studies to confirm that effective treatment for interictal spike-waves provides real improvement of cognitive functions. Our open study confirms previous reports suggesting that specific cognitive functions might be deficient in children with BECTS. It is estimated that such dysfunctions affect 15-30% of children with rolandic epilepsy. When such a deficit is suspected, neuropsychological assessment is indicated. It may identify specific deficits allowing the application of the most appropriate educational support and care. □

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