Seizure 18 (2009) 320–326

Contents lists available at ScienceDirect

Seizure

journal homepage: www.elsevier.com/locate/yseiz

Speech and school performance in children with benign partial epilepsy with centro-temporal spikes (BCECTS)

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ARTICLE INFO

Article history:
Received 13 June 2008
Received in revised form 21 October 2008
Accepted 20 November 2008

Keywords:
Rolandic epilepsy
Language
Speech performance
CBCL
Childhood

ABSTRACT

Purpose: To assess prospectively language and speech ability in children with benign partial epilepsy with centro-temporal spikes (BCECTS). To evaluate academic performance and social competencies both during the active disease and after remission.

Methods: Right-handed school children with typical BCECTS and a control group matched by age, sex, handedness and socioeconomic status were examined. The German version of WISC-R, the "Tübinger Luria Christensen Neuropsychological Test Set for children" (TÜKI), the "Verbal Learning Memory Test" (VLMT), the "Heidelberger Speech Development Test-second edition" (HSET), and the "Salzburger reading and writing test" (SLRT) were applied. The Child Behavior Checklist (CBCL) and a semi-structured interview were performed with the children's teachers to gain insight into school performance, behavioral and emotional problems.

Results: During the active phase, the patient group – despite normal global intellectual abilities – showed few, but significant impairments both in expressive speech and in receptive and expressive vocabulary. A significant deficit in the ability to recognize and express interpersonal relations was also found. Patients' teachers stated deficits concerning academic performance and complained about disturbing behavior. Parents reported significantly more psycho-pathological features in the subscales "Aggressive Behavior", "Attention Problems" and "Anxious/Depressed" of the CBCL. Results were independent of medication and spike localization.

After complete recovery from BCECTS, these problems were not found any more.

Conclusions: Both deficits of speech-related abilities (in both expressive and receptive vocabulary) and behavioral disturbances can be detected in children with typical BCECTS, but are no longer apparent after remission of the seizure disorder.

1. Introduction

Accounting for 24% of all children with epilepsy between age 5 and 14, benign childhood epilepsy with centro-temporal spikes (BCECTS) is the most common idiopathic localization-related epilepsy syndrome. An even higher percentage – 1.5–2.4% of all school-age children within the susceptible age range – exhibit the characteristic "rolandic sharp waves" in the EEG.[1,2] Both seizures and EEG discharges usually remit before the age of 16. Complete recovery occurs even where there has been no systematic anti-epileptic drug (AED) treatment and also in patients whose seizures are difficult to control.[3–6]

Although the absence of both intellectual and behavioral abnormalities is part of the original International League against Epilepsy's defining criteria for BCECTS,[1] a diversity of sectorial cognitive deficits have been reported, even in those children without overt seizures.[7–21] Below-average school performance and various behavioral co-morbidities, e.g. attention deficit hyper-activity disorder (ADHD) have also been described.[11,22–24]

Studies specifically investigating language/speech performance in children with BCECTS found atypical (modified) hemispheric specialization, impairments in phonological production and verbal fluency, (i.e. naming, reading, spelling, expressive grammar), syntactic comprehension and literacy skills including orthography and dyslexic-type errors.[9,25–33]

Despite this growing body of literature, the role of selection bias (i.e. different inclusion criteria, especially inclusion of "more or less atypical" cases and wide age ranges), the immediate and long-term effect of interictal EEG discharges and antiepileptic drug (AED) treatment are subjects of ongoing controversial debate.

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doi:10.1016/j.seizure.2008.11.011

(1) Most of the studies found language delay which is exclusively related to an atypical phenomenology and/or frequent subclinical epileptic discharges, primarily occurring during non-rapid eye movement (NREM) sleep.[7,17,34,35] Contrary, some authors reported only minimal association between EEG features and cognition.[25,36]
(2) In some studies remarkable improvement of language deficits was noticed after remission,[15,37] whereas others reported the persistence of significant impairment, suggesting possible long-term effects.[17,24,26,29,38,39]
(3) Published information about the effect of specific AED treatment ranges from "no effect at all"[19,40] to "beneficial effect"[7] to aggravation [e.g. Ref. 40].

The present prospective study was therefore designed: (1) to delineate the extent and characteristics of speech and/or language impairment in children with typical BCECTS, (2) to examine potential effects of age at seizure onset, seizure type and frequency, "spike burden" in the EEG and AED treatment, (3) to evaluate school performance and/or behavioral problems during active BCECTS and after complete recovery. The following hypotheses were stated:

(1) Compared to healthy controls, school children with active typical BCECTS display significant impairment in language and/or speech. This assumption is based on the knowledge that oromotor symptoms, speech arrest, dysarthria and anarthria are part of the seizure semiology, and the EEG displays sharp waves located primarily in the peri-sylvian region[7] as well as on the neuropsychological data published so far (see Section 1 of this paper).
(2) Based on the assumption that BCECTS is a "bilateral thalamocortical disorder", with EEG discharges typically seen independently on both sides either during one and the same EEG or in subsequent EEG recordings of the same patient,[7,17] impairment should be independent of spike lateralization in the EEG recorded at the time of testing.
(3) Age at seizure onset, the frequency and/or type of seizures and the extent of sub-clinical EEG discharges ("spike burden") during sleep have a significant impact on language and/or speech abilities.
(4) Speech/language deficits are a consequence of the disorder "per se" and are therefore found in both children who are and those who are not treated with sulthiame (STM). STM is the first-line AED for treatment of BCECTS in Austria and is known to have no aggravation potential. STM, however, abolishes EEG discharges only initially, but not permanently.[5,40,41]
(5) Compared with healthy controls, the results of neuropsychological testing show that children with BCECTS not only display significant deficits, but also exhibit significantly more learning and/or behavior problems which require intervention.
(6) BCECTS not only causes immediate deficits during the active period, but – as a result of aberrant speech/language development – permanent impairment of academic performance and social competencies can be observed, even after remission of seizures and epileptic discharges in the EEG.

2. Methods

2.1. Subjects

Consecutive right-handed children diagnosed with active typical BCECTS according to ILAE criteria[1] were selected from the pediatric epilepsy outpatient department. Due to the neuropsychological tests administered, children had to be between 6 and 15 years of age and to attend normal schools.

Children who had participated in our former study[19] were not included. Also excluded were children presenting with atypical features according to the results of diagnostic workup, children with abnormal MRI findings and children with global intellectual impairment (details see below). The "best buddy" method was used to recruit control subjects matched for age, sex, handedness and socioeconomic status. The control group was put through the same examinations as the patient group in order to rule out global intellectual, neurological and sensory impairment, as well as undiagnosed seizure disorders or epileptic EEG activity.

All children included had to be native German speakers.

2.2. Diagnostic workup

All children underwent clinical neurological examinations, high-resolution magnetic resonance imaging (MRI) and EEG studies including sleep. In addition, they were put through audiological-phonetic and ophthalmologic examinations.

EEGs were analyzed visually by the senior author and scored according to Bast et al.[41] and Aeby et al.[42] Five grades were defined (grade 0 = normal EEG; grade 1 = normal background, unilateral centro-temporo-parietal sharp wave focus; grade 2 = normal background; bilateral independent sharp waves located in the centro-temporo-parietal electrodes; grade 3 = destructured background, intermittent slow wave focus, sharp waves diffusing to one hemisphere or multiregional sharp waves; grade 4 = destructed background, intermittent slow wave focus, sharp waves diffusing to both hemispheres). Only patients with EEGs grades 1 and 2 were included, those with grades 3 and 4 were considered atypical and therefore excluded from the study. Controls were only included in case of EEG grade 0. In order to be able to detect a possible association between "spike density" and the extent of language/speech impairment, a sharp-wave index (SWI) during the first 30 min of NREM stages of the first and last sleep cycles was calculated by dividing the number of seconds presenting one or more sharp waves in the two 30 min periods, divided by 3.600 and then multiplied by 100, to express the results in percentage.[42,43]

2.3. Instruments

2.3.1. Assessment of cognitive performance

Global intellectual functioning was examined using the HAWIK-III and the German version of the WISC-III.[44] Children with Full Scale IQ below 85 were excluded from the study. Specific neuropsychological assessment was performed using a set of standardized tests (Table 1): the "Tübinger Luria Christensen Neuropsychological Test Set for children" – TÜKI,[45] the Auditory Verbal Learning Test of Rey, the "Verbal Learning Memory Test" – VLMT,[46] the "Heidelberger Speech Development Test – 2nd Edition" – HSET[47] and the "Salzburger reading and writing test" – SLRT.[48]

Different aspects of language/speech were examined, namely:

- Auditory discrimination of real words. This was tested using one subtest of the TÜKI where the child is asked to identify a word presented orally by selecting the corresponding picture from a set of two. The words describing the picture differ only by one phoneme, for example "house–mouse".
- Auditory verbal memory, which was measured using the VLMT. In contrast to many other tests, which assess only one aspect of memory, the VLMT permits the evaluation of "learning capacity" (number of correct responses in five trials), "retention" (correct responses after 30 min) and "recognition" (correct minus false positive answers).
- Receptive grammar, which was assessed using various subtests from the HSET which examine the child's ability to understand orally presented grammatical structures and

Table 1
Assessment of cognitive performance.

Test procedure	Abilities examined/assessed
HAWIK-IIII (German Version of WISC-III)	Intellectual abilities
Verbal Learning and Memory Test (VLMT)	Auditory verbal memory
Tübinger Luria Christensen Neuropsychological Test Set for Children (TÜKI)	Auditory discrimination, Receptive and expressive vocabulary
Heidelberger Speech Developmental Test – 2nd Edition (HSET)	Receptive and expressive grammar
Salzburger Reading and Writing Test (SLRT)	Reading and orthographic abilities

contrasts (''Sentence structure'', ''Morphological structure'', ''Meaning of sentence'').
- Expressive grammar, which was evaluated using various subtests from the HSET, assessing the child's ability to formulate simple and complex sentences.
- Receptive and expressive vocabulary were measured using various subtests from the HSET and two subtests from the TÜKI, which examine comprehension and naming of words, as well as comprehension and use of semantic features, relations and fields.
- Reading and orthographic abilities were measured using the SLRT. Reading ability was assessed via synthetic reading, evaluation of automatic direct recognition and by reading texts.

2.3.2. Assessment of school performance and social competencies

Parents completed the Child-Behavior Checklist (CBCL/4–18),[49] a widely used tool to measure child behavior problems with well-established reliability and validity. The CBCL consists of 118 items related to behavior problems which are scored on a 3-point scale of 0 (no true), 1 (sometimes true), and 2 (very true). Using a computer-scoring algorithm, the child's behavior problems are used to create the following subscales: two broadband dimensions (internalizing and externalizing) and eight core syndrome subscales (social Withdrawal, Somatic Complaints, Anxiety/Depression, Social Problems, Thought Problems, Attention Problems, Delinquent Behavior, and Aggressive Behavior) plus DSM-oriented scales. There are also 20 social competency items used to obtain parents' reports of the amount and quality of their child's participation in sports, hobbies, games, activities, organizations, jobs and chores, friendships, how well the child gets along with others and plays and works by him/herself, and school functioning. Norms based on age and genders are available for all scales. Although the CBCL was not designed for diagnosing psychopathology in children with chronic illnesses, they have become extensively used research tools to assess behavioral problems in pediatric populations, including children with epilepsy.[28,50–53]

After parents had given their informed consent a semi-structured interview with the teachers of both the patients and controls was performed to assess academic performance, adaptive functioning, and behavioral and/or emotional problems.

2.4. Study visits

Children with BCECTS were evaluated 1 year after disease onset, in order to guarantee exact syndrome diagnosis (T1) and again 1 year after remission, after tapering of AED treatment (T2).

At T1 complete diagnostic work up, complete cognitive assessment and parent interviews including the CBCL 4–18 and semi-structured interviews with the teachers were performed. Reevaluation at T2 consisted of complete clinical neurological and psychiatric investigations, as well as EEGs including sleep and the CBCL 4–8.

Controls followed the same investigation procedure at T1, but were not seen at T2 for ethical reasons. Only their parents answered the CBCL 4–18 a second time during a structured telephone interview.

2.5. Statistical analyses

All statistical analyses were conducted using the Statistical Package for the Social Sciences (SPSS, Version 15). For descriptive purpose the means and standard deviations for individual subtests and composite scores were calculated.

Group differences were assessed using independent samples *t*-tests or ANOVA models in case of normality and homogeneity of variances or Mann–Whitney *U*-tests or Kruskal–Wallis tests otherwise.

The effects of age at seizure onset, seizure type and frequency, AED treatment and EEG (''spike load'') on test performance were examined by further independent sample *t*-/Mann–Whitney *U*-tests.

Changes in the CBCL- scores between T1 and T2 were tested using paired sample *t*-tests or ANCOVA models with the time between T1 and T2 serving as covariate.

The semi-structured interviews with the teachers were analyzed according to the principle of inductive qualitative content analysis.[54] Categories were only taken for further analysis when the inter-rater reliability of two independent-raters was sufficiently high ($\kappa \geq 0.7$). The global error type I was set to 0.05; Due to multiple testing only p-values ≤ 0.002 (Bonferroni correction) were accepted.

3. Results

3.1. Intake data (T1)

A total of 30 patients was screened, 10 were excluded because of atypical features (see inclusion/exclusion criteria). 20 patients (11 boys and 9 girls aged 6.0–14.11 years) and 20 healthy controls (matched by age and gender) were included in the statistical analysis.13 patients (65%) were $\leq$10, 7 (35%) were >10 years of age. All patients had active EEG-foci. Within the 12 months prior to testing, 5 patients (25%) had experienced $\leq$5 seizures, 11 (55%) had experienced between 5 and 10 seizures and 4 (25%) had experienced >10 seizures. The majority of the children experienced exclusively partial seizures; secondary generalized seizures were reported in only 7 cases (40%).

EEG recordings were grade 1 in 11 patients (55%) with right-hemispheric foci in 30% and left-hemispheric foci in 25%. Independent bilateral foci (EEG grade 2) were found in 9 patients (45%). The SWI ranged between 0.5 and 43 (mean 36), and was $\leq$25% in 7 (35%) and >25% in 13 (65%).

A total of 8 patients (40%) had never received AED treatment, 12 (60%) were treated with STM 5–10 mg/kg body weight.

3.2. Cognitive performance

3.2.1. Global intelligence

Full scale IQ of the 20 patients ranged from 85 to 114 and from 92 to 119 for the control children, implying a significant difference between patients and controls only before Bonferroni correction (t (38) = −2.506, p = 0.017); this tendency appeared to be independent of AED-treatment, age of onset, seizure type and

Table 2
Intelligence (HAWIK-III).

	Patients ($n = 20$)		Controls ($n = 20$)		
	Mean	SD	Mean	SD	p
HAWIK-III					
Full-Scale IQ	43.65	26.95	64.30	25.14	0.017*
Verbal IQ	55.70	29.40	68.40	22.57	0.134
Performance IQ	34.10	28.67	6.00	6.59	0.017*
HSET					
Adjective derivations	42.35	21.03	65.11	19.73	0.001**
Correction of semantically inconsistent sentences	37.75	31.20	63.61	21.92	0.004**
Descriptive flexibility	44.94	22.19	73.92	23.64	<0.001***
Relating to verbal and non-verbal information	36.64	32.05	69.30	16.02	<0.001***

Note. The mean of each IQ-score is shown as a percentile rank; *≤0.05 (not significant after Bonferroni correction, **≤0.01 (not significant after Bonferroni correction), **≤0.002 (significant after Bonferroni correction, *** ≤0.001 (significant after Bonferroni correction); HAWIK-III = Hamburger Wechsler Intelligenztest für Kinder – 3rd Edition.

frequency and spike burden. There was no significant difference with regard to the Verbal IQ, but – again only before correction – in the Performance IQ (Table 2), particularly in the subtests "Block Design" (t (38) = 2.139, p = 0.039) which measures spatial perception, and the subtest "Object Assembly" (t (38) = 2.048, p = 0.048) which examines "visual analysis". Patients with right sided foci tended to obtained slightly lower Verbal IQ results (t (18) = 2.224, p = 0.039) but no significant difference was found between patients with right-unilateral, left-unilateral or bilateral foci.

Additionally there were no differences between the patient-groups when controlling for AED-treatment, age of onset, seizure type and frequency and spike burden.

3.2.2. Expressive grammar/speech

No significant differences were found between patients and controls with regard to repeating single, phonemically similar words (t (38) = 0.592, p = 0.558) and also to the imitation of grammatical structural forms (t (38) = 1.006, p = 0.321) or morphological structure when creating plural and singular forms (t (38) = 0.539, p = 0.594). However, concerning both the derivation of adjectives and the correcting of semantically wrong/illogical sentences, patients produced significantly lower results than their controls (see Table 2). Nevertheless, patients of the younger age group (6–10) tended slightly to perform worse in the subtests imitation of grammatical structural forms (t (18) = 1.798, p = 0.089), morphological structure (t (18) = 1.775, p = 0.093) and correcting of grammatically wrong sentences (t (18) = 1.885, p = 0.076) compared to the older age group (11–14).

All further stratifications – EEG focus, AED-treatment, seizure type and frequency and spike burden – showed no effect on these subtests.

3.2.3. Receptive and expressive vocabulary

Patients displayed significantly lower results than their controls in their ability to recognize and express interpersonal relations, measured by the subtest "termination flexibility" (t (38) = 2.774, p = 0.008). They did not differ according to their EEG focus, AED-treatment, age of onset, seizure type and frequency and spike burden.

3.2.4. Receptive grammar

Neither significant differences between patients and controls in understanding the basics of logical-grammatical structures in recording prepositions (t (38) = 0.360, p = 0.721) and comparative constructions (t (38) = 0.668, p = 0.508), nor in the comprehension of grammatical structures (t (38) = 1.490, p = 0.144) could be found. Patients, however, showed lower results than their controls regarding the ability to analyze the emotional content of a statement; both the verbal and non-verbal elements (see Table 2). No differences according to their EEG focus, AED-treatment, age of onset, seizure type and frequency and spike burden could be found for these subtests.

3.2.5. Auditory discrimination

No significant difference between patients and controls could be stated according to auditory discrimination (t (38) = 0.448, p = 0.657). Once more, patients did not differ according to their EEG focus, AED-treatment, age of onset, seizure type and frequency and spike burden.

3.2.6. Auditory verbal memory

Patients did not differ significantly from the control children with regard to short-term memory for verbal material (t (38) = 0.605, p = 0.549) and there was also no significant difference concerning the consolidation of learning materials into long term memory (t (38) = 1.308, p = 0.199). Furthermore, groups did not differ in their ability to recognize memorized content (t (38) = 1.156, p = 0.255). Again no significant differences were found according to the EEF focus, AED-treatment, age of onset, seizure type and frequency and spike burden.

3.2.7. Reading and orthographic abilities

There was no significant difference in orthographic abilities between patients and controls (t (20) = 0.823, p = 0.420) and in reading abilities. However, children of the lower age group (6–11) tended to perform worse (6–11: t (18) = 1.769, p = 0.094) than children in the higher age group (11–14).

Even patients with bilateral foci produced no lower results in their synthetic reading abilities (subtest "word-dissimilar pseudo-words") (F (2, 17) = 4.033, p = 0.061).

In summary, significant differences between patient and control groups were found in expressive and receptive grammar: patients showed deficits in the derivation of adjectives and the correcting of semantically incorrect sentences, in the ability to analyze the emotional content of a statement and in recognizing and expressing interpersonal relations. According to auditory discrimination, auditory verbal memory and to reading and orthographic abilities, no differences were found.

Also, no further differences were found according to age (children ≤10 and those >10 years of age) or "spike burden" (SWI ≤25% versus >25%).

3.3. School performance and social competencies

For the semi-structured interviews with the teachers, four main categories – resulting from the procedure of inductive qualitative content analysis – were identified, namely (1) academic performance (referring to the major subjects German, English, Mathematics), (2) work habits, (3) social behavior and (4) disturbing behavior. Patients were considered more conspicuous concerning work habits, social behavior and academic performance, with teachers especially stressing the exhibition of disturbing behavior.

According to CBCL 4–18, the parents of children in the patient group indicated the presence of problems significantly more often than the parents of children in the control group (t (38) = 3.783, $p < 0.001$). Significantly more problems were indicated in the scales "Aggressive Behavior", "Attention Problems" and – only before correction – "Anxious/Depressed" (Table 3).

The results of the teacher interview showed that 35% of the patients' teachers, but only 13% of the teachers in the control group, indicated performance problems in the main subjects German, Mathematics and English.

Table 3
Child Behavior Checklist (CBCL) – T1 and T2.

	Patients (n = 20)		Controls (n = 20)		
	Mean	SD	Mean	SD	p
CBCL-T1					
Total problems	67.65	4.53	46.40	7.47	<0.001***
Aggressive	16.20	7.39	11.90	3.88	0.028*
Anxious/depressed	11.05	6.60	4.80	2.42	<0.001***
Attention problems	8.60	4.97	4.90	1.55	0.003**
Delinquent	6.60	3.97	5.65	2.94	0.395
Social problems	4.70	2.75	3.55	2.21	0.153
Somatic complaints	3.35	3.59	3.00	1.95	0.704
Thought problems	2.60	2.23	1.75	1.77	0.180
Withdrawn	6.80	3.14	6.25	2.29	0.531
CBCL-T2					
Total problems	47.75	6.39	46.45	7.47	0.558
Aggressive	12.15	2.38	10.90	2.88	0.817
Anxious/Depressed	4.89	1.85	4.60	2.22	0.896
Attention problems	5.75	2.36	4.40	1.75	0.186
Delinquent	6.25	3.97	5.65	2.94	0.605
Social problems	4.10	1.89	3.35	2.11	0.403
Somatic complaints	3.10	2.59	2.95	1.95	0.891
Thought problems	2.65	2.10	1.75	1.77	0.518
Withdrawn	6.85	3.26	6.25	2.29	0.505

Note. *$\leq$0.05 (not significant after Bonferroni correction, **$\leq$0.01 (not significant after Bonferroni correction), **$\leq$0.002 (significant after Bonferroni correction, ***$\leq$0.001 (significant after Bonferroni correction).

3.4. Results at T2

One year (12.4 $\pm$ 1.1 mo) after remission (determined by clinical aspects and the EEG), all patients were off medication. Not one child showed an atypical evolution (based on the observations of the regular clinical evaluation and EEG control recordings). A total of n = 7 (35%) children had to receive psychological interventions (neuropsychological training, behavioral therapy, etc.) due to their test results at T1.

At T2 no significant differences, either in total or on individual scales of the CBCL 8–14, were found compared to the scores of healthy controls.

Neither parents nor teachers complained about significant problems in school life or general education requiring any further intervention or treatment.

4. Discussion

The results concerning global intellectual functioning obtained in this study are in concordance with previous studies and demonstrate that children with active typical BCECTS have normal levels of intelligence, although on average their global IQ scores were slightly below those of a healthy control group. The lower Performance IQ scores in the patient group, but equal Verbal IQ scores, also correspond with the literature.[19,20]

Although not an issue of this study, we noticed a difference between patient and control group with respect to the subtests ''Block Design'', which measures spatial perception, and ''Object Assembly'', which examines ''visual analysis'', thus replicating and corroborating our previously published results in another sample of children with BCECTS.[19]

In contrast to the study results of Pinton et al.,[31] mild deficits in both receptive and expressive grammar and in vocabulary were found, especially with respect to the verbal expression of emotional relations: patients performed worse than their controls when analyzing the emotional content of a statement, both for verbal and non-verbal elements. As the Heidelberger Speech Development-Test (HSET) may be influenced by social skills, our result may correspond to that of Eggers[55] who described social withdrawal tendencies in children with active epilepsy.

Furthermore, and in contrast to the study performed by Northcott et al.,[26] our patient group performed more poorly in the derivation of adjectives and the correcting of semantically wrong/illogical sentences.

This stresses the importance of a differentiated neuropsychological examination which also takes into account complex speech abilities.

Contrary to our expectations, we were not able to demonstrate differences according to age (children $\leq$10 and those >10 years of age) which may be primarily due to the small sample size.

Finally, there was no difference between children with SWI $\leq$25% compared to those with >25%). In our opinion, this may be due both to the small sample size and to the selection criteria chosen for this study (SWI was below 45% in all children, there were no atypical features).

In accordance with some previous studies,[29,33] we found no significant differences between groups in auditory discrimination and in reading and orthographic abilities. Our study did not confirm the findings of Northcott et al.[28] which referred to deficits in verbal memory. The patient group did not exhibit any apparent differences compared to healthy controls with respect to verbal memory, auditory discrimination, simple grammatical structural forms and the repetition of single, phonemically similar words.

Our results from two studies in different samples with typical BCECTS suggest that during the active disease these children are prone to develop deficits in spatial-awareness,[19] whereas speech dysfunctions do not seem to be predominant.

An important study result is that parents of patients with typical (''uncomplicated'') BCECTS described their children as exhibiting attention problems and aggressive behavior more frequently (CBCL 4–18). Of special importance in this context is our finding that significant school and/or behavior problems were no longer reported after disease remission.

In contrast to the results published by Wirrell et al.,[56] who showed that six children with BCECTS who were treated with sulthiame showed a significantly reduced spike frequency associated with deterioration in cognitive functions, especially reading ability, general memory, attention skills and mathematical ability, our study showed no significant difference between patients with and without AED treatment concerning speech and school performance. Therefore, neither an improvement nor deterioration in the cognitive abilities examined can be explained as the result of AED treatment.

4.1. Limitations of the study

The study results are limited due to the relatively small sample size, which did not allow further stratification, and the highly selective group of school-age children with typical BCECTS (no atypical features, EEG grade 1 or 2, SWI < 50, no AEDs or moderate dosages of STM, complete recovery before age 16).

However, in our experience, this sample is representative for the vast majority of children with BCECTS, and further prospective long-term studies in well defined subgroups of the BCECTS spectrum will need to evaluate children with atypical features (i.e. EEG grades 3 and 4, SW index > 50%) compared to those with typical BCECTS. Another limitation is that no test data are available for the controls at T2, which does not allow for the assessment of any possible longitudinal confounding factors. As we tried to keep down the strain at least for the healthy children we decided to skip a complete re-examination for the controls at T2.

4.2. Possible clinical implications

(1) In children with typical BCECTS, spatial deficits seem to be of higher relevance than deficits in language and speech, although

longitudinal data are still missing. Maybe these deficits can be compensated when the child grows up, as no serious school problems were reported to the study authors at T2.

(2) Behavioral abnormalities, especially those concerning attention and impulse control, were only seen during the active period; the children's further development did not seem to be influenced negatively.

(3) We were not able to find any effects resulting from AED treatment with STM.

(4) It is important that children with typical BCECTS undergo regular clinical investigations (including parents' and teachers' reports about cognitive performance and behavior) in order to start necessary interventions as early as possible.

(5) Long-time prognosis, not only of seizures and EEG spikes but also of cognitive and behavioral abnormalities, seems to be excellent, and children with typical BCECTS should not be considered more ill than they are.

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