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Cognitive Epileptiform Disintegration in Children

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Abstract

Cognitive epileptiform disintegration is a specific disease associated with severe epileptiform activity on the EEG in the absence of epileptic seizures accompanied by different developmental cognitive disorders in children. The quantitative and qualitative characteristics of these developmental disorders have changed in the last 10 years. The article presents the analysis of cognitive epileptiform disintegration specificity and the results of the survey that included 57 children aged 2–7 with developmental disorders caused by epileptiform activity that was revealed during video EEG monitoring with sleep deprivation. The children were assessed with 19-channel EEG, synchronized with a video-monitoring system. Recording time was 1–2 hours. The psychometric methods used for the assessment of cognitive functions were three batteries of tests for children according to their age (2 years – MacArthur Communicative Development Inventories, 3–5 years – subtests created in Kazan' Federal University, 6–7 years – T.V. Akhutina's methods of neuropsychological diagnosis). The results of the research show that cognitive impairment in children with cognitive epileptiform disintegration aged 2–7 are complex, and predominantly they are the results of general developmental disorders associated with constant strong electrical impact on the functional blocks of the brain. Speech is most severely affected, both expressive and impressive, but speech disorders are always accompanied by the deficit of visual-spatial and kinesthetic analyzers, the deficit of the third functional block development and behavioral (predominantly autism-like) disorders.

Keywords: cognitive epileptiform disintegration, developmental disorders, speech disorders, video EEG monitoring, higher cortical functions, assessment of speech, assessment of cognitive functions

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1. Introduction

Cognitive epileptiform disintegration is a disease associated with severe epileptiform activity on the EEG in the absence of epileptic seizures accompanied by different developmental cognitive disorders in children, accompanied by speech and behavioral disorders. It is necessary to underline that cognitive epileptiform disintegration is a disease *without* seizures, and there is a significant difference between epileptic encephalopathy and cognitive epileptiform disintegration. The presence of single seizures in anamnesis of patients with cognitive epileptiform disintegration is allowed, but these single seizures must be carefully analyzed.

The mechanism of cognitive epileptiform disintegration is the pathological effect of prolonged epileptiform activity on higher mental functions. Deterioration of cognitive functions does not arise from birth, but appear in the process of brain maturation [1]. This problem is actively discussed in scientific literature from the end of the 20^{th} century. In 1997 R.F. Tuchman and I. Rapin presented a range of different types of cognitive disintegration in pediatric practice and drew attention to the frequent combination of disintegrative disorders with epileptiform activity on the EEG [2, 3]. In 2002 G. Gobby offered the term *cognitive epileptiform disintegration* for the acquired impairment of cognitive functions as a result of continuous epileptiform activity on the EEG [4]. K.Yu. Mukhin introduced this term into neurological practice in Russia [1], though some authors (i.e., L.R. Zenkov) offered the terms *epileptic brain dysfunction* and *acquired epileptic neuropsychological syndrome* [5]. The term "cognitive epileptiform disintegration" is widely used nowadays in the works of specialists in neurology and epilepsy [6–9].

Insufficient information on higher cortical functions development in cognitive epileptiform disintegration in Russia is due to the fact that EEG monitoring with sleep deprivation is not included into basic diagnostic protocols, and in most cases cognitive epileptiform disintegration is revealed accidently, sometimes very late or not revealed at all in the situation of severe mental retardation of development in a child. The presence of cognitive epileptiform disintegration influences sufficiently the tactics of treatment, because in this disease, a number of drugs that are commonly used in speech and cognitive disorders therapy are contraindicated. So, as a consequence, there are no universal protocols of cognitive functions assessment worked out for different age groups, rehabilitation programs also differ and are not specified for children with cognitive epileptiform disintegration. The laboratory of Clinical linguistics at Kazan' Federal University is working out the diagnostic materials which help

to assess the cognitive functions, especially speech, by means of collaboration of neuropsychologists, neurologists, specialists in epilepsy and clinical linguists.

2. Methodology

The research included instrumental (functional diagnosis) and psychometric (batteries of tests) methods. Children aged 2–7 with developmental problems were subjected to an electroencephalographic study according to a standard procedure using the international electrode arrangement "10–20" in bipolar installation. The recording of the electroencephalogram (EEG) was performed in wakefulness and daytime sleep on the 19-channel computer electroencephalograph EEG-A-21/26 Encephalan-131 M (research and production company Medikom-MTD, Taganrog). Recording time was 1–2 hours.

The development of cognitive functions was assessed with reliance on the principles of neuropsychological diagnosis worked out for children aged 6–7 by T.V. Akhutina and her colleagues [10] and with the help of subtests created in Kazan' Federal University (children aged 3–5). Little children (2–2.11) were assessed with the help of MacArthur Communicative Development Inventories (MacArthur CDI [11] – the Russian version by M.B.Eliseeva, E.A. Vershinina, V.L. Ryskina [12].

The research was conducted at A.Yu. Ratner Pediatic Clinical Hospital №8 (Kazan', Russia), the data were processed in the laboratory of Clinical linguistics (Kazan' Federal University).

During the period from August 2016 till February 2018 the authors observed 57 children aged 2–7 with developmental disorders caused by epileptiform activity which was revealed with the help of video EEG monitoring with sleep deprivation (9 children aged 2–2.11, 35 children aged 3–5.11, 13 children aged 6–7.11). The authors assessed impressive speech, expressive speech; functions of programming, regulation and control; visual gnosis; visual-spatial gnosis; functions of auditory analyzer; functions of kinesthetic analyzer; general adaptive skills; development of motor skills and hand motor skills; executive functions; emotional sphere and the specificity of behavior.

3. Results

The results of psychometric tests are summarized in Table 1 and Table 2, which show that predominantly the disorders are systemic, they do not touch upon speech development only – but speech suffers most of all. Cognitive impairments in general can

Table 1: Speech disorders.

Type of decline	Age	Degree of decline	Number	Percentage
Total absence of speech, both expressive and impressive	2–2.11	severe	6 of 9	66.7%
	3–5.11	severe	9 of 35	25.7%
	6–7.11	severe	2 of 13	15.4%
Total absence of expressive speech only	2–2.11	severe	1 of 9	11.1%
	3–5.11	severe	4 of 35	11.4%
	6–7.11	severe	1 of 13	76.9%
General speech impairment (in Russian terminology; international synonyms are *language delay* or *developmental dysphasia* [13. 14]	2–2.11	moderate	1 of 9	11.1%
	3–5.11	severe moderate mild	5 of 35 8 of 35 7 of 35	14.3% 22.9% 20%
	6–7.11	severe moderate mild	4 of 13 5 of 13 1 of 13	30.8% 38.5% 76.9%
Echolalia	2–2.11	severe moderate mild	3 of 9 2 of 9 2 of 9	33.3% 22.2% 22.2%
	3–5.11	severe moderate mild	6 of 35 6 of 35 8 of 35	17.1% 17.1% 22.9%
	6–7.11	severe moderate mild	– 2 of 13 5 of 13	– 15.4% 38.5%
Logorrhea	2–7.11	severe moderate mild	11 of 57 16 of 57 8 of 57	19.3% 28.1% 14%
Non-communicative speech	2–7.11	severe moderate mild + impossible to assess in non-speaking	11 of 57 6 of 57 2 of 57	19.3% 10.5% 3.5%
Delayed answers to the questions	2–7.11	moderate mild	9 of 57 2 of 57	15.8% 3.5%

be moderate or even mild, as well as behavioral disorders, but speech impairment is registered in 100% of the children from the observed group.

93.7% of children demonstrated at least two of these features:

1. non-stable response to the name (in the presence and in the absence of speech);

Table 2: Cognitive functions, motor and self-care skills.

Type of decline	Percentage
retardation of higher mental functions development:	
– functions of programming, regulation and control	71%
– visual gnosis	13%
– visual-spatial gnosis	52%
– functions of kinesthetic analyzer	43%
high index of hyperactivity and attention deficit	62.50%
deficit of major motor skills	33.60%
low level or absence of self-care skills (tidiness – visiting the toilet, washing, dressing, eating without help, etc.)	79%

2. verbal and non-verbal communication disorders (non-stable eye-to-eye contacts, a child is not able to play together with somebody, a child does not seek help from an adult);
3. autism-like games (infinite alignment of toys or other objects in rows, their sorting; the construction of "own worlds" from the objects, in which nothing can be changed; running around in a circle, jumping in one more than half an hour, etc.);
4. eating disorders;
5. stereotype behavior;
6. "childish" behavior (evaluated in children from 4 to 7 years old);
7. aggressive behavior (including self-aggression);
8. disorders of the emotional sphere;
9. absence of index gesture (in some cases the child takes the hand of an adult, in some cases the child points with the thumb or the entire palm).

4. Discussion

Cognitive epileptiform disintegration develops as a result of (a) genetically determined impairment of brain maturation processes; (b) disturbance of neuronal connections due to persistent continuous epileptiform activity on EEG; (c) morphological changes in the brain due to the pathology of prenatal development [1].

The age at which epileptiform activity first appears is very important. If this occurs at the very early stages of speech development, it leads to the total absence of speech as the process of brain maturation becomes perverted or even stops. If this occurs in a

child with partially developed speech, severe deterioration is usually not observed, but the development is significantly slowed down or suspended. The main features of EEG sleep in patients with cognitive epileptiform disintegration are: severe multiregional activity – benign childhood centrotemporal spikes (BECTS) – with a sharp increase in the phase of slow sleep and (or) frequent high-amplitude diffuse low-synchronized peak-wave discharges [1]. The index of epileptiform activity is no less important. If it reaches 50–85% of the sleep record, it significantly affects the possibilities of higher cortical functions development. The constant strong electrical impact on the functional blocks of the brain leads to their overexcitation, and then to the blocking [15].

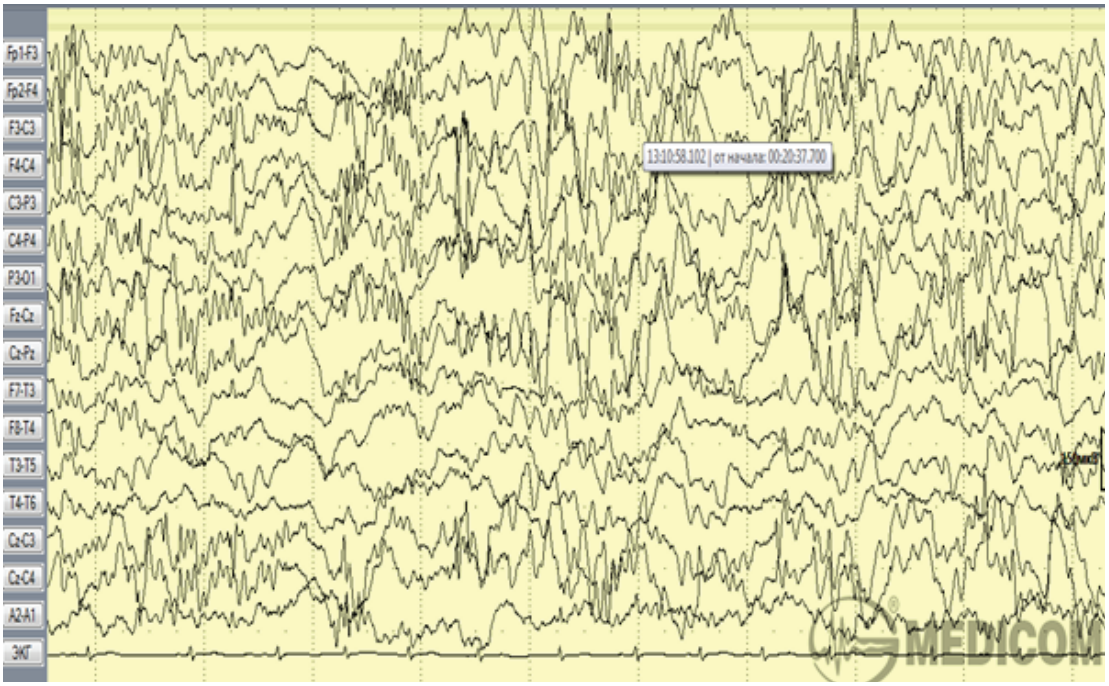

Figure 1: Patient V.D., 5 years old, sleep EEG.

For instance, a girl with the activity presented on EEG has total absence of impressive and expressive speech, psychomotor disorders, severe deficit of hand skills, severe spatial-visual disorders and specific "childish" behavior (behaves as a two-year-old baby). For three years there were no changes in her cognitive development (in the absence and in the presence of treatment) because of the strong diffuse epileptiform activity without seizures.

So, the registration of epileptiform activity – benign childhood centrotemporal spikes (BECTS) – on the EEG in combination with developmental problems can be a predictor of severe speech and cognitive disorders, and these children need adequate treatment. But the tactics of treatment is currently still discussible. The low effectiveness of oral antiepileptic drugs for continued epileptiform activity in the EEG and the

intrinsic inhibitory effect of antiepileptic drugs on cognitive functions require a careful selection of medications for this pathology. At the same time, the treatment should be prescribed timely in order not to miss the time of the formation of the speech function. Unequivocal is the decision to prescribe antiepileptic drugs in the presence of epileptic seizures and continued epileptiform activity. Indication for the appointment of antiepileptic drugs is also a combination of progredient cognitive impairment in combination with a high index of continued epileptiform activity. At the same time, in the absence of epileptic seizures with single epileptiform patterns on the EEG, the appointment of antiepileptic drugs can contribute to worsening of the child's cognitive performance.

5. Conclusions

We revealed typical for children with cognitive epileptiform disintegration speech and communication disorders, such as (a) total absence of speech, both expressive and impressive; (b) total absence of expressive speech only; (c) general speech impairment; (d) echolalia; (e) logorrhea; (f) non-communicative speech; (g) delayed answers to the questions.

The children with CED show deficit of programming, regulation and control, have a delay in the development of visual and visual-spatial gnosis, the deficit of auditory analyzer functions and kinesthetic analyzer functions.

93.7% of children with cognitive epileptiform disintegration demonstrate at least two features of autism-like behavior and behavioral disorders: (a) non-stable response to the name; (b) verbal and non-verbal communication disorders; (c) autism-like games; (d) eating disorders; (e) stereotype behavior; (f) "childish" behavior; (g) disorders of the emotional sphere; (h) absence of index gesture.

The results of our research proves A.Yu. Mukhin's cognitive epileptiform disintegration theory and show that cognitive impairment in children with cognitive epileptiform disintegration are complex, but the severity of speech disorders should be at the first place followed by the deficit of visual-spatial and kinesthetic analyzers, the deficit of the third functional block development and behavioral disorders.

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