

Benign rolandic epilepsy: neuropsychological findings

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Benign rolandic epilepsy (BRE) is a partial idiopathic epilepsy of childhood presenting with a nocturnal seizure and with a typical EEG showing centrotemporal spike and multifocal or generalized sharp slow waves. Although normal neurological and intellectual development are expected in BRE, it is not infrequent to detect subtle defects in neuropsychological functions and neuromotor development. This study included 20 cases of BRE diagnosed according to the criteria of ILAE. The patients underwent several tests of neuropsychological functions as well as detailed neurological examination and the results were compared statistically to normal controls. In the patient group, a family history of language delay or learning disability ($P < 0.005$), presence of consanguinity ($P < 0.05$), dyspraxia in the lower extremities (to imitation) ($P < 0.05$), difficulties in go-no-go test ($P < 0.001$), as well as some problems related to language such as dysprosody ($P = 0.05$), minor motor deficits in the left ($P < 0.05$) and right upper extremity ($P < 0.05$) were significantly more frequent compared to the control group. One should be rather guarded against the prognosis in BRE with respect to the higher cortical functions and neurodevelopmental problems.

Key words: neuropsychology; child; rolandic epilepsy.

INTRODUCTION

Benign rolandic epilepsy (BRE) classically occurs in neurologically and cognitively normal children who usually present with a nocturnal partial seizure and with a typical EEG showing centrotemporal and multifocal or generalized sharp slow waves¹. BRE accounts for between 6% and 16% of all non-febrile seizures in children². However, BRE does not constitute a serious problem as the seizures are rare and nocturnal.

Considering the clinical and EEG features, there seems a nosological boundary where Landau–Kleffner syndrome, epilepsy with continuous spikes and waves during slow sleep, atypical benign partial epilepsy and BRE merge into each other³. It is a fact that epileptic seizures in these syndromes are responsive to medical treatment with a benign and age-dependent course, while EEG abnormalities (with the exception of BRE) and developmental cognitive problems may well persist.

Therefore although normal neurological and intellectual development are accepted as criteria of BRE, it is not unlikely to find out some subtle cognitive

behavioral deficits. Indeed, there are few studies where atypical clinical and laboratory features and forms of BRE with some neuropsychological dysfunctions were reported^{4,5}.

This study aims to search some of these neuropsychological deficits in the BRE patients with emphasis on minor neurological dysfunction, visuo-spatial skills and frontal functions.

MATERIALS AND METHODS

The patient group (P) included 20 children, 13 males (65%) and 7 girls (35%) diagnosed as having BRE, according to the criteria defined by the international classification of epileptic syndromes (ILAE, 1989)⁶ and a control group which included 9 males and 6 girls. The mean age of the patients was 10.62, the age interval was 4.5–16. The control group consisted 5–14 patients with a mean age of 9.93. These patients were identified by reviewing all EEG reports obtained in the Neurology Department, EEG section of İstanbul Cerrahpaşa Medical Faculty between 1995–97. All patients who had a normal IQ were followed for at least 1 year with seizure control on carbamazepine

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during this time interval. The clinical parameters evaluated were detailed neurological examination including oromotor functions, minor neurological dysfunctions as defined by Touwen⁷ and Denckla⁸, praxis tests, frontal lobe tests, attention, visual-spatial tests as well as seizure variables and EEG features. Praxis evaluation included items for nonrepresentational movements, facial praxis, intransitive and transitive movements and ideational apraxia. Praxis tests are done to command, imitation, and where applicable, with objects^{9,10}.

Table 1: Items of assessment.

ITEMS OF MINOR NEUROLOGICAL DYSFUNCTION	
Astereognosia	
Agraphesthesia	
Adiadochokinesia	
Dysarthria	
Word finding difficulty	
Oro-motor apraxia	
Sialorrhea	
Change in jaw reflexes	
Change in deep tendon reflexes	
Asymmetry in deep tendon reflexes	
Asymmetry in the associated movements	
Hypokinesia	
Motor coordination deficits	
Changes in muscle tone	
Tremor	
Choreiform movements	
Nystagmus	
Strabismus	
PRAXIS EVALUATION	
Face	
to order	
to imitation	
to real object use	
Upper extremity	
to order	
to imitation	
to real object use	
Lower extremity	
to order	
to imitation	
to real object use	
INTELLIGENCE	
Cattell	
WISC	
FRONTAL LOBE FUNCTIONS (EXECUTIVE FUNCTIONS)	
COWAT	
Go-no-go test	
Luria's alternating sequence of drawing	
VISUO-SPATIAL PERCEPTION	
Raven's colored matrices	
LANGUAGE	
Articulation (a test developed by one authors)	
Prosody (clinical)	
Resonance (clinical)	
Vocabulary (Griffiths)	
Expression (grammar, fluency, spontaneity, picture telling) (Griffiths)	

The Cattell Culture Free Test¹¹ and WISC¹² were administered for IQ assessment. Language evaluation included screening of hearing and taped examination of oral-motor speech skills. Receptive and expressive language were assessed by items selected from Griffiths' test¹³. The articulation was tested according to a non-standardized test of articulation which included the main sounds of Turkish.

Attention was tested by backward naming of days of the week and of the months, counting after 20 in 3s, counting backward from 50 in 7s. Selective attention was assessed by asking the place of the things in the room and the color of them after eyes closed without prior warning. In the second test of attention, the child is asked to tap his finger every time A is repeated while the examiner recited a series of letters¹⁴.

Frontal tests included generation of word lists (COWAT: the number of animal names starting with a definite letter within one minute.)¹⁵, response inhibition (go-no-go test)¹⁶, and Luria's alternating sequence of drawing¹⁷. For complex visual functions, Raven's colored matrices¹⁸ were introduced. Items of examination and test battery are shown in Table 1.

All patients but five had sleep EEGs while all had awake EEGs. EEG recordings were evaluated by two independent raters, both neurologists. All patients had radiological evaluation, including 18 cranial MRIs and two cranial CTs.

The statistical evaluation consisted of comparison of groups according to the chi-squared test.

RESULTS

The seizures started earliest at the age of three, latest at the age of 12. In 15 patients (75%), seizure onset was at the age of between 3–10 while in five of them (25%) it was after age 10. No status epilepticus or Todd paresis are defined. Main features of seizures are shown in the Table 2.

All patients had at least one EEG characteristic for the BRE. The last recordings obtained included 15 patients with centrottemporal spikes, 10 (66.6%) had

Table 2: Seizure features.

	n of patients	(%)
Hemifacial clonic spasms	20	100
Speech arrest during ictal phase	19	95
Siyaloerrhea	19	85
Unilateral convulsions	15	70
Sensorial findings before the seizure	10	50
Dysphagia	8	40
Secondary generalization	6	30
Involvement of consciousness	6	30
Awakening with fear	5	25

Table 3: Comparison of patient and control groups.

Parameter evaluated	Patient (20)	Normal (15)	Chi-squared <i>P</i>	Fisher <i>P</i>
Family history of neurodevelopmental problem	11	1	chi-squared = 8.887 <i>P</i> < 0.005	
Consanguinity	6	0		<i>P</i> < 0.05
Dyspraxia in the leg to imitation	6	0		<i>P</i> < 0.05
Vocabulary	12	2	chi-squared = 7.77778 <i>P</i> < 0.01	
Dysprosody	8	1		<i>P</i> < 0.05
Failure in go-no-go test	11	0		<i>P</i> < 0.001
Minor motor deficit in the upper right extremity	6	0		<i>P</i> < 0.05
Minor motor deficit in the upper left extremity	7	0		<i>P</i> < 0.05

right, five (33.3%) had left sided while five had bilateral paroxysmal changes. The number of spikes and other sharp elements per minute in awake EEGs were calculated as 1.05–34.80 while in non-REM II they were found to be 3.97–39.90.

Considering developmental milestones, one patient (5%) had a delay in speech onset, four patients (20%) had a delay in walking, nine patients (45%) had a delay in bladder training. Family history included six patients (30%) with epilepsy, six patients (30%) with febrile convulsions and 10 patients (50%) with a language delay or learning disability. Five patients (25%) had parental consanguinity, 18 patients (90%) had term birth, three cases (15%) had a birth weight under 2.5 kg.

Psychiatric evaluations of all patients, which were performed by a child psychiatrist, were normal. Academic achievement as determined according to the school marks (except two patients who had not started primary school) showed that 11 had good school success, three were in the medium range, four failed. In the control group eight had good success while six had medium success. Two children were given WISC-R, 18 children had the Cattell A-B culture free test. Three (15%) patients had high intelligence while 17 (85%) had normal findings. IQ test scores were between 88–165. All patients in control group had a normal IQ. As regards some of the language features, naming and articulation were normal in all. The mean value of word generation during 1 minute was 13.4. Three children (15%) had grammar disorder, four (20%) had dysfluency in speech, four had problems in spontaneous speech (20%), nine (45%) had difficulties in picture telling, three (15%) had resonance deficit, 11 (55%) patients had slightly limited vocabulary problems.

Seventeen patients (85%) had right hand dominance while three patients (15%) had left hand dominance. In one or more of the praxia items, 15 patients (75%) were unsuccessful. Praxis tests performed to command were unsuccessful in 14 (70%) patients while there were some problems in 11 patients (55%) when they were

performed to imitation and nine patients (45%) had dyspraxia when real objects were used. Considering Test of Alternating Sequence of Drawing was normal in all patients. Ten patients failed in Raven's coloured matrices. Eleven (55%) patients were unsuccessful in go-no-go test.

In the patient group, a family history of language delay or learning disability (*P* < 0.005), presence of consanguinity (*P* < 0.05), apraxic deficits in the lower extremities (*P* < 0.05), difficulties in go-no-go test (*P* < 0.001) were significantly more frequent compared to the control group. In addition to that, language problems related to prosody (*P* < 0.05), vocabulary (*P* < 0.001) and minor motor deficits in the right upper extremity (*P* < 0.05) and left upper extremity (*P* < 0.05) were significantly high in the patient group. Significant findings as regards comparison of normal and patient groups are shown in the Table 3.

DISCUSSION

Although epilepsy is a chronic condition, in most cases seizures are either responsive to treatment or remit spontaneously. The overall impact of epilepsy on the developing brain is still unknown^{19,20}; however, there is some evidence that subtle effects of seizures on brain, regarding neurocognitive development may occur even in benign idiopathic partial epilepsies^{21–24}.

In this study, some neurological and neuropsychological dysfunctions such as language problems related to vocabulary and prosody and frontal dysfunction (go-no-go test, motor deficits, dyspraxia) and a family history of consanguinity and neurodevelopmental problem were detected in the patient group. While the latter two findings may be due to the possibly genetic inheritance of this epileptic syndrome, motor and language problems may be interpreted as related to a likely neuropathology in the sylvian area in BRE.

The results indicated that BRE has some phenotypic variability regarding the neuropsychological status oc-

curing in a range of 5%–75% of patients in one of the tests administered. Indeed, Doose and Baier have suggested the BRE has a multifocal pathogenesis with diverse phenotypes²⁵; which might be the basis for an explanation of the deficits found in these patients.

The study had some limitations regarding the insufficient number of patients as well as the lack of control group which did not include age- and sex-matched normal subjects. Indeed, the main shortcoming of the study was the lack of standardized neuropsychological tests in Turkey so that the results may be criticized as not having enough reliability and validity although most test items applied actually required clinical assessment by scoring. Obviously, the main difficulty was with the language tests as there have been no standardized test for the Turkish language hitherto.

As some atypical clinical and EEG findings might be more frequent than expected in BRE, BRE may be accepted as a heterogeneous syndrome. Therefore it seems that the term benign should be limited to seizure prognosis and global neurological status in BRE while one should be guarded against the neuropsychological and neurodevelopmental deficits.

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