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## West syndrome and pschycomotor retardation

### To the Editor,

We have read the research titled “Autistic disorder in West syndrome” by Haçerli et al. (1) with interest. As mentioned in the introduction of the article, the three main characteristics of West syndrome include :1) infantile spasms, 2) regression and retardation in psychomotor development and 3) appearance of hypsarryhtmia on electroencephalogram. It is understood that regression and retardation was present in psychomotor development in 100% of 267 subjects who were diagnosed with West syndrome and in 90 subjects who were included in the research from this group. In approximately 95% of the subjects, seizures began in the first 12 months of life and 90 of these were examined after the age of three years. Psychomotor retardation was found in 78 subjects (87%) as a result of examination performed by the Denver developmental screening test. Thus, the rate of regression and retardation in psychomotor development which was 100% at the baseline was found to be decreased to 87% in approximately two years. Improvement of the psychomotor state with treatment in a severe condition like West syndrome is a significant success.

It is known that cases with seizures assessed to be infantile spasms which are one of the three characteristics of West syndrome used to be divided into three groups (symptomatic, cryptogenic and idiopathic). Currently, however, the cases are classified mainly in two groups (symptomatic and cryptogenic) (2). The subjects in whom the cause leading to seizures can be demonstrated and/or who have significant developmental retardation before seizures start are qualified as symptomatic subjects. The subjects in whom the cause is not known and who have normal development before seizures start are named as cryptogenic subjects (2,3). Approximately 20% of the patients are in the cryptogenic group and 80% are in the symptomatic group. The patients in the cryptogenic group have a better general well-being (4). Only 16% of the subjects with infantile seizures who had a mean follow-up period of 31 months had normal development in 67

studies published (3). In another article, it was reported that retardation in development was observed with a rate of 50% even in cryptogenic subjects who responded to treatment in a short time (5).

It is known that medical and surgical treatment in epilepsy may affect cognition positively (6-8). In fact, it was reported that even epilepsy treatment administered in patients who had cognition disorders and EEG disorders without seizures had useful effects on cognition (9). Improvement in cognition in infantile spasm has not been mentioned much except for papers which reported improvement in cognition and behavior with vigabatrin treatment, when the cause of infantile spasm was tuberous sclerosis (10). Therefore, these results in the study of Haçerli et al. (1) are pleasing in terms of the prognosis of the disease, though the reasons were not expalined.

In addition, it will be useful to correct the contradictions related with some information in the “Summary” and “Results” parts of the study. In the summary, it was reported that psychomotor retardation was found in 86% of 267 subjects with West syndrome. On the other hand, it was stated that psychomotor retardation was found in 78 (87%) of a total of 90 subjects (not 267 subjects) with the Denver developmental screening test in the “Results” part.

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#### Dear Editor;

We thank to Dr. Betül Aydın for her contribution to our article. In the study we performed in 2007, the files of 267 patients who presented to Istanbul University, Istanbul Medical Faculty, Division of Pediatric Neurology between 1995 and 2007 and were diagnosed with West syndrome were examined retrospectively. The diagnosis was made with the ages of the patients, description of seizures by the families and/or observation of seizure during examination and hypsarrhythmia finding on EEG. Electroencephalograms were taken by Micromed 18 channel digital video EEG device found in the Division of Pediatric Neurology using international 18-20 electrode system. Electroencephalograms were evaluated by the teaching staff of the Division of Pediatric Neurology.

At the first presentation, the age, gender, age at presentation, follow-up period, prenatal and postnatal histories, psychomotor development stages, physical and neurological examinations, spasm forms, cranial imagings and laboratory findings of the subjects were examined. Metabolic screening tests (tandem ms, urinary organic acids by thin layer chromatography, biotinidase screening test), biochemical tests (thyroid hormones, serum ammonia and lactate levels) were performed. Among cranial imaging methods, computed tomography (CT) and/or magnetic resonance imaging (MRI) were performed. The psychomotor development of the subjects was evaluated by clinical findings and Denver developmental screening test.

The subgroups of West syndrome were determined according to the 1991 ILAE (International League Against Epilepsy) classification. The patients whose psychomotor development was normal, whose head circumference percentiles were within normal limits and whose neurological findings, laboratory tests and radiological assessments were normal before the spasms started were included in the cryptogenic WS group. It was understood that 43 of 267 patients with WS who were examined retrospectively were lost. When 267 patients were evaluated after the follow-up period (1 month-180 months, mean: 12 months), it was found that spasms stopped in 27.3%, spasms and/or other seizure types continued in 56.5% and 16.1% were lost. The mortality rate was found to be 8% in cryptogenic subjects and 16.8% in symptomatic subjects.

The psychomotor development is poor in West syndrome (1-3). In our study, the patients were assessed by the Denver developmental screening test and psychomotor retardation was found in 232 patients (86%) and normal psychomotor development was found in 32 patients (2%). Considering that 91% of our patients were symptomatic, these data were found to be compatible with the literature.

It was understood that 43 of 267 patients with WS who were examined retropectively were lost. Among 224 patients, autism behavioral checklist was applied in a total of 90 patients who could be reached and presented to our outpatient clinic above the age of three after obtaining consent from the families. 57 items were asked to the mother and/or father of these patients by the same person (assistant researcher). In the evaluation of a total of 90 patients by the Denver developmental screening test, psychomotor retardation was found with a rate of 87% (78 patients). 100% of our patients with a high potential of autism had a score above 67. However, this result was not statistically significant ( $p:0.08$ ,  $p>0.05$ ).

In our study, autisms was considered with a high probability in 17 (18.9%) patients who scored 68 and above among 90 patients who were tested. When we added 14 patients who scored between 54 and 67, a high rate of 34.5% was reached. However, it is known that prevalent characteristics specific for autism may be present in a portion of the individuals with a score between 54 and 67 or these individuals may have high function. Clinical evaluation should be performed according to diagnostic test results, the developmental history of the child and observational data. Assessment should be repeated at regular intervals especially in children with a borderline score. These children who require a more detailed examination were referred to pediatric psychiatry outpatient clinics.

In West syndrome, autistic disorder is observed with a high rate which can not be ignored. Since autism is a life-long disorder, the type of treatment varies according

to the age and development of the individual. As a result of many studies conducted on autism, the best treatment was found to be education. Therefore, it is very important to start education at the earliest age possible. Therefore, patients with West syndrome should be screened during their follow-up in terms of autistic disorder. Patients with a high and borderline potential of autism should be referred to pediatric psychiatry. In addition, more studies on neurological diseases accompanying autism are needed.

“Multi-center” institutions related with divisions of pediatric neurology in faculties which can be directive in any area required by children with West syndrome are needed. Medical care, education and rehabilitation of the child will be provided by the teams and assistants found in such centers.

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