Clinical, neuroimaging and developmental outcomes of West syndrome

Abstract. Background. West syndrome is characterized by a hypersarrhythmia pattern on electroencephalography, spasm type seizures, and psychomotor regression triad. In this study, we aimed to document demographic characteristics, clinical and laboratory findings, treatment responses, neurodevelopmental outcomes, and risk factors developed during long-term follow-up with the diagnosis of West syndrome. Materials and methods. The study included patients who were diagnosed with West syndrome between July 2011 and December 2012 in the Department of Pediatric Neurology of Atatürk University. The following data were collected from each patient’s history. Systemic and neurological examinations, cerebral imaging, and electroencephalography were reviewed. Biochemical tests were performed from laboratory tests. The development of each child was assessed using the Denver 2 developmental screening test, the Ankara Developmental Screening Inventory test. Results. The ratio of male/female of our patients was found 2.28/1 and the mean age of the referral was 8.62 ± 7.20 months (median: 8.0). 59 (85.51 %) of the patients were in the symptomatic group and 10 (14.49 %) were in the idiopathic group. The most common factors in the symptomatic group were anomalies of the congenital central nervous system (45.7 %) and hypoxic-ischemic encephalopathy (28.8 %). There was a significant difference between the idiopathic and symptomatic groups in terms of relapse rates, radiological findings, and prognosis rates (p: 0.035/p < 0.001/p < 0.001). Relapse was detected in 43.5 % of the patients. While 83 % of patients treated with adrenocorticotropic hormone, treatment was responded to, 17 % had resistant seizures. There was no statistically significant difference between the treatment responses with adrenocorticotropic hormone and other treatments (p = 0.093). 46 of 55 children (83.6 %) were found to be a developmental delay. Conclusions. Early diagnosis and treatment, the provision of appropriate and convenient treatment in the West syndrome can make a positive contribution to prognosis according to etiology. Keywords: West syndrome; infantile spasms; developmental delay; epilepsy
three drugs has so far been proven; corticotropin (ACTH),
vigabatrin, and corticosteroid. The first choice is adreno-
corticotropic hormone (ACTH) or oral steroids, and the
second choice is vigabatrin or other antiepileptic drugs. In
cases resistant to these three drugs used as the first choice,
pyridoxine phosphate, valproic acid, topiramate, clobazam,
zonisamide, levetiracetam, the ketogenic diet can be added to
the treatment [7–9].

The purpose of this study is to document demographic
characteristics, clinical and laboratory findings, treatment
responses, neurodevelopmental outcomes, and risk factors
developed during long-term follow-up with the diagnosis of
WS.

Materials and methods

Patients who were diagnosed with WS between July 2011
and December 2012 were included in the study. Ethical
approval has been obtained for the use of file information.
The following data were collected from each patient: ages,
gender, findings associated with spasms, seizure frequency,
duration of follow-up, prenatal, natal, and postnatal stories,
family history. Systemic and neurological examinations, ce-
rebral imaging (brain magnetic resonance imaging (MRI)
and/or computed tomography (CT) and laboratory fin-
dings, and EEG were reviewed. Biochemical tests (uric acid,
creatine kinase, blood urea nitrogen, creatinine, thyroid
hormones, serum ammonia, lactate) were performed from
laboratory tests.

The development of each child was assessed using the
Denver 2 developmental screening test, the Ankara De-
velopmental Screening Inventory test. Following the onset
of spasms, follow-up types, neurological examination, de-
velopmental tests, applied treatments, and side effects and
neurological sequelae were recorded. Age at onset of spasms
are classified: early-onset: < 3 months (adjusted age for pre-
mature), classical onset: 3 months to 12 months, late-onset:
12 months and later.

Etiological classification

The patients are divided into two groups according to a
definable cause: the symptomatic group and the idiopathic
group. The patient’s head circumference measurements were
assessed according to age and gender. The time between the
start of treatment and the delay of treatment was recorded.
The time between the first observation of the spasms and the
beginning of the treatment was considered as early if it is
lower than 2 months and as late if it is more than 2 months.

Treatments. Adrenocorticotropic hormone (ACTH), vi-
gabatrin (VGB), and corticosteroids are the first-line treat-
ments for WS. Synthetic ACTH preparation (syna-
chent depot® ampoule) was administered 0.5 mg twice a week
for younger than 12 months or under 10 kg; 1 mg twice a week
for older than 12 months or more than 10 kg, intramuscularly
for 12 weeks. Vigabatrin (75–100 mg/kg/day) was started for
those who were unable to respond and who had tuberous scle-
rosis in the etiology. When side effects of ACTH treatment
resulted in hypertension, infection, hyperglycemia, ACTH
was discontinued and continued treatment with clona-
zapam. In cases of resistant hypertension, frequent recurrent
infections, ACTH therapy was discontinued and vigabatrin
therapy was started. Treatment response was evaluated by
clinical and EEG findings.

Clinical response. Spasms were defined as disappearance
within 14 days of initiation of treatment and not to repeat
spasms at least 28 days after the last spasm.

EEG response. The EEG took approximately 4 weeks
after treatment; was defined as complete disappearance of
hypsarrhythmia, followed by any spasm period seen as a
cluster after the recovery of the spasms, and observation of
vague spasms accompanied by EEG changes. The prognosis
of the patients was evaluated according to their seizure, mo-
tor, and cognitive/neuromotor developmental status after at
least 12 months of follow-up. The absence of seizures du-
during the last 6 months, the ability to have motor skills, and
developmental characteristics consistent with the patient’s
age, was evaluated as a good prognosis.

Statistical analysis

SPSS for Windows 21 package program was used for sta-
tistical analysis of the data. Mean ± standard deviation for
data with appropriate distribution in the study and median
(minimum–maximum) for data with inadequate distribution
were specified. The chi-square test was used for the com-
parison, the t-test was used for the independent samples, and
Pearson correlation analysis was used for the correlations.
A p-value of less than 0.05 was considered statistically sig-
nificant.

Results

Clinical and demographic profile

A total of 69 children were screened. Forty-eight (69.6 %)
were male and twenty-one (30.4 %) were female. The mean
age of the referral was 8.62 ± 7.20 months (median — 8.0).
Fifty-six children (81.6 %) were delivered at term. Eighteen
children (26.1 %) were small for gestational age. Table 1
summarizes the features of our cases.

Table 1. Demographic characteristics of patients
with West syndrome

|                      | Total, n (%) | Female, n (%) | Male, n (%) | Age of onset of seizures in months, Mean ± SD | Referral age, month, Mean ± SD | The time until starting treatment in month, Mean ± SD |
|----------------------|--------------|---------------|-------------|---------------------------------------------|-------------------------------|---------------------------------------------------|
| Total, n (%)         | 69 (100)     | 21 (30.4)     | 48 (69.6)   | 6.8 ± 4.9                                   | 8.62 ± 7.20                   | 3.90 ± 6.09                                       |

Note: SD — standard deviation.

Seizure profile at onset. The average age at seizure onset
was 6.8 ± 4.9 months. The mean time between the onset
of seizures and the initiation of treatment was 3.90 ± 6.09
months (table 1). West syndrome seizures appeared as a
single or cluster. It was observed that 27.5 % (19/69) of
the patients presented as single seizures and 72.5 % (50/69)
of them cluster seizures. Spasm count was at least 2/day and
at most 35/day, 43 patients (62.3 %) had flexor, 19 patients
(27.5 %) mixed, and 6 patients (8.7 %) had extensor-type
spasms. In most patients, seizures were seen while waking up or awake (table 2).

**Neuroimaging profile.** While symptomatic causes were considered in 60 % of cases before cranial imaging, an increase was observed in the number of symptomatic cases with cranial imaging. Cranial imaging studies were performed at least once in all the patients. 55 (80 %) patients had MRI and others had CT. While pathology was detected in 52 (75 %) of them, imaging was normal in 17 patients (24.6 %). The cranial imaging findings of the patients are shown in table 3.

Mean EEG recovery time was 1.17 ± 1.43 months in the symptomatic group and 2.00 ± 2.88 months in the idiopathic group. There was no significant difference between idiopathic and symptomatic groups in terms of incidence of EEG types ($p = 0.308$).

**Table 2. Seizure characteristics of patients with West syndrome**

| Number of seizures | n = 69 | %   |
|--------------------|--------|-----|
| Per day 1–5 seizures | 37     | 53.6|
| In a day > 5 seizures | 13     | 18.8|
| One intermittent seizure | 19     | 27.5|

**Seizure type**

| Flexor | 43 | 62.3 |
| Extensor | 6 | 8.7 |
| Mixed | 19 | 27.5 |
| Other types spasms | 1 | 1.4 |

**Seizure features**

| Awake | 25 | 36.2 |
| In sleep | 5 | 7.2 |
| While about to wake up | 28 | 40.6 |
| Awake + in sleep | 11 | 15.9 |

**Table 3. Cranial imaging findings of patients**

| Indicator | n = 69 | %   |
|-----------|--------|-----|
| Normal | 17 | 24.6 |
| Cortical atrophy | 12 | 17.4 |
| Subdural effusion | 5 | 7.2 |
| Delay in myelination | 4 | 5.8 |
| Lissencephaly | 4 | 5.8 |
| Cystic encephalomalacia | 4 | 5.8 |
| Periventricular leukomalacia | 3 | 4.3 |
| Secondary changes in hypoglycemia | 2 | 2.9 |
| Subdural hygroma | 2 | 2.9 |
| Arachnoid cyst | 2 | 2.9 |
| Cerebral, lacunar infarct | 2 | 2.9 |
| Previous intracranial hemorrhage | 2 | 2.9 |
| Band heterotopia | 1 | 1.4 |
| Corpus callosum agenesis | 1 | 1.4 |
| Corpus callosum disгенезис | 1 | 1.4 |
| Cortical tuber | 1 | 1.4 |
| Dysmyelinizing disorder | 1 | 1.4 |
| Bazel ganglion lesions | 1 | 1.4 |
| Hypoxic ischemic encephalopathy | 1 | 1.4 |
| Cerebral edema | 1 | 1.4 |
| Chiari malformation | 1 | 1.4 |
| Hydrosefalia | 1 | 1.4 |

**Notes:** CT — computed tomography; MRI — magnetic resonance imaging.
Etiology

59 (85.51 %) of the patients were in the symptomatic group and 10 (14.49 %) were in the idiopathic group. Congenital CNS malformations (45.7 %) and sequential HIE (28.8 %) were the most frequent causes in the symptomatic group (table 4). There was a significant difference between the idiopathic and symptomatic groups in terms of relapse rates, radiological findings, and prognosis rates (p: 0.035/ p < 0.001/p < 0.001). In the idiopathic group, the prognosis was better, relapse rates were low, and radiological findings were less. The prognosis was better in the advanced EEG findings group (p < 0.001). There was no difference between idiopathic and symptomatic groups in terms of seizure type distribution (p: 0.721).

Table 4. Etiological causes of patients in West syndrome

| Etiology                                      | n=59 | %    |
|----------------------------------------------|------|------|
| HIE sequelae                                  | 17   | 28.80|
| CNS malformations                            | 27   | 45.7 |
| Intracranial hemorrhage, effusion             | 7    | 11.8 |
| CNS infections                                | 6    | 10.16|
| Perinatal factors                             | 6    | 10.16|
| Neurocutaneous diseases                       | 3    | 5.08 |
| Congenital infections                         | 3    | 5.08 |
| Bilirubin encephalopathy                      | 2    | 3.38 |
| Intracranial cyst, tumor (glial tumor)        | 2    | 3.38 |
| Cerebral damage secondary to hypoglycemia     | 2    | 3.38 |

Notes: HF — hypoxic ischemic encephalopathy; CNS — central nervous system.

Treatment profile

Before the onset of ACTH therapy, 52 % of patients used antiepileptic drugs, the most frequent was phenobarbital (26.1 %). While 83 % of patients treated with ACTH, treatment was responded, 17 % had resistant seizures. In 64 patients treated with ACTH, side effects developed in various clinical forms. The most common side effect was restlessness. There was no statistically significant difference between the treatment responses with ACTH and other treatments (p = 0.093).

Early response was assessed as partial or complete loss of seizures after ACTH treatment at 1 week. 44 patients (63.8 %) had an early response to ACTH treatment. The prognosis was much better in the patients that started an early treatment (p < 0.001). Relapse was found in 43.5 % of patients after treatment. Clinical response to ACTH treatment was found to be effective regardless of etiology.

Developmental outcomes

55 children were assessed using the Denver 2 developmental screening test, the Ankara Developmental Screening Inventory test [10, 11]. Denver II consists of 134 items evaluating four developmental areas: personal-social, fine motor-adaptive, language, and gross motor. After calculating the age of the child, it is seen what skills the child should be able to draw by drawing the age line, the tester evaluates the compliance of the child’s development with the age.

Ankara Developmental Screening Inventory test provides an opportunity to identify developmental delay and to recognize the developmentally at-risk babies and children at an early stage and take early interventions. It consists of 154 questions asked to primary caregivers about the development of children. For the other patients for whom these tests could not be performed, clinical observations and family information were used. In our study, 46 (83.6 %) were found to be a developmental delay.

Discussion

A comprehensive clinical evaluation, EEG evaluation, cranial and genetic and metabolic tests aid early diagnosis. In 277 incidence series of Lombroso [12], the incidence of WS cases with an onset age of 12 months or more was reported as 10 %. Consistent with the literature, the rate of spasms that started after 12 months in our study was 12.5 % and the age of onset of WS was between 3 months and 12 months in 67.9 % of the patients. In our study, patients applied to a health center after an average of 3.9 months after the spasms started. In developed countries, the time from the onset of spasms to diagnosis is reported between 1 and 1.5 months [13–15], while in developing countries this time can be extended to 7.9 months [16]. In our study the percentage of males was 69.56 % and the male: female ratio was 2.28: 1. As reported in our literature, WS is reported to be more frequent in males [5, 16, 17].

In WS, the difference between the symptomatic and idiopathic groups is of practical importance. This classification helps to predict prognosis. Patients in the idiopathic group respond very well to treatment and their prognosis is much better. In our study, the symptomatic group rate was higher (85.51 %). Congenital CNS malformations (45.7 %) and sequential HIE/perinatal asphyxia (28.8 %) were the most frequent causes in our study. While perinatal asphyxia is the most common cause in developing countries, perinatal causes such as cortical malformation, neurocutaneous diseases, genetic-metabolic diseases occur in developed countries. The most common risk factor is perinatal asphyxia in our study. In Gupta et al study, asphyxia and other adverse perinatal events are the most common causes [18]. Perhaps this will change with the improvement of prenatal services, institutional distribution, guidance system, and perinatal care [19].

Seizures may be hardly noticeable in the form of a slight, or in the form of a tonic contraction that can last for 10 seconds following a rapid spike of 1–2 seconds [2]. Sometimes there is no tonic phase seen afterward. As the number of seizures can be 1–2 per day, the number of seizures can be up to 100. The duration of seizure clusters may vary from 1 minute to 10 minutes. In our cases, flexor spasm was observed in 62 % of the patients, extensor spasms were seen in 8.7 % and mixed type spasms were observed in 27.5 % of the patients. In the study of Yilmaz et al. [17] studied 79 % of patients with flexor spasms that is similar to our study.
One of the diagnostic parameters of WS is hypsarrhythmia in the EEG. In our study, there was 43.6% classic hypsarrhythmia and 11.6% modified hypsarrhythmia in EEG. EEG findings were consistent with the literature [20]. We observed hypsarrhythmia or modified hypsarrhythmia in EEG (respectively 81.3, 83.3%) in the symptomatic group. In some idiopathic cases in the literature, the first EEG may be normal or borderline abnormal. In this case, the EEG is required to be withdrawn after 7–10 days [1]. The higher incidence of EEG pathology in the symptomatic group may be associated with the late manifestation of EEG findings in idiopathic cases. We observed 40% of other seizure types in our study. The most common was myoclonic seizures of the newborn period with 20.3%. The most common abnormal neurological examination findings were tonus changes, mental retardation, increased deep tendon reflexes, and spasticity. The cerebral palsy rate was 47.3% at admission. The literature has also been reported between 44 and 67% [17, 18].

Imaging studies have significantly contributed to the etiology of WS. Abnormal imaging findings are associated with 80% of WS patients. In literature, cerebral atrophy is the most commonly identified, whereas our study rate was 74.6% similar to the literature. As is known, the prognosis in the symptomatic group is poor. Structural cerebral anomalies have been reported to significantly affect the prognosis adversely [21]. It is not surprising that the prognosis is poor in this group since the presence of abnormal neurological findings indicates that the cases are symptomatic. Tuberous sclerosis and epilepsy are frequently encountered in studies related to etiology [22]. In our study, tuberous sclerosis was seen in 3.89%.

Seizures in WS are generally resistant to antiepileptic drugs. The most important cause of the difficulties in treatment is the differences in etiology. ACTH is the best medication proven in treatment. Vigabatrin is preferred in cases with tuberous sclerosis and in cases where ACTH is unresponsive [8]. Lux et al. received treatment response in 76% of patients receiving ACTH [2]. Hrachovy et al. reported that 75% of patients using ACTH received a response within the first 2 weeks and relapsed in 31% of long-term follow-up [23]. In our study, the response rate was 83% and the relapse rate was 40%. 63.8% of the patients had an early response to ACTH treatment. In our study, we found that there was no significant difference in response to ACTH and vigabatrin treatment modalities (p > 0.05) as in the study of Ibrahim et al. [24].

Idiopathic and symptomatic groups had similar responses to ACTH treatment. In both groups, spasms were diminished or disappeared at similar rates. In our study, the rate of response to treatment was high at the early onset of ACTH treatment. Partikian and Mitchell reported that 159 WS patients who were followed for at least one year were seen resistant epilepsy in 53.2% [25]. In our study, it was 49.2% were seen as resistant to epilepsy. Koo et al. found that the rates of conversion to neurological sequelae and other epilepsy in the cryptogenic group were lower than in the symptomatic group [26]. In our study, the symptomatic group prognosis (resistant epilepsy recurrence, psychomotor retardation, neurological deficit) and relapse rates were significantly higher than in the idiopathic group (p < 0.001/ p = 0.035/p < 0.001). The epileptic prognosis was worse in relapsed patients after treatment (p = 0.029). The treatment response did not have any effect on relapse and epileptic transformation.

Hrachovy and Glaze [27] reported that spasms were accompanied by autonomic and non-spasm findings at varying rates. In our study, 42% of the patients had abnormal eye movements and 34.8% of the patients had pre-seizure tremors; were the most common accompanying symptoms.

46 of 55 children (83.6%) had developmental delay. Consistent with our study, Gupta et al. found that 90.1% of children had a developmental delay [18]. In the study of Yilmaz et al. With 269 patients, the developmental prognosis of the cases was assessed by Ankara Developmental Screening Inventory; It was shown that 40 (19%) patients had borderline, 91 patients (42%) had mild/moderate delay and 57 patients (28%) had severe development delay [28]. The presence of developmental delay at first admission in WS is a poor prognostic criterion. Developmental delay is more prominent in those with a young onset age of spasm. In our study, intellectual disability was detected in 54.2% of patients in the symptomatic group in which ACTH treatment was started early and in 74.2% of those who were late in the treatment. In a study of Nasiri et al., 67 patients with WS was associated with poor neurodevelopmental outcome. Most developmental delays have also been shown to be due to the presence of symptomatic WS and resistant seizures [29]. Singh and Ray reported 46.7% cerebral palsy in the 165 WS patients [30]. The incidence of cerebral palsy in our study was 37.2% in cases with neurological sequelae. This difference may be since our records are not well maintained, especially in terms of cerebral palsy.

**Conclusions**

As a result, WS not only causes resistant seizures but also causes severe neurological sequelae affecting the mental, motor, psychological and sensory development of patients. Spasms can cause more damage to the developing central nervous system. Treatment can reduce psychomotor regression. However, there is currently no optimal treatment for WS. Optimal treatment should be done with at least current treatment protocols. For this reason, early diagnosis, appropriate and adequate treatment may be a positive contribution to the prognosis. Early control of spasms has a positive effect on intellectual development.

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Синдром Веста: клінічні, нейровізуалізаційні симптоми та показники розвитку

Резюме. Актуальність. Синдром Веста характеризується картиною гіпсаритмії на електроенцефалографії, судомами спазмового типу і триадою психомоторної регресії. Метою цього дослідження було визначення демографічних характеристик, клінічних та лабораторних показників, відповіді на лікування, впливу на формування нервової системи та факторів ризику, що розвинулися протягом тривалого спостереження пацієнтів із діагнозом синдрому Веста.

Матеріали та методи. У дослідження були включені пацієнти, у яких був діагностований синдром Веста в період з липня 2011 року по грудень 2012 року на кафедрі дитячої неврології університету Ататюрка. З анамнезу кожного пацієнта були зібрані наступні дані. Були розглянуті показники загального та неврологічного дослідження, церебральної томографії та електроенцефалографії. Біохімічні аналізи проводилися за результатами лабораторних досліджень. Розвиток кожної дитини оцінювався за допомогою тесту для скрінінгу на розвиток Denver 2, тесту для контролю та скрінінгу розвитку Ankara.

Результати. Співвідношення хлопчиків і дівчаток серед наших пацієнтів становило 2,28 : 1, а середній вік становив 8,62 ± 7,20 місяця (медіана: 8,0). 59 (85,51 %) пацієнтів були в симптоматичній групі й 10 (14,49 %) — у групі з ідіопатичним генезом захворювання. Найбільш частими факторами в симптоматичній групі були врожені аномалії центральної нервової системи (45,7 %) і гіпоксично-ішемічна енцефалопатія (28,8 %). Спостерігалася вірогідна різниця між ідіопатичною та симптоматичною групами за частотою рекуренцій, рентгенологічними даними та прогнозом (р = 0,035/p < 0,001/p < 0,001). Рецидив спостерігався в 43,5 % хворих. Серед пацієнтів, які отримували адренокортикотропний гормон, 83 % особи відповіли на лікування, у 17 % реєстрували резистентні судоми. Статистично значущою різницею між відповідями на лікування адренокортикотропним гормоном та іншими методами лікування не було (р = 0,093). У 46 з 55 дітей (83,6 %) була виявлена затримка розвитку. Висновки. Рання діагностика і лікування, надання належного і зручного лікування при синдромі Веста можуть чинити сприятливий вплив на прогноз за рахунок етіології. Ключові слова: синдром Веста; інфантильні спазми; затримка розвитку.