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Neurodevelopmental outcomes of the West syndrome in pediatric patients: The first report from the Middle-East

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ABSTRACT

Background: This study aimed to investigate the clinical characteristics and neurodevelopmental outcomes of children with West syndrome (WS) by using the Bayley-III scale of infant development, as the first report from the Middle-East.

Methods: Between January 2013 and February 2016, we prospectively enrolled 67 consecutive patients with a confirmed diagnosis of WS from Isfahan, Iran. Cognition, language and motor outcomes of the studied subjects were evaluated with the Bayley Scales of Infant and Toddler Development-Third Edition (Bayley-III).

Results: Overall, 67 cases, including 34 (50.7%) boys and 33 (49.3%) girls (a male/female ratio of 1.03), were enrolled for the study. The mean age was 26.7 ± 12.9 months. Among the subjects, 50 (74.6%) patients had symptomatic WS, and 17 (25.4%) patients had cryptogenic WS. "Severe delay" was found in 76.9% of the patients regarding cognitive evaluation, 67.7% for language and communication abilities, and, 81.5% for motor function. The patients with cryptogenic WS were significantly more likely to have more favorable outcomes in motor ($p = 0.035$), cognitive ($p = 0.035$) and receptive language ($p = 0.043$) in comparison to those who had symptomatic WS. The patients with controlled seizures were significantly more likely to have more favorable outcomes in motor ($p = 0.027$) and cognition ($p = 0.011$) as compared to those with uncontrolled seizures.

Conclusion: WS was associated with poor neurodevelopmental outcome in our study. Severe developmental delay was associated with two major factors: (i) presence of a specific underlying abnormality (symptomatic WS) and(ii) persistent seizures as a result of the former.

What this paper adds?

Developmental outcome in patients with West syndrome, in addition to underlying abnormality (symptomatic WS), greatly depends on the persistence of seizures. Effective treatment is the most important and even in most of these children, the only way to prevent developmental deterioration.

1. Introduction

Infantile spasm is a convulsive disease characterized by sudden bilateral symmetrical contraction of the muscles of the neck, trunk

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and extremities (Pavone, Falsaperla, Ruggieri, Praticò, & Pavone, 2013). Infantile spasm, also known as West syndrome (WS), was originally described by Dr. William West, who observed the seizures in his son in 1841. The clinical presentation of WS includes the triad of infantile spasms, neurodevelopmental regression, and a characteristic electroencephalogram (EEG) known as hypsarrhythmia (Wong & Trevathan, 2001). Its incidence is estimated to be between 0.25 and 0.60 per 1000 live births annually, and the prevalence ranges from 0.14 to 0.52 per 1000 children in different studies. The onset is predominantly in the first year of life, with the maximum incidence reported between 3 and 7 months of age (Pavone, Striano, Falsaperla, Pavone, & Ruggieri, 2014). Based on the suspected etiology, WS can be divided into that of symptomatic origin and non-symptomatic (cryptogenic) origin. A diagnosis of symptomatic WS is made when a specific underlying etiology including brain structural abnormality, hypoxic-ischemic brain injury or metabolic disorders can be identified. On the other hand, when no identifiable cause is found in a child with developmental delay or other neurologic impairment before the onset of spasms, the term cryptogenic WS is used (Dulac, 2001; Taghdiri & Nemati, 2014).

The overall long-term prognosis of WS is poor as most of the underlying etiologies are generally associated with great risk of neurodevelopmental delay and mortality. Several neurodevelopmental areasincluding cognitive, behavioral and motor domains might be affected. Visual and auditory deficits can be found in 30–50% of the affected children (Turkdogan, Us, & Akyuz, 2003). Mental retardation is observed in 70% of patients, which is often associated with psychiatric problems including autistic spectrum disorder and attention deficit hyperactivity disorder. Seizures may persist until adulthood, and 50–70% of the patients may develop other types of seizures (Riikonen, 1982). Unfavorable prognostic factors include the onset of the disease earlier than three months, the presence of other types of seizures before the onset of spasms, and symptomatic etiology. On the other hand, onset at more than four months of age, the absence of partial seizures and cryptogenic etiology are suggested to be associated with a favorable outcome (Hrachovy & Frost, 2003).

To our best of knowledge, there are limited recently published studies in the literature on

the comprehensive evaluation of neurodevelopmental domains including cognition, memory, language and motor development in children with WS. This study aims to investigate the clinical characteristics and neurodevelopmental outcomes of WSby using the Bayley-III scale of infant development, as being the first report from Middle-East.

2. Materials and methods

2.1. Study population and setting

BetweenJanuary 2013 and February 2016, we prospectively enrolled 67 consecutive patients with a confirmed diagnosis of WS, who were referred to Emam-Hossein University Hospital in Isfahan, Iran. This hospital is the leading tertiary care pediatric neurology center for pediatric neurology in Isfahan province. The patients inclusion criteria were: (i) confirmed diagnosis of WS according to“International League Against Epilepsy” (ILAE) definitions (Berg et al., 2010) and based on the presence of infantile epileptic spasms, hypsarrhythmia or modified hypsarrhythmia on EEG and developmental regression; (ii) being lower than 42 months of age. The age limit of “up to 42 months” was considered in the present study since the Bayley-III scale is designed to evaluate the developmental abilities of children from birth up to 42 months of age (Bayley, 2006).

A qualified pediatric neurologist performed neurological examination. The following data were collected from each patient: age, gender, and the age of onset for infantile spasms, age at diagnosis of WS, the presence of developmental delay before the onset of spasms and the presence of other types of seizures before the onset of spasms. All the subjects underwent brain magnetic resonance imaging (MRI) and biochemical screening (blood and urine) for inborn errors of metabolic diseases. Patients with a definite underlying etiology were classified the symptomatic WS and in cases in which the medical history and clinical evaluations did not lead to a known etiology,classified with symptomatic WS.The study did not interfere with normal clinical management and treatments.The patients were treated according to our Institution’s protocol, which consisted of vigabatrin, nitrazepam, and pyridoxine. Each patient had at least one monthly visitduring which the investigator assessed his/her response to the treatments and investigated the developmental outcomes based on the study tools.For patientswho did not have an appropriate response to the mentioned treatments within three weeks,ACTH was commenced. The appropriate response was defined as the termination of spasms and normalization of EEG. The protocols of the study were completely explained and written informed consent was obtained from parents/caregivers. The study protocol was approved by the ethical committee of Isfahan University of Medical Sciences.

2.2. Research tools and techniques

Cognition, language and motor outcomes of the studied subjects were evaluated with the Bayley Scales of Infant and Toddler Development–Third Edition (Bayley-III) (Bayley, 2006). The Bayley-III is one of the most commonly used tests for assessing the developmental function of infants and children in the age group of 1–42 months. It provides standardized measures of infant development in five areas: cognition, language, motor skills, social-emotional, and adaptive behaviors. It typically takes 30–90 min to complete the Bayley-III scale depending on the children’s age and level of function. Each test subscale produces a total raw score based on the number of items given; the raw score is then converted to a scaled score based on normative data. Each scaled score has a mean of 10and a standard deviation (SD) of 3. The scaled scores for the five subscales are transformed into composite scores. Normal within 1 SD of the mean (≥ 85),mild developmental delay in any of the three areas is defined as a composite score of -1 to -2 SD (≥ 70 and < 85), moderate delay as a score between -2 to -3 SD (≥ 55 and < 70), and severe delay as greater than 3 SD below the mean standard scores (< 55). We assessed the cognitive, language, and motor scales by direct testing of the infants. The social-emotional and adaptive behavior scales were filled via face-to-face interviews with parents or primary caregivers. All the mentioned

Table 1
Sample sizes, means, SD, and 95% confidence intervals and median scores and ranges.

	N	Mean	SD	95% CI	Min	Median	Max
Cognitive							
Raw	67	25.0	23.5	19.97–31.62	0	25	79
Scale	67	2.75	3.12	1.98–3.52	1	1	12
Composite	67	63.7	15.61	59.90–67.63	55	55	110
Language: receptive and expressive							
Scale	67	7.78	6.79	6–9.46	2	5	27
Composite	67	64.21	20.18	59.21–69.21	46	56	121
Receptive language							
Raw	67	12.1846	10.43	9.5–14.7	0	12	41
Scale	67	4.03	3.42	3.21–4.97	1	3.00	14
Expressive language							
Raw	67	11.38	10.26	8.84–13.92	0	9	43
Scale	67	3.69	3.60	8.73–10.43	1	2	17
Motor: fine and gross							
Scale	67	5.6	6.69	19.71–23.65	2	2	31
Composite	67	56.84	20.05	51.87–61.81	46	46	133
Fine motor							
Raw	67	17.44	15.74	13.54–21.34	00	15	59
Scale	67	2.80	3.27	1.98–3.61	5	1	15
Gross motor							
Raw	67	21.24	21.9	15.81–26.67	0	17	70
Scale	67	2.8	3.57	1.91–3.68	1	1	17

scales were administered by a pediatric neurologist.

2.3. Statistical analyses

The quantitative and categorical data were presented as mean (SD)/or median (minimum-maximum) and frequency (percentage). The normality of data was evaluated by using the Smirnov-Kolmogorov test and Q-Q plot. The normality distributed data were compared betweenthe groups by using independent samples *t*-test and the non-normal data by using the Man-Whitney U test.

Statistical analyses were performed with SPSS version 20 (SPSS Inc., Chicago, IL, USA) and a $P < 0.05$ was considered as the significance threshold.

3. Results

3.1. Clinical and demographic data

For our prospective data collection over a period of three years,67 patients with a confirmed diagnosis of WS were recruited. They consisted of 34 boys (50.7%) and 33 (49.3%) girls (male/female ratio of 1.03). The mean age was 26.75 ± 12.9 months (range 7-42months). The mean age at the onset of spasms was 7.2 ± 6.3 months (range 1–34 months). Developmental delay prior to the onset of spasms was present in 39(58.2%) patients. Table 1 summarizes the features of our cases.

3.2. Etiology of west syndrome

Overall, 50 (74.6%) cases had symptomatic WS,including 15 (22.4%) patients with hypoxic-ischemic encephalopathy, 13 (19.4%) patients with brain dysgenesis, six (7.5%) patients with neurometabolic disorders, five (7.5%) patients with tuberous sclerosis complex, five (7.5%) patients with genetic abnormalities, three (4.5%) patients with TORCH-related infections, and three (4.5%) patients with brain hemorrhage. A definite etiology could not be ascertained for the remaining 17 (25.4%) patients, who were classified as having cryptogenic WS.

3.3. Bayley-III developmental assessments

Regarding the cognition scale, eight (12.3%) patients were within the normal range. Two (3.1%) patients scored 1–2 SD below the mean (mild delay);five (7.7%) patients scored 2–3 SD below the mean (moderate delay), and 52 (76.9%) patients scored lower than 3 SD below the mean (severe delay). In the language scale and communication abilities, six (9.2%) patients scored within the normal limits, six (9.2%) patients had a mild delay, nine (13.8%) patients had a moderate delay,and 46 (67.7%) patients had a severe delay. Motor scale assessmentrevealed thatfour (6.1%) patients had normal scores, three (4.6%) patients had a mild delay,five (7.7%) patients had a moderate delay, and the remaining 55 (81.5%) patients had a severe delay.

Table 2
Developmental outcome in cryptogenic and symptomatic infantile spasm.

	Cryptogenic N = 17	Symptomatic N = 50	P
Cognitive			
Raw mean(SD)	35.3(29.3)	22.4(20.7)	0.050
median(min–max)	44(0–79)	23(0–77)	
Scale mean(SD)	4.1(3.7)	2.3(2.8)	0.035
median(min–max)	2(1–10)	1(1–12)	
Composite mean(SD)	70.5(18.6)	61.6(14.0)	0.035
median(min–max)	60(55–100)	55(55–110)	
Language: (receptive and expressive)			
Scale mean(SD)	10.0(7.4)	7.1(6.5)	0.109
median(min–max)	9(2–21)	5(2–27)	
Composite mean(SD)	70.5(18.6)	61.3(13.7)	0.110
median(min–max)	60(55–100)	55(55–110)	
Receptive language			
Raw mean (SD)	18(12.7)	10.1(8.7)	0.006
median(min–max)	19(0–41)	10.5(0–33)	
Scale mean(SD)	5.5(4.4)	3.5(3.0)	0.043
median(min–max)	6(1–14)	2.0(1–12)	
Expressive language			
Raw mean(SD)	15.4(13.1)	9.95(8.7)	0.059
median(min–max)	10(0–43)	9.0(0–30)	
Scale mean(SD)	4.47(3.4)	3.41(3.6)	0.304
median(min–max)	3.0(1–12)	2.0(1–17)	
Motor: fine and gross			
Scale mean(SD)	8.5(8.8)	4.5(5.4)	0.031
median(min–max)	3.0(2–31)	2.0(2–29)	
Composite mean(SD)	65.7(26.6)	53.6(16.3)	0.032
median(min–max)	49(46–133)	46(46–127)	
Fine motor			
Raw mean(SD)	21.9(17.2)	15.8(15.0)	0.173
median(min–max)	20(0–59)	13.5(0–58)	
Scale mean(SD)	3.7(3.8)	2.4(3.0)	0.160
median(min–max)	2.0(1–14)	1.1(1–15)	
Gross motor			
Raw mean(SD)	31.0(26.6)	17.79(19.0)	0.032
median(min–max)	34(0–70)	15.5(0–58)	
Scale mean(SD)	4.8(5.1)	2.1(2.5)	0.06
median(min–max)	2(1–17)	1(1–14)	

3.4. Factors predicting favorable outcome

The patients with cryptogenic WS were significantly more likely to have a more favorable outcome in the scale ($p = 0.031$) and for composite fine & gross motor ($p = 0.032$), cognitive ($p = 0.035$) and receptive language ($p = 0.043$) in comparison with those who had symptomatic WS (Table 2).Epileptic seizures were under control in 11 (64.7%) patients in the cryptogenic group and persisted in the remainingsix (35.3%). The patients with controlled seizure more likely to have favorable outcome in the general/overall motor sub-scale (fine and gross combined, $p = 0.027$) and within the fine (0.031) and gross (0.026) sub-scales specifically and cognition ($p = 0.011$) in comparison to those who had uncontrolled seizures (Table 3).

Girls showed better performance in neurodevelopmental assessments in motor (6. 6 ± 24.7 vs. 4.5 ± 4.4), language (66. 36 ± 22.4 vs. 62 ± 17.6), and cognition (66.9 6 ± 18.1 vs. 60.4 ± 11.8); however, the difference did not reach the significant threshold ($p > 0.05$).

4. Discussion

To the best of our knowledge, our study is the first report from the Middle-East that presents a comprehensive evaluation of the neurodevelopmental outcomes of children. Based on our observations, a vast majority of the children with WS had a poor developmental outcome.Notably, “severe delay” was found in as high as 76.9% of the patients in cognitive evaluation, 67.7% of the patients in language and communication abilities, and, 81.5% in motor function assessment. The high frequency of abnormal development observed in the present studyis comparable with the data available in the literature. In a systematic review byHrachovy et al. (Hrachovy & Frost, 2003) reported that roughly 16% of children with WS had normal development. In a retrospective study from Belgium on children with WS, up to 75% of the subjects had a delay in their psychomotor developments (Lagae et al., 2010). A sizeable population-based survey on children with WS syndrome in Atlanta, USA, revealed that 83% of the patients had varying degrees of mental retardation (i.e.,IQ score < 70) (Trevathan, Murphy, & Yeargin-Allsopp, 1999). Finally, a retrospective analysis of patients from the UK reported that 87.5% of subjects had abnormal neurodevelopmental outcome (Mohamed, Scott, Desai, Gutta, &

Table 3
Developmental outcome in cryptogenic IS with controlled and uncontrolled seizure.

	Cryptogenic Seizure not controlled N = 6(35.3%)	Cryptogenic seizure under control N = 11(64.7%)	P
Cognitive			
Raw mean (SD)	19.5(24.2)	22.4(20.7)	0.101
median (min–max)	9(0–51)	23(0–77)	
	N = 17	N = 48	
Scale mean (SD)	1.1(0.4)	2.3(2.8)	0.011
median (min–max)	1(1–2)	1(1–12)	
Composite(SD)	55.8(2.0)	44.0(29.1)	0.011
median (min–max)	55(55–60)	54(00–79)	
Language: (receptive and expressive)			
Scale mean (SD)	5.5(5.3)	12.5(7.3)	0.059
median (min–max)	2.5(2–15)	11(2–21)	
Composite mean(SD)	70.5(18.6)	61.3(13.7)	0.059
median (min–max)	60(55–100)	55(55–110)	
Receptive language			
Raw mean (SD)	8.1(11.4)	19.4(12.2)	0.542
median (min–max)	9(0–34)	22(0–41)	
Scale mean (SD)	3.5(3.8)	6.7(4.4)	0.160
median (min–max)	1(1–9)	6(1–14)	
Expressive language			
Raw mean (SD)	8.1(11.4)	19.3(12.6)	0.093
median (min–max)	4.5(0–30)	22(0–43)	
Scale mean (SD)	2.0(2.0)	5.8(3.4)	0.024
median (min–max)	1.0(1–6)	5.0(1–12)	
Motor: fine and gross			
Scale mean (SD)	2.3(0.5)	12(9.4)	0.027
median (min–max)	2.0(2–3)	9(2–31)	
Composite	47.0(1.5)	76(28.4)	0.027
	46(46–49)	67(46–133)	
Fine motor			
Raw mean (SD)	8.5(12.4)	29.2(15.2)	0.012
median (min–max)	2.5(0–31)	33(0–59)	
Scale mean (SD)	1.1(0.4)	5.1(4.0)	0.032
median (min–max)	1.0(1–2)	4(1–14)	
Gross motor			
Raw mean (SD)	9.8(20.2)	42.5(22.7)	0.010
median (min–max)	1.5(0–51)	53(4–70)	
Scale mean (SD)	1.1(0.4)	6.8(5.4)	0.026
median (min–max)	1(1–2)	5(1–17)	

Patil, 2011).

In the present study, we observed better developmental and cognitive outcomes among children with cryptogenic than those with symptomatic WS. This observation was in line with the results of several studies in this field (Lagae et al., 2010; Riikonen, 2001, 2010), including the systematic review by Hrachovy et al. (Hrachovy & Frost, 2003),which suggested thatchildren with cryptogenic WS had the most favorable prognostic factor for developmental outcomes. However, this notion has not been confirmed in other similar studies in this field including a survey among American children in which the authors observed no difference in developmental outcomes between children with cryptogenic and those with symptomatic WS (Trevathan et al., 1999). Currently, there is an ongoing debate in the literature regarding the extent of the investigations needed to be carried out to confirm the diagnosis of cryptogenic WS in patients for whom history, examination, and imaging have failed to identify an underlying etiology. At present, there are at least 200 suggested etiologies associated with WS in the literature (Riikonen, 2010); however, investigations for many of these etiologies require expensive or invasive methods. It is clear that as the diagnostic methods become more precise, the proportion of cryptogenic WSwould decrease. Therefore, the discrepancies in extant studies might probably be due to methodological, definitional, and diagnostic differences.

Recently, Osborne et al. (2010) took an interesting and refreshing approach to classify WS and challenged the traditional terminology of “cryptogenic” and “symptomatic”WS. They introduced a novel classification system: “proven etiology” (if a specific underlying disease was identified),“no identified etiology” (if no evidence of underlying disorder was found after investigation), and “not fully investigated” (if a major piece of patients data was missing or not available). They argued that this new classification system would enable better comparison of the different groups of patients, better understanding and analysis of the results of trials and of cohorts, and also better evaluation of response to a particular therapy. Taken altogether, as proposed by Appleton (2015), as more of the “iceberg-tip of cryptogenic”of WSwould melt with advancements in molecular genetics and higher-resolution neuroimaging techniques, more light would be shed on the prognostic value of the etiology of WS in the evaluation of the disease outcome.

The treatment protocol in our study consisted of vigabatrin, nitrazepam, and pyridoxine,which was started immediately after diagnosis for all the patients. For the patients who did not respond to the first line treatment within three weeks, ACTH was started. Based on our treatment results, we found that patients with controlled seizures showed better motor and cognitive outcomes than the

patients with uncontrolled spasms. Similar results were reported by Mohamed et al. (2011) and Matsuo et al. (2001), who indicated that children who responded to first-line treatments had a more favorable long-term developmental outcome than the other groups. Several authors (Hamano et al., 2007; Karvelas et al., 2009; Kivity et al., 2004) have also addressed the early treatment of infantile spasms and shorter treatment lag after the diagnosis was associated with more favorable long-term cognitive outcomes.Since rapid standard treatment was started for most of our patients, our study was not conclusive enough regarding the effects of delay in treatment on the neurodevelopmental outcome of these patients.

5. Conclusion

In conclusion, our study is the first report on neurodevelopmental outcomes of Iranian children with WS. WS was associated with poor neurodevelopmental outcome among the studied subjects. Developmentwas associated with two major factors: (i) presence of a specific underlying abnormality (symptomatic WS) and (ii) persistent seizures. Further studies are needed to answer crucial issues affecting neurodevelopment outcome in this population.

Disclosure

The authors have no proprietary interest in the materials presented here.

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