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Clinico-aetiological Profile and Outcome of West Syndrome from North India

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Abstract ***Aim*:** West syndrome is an age dependent epileptic encephalopathy and there is paucity of data regarding its clinic-aetiological profile. ***Methods*:** We studied 76 patients of West syndrome, carrying out a detailed aetiological work-up of all. ***Results*:** Majority of patients were males (57/76), having flexor infantile spasms (66/76), usually occurring in series (45/76) and seen mostly on awakening (63/76). We found a good seizure outcome in 40.8% and a good development outcome in 8.1% cases. ***Interpretation*:** West syndrome in India also, has almost a similar clinico-etiological profile as in other parts of the world where it has been studied.

Key words Clinico-aetiological; Epileptic encephalopathy; Infantile spasms; Profile; West syndrome

Introduction

West syndrome is an age dependent epileptic encephalopathy characterised by a clinico-electrical triad: infantile epileptic spasms, arrest or regression of psychomotor development and hypsarrhythmia on EEG (Electro-encephalogram), although the latter element maybe missing.[1]

The present study was done to study the clinico-aetiological profile of children, with West syndrome, attending a tertiary care hospital in North India and to study the factors determining its outcome.

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Participants and Methods

Consecutive children aged 3 months-5 years, attending the general paediatric outpatient clinic or Paediatric Neurology clinic or admitted to the paediatric wards of a tertiary care teaching hospital, over a period of 1 year, who satisfied the inclusion criteria, comprised the study population. Inclusion criteria comprised the presence of any 2 of the following; presence of infantile spasms, presence of hypsarrhythmia on EEG and psychomotor retardation or regression (as used by Kalra et al[2]). Ethical approval was taken by the institutional ethics committee.

Details of clinical history, physical examination, investigations and follow-up were recorded on a pre-designed data collection proforma, (after taking informed consent), which was updated on every visit/telephonic interaction with the patient. Socioeconomic status was judged by the Kuppuswamy scale.[3] Valproate was used as the first line drug, while ACTH (Adreno-corticotropic hormone) or prednisolone were used as the second line and Lamotrigine/Topiramate were used as the third line drug.

Neuro-developmental assessment was done at the time of enrollment and at the end of 6 months. Standard psychometric tests like Developmental Scale for Indian Infants (<3 years of age),[4] Binet Kamat test (3-5 years of age) and Vineland Social Maturity Scale, were used for the

same and they were administered by a trained child psychologist, initially and at the end of 6 months. Primary outcome measure was seizure outcome at 6 months of starting treatment. Outcome was considered good if there was more than 80% control of spasms at the end of 6 months without relapse or occurrence of other seizure types. Secondary measure was neurodevelopmental status at 6 months and was considered good if both DQ (Developmental Quotient) and SQ (Social Quotient) were more than 70. Neuroimaging and EEG (Electroencephalogram) were done.

Statistical Analysis

Statistical analysis was done using Epi info software version 3.5.1. Mean and standard deviation were calculated for the numerical data.

Results

A total of 76 patients were enrolled in 12 months. Of these 69, were freshly diagnosed cases. Majority of patients had urban residence (55.3%), belonged to the middle socio-economic class (63.2%) and were delivered by normal vaginal delivery (81.6%). Clinical characteristics of the patients are as mentioned in Table 1. Major pre-disposing adverse event was perinatal asphyxia (64.4%). Aetiological profile of patients was as given in Tables 2 and 3.

Forty-nine patients who completed 6 months follow-up irrespective of the therapy taken, were studied for final outcome. Of these only 20/49 (40.8%) patients had a good seizure outcome and only 4/49 (8.1%) had a good developmental outcome, at the end of 6 months of therapy. Details of the 4 patients with good developmental outcome have been mentioned in Table 4. Twenty-two of 76

Table 1 Clinical profile of patients with West syndrome

Factor		**Mean**	**Standard deviation (SD)**
Age at onset (in months)		5.69	5.96
Age at presentation (in months)		15.44	11.31
Delay in presentation (in months)		9.88	9.47
Clinical feature		**No. of patients (n=76)**	**Percentage**
Male sex		57	75
Weight for age	Wasted	49	64.5
	Normal	26	34.2
	Overweight	1	1.3%
Abnormal motor examination		61	80.3
Microcephaly		45	59.2
Any other dysmorphic feature		17	22.4
Motor development at presentation	Normal	6	7.8
	Delay	66	86.8
	Regression	4	5.2
Mental development at presentation	Normal	16	21.1
	Delay	56	73.7
	Regression	4	5.3
Social development at presentation	Normal	17	22.4
	Delay	55	72.4
	Regression	4	5.3
Types of spasms	Flexor	66	88
	Extensor	6	8
	Mixed	2	2.7
Features associated with spasms	None	36	47.3
	Cry	36	47.3
	Laugh	4	5.2
Spasms on awakening		63	82.9
Spasms in series		45	59.2
Other seizures at presentation		22	28.9

(28.9%) patients developed other seizure types during the period of study.

Discussion

The study comprised of relatively larger number of patients compared with most of the other studies; 47 children by Matsuo et al,[5] 26 children by Sharma and Vishwanathan[6] and 28 children by Goldstein and Slomski.[7] Kaushik et al[8] did a retrospective study of 148 patients admitted over a duration of 4 years.

The mean age at onset of spasms was almost similar to that found in other studies. It was 5.69 months compared with 5.5 months in a study by Cohen-Sadan et al.[9] The mean delay in presentation of patients to us (9.88 months) was, however, found to be significantly more. This is because majority of our study population is illiterate. They resort to superstitious practices before presentation to a tertiary care hospital. Because of the nature of the seizures, they often go unrecognised by the family unless the physician specifically asks for 'startling'.

Perinatal insults formed the major group of aetiology compared with the major group of aetiology compared with the antenatal and postnatal timimg of insult. This has been the conclusion in some of the previous studies as well (Matsuo et al,[5] Kalra & Passi,[10] Mackay et al,[11] Tsuji et al,[12] Kaushik et al[8]). A slightly larger number of patients were found to have symptomatic aetiology, 88.2% compared with 83% by Matsuo et al[5] and 66% by Kalra & Passi.[10]

We inquired about typical and atypical features of infantile spasms in our patients. Typically, infantile spasms are brief jerks – either flexion or extension type which occur in series, specially on awakening. Not all patients have typical spasms and such spasms were seen in 73% of patients, percentage being similar to the one observed by Singhi and Ray[13] of 74%.

Twenty-two of our initial 76 patients (28.9%) developed other seizure types during the period of follow up. These were generalised tonic seizures, atypical absence and minor motor type seizures or any combination of these. Other types of seizures may occur upto 2 years of initial treatment of West syndrome. Hence, the actual number of patients who might have developed other seizures cannot be commented on, since the total duration of the study was 1 year.

We used valproate for management of infantile spasms as has also been done by a few previous studies (Seimes et al,[14] Pratz et al,[15] Ohtsuka et al[16]). A relatively larger percentage of patients, 59.1% patients, were found to have a good seizure outcome at the end of 6 months follow-up. Singhi and Ray,[13] found complete cessation of seizures in 42.4% patients, but the duration of follow-up has not been specified by them. Thus, our study protocol was useful in determining the final seizure outcome. Some of this better outcome as seen in our study may be accounted for by the use of valproate as the first line treatment. Kalra et al[2] found a good final seizure outcome in 36.6% in those treated with hormonal therapy. Kaushik et al[8] found favourable outcome in 45 (30.4%) children with spasm cessation rate of 25.4% with prednisolone.

Table 2 Aetiological profile of patients with West syndrome

Aetiology	Number (n=76)	Percentage (%)
Prenatal	3	3.9
• Tuberous sclerosis	1	1.3
• Pachygyria	1	1.3
• Corpus callosal agenesis	1	1.3
Perinatal/Neonatal	55	72.3
• Perinatal asphyxia	31	40.3
• Low birth weight	19	25
• Neonatal illness	42	55.2
Poat neonatal	10	13.1
• Meningoencephalitis	8	10.5
• Head injury	2	2.6
Cryptogenic	8	10.5

Table 3 Developmental profile of patients with good neuro-developmental outcome at 6 months follow-up

Patient	Development Quotient (Motor)		Development Quotient (Mental)		Social Quotient		Neuroimaging
	On admission	On follow-up	On admission	On follow-up	On admission	On follow-up	
1	66	74	65	70	68	106	Normal
2	66	71	66	78	54	83	Normal
3	68	84	67	83	57	81	Normal
4	65	78	65	86	60	98	Normal

The developmental outcome was very poor in both symptomatic and cryptogenic West syndrome. The mean developmental quotient (DQ) in all patients was 25, and only 4 patients (11%) had a normal DQ (>70%). However, since the period of follow-up was only 6 months, much importance cannot be given to this.

Limitations

A still larger number of patients and a longer period of follow-up, should be studied to make any definite conclusion about the outcome, specially developmental outcome.

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Conflict of Interests

None

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