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Original article:

Study of clinical profile of West syndrome in India

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Abstract:

Introduction: West syndrome is a constellation of symptoms characterized by epileptic/infantile spasms, abnormal brain wave patterns called hypsarrhythmia and intellectual disability. West syndrome is a type of epilepsy that affects babies. This condition is rare. It affects fewer than 6 babies out 10,000. Most infants get it before they're a year old, usually between months 4 and 8. A little over half of babies who have West syndrome are boys.

Material and methods: This was a prospective Observational Study. In this observational study we identified diagnosed cases of West syndrome that attended Pediatric Neurology OPD or were admitted in IPD in Dr D.Y. Patil Medical college hospital and research centre, Pune during 2 years between August 2018 to September 2020. There were 25 patients who met the inclusion criteria and were enrolled in the study. We did the necessary evaluation and recorded data on a proforma.

Results : The average duration of each cluster of spasms was 3.7 minutes. The average frequency was 8-10/day. 22 (88%) of the patients had hypotonia at presentation, whereas the other 3 had spasticity. Other seizures types occurring in the patients was noted; 6 patients (24%) had myoclonic seizures, 4 patients(16%) had GTCS type seizures, 2 patients (8%) had focal seizures.

Conclusion: Associated with this, taking the clinical profile in isolation, there are no significant differences between the clinical profile in India as opposed to other countries studied more globally.

Introduction:

West syndrome is a constellation of symptoms characterized by epileptic/infantile spasms, abnormal brain wave patterns called hypsarrhythmia and intellectual disability. West syndrome is a type of epilepsy that affects babies. This condition is rare. It affects fewer than 6 babies out 10,000. Most infants get it before they're a year old, usually between months 4 and 8. A little over half of babies who have West syndrome are boys.

The term 'West syndrome' was coined after the name of the doctor, Dr William J. West, who brought it to the attention of the medical profession in 1841 after his four month old son displayed what is now known as infantile spasms and was subsequently diagnosed with it. [1]The first written description of what is now known as infantile spasms appeared in 1841. William James West, a general practitioner in Tonbridge, Kent described in great detail, in a letter to the *Lancet,* episodes of repeated head nodding in his son, James Edwin West. [2]The letter was as much an appeal to specialists and would go on 'to become a classic of childhood epileptology' [1]. Approximately about a third of all children with West syndrome go on to develop ongoing epileptic seizures as they get older and may develop what is known as Lennox-Gastaut syndrome, which present with a variety of kinds of seizures that are not easy to control and are linked too with cognitive retardation. About another third will carry on having epileptic spasms as they age. The final third will have spasms that resolve with time [3].

Material and methods:

This was a prospective Observational Study. In this observational study we identified diagnosed cases of West syndrome that attended Pediatric Neurology OPD or were admitted in IPD in Dr D.Y. Patil Medical college hospital and research centre, Pune during 2 years between August 2018 to September 2020. There were 25

patients who met the inclusion criteria and were enrolled in the study. We did the necessary evaluation and recorded data on a proforma. They were followed up again at 6 months and 12 months to study the course of the disease, and ascertain the response to therapy. The study has been carried out over a period of 2 years after the approval from institutional ethical committee, from August 2018 to September 2020**.**

Patients had attended the pediatric outpatient clinic or the neurology clinic or the pediatric wards of a tertiary care teaching hospital over a period of 2.5 years and met the inclusion criteria. Patients were informed about the study and of their rights of inclusion/participation as well as their right to withdraw. The matters of the confidentiality and anonymity of their records was also assured.

Inclusion criteria:

Children, M and F, with West syndrome, aged between 6 months to 30 months. West syndrome here was taken to mean the constellation of symptoms in the triad namely 1) infantile epileptic spasms, 2) hypsarrhythmia 3) psychomotor retardation.

Patients who willingly gave their written informed consent for the study.

Patients who maintained records of spasms and came for regular follow up were included in the study.

Exclusion criteria:

Patients who did not give consent.

Medical records of patients who were followed for <6 months were excluded.

Patients who did not maintain records were excluded from the study

Patients with severe malnutrition or chronic system illnesses like Tuberculosis.

Results and Observations :

A total of 32 children were screened. 3 children were excluded due to the non availability of primary caregiver, 2 due to severe malnutrition, and refusal to give consent, in 2 cases. 25 patients were found to be eligible for enrolment. There were 16 males and 9 females. The mean age of spasm onset was 7 months for males and 7.6 months for females. The mean age at enrolment was 11.4 months. The youngest child was 7 months old and the oldest one was 24 months old. Mean age of enrolment was 11.9 months for males and 12 months for females.

In 16 (64%) children, there was significant motor delay at presentation. Motor regression was seen in 3(12%) patients.

6 patients (24%) had normal motor development for their age.

The most common semiology was flexor spasms in 18 children (72%), followed by mixed in 6(24%), and extensor spasms in 1 (4%).

In all the patients, spasms occurred in clusters. They were associated with cry in 17 (68%), and laugh in 2 (8%). They occurred on awakening in 15(60%), and on/after feeding in 16 (64%) of children.

The average duration of each cluster of spasms was 3.7 minutes. The average frequency was 8-10/day.

22 (88%) of the patients had hypotonia at presentation, whereas the other 3 had spasticity.

Other seizures types occurring in the patients was noted; 6 patients (24%) had myoclonic seizures, 4 patients(16%) had GTCS type seizures, 2 patients (8%) had focal seizures.

CLINICAL PROFILE		
Abnormal motor examination	25(100)	
Microcephaly	13(52.0)	
Motor development at presentation	Normal	6(24.0)
	Delay	16(64.0)
	Regression	3(12.0)
Tone	Hypotonia	22(88.0)
	Spasticity	3(12.0)
Type of spasms	Flexor	18(72.0)
	Extensor	1(4.0)
	Mixed	6(24.0)
Features associated with spasms	None	6(24.0)
	Cry	17(68.0)
	Laugh	2(8.0)
Spasms on awakening		15(60.0)
Spasms with feeding		16(64.0)
Spasms in Clusters		25(100)
Other seizures at presentation	Myoclonic	6(24.0)
	GTCS	4(16.0)
	Focal	2(8.0)
	None	13(52.0)

Discussion:

West syndrome is a specific kind of epilepsy seen developing in early infancy. This study was conducted in the resource constrained settings of India. This observational study was conducted for 25 patients with diagnosed

West syndrome, attending a tertiary health care centre of Pune for determining the clinical profile, investigative findings and treatment outcomes of this rare disorder. The first discussion point concerns male prevalence is seen in the patients we studied, a finding which is consistent with other paper [4,5]. In the context of India society this could be explained because of mainstream societal bias towards male bodies. This trend has been changing in society at large due to social and educational reforms but it still prevails as a whole. If it was a cultural issue though we might expect to see different findings in the West, or other countries where this male bias isn't entrenched. Such studies have also reported a similar gender ratio, about 60:40 which suggests a possible biological proneness of the male gender for this epilepsy which needs to be further explored[6,7,8.]

Most of the patients in our study had the onset of spasms from 5 months onwards, to 8 months, and this was irrespective of the gestational age. Earlier studies claimed, it has been hypothesized that West syndrome develops from the time of conception, and that the development in West syndrome need not be connected to the point of brain insult but on the process of brain maturation[8.] Seizures that occurred during the neonatal period were not recognized (either overlooked or not even noticed) hence explaining the delay between onset of spasms/seizures, acknowledgement of the disease and subsequent treatment. This time lag, which averaged at 5.4 months in this study between the onset of the disease and presentation to a specialist was because caregivers and also general physicians in some cases accounted for the spasms with the explanation of startle. This lack of knowledge about the disease among the medical fraternity is also a contributing factor in late diagnosis and thus poor management of this condition. [9]

In this study, 76.4% of cases had the onset of spasms by the age of 8 months. This finding matches with others[10]. In India, especially in rural and semi urban settings with low literacy rates and awareness many times the children are believed to be possessed by evil spirits and taken to soothsayers and quacks for a quick remedy. This wastes valuable time [11].

Some studies suggest that the sooner therapy is given the better the response may be especially if it is administered within thirty days since identifying the onset of spasms. However, that has no bearing a better response when considering psychomotor issues [54]. This point remains controversial. In the West, the mean lag time is shorter, reportedly about 1-1.5 months in most centres[12]. The spasms are commonly overlooked by the family and physician since they resemble so many benign conditions like the hiccup or startle reflex, or the Moro reflex. The slightness of their articulation may mean that they are missed entirely. In this study a positive correlation was made between the lag of treatment and seizure response. The mean lag time for our patients was 4.65 months. The difference between males and females in this regard was insignificant. A shorter lag time correlated to better outcome in terms of spasm control, motor development, and hypsarrythmia resolution [13,14] . The data available in this cohort was not sufficient in the sense of size or detailed enough to apply this system of spasm classification. However, its merits were reflected on, and future studies would warrant use of this system.

Conclusion:

Associated with this, taking the clinical profile in isolation, there are no significant differences between the clinical profile in India as opposed to other countries studied more globally.

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