

## West Syndrome: A Rare Epileptic Syndrome of Infancy

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**Abstract:** We report a case of 9 month old boy who presented with complaint of irritability, jerky head nodding and developmental delay. Neuro imaging was normal but EEG showed hypsarrythmia. Later diagnosed as a case of West syndrome and managed with ACTH and prednisolone therapy.

**Keywords:** West Syndrome, irritability, jerky head nodding, ACTH

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### INTRODUCTION

West syndrome is a rare form of epilepsy usually affecting children in early infancy. The peak age of onset is between 3 and 7 months; onset after 18 months is rare, though onset up to 4 years of age has been reported [1]. It was first described by Dr. Williams James West 75 years ago in his own son but still its diagnosis, evaluation and management remains to be a challenge for medical professionals [2]. Incidence of West syndrome in children ranges from 2 to 3.5 per 1000 thousand live births [3]. West syndrome comprises of characteristic triad of infantile spasms, developmental delay and hypsarrythmia on EEG [4]. Prognosis of children is usually poor despite proper treatment [5].

### CASE REPORT

A nine month boy who is the second issue of non consanguineous parents completely immunized to EPI was admitted in our hospital with complaint of irritability and head nodding several times a day since 1 month. He was born as a result full term normal delivery after an uneventful antenatal period.

On general examination his vitals were within normal limits, pallor was present and no other significant finding was noticed. There was gross developmental delay with developmental quotient of 58%. On investigation complete blood count, liver function test, renal function test, serum electrolytes was normal. EEG showed bilaterally asynchronous high-voltage polyspikes, and slow wave discharges interspersed with multifocal spikes and slow waves. MRI scan revealed normal brain. He was diagnosed as a case of West syndrome.

Primarily management was started with phenobarbitone 35 mg daily for 2 days and then phenytoin was added in a dose of 35 mg daily for next two days. After 4 days when there was no improvement sodium valproate was started 140mg daily and then increased up to 300mg daily. No significant response was appreciated. On 9<sup>th</sup> day we started ACTH therapy in dose of 20 U/day for 2 weeks and then increased to 30 U/day and then to 40 U/day intramuscular for additional 4 weeks. After completion of ACTH therapy prednisolone was given in a dose of 2mg/kg/day for 2 weeks.

On completion of ACTH therapy there was significant decrease in frequency of head nodding but EEG continue to show intermittent generalized rapid spike bursts. There was no improvement in developmental quotient.

### DISCUSSION

We described a case of 9 month old boy with West syndrome. Infantile spasm, developmental delay and hypsarrythmia in EEG are the characteristic triad of West syndrome when found in infants as in our case.

Infantile spasms are resistant to routine anti epileptic drugs eg: Phenobarbitone, phenytoin and sodium valproate but responded to ACTH therapy [6]. Although EEG and neuro developmental behavior did not show much changes even after completion of ACTH therapy. These findings were similar to the results of Richard et al who studied west syndrome outcome in 214 children. On the contrary, Riikonen in 1996 and Kimura et al in 1999 when managed there cases with low dose ACTH therapy showed significant improvement in controlling of seizures and developmental delay [7,8]. Later Sharma et al in 2008

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suggested that if ACTH therapy is given in first month of occurrence of infantile spasm outcome can be not as bad as it has been known [9].

Even though some authors have reported significant findings in MRI brain in West syndrome, normal MRI scan is not an uncommon finding as found in our case. Abnormal MRI if reported leads to confirmation of etiology [10]. Etiology in most of the cases is known now days. Hypoxic ischemic encephalopathy, neonatal sepsis and neonatal meningitis remain the predominant cause of West syndrome [11].

## CONCLUSION

West syndrome is a rare form of epilepsy with poor outcome. Infantile spasm is resistant to phenytoin and sodium valproate. ACTH therapy is effective means to control spasms but has little role in improvement of neuro developmental outcome. There is an absolute need for further studies which can discover the early diagnostic measures and more effective treatment.

## REFERENCES

1. Hrachovy RA, Frost JD; Severe encephalopathic epilepsy in infants: infantile spasms. *Pediatr Epileps*. 2008; 1: 249–268.
2. West WJ; peculiar form of infantile convulsions. *Lancet*. 1841; 1:724–725.
3. Agarwal V, Tripathi A, Sivakumar T; Treatment resistant attention deficit hyperactivity disorder an outcome of West syndrome: A Case Report. *Annals of Neurosciences*. 2008; 15: 3
4. Infantile Spasms; Jean Bacardi: Disease of Nervous system in childhood. 3rd ed. Mac Keith pres; 2009; 593-7.
5. Richard E Appleton. West syndrome: long-term prognosis and social aspects. *Brain and development*. 2001; 23(7): 688-91.
6. Hamouda HB, Mnasri H, Ghanmi S, Soua H, Sakli G, Dogui M, Sfar MT; West syndrome associated with Down syndrome: Case report and literature review. *Pediatr Polska*. 2014; 89: 203-6.
7. Kimura M, Miyamoto S, Sejima H, Yamaguchi S; A case of West syndrome well controlled by short and very low dose ACTH therapy. *Psychiatry and clinical neurosciences*. 1999; 53: 67-70.
8. R. Riikonen. Long-Term Outcome of West Syndrome: A Study of Adults with a History of Infantile Spasms. *Epilepsia*, 1996; 37(4): 367–72.
9. Sharma NL, Vishwanthan V; Outcome in West Syndrome. *Indian Pediatr*. 2008; 45(7): 559-63.
10. Taghdiri MM, Nemati H; Infantile Spasm: A Review Article. *Iran J Child Neurol*. 2014; 8(3): 1-5.
11. Kaushik JS, Patra B, Sharma S, Yadav D, Aneja S; Clinical spectrum and treatment outcome of West Syndrome in children from Northern India. *Seizur*, 2013; 22(8): 617-21.