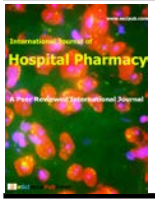
Case Report **IJHP 2017, 2:4**



International Journal of Hospital Pharmacy (ISSN:2574-0318)

CASE REPORT OF WEST SYNDROME: A RARE SEVERE EPILEPSY IN INFANTS

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ABSTRACT

West syndrome (WS) also known as infantile spasms, is severe form of epilepsy of early childhood was first describe in 1840s. WS presents with myoclonic-tonic seizures (spasms) characterized by flexor, extensor or mixed movements with distinct electroencephalogram (EEG) pattern of hypsarrthmia and psychomotor arrest. Prevalence rate is ~ 4.0 cases per 10,000 children. Peak age of beginning is between 3 and 7 months; onset after 18 months is rare. Recent guidelines from the American Academy of Neurology and the Child Neurology Society for medical treatment of WS, recommends that ACTH is probably effective and vigabatrin is possibly effective in cessation of spasms of hypsarrhythmia. Although discovered 160 years ago, still its diagnosis, assessment and management continue to create many challenges to health care professionals and affected families. In this case of a 3 yr old male child was admitted for long period of almost 2 months before being discharged. This case was confirmed early but due to its complexity it took long time to get complete resolve of situation. ACTH with Valproate was found to effective; though ACTH showed some reactions still it was considered the best available medicine for WS. In conclusion, early detection and referral to a pediatric neurologist for clinical evaluation and prompt effective treatment is strongly recommended as it may improve prognosis.

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How to cite this article:
G. Pravalika, Sushanta Kr Das, Saurabh Gupta. CASE REPORT OF WEST SYNDROME: A RARE SEVERE EPILEPSY IN INFANTS.. International Journal of Hospital Pharmacy, 2016,1(1): 0001-0005.

eSciencePublisher
eSciPub LLC, Houston, TX USA.
Website: http://escipub.com/

INTRODUCTION

West syndrome (WS) also known as infantile spasms, is a severe form of epilepsy of early childhood. Dr. William James West, in 1840s first described about it [1]. WS presents with myoclonic-tonic seizures (spasms) characterized by flexor, extensor or mixed movements with distinct electroencephalogram (EEG) pattern of hypsarrthmia and psychomotor arrest [2]. Its prevalence rate is ~ 4.0 cases per 10,000 children. Peak age of beginning is between 3 and 7 months; onset after 18 months is rare, though onset up to 4 years of age has been reported in few reports [3]. About 2/3 of babies with WS have some known cause for seizures. Etiologic classification comprises the categories of unknown and symptomatic. In infants with unknown WS, no underlying cause is recognized and children have normal development prior to the onset of disease [4]. Causes of WS may be prenatal, perinatal or postnatal. Roughly 50% of cases have a prenatal cause, including central nervous system malformations, intra-uterine insults, neuro-cutaneous syndromes such as tuberous sclerosis complex (TSC), metabolic disorders, genetic syndromes; Down syndrome [5]. Perinatal causes includes; neonatal (hypoxic-ischemic) encephalopathy and postnatal causes include; trauma, infection, and rarely tumors. The majority infants diagnosed with west syndrome need other confirmatory diagnosis like MRI of brain, blood tests including FBC, LFTs, renal function tests, glucose, calcium, magnesium. In addition to blood test Urine tests including amino acids and organic acids also plays a crucial role to confirm the disease. Defining features of WS include hypsarrhythmia (a specific EEG pattern) and developmental degeneration. Even a brief EEG recording may confirm the diagnosis, but if WS is suspected, a prolonged awake and sleep video-EEG study is recommended [6]. For 30% of infants with WS, pyridoxine dose 100 mg IV may be administered to screen for pyridoxine-dependent seizures. Administration of pyridoxine should be done immediately before monitoring EEG. The most recent followed guidelines from the American Academy of Neurology and the Child Neurology Society for medical treatment of WS, which reviewed the offered evidence as of 2004, state that ACTH is probably effective and vigabatrin is possibly effective in the cessation of spasms of hypsarrhythmia [7]. Additional anti-epileptics medications that may appear helpful for WS includes; Valproate, Topiramate, Pyridoxine, Zonisamide, Lobazam or Clonazepam. Occasionally multiple therapy is used at a time - for example, ACTH and Vigabatrin or Hydrocortisone and Valproate. A high-fat, adequate-protein, low-carbohydrate (ketogenic), diet has shown good outcomes in some studies but the role of ketogenic diet in the management of infantile spasms has not been established yet [6].

CASE PRESENTATION:

A 3 year male child was admitted in Pediatrics Department with chief complaints of seizures lasting for 1-2minutes which is of generalized type.

On examination (O/E):

Child was conscious & irritable, PR was 78bpm, RR was 24/min CVS and per-abdomen was found to be NAD.

Past medical/medication history:

History of 1st episode of seizure at 9months of age and history of seizure disorder with Global development delay for which he was treated with anti-epileptics; Sodium valproate, Leviacetram, Clobazam, Clonazepam, Vigabatin, Lamotrigine & Topiramate in time to time.

Lab tests:

Complete blood picture: WBC: 7200 (4500-11500), HGB: 10g/dl (11.0-16.5), Polymorphs-68% (65-75), Lymphocytes-28% (20-25), Eosinophils-2% (2-5), Monocytes-2% (1-2).

Liver function test: TSB: 0.2 mg% (0.3-1.0 mg %), SGPT: 12 IU/L (0-45 IU/L), ALP: 221 IU/L (44to 147 IU/L).

Renal Funtion test: (80-160 mg/dl), BUN: 27mg/dl (10-45 mg/dl), Sr.Cr: 0.7 mg/dl (0.5-1.5mg/dl).

Serum Electrolytes: Na: 147 mEq/L (135-150),K: 3.4 mEq/L (3.5-5.5), Cl:96 mEq/L (95-105).

RBS: 132 mg/dl

Differential Diagnostic test:

EEG: Shows multifocal epileptiform activity over both hemispheres.

MRI: Shows periventricular white matter hyper intensities. Evidence of glosis in right posterior parietal and occipital region with mild dilation of occipital horn of right lateral ventricle.

Based on medical history, clinical examination and laboratory findings; confirmatory diagnosis was "**WEST SYNDROME''.**

Upon admission, baby was prescribe with the following prescription

1. Injection Sodium valproate 120mg in 20cc NS over 20min

2. Tablet Clobazam 5mg BD

3. Tablet Clonazepam 0.5mg BD

4. Tablet Lamotrigine 25mg BD

5. Tablet Topiramate 25mg BD

6. Syrup Valproate 5ml TID

7. Syrup Leviacetram 2.5ml BD

8. Injection Midazolam 1.2cc+2cc NS SOS

On day 2(30/1/16) - 10(1/2/16): Child condition was same & same medication was continued.

On Day 11(2/2/16): Child condition was same & continued same medication with addition of Tablet Vigabatrin 500mg BD.

On day 12(3/2/16): Child was conscious & coherent, afebrile, per-abdomen was soft, PR-92bpm, BP 110/90 mmHg. Complaint of myoclonic jerks, cry during night. Same treatment was continued.

On day 13(4/2/16): Child was conscious & coherent, afebrile, PR-82 beats/min, BP-110/90 mmHg. Continued the same treatment with addition of Tablet Paracetamol 250mg BD, and Syrup Chlorphenaramine maleate 5ml OD.

On day 14(5/2/16)-17(8/2/16): Child vitals were normal. Same treatment was continued during the period.

On day 18(9/2/16) Child condition was same .He was freshly prescribed with

1. Tablet Vigabatrine 500mg BD

2. Tablet Topiramate 25mg TID

3. Tablet Lamotrigine 25mg BD

4. Syrup Sod.valproate 5ml TID

5. Syrup Levipil 2.5ml BD

6. Injection ACTH 75 IU SC QID

On day 19(10/2/16): Child condition was same & same treatment continued with ACTH dose decreased to 40 IU and Syrup MVT 5ml was added.

On day 20(12/2/16) - day 33(25/2/16) Child vitals were normal. Continued same treatment

On day 34(26/2/16): Child was conscious & coherent. Complaint of unrolling of eye balls. Continued same treatment with addition of Syrup .Zincovit BD.

On day 35(27/2/16) - day 37(29/2/16): Child became dull with complaint of dark pigmentation of lips and face. It was suspected ACTH induced skin discoloration and continued same treatment.

On day 38(1/3/16): Child was conscious & coherent, afebrile, PR-100/min. with complaint of 1episode of seizure, continued same treatment by adding Syrup Lactulose 5ml BD.

On day 39(2/3/16): Child vitals were normal & continued same treatment with addition of Syrup Chlorphenaramine malaete 5ml OD.

On day 40(3/3/16): Child vitals were normal, same treatment continued with addition of, Tablet Clobazam 5mg BD.

On day 41(4/3/16) - day 43(6/3/16) Child vitals were normal & continued same treatment.

On day 44(7/3/16): Child vitals were normal, continued same treatment with ACTH administer on alternate days.

On day 45(8/3/16): Child vitals were normal and he was freshly prescribed with

1. Injection ACTH 0.6ml IM OD on alternate

days.

2. Syrup Valproate 6ml TID

3. Syrup Lactulose 5ml OD

4. Tablet Vigabatrine 500mg BD

5. Tablet Topiramate 25mg BD

6. Tablet Lamotrigine 25mg BD

7. Syrup Levipil 2.5ml BD

8. Syrup Zincovit 5ml BD (Multivitamine with Zinc)

9. Syrup Chlorphenaramine malaete 5ml BD

10. Tablet Clobazam 5mg BD

On day 46(9/3/16) - day 57(20/3/16) Child vitals were normal & continued same treatment.

On day 58(21/3/16) Child was fit to discharge and was discharged with following medication:

1. Syrup Prednisolone 8ml BD

2. Syrup Valproate 6ml TID

3. Tablet Vigabatrine 500mg BD

4. Tablet Topiramate 25mg BD

5. Tablet Lamotrigine 25mg BD

6. Syrup Levipil 2.5ml BD

7. Tablet Clobazam 5mg BD

8. Syrup .Zincovit 5ml BD

DISCUSSION:

Although WS was first described over 160 years ago, its diagnosis, assessment and management continue to create many challenges to health care professionals and affected families [1] similar condition was found in this case, where baby developed seizure in 9th month of age and was admitted for WS in 3rd Yr of age. WS occurs in children from all racial groups. Male child are affected slightly higher than girls (ratio of 60:40) [8]. The stress/corticotropin-releasing hormone (CRH) suggestion tells that the common mechanism in all the etiologies of WS can increase in the release of stress-activated mediators in the brain, especially the neuropeptide CRH in the limbic and brain stem regions in children with WS. Adrenocorticotropic hormone (ACTH) suppresses the synthesis of CRH [9] similarly in this case ACTH was useful to overpower the disease condition to provide ultimate care. In some children, WS respond easily to treatment, whereas in others they keep on episodic. Similar was found in this case; as it took quite a long time to overcome from the situation and to bring the patient in normal condition. Previous studies in children with WS, which uses Valproate as monotherapy, proved effective in controlling either hypsarrhythmia or epileptic spasms [10, 11]. Besides Valproate, Topiramate and Nitrazepam (monotherapy or in combination) showed good responses in WS [12]. Short-term treatment with hormonal therapy (i.e., Adrenocorticotropic hormone ACTH), has been reported to succeed in 60–80% of the infants with WS [13]. First line treatment of WS and more in general of infantile spasms includes ACTH as well as Vigabatrin (the latter being effective especially in infantile spasms in the setting of tuberous sclerosis) [14].

CONCLUSION:

Children with WS often require an evaluation for early intervention programs for developmental impairment. Because available resources vary by community. Early detection and referral to a pediatric neurologist for clinical evaluation and prompt effective treatment is strongly recommended as it may improve prognosis. It is essential that malignancies be discarded and that the patient follow-up is maintained.

CONFLICT OF INTEREST: None declared.

SOURCE OF FUNDING: Nil.

ETHICAL PERMISSION: Not Required.

LIST OF ABBREVATIONS:

ACTH - Adenocorticotrophic hormone

ALP - Alanaine phosphatase

BD - Twice a day

BU – Blood urea

CRH - Corticotrophin releasing hormone

EEG - Electroencephalogram

IU - International units

K - Potassium

Na - Sodium

NAD - Normal at diagnosis

NS - Normal saline

MRI - Magnetic resonance imaging

OD - Once daily

PR - Pulse rate

QID - Four times a day

RR - Respiratory rate

RBS - Random blood sugar

SGOT - Serum glutamic oxaloacetic transaminase

SGPT - Serum glutamate pyruvic transamainase

TSB - Total serum bilirubin

TSC - Tuberous sclerosis complex

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