Int J Ayu Pharm Chem

REVIEW ARTICLE

www.ijapc.com

e-ISSN 2350-0204

West Syndrome and its Ayurvedic Management

Vivek B Singh[1*], Supriya B Kurane[2] and Deepnarayan V Shukla[3]

[1,2] Sharir Rachna Dept., R. A. Podar Medical College (Ayu), India

Abstract

West syndrome is a rare epileptic disease having the triad of infantile spasms, a pathognomonic EEG pattern (called hypsarrhythmia), and developmental regression - although the international definition requires only two out of these three elements. This means that it is an electroclinical epileptic syndrome. The purpose of this study was to find out an effective remedy to reduce the number of seizures and also help the child to attain development. The concept of using ghruta preparation in the this syndrome has been studied and various other ayurvedic panchkarma treatment which is useful in treating the disease.

Keywords

West syndrome, Ayurveda, *Apasmara*

Greentree Group

Received 15/6/15 Accepted 16/6/15 Published 10/7/15

INTRODUCTION

Wests' syndrome[1] is an uncommon-to-rare epileptic disorder in infants**,** children and adults. It is named after the English physician, William James West (1793–1848), who first described it in an article published in The Lancet in 1841[2]. The original case actually described his own son, James Edwin West (1840–1860). Other names for it are "Generalized Flexion Epilepsy", "Infantile Myoclonic Encephalopathy", "Jackknife convulsions", and "Salaam spasms" because the appearance of the seizures is like a bowing forward and backwards movement. West syndrome in modern usage is the triad of infantile spasms, a pathognomonic EEG pattern (called hypsarrhythmia), and developmental regression[3] - although the international definition requires only two out of these three elements. The syndrome is age-related, generally occurring between the third and the twelfth month, generally manifesting around the fifth month. The syndrome is often caused by an organic brain dysfunction whose origins may be prenatal, perinatal (caused during birth) or postnatal. This type of epilepsy occurs in about one in 2500-3000 children[4].

Etiology (causes)

The causes of this syndrome is still unknown which bio-chemical mechanisms lead to the occurrence. It is conjectured that it is a malfunction of neurotransmitter function, or more precisely, a malfunction in the regulation of the GABA transmission process. Another possibility being researched is a hyper-production of the Corticotropin releasing hormone (CRH). It is possible that more than one factor is involved. Both hypotheses are supported by the effect of certain medications used to treat West syndrome.

Cases of epilepsy have been historically divided into three different groups: symptomatic, cryptogenic, and idiopathic. The International League Against Epilepsy (ILAE) recommended in 2011 to abandon these terms[5,6] for reasons of clarity and instead try to place individual cases into one of the following three groups: genetic, structural/metabolic, and unknown. In a study it is said that out of 207 infants studied, 127(61%) had proven etiology, 68(33%) had no identified etiology, and 12(6%) were not fully investigated. Etiologies were prenatal in 63, perenatal in

38, and postnatal in 8 and 18 other. The most common etiologies were hypoxic-ischemic encephalopathy (HIE) 21(10%), chromosomal 16 (8%), malformations 16(8%), stroke 16(8%), tuberous sclerosis complex 15(7%), and periventricular leukomalacia or haemorrhage 11 (5%). The remaining 32 etiologies were all individually uncommon[7].

Signs and Symptoms

The epileptic seizures which can be observed in infants with West syndrome fall into three categories, collectively known as infantile spasms. Typically, the following triad of attack types appears; while the three types usually appear simultaneously, they can also occur independently of each other:

1) Lightning attacks: Sudden, severe myoclonic convulsions of the entire body or several parts of the body in split seconds, and the legs in particular are bent (flexor muscle convulsions here are generally more severe than extensor ones).

2) Nodding attacks: Convulsions of the throat and neck flexor muscles, during which the chin is fitfully jerked towards the breast or the head is drawn inward.

3) Salaam or jackknife attacks: a flexor spasm with rapid bending of the head and torso forward and simultaneous raising and bending of the arms while partially drawing the hands together in front of the chest and/or flailing. If one imagined this act in slow motion, it would appear similar to the oriental ceremonial greeting (Salaam), from which this type of attack derives its name.

Symptomatic

Almost any cause of brain damage could be associated with West syndrome and these are divided into prenatal causes, perinatal causes and post-natal causes. The following is a partial list:

In around one third of the children, there is evidence of a profound organic disorder of the brain. This includes:

1) microcephaly
2) cortical dysplasia
3) cerebral atrophy
4) lissencephaly
5) bacterial meningitis
6) phakomatoses (e.g. tuberous sclerosis)
7) Aicardi syndrome
8) cephalhematoma and
9) vascular malformation.

There are known cases in which the spasms occurs for the first time after vaccination against Measles, Mumps and Rubella or Tetanus, Pertussis, Diphtheria, Polio, Hepatitis B and Haemophilus influenzae Type B. However, there is no

causal relationship between immunization and West syndrome, since stress of any kind is a common trigger for seizures and the immunization occurs during the time-frame in which many typical cases become conspicuous.

Treatment

Compared with other forms of epilepsy, West syndrome is difficult to treat. To raise the chance of successful treatment and keep down the risk of long-lasting effects, it is very important that the condition is diagnosed as early as possible and that treatment begins straight away. However, there is no guarantee that therapy will work even in this case.

Based on what is known today, the prognosis depends mainly on the cause of the attacks and the length of time that hypsarrhythmia lasts[8]. In general it can be said that the prognosis is worse when the patient does not react as well to therapy and the epileptic over-activity in the brain continues. Treatment differs in each individual case and depends on the cause of the West syndrome (etiological classification) and the state of brain development at the time of the damage.

Drugs used for

1) Prednisolone

2) ACTH

3) Vigabatrin

Vigabatrin (Sabril) - Approved in several countries, including most of Europe, Canada, Mexico, and more recently the United States.

Side effects are: Somnolence, headache, dizziness, fatigue, weight gain, decreased vision or other vision changes

Vigabatrin is known for being effective, especially in children with tuberous sclerosis, with few and benign side effects. But due to some recent studies[9] showing visual field constriction (loss of peripheral vision), it was not approved in the United States until mid-2009. It is currently debated that a short use (6 months or less) of Vigabatrin will not affect vision. Also, considering the effect of frequent seizures on day to day life and mental development, some parents prefer to take the risk of some vision loss.

Other

Other drugs may be used in conjunction or alone. In Japan, there is a good experience with pyridoxine therapy. Further, topiramate (Topamax), lamotrigine (Lamictal), levetiracetam (Keppra) and zonisamide

(Zonegran) are amongst those drugs most widely used. The ketogenic diet has been shown to be effective in treating infantile spasms[10], up to 70% of children having a 50% or more reduction in seizure.

The role of ketogenic diet in the management of West Syndrome

The **ketogenic diet** is a high-fat, adequate-protein, low-carbohydrate diet that in medicine is used primarily to treat difficult-to-control (refractory) epilepsy in children. The diet forces the body to burn fats rather than carbohydrates. Normally, the carbohydrates contained in food are converted into glucose, which is then transported around the body and is particularly important in fuelling brain function. However, if there is very little carbohydrate in the diet, the liver converts fat into fatty acids and ketone bodies. The ketone bodies pass into the brain and replaces glucose as an energy source. An elevated level of ketone bodies in the blood, a state known as ketosis[11], leads to a reduction in the frequency of epileptic seizures.

The original therapeutic diet for pediatric epilepsy provides just enough protein for body growth and repair, and sufficient calories to maintain the correct weight for age and height. This classic ketogenic diet contains a 4:1 ratio (although a 3:1 ratio has also been used) by weight of fat to combined protein and carbohydrate. This is achieved by excluding high-carbohydrate foods such as starchy fruits and vegetables, bread, pasta, grains and sugar, while increasing the consumption of foods high in fat such as nuts, cream and butter[12]. Thus, an individual's diet is composed of 90% and 86% of calories coming from fat, respectively. Most dietary fat is made of molecules called long-chain triglycerides (LCTs). However, medium-chain triglycerides (MCTs)—made from fatty acids with shorter carbon chains than LCTs—are more ketogenic. A variant of the classic diet known as the MCT ketogenic diet uses a form of coconut oil, which is rich in MCTs, to provide around half the calories. As less overall fat is needed in this variant of the diet, a greater proportion of carbohydrate and protein can be consumed, allowing a greater variety of food choices[13,14].

Almost half of children and young people with epilepsy who have tried some form of this diet saw the number of seizures drop by at least half, and the effect persists even after discontinuing the diet[15].

Ayurvedic management of West Syndrome

West syndrome according to Ayurveda is considered as *Apasmara* which is defined as

Smrutibhootarth vijnyanamapash parivarjane |

Apasmara iti proktaastatoayam vyadhirantkrut ||

Su uttartantra 61/1.

Smruti means to remember anything and *apshabd* is known to as to go so ultimately the meaning of the word becomes as *smrutivinaash* not to remember anything. The disease causes even death on fall of the patient so name as *vyadhi antkrut*.

Apasmaar treatment –

Tasya karryo vidhi: sarvo ya unmaadeshu vakshyte |

Puraansarpish: paanamabhyangashescheva poojita: ||

Su uttartantra 61/ 22

Taeravryttanaam hrutstrotomanasam samprabhodhanam |

Tishkshaneraadau bhishak kuryat karmabhirvamanadibhi:||

Ch chi 10/ 14

According to Shusruta acharya it is said the treatment of *Apasmara* is as same as that of *Unmada*. He has advocated the use of *puraan sarpi* (aged ghee) for all the purpose such as *paan, abhyanga, avsechana* etc.

Acharya Charak in chikitsa sthama has mentioned to peform all *tikshana* karma like *vamana* in order to free the *hrudya, stroto, manasa* for all the *avarana*.

Drugs described in the treatment of *Apasmara*(West syndrome)

1) Vachadi ghruta
2) Amlakyadi ghruta
3) Katbhyadi ghruta
4) Panchgavya ghruta
5) Mahapanchgavya ghruta
6) Bhramhi ghruta[16]
7) Kushmanda ghrutam

The drug administered here is all sneha. Manda guna of sneha has the ability to pacify the doshas especially the vata dosha. The distribution of drug in blood is chiefly influenced by its lipid solubility. The lipid soluble drugs are rapidly distributed through intra and extra cellular spaces. The lipid soluble drugs also have the capacity to cross the blood brain barrier thus increasing the availability of the active principles in the brain even more.

In many patients the basti treatment is also used effectively as described by kashyapa samhita khilsthana 8th chapter.

Panchadau karmani shnehaschatvaroante tathaanyo: |

Madhye shannanam niruhanaamantareshu trayastraya ||

Aadavanteantare cheva kale snehastrayatraya: |

Yoge niruhantritastrayoante dvaviti kramat ||

In this above given shloka kashayapa have described the order in which the karma Basti should be used. In starting five Sneha Basti and in the end four Sneha Basti should be given and only six Niruha Basti should be given in between at the interval of every three Sneha Bastis. In total twenty four Sneha Basti and only six Niruha Basti should be used.

Apasmara is a disease in which manas afflicted and the major dosha involvement is of vata dosha[19]. Role of Basti in treatment of West syndrome is that the main cause of seizures i.e., chestha is the vata dosha and as mentioned in text the main treatment for the vata dosha is Basti.

Ras kalpas like smrutisagar, suvarnmalini vasant also provides good results in improving development of the child and also improving the intellect of the child.

Panchakarma like virechana and nasya are also used affectively used in treating West syndrome. Nasya karma or shirovirechana is also prescribed because nose is said to be the gateway of shira as it is said *nasa hi shirso dwaram.* Therefore, drugs to act on brain have to be given through nose. Vacha churna pratimarsh nasya was being used in the patient as vacha acts.

The role of Virechana in these patients is, in Apasmara the *pitta* and the *kapha* doshas were vitiated exclusively and they remain lying in the body. Virechana has the quality to remove both pitta and kapha. By eliminating the kapha dosha the obstruction (avarana) of the path of vata dosha was removed. The virechana karma clears the margavarodha, eliminate the morbid doshas from rasa rakta and regulate the activity and movement of vata[20].

CONCLUSION

The role of modern drugs in West syndrome is to reduce the number of seizure episodes of the patients with risk of loss of vision. On the other hand Ayurveda treatment gives patients a healthy life without any side effects. It also reduces the life long dependency of the allopathic drugs by giving a potential option of ayurvedic medicine. Many *ghruta kalpas* were described by the acharyas which means that

they have followed the treatment using ketogenic diet including those who treated themselves with some herbal medicine to provide added effect of the medicine.

REFERENCES

1) Lachhwani et al (2006). The treatment of epilepsy: principles and practice (4 th ed ed). Philedelphia: lippincot Williams and Wilkins page 333.
2) West, W.J. (1841). "On a peculiar form of infantile convulsions". The Lancey 35 (911): 724.
3) NINDS infantile spasms Information Page retrieved January 10,2012
4) Pellock J M, et al. infantile spasm: a US consensus report. Epilepsia. 2010;51(10): 2175-89.
5) http://www.ilae.org/visitors/centre/cff/documents/New concepts_classification_2011_000.cfm.
6) http://www.ilae.org/visitors/centre/cff/ctfoverview.cfm
7) Osborne JP, et al, The underlying etiology of infantile spasm, Epilepsia 2010. 2010;51(10): 2168-74
8) Treatment of epileptic encephalopathies. CNS drugs 27 (3): 175-84. Mar 2013.
9) Iannetti, et al, October 2000. Visual fields constriction in children with Epilepsy on Vigabatrin Treatment. Paediatrics 106 (4): 838-42.
10) Nordli Jr et al (2002). Experience with the ketogenic diet in infants. Paediatrics 108, 129-133.
11) Freeman JM et al. The ketogenic diet: one decade later. Paediatrics 2007 mar; 119(3): 535-43
12) Liu YM. Medium chain triglyceride (MCT) ketogenic therapy. Epilepsia 2008Nov ; 49 suppl 8:33-6
13) Zupec- cania et al. an overview of ketogenic diet for pediatric epilepsy. Nutr Clin pract . 2008 dec-2009 jan;23 (6); 589-96.
14) Levy, et al, ketogenic diet and other dietary treatments for epilepsy. Cochrane database syst rev. mar 2012 14;3:CD001903.
15) Pandit kashinath shastri, dr gorakhnath, Charak Samhita, Chikitsa sthana, Varansi: chaukhamba bharti: reprint 2002; p- 705.
16) Shetty sk. Efficacy of an ayurvedic formulation (bhramhi ghruta) in epilepsy. Germany; Lambart Academic publishing 2013.
17) Ambika dutta shashtry, shushruta samhita with Tatva Sandeepika Commentry by Kaviraj vol 1 . Varansi chaukhamba Sanskrit Sansthana; 2003: p-173
18) Satyapal bhishakacharya , kashyap samhita with vidyotini hindi commentary by Pandit hemraja Sharma, varansi Chaukhambha Sanskrit Sansthsana , reprint 2010; page 278.
19) Chaudhari k et al, Evaluation of diet and lifestyle in etiopathogenesis of senile

dementia. A survey study. Ayu 2011; 32:171-6.

20) Shukla G et al. efficacy of virechana and basti karma with shaman therapy in the management of essential hypertension. A comparative study. Ayu 2013; 34(1) : 70-76.