ISSN (Online): 2455-3662
EPRA International Journal of Multidisciplinary Research (IJMR) - Peer Reviewed Journal
Volume: 8| Issue: 9| September 2022|| Journal DOI: 10.36713/epra2013 || SJIF Impact Factor 2022: 8.205 || ISI Value: 1.188

WEST SYNDROME, UPDATED INFORMATION FROM 2021 UNTIL 2022

Bryam Esteban Coello García[1], Diego Javier Cañar Calderón[2], Luz Elena Suqui Belesaca[3], Jeannette Rosmery Zari Tuapante[4], Erika Sofía Aguilar Paredes[5], María Gabriela Racines Narváez[6], Kelly Daniela Vintimilla León[7]

[1]*General Practitioner in independent practice, Faculty of Medical Sciences, Universidad de Cuenca. Azuay - Ecuador. ORCID: https://orcid.org/0000-0003-2497-0274*

[2]*General Practitioner in independent practice, Faculty of Medical Sciences, Universidad de Cuenca. Azuay - Ecuador. ORCID: https://orcid.org/0000-0002-1496-3663*

[3]*Internal Medicine Resident. Hospital Básico Misereor - Gualaquiza. Morona Santiago. Ecuador.*

[4]*General Practitioner in independent practice, Faculty of Medical Sciences, Universidad de Cuenca. Azuay - Ecuador. ORCID: https://orcid.org/0000-0002-3743-406X*

[5]*General Practitioner in independent practice, Faculty of Medical Sciences, Universidad de Guayaquil. Guayas - Ecuador. ORCID: https://orcid.org/0000-0001-7447-889X*

[6]*General Practitioner in "Empresa Universitaria de Salud - Universidad de Cuenca. Azuay - Ecuador. ORCID: https://orcid.org/0000-0003-3457-9033*

[7]*General Practitioner in "CETAD Municipal de Azogues" - Universidad Católica de Cuenca. Azuay - Ecuador. ORCID: https://orcid.org/0000-0002-4014-2017*

Article DOI: https://doi.org/10.36713/epra11326
DOI No: 10.36713/epra11326

SUMMARY

Congenital Zika infection has been linked to phenotypic features including neurological sequelae. In relation to West syndrome, it has been shown that of 147 children with congenital Zika infection, 7.5% suffer from West syndrome. Furthermore, in the management of this syndrome, the use of vigabatrin has been shown to be an appropriate therapy for these patients (1). This is one of the studies with which we want to demonstrate the variations in the current treatment of West syndrome.

The aim of this literature review is to demonstrate how varied the treatment of this pathology is so far, to show what the medical literature says about the most adequate treatment in the last 2 years.

KEYWORDS: ***Nervous System, West Syndrome, Infantile Spasms***

INTRODUCTION

In a case reported in 2021 of an eight year old female patient from New Delhi, poor psychomotor development and anxiety were evident in this patient. The prenatal history indicates prematurity, in addition to a maternal history of dehydration, fever and vaginal infection during delivery. The patient presented with seizures from an early age up to the age of 20 years, which was more characteristic at the time of awakening. The seizures were accompanied by jerking movements of the upper and lower extremities. The electroencephalogram showed hypsarrhythmias and diffuse cerebral atrophy on magnetic resonance imaging. This patient was initially given adrenocorticotropic hormones, but was replaced by anticonvulsant medication. Since the change of medication to anticonvulsant drugs earlier, the patient has had no new seizures (2).

ISSN (Online): 2455-3662
EPRA International Journal of Multidisciplinary Research (IJMR) - Peer Reviewed Journal
Volume: 8| Issue: 9| September 2022|| Journal DOI: 10.36713/epra2013 || SJIF Impact Factor 2022: 8.205 || ISI Value: 1.188

A study of 26 patients with West syndrome who were given short-course therapy in 2020 found that idiopathic West syndrome is self-limiting and has a good response to treatment. Another feature found in the study was that treatment with vigabatrin and adrenocorticotropic hormones are the first line of treatment. Another element to consider is that idiopathic West syndrome has a genetic component which should be studied; and that generally patients with idiopathic West syndrome with excellent prognosis of treatment is usually of short duration (3).

In a study conducted in 2021, it became clear that the use of high-dose prednisolone, typically a dose of more than 4 mg per kilogram, offers electro-clinical resolution and is a great cost-effective strategy for the treatment of West syndrome. There is also evidence that high-dose prednisolone has efficacy equal to that of adrenocorticotropic hormone therapy. Finally, the safety and tolerability of methylprednisolone followed by oral prednisolone has been found to be greater than that of adrenocorticotropic hormone-based treatment (4).

It should be remembered that West syndrome is often accompanied by hypsarrhythmias which are the most important features in the presence of developmental delay. These two features together with the presence of infantile spasms lead to the diagnosis of this syndrome (5). In addition, West syndrome includes hormone therapy, which includes hydrocortisone, prednisolone or Pina adrenocorticotropin. To this is added vigabatrin as a first-line treatment. A combination of hormone therapy and vigabatrin has shown greater efficacy than hormone-only treatment in patients aged two to 14 months with new-onset West syndrome (6).

A study conducted in Japan in 2021 showed the following results: Seizures occurred daily in 69.3%; seizure frequency of less than one per year was observed when the etiology was unknown (22.6%). Genetic aetiology was found in 23.8% and cortical developmental formation in 19.1%. Neurological abnormalities were absent in 37% but a high percentage of abnormalities were seen when the patient presented hypoxic ischaemic encephalopathy or Aicardi syndrome. Electroencephalogram abnormalities were found in 96.7% and MRI was altered in 62.7%. Treatment included hormone therapy, anti-epileptic therapy and a specific diet. Importantly, intellectual developmental delay was present in 88.4% and was more severe in patients with Aicardi syndrome, genetic aetiology and hypoxic ischaemic encephalopathy (7).

The metabolic alterations found in West syndrome were: obesity in 27.3%, diastolic hypertension in 48.8%, hypertriglyceridaemia in 71%, decreased HDL in 54.2%, weight gain and increased triglyceride levels in patients treated with ACTH (8).

A study conducted in 2021 looked at the relationship between BCG vaccination and administration of adrenocorticotropic hormones for West syndrome. This is studied because hormone therapy has caused immunosuppression and could result in infection after BCG vaccination. As a result of this study it was shown that 33 patients received hormone therapy prior to BCG vaccination. The interval between the administration of hormone therapy and vaccination was very short, almost 14 days. The evidence showed that none of the patients developed a BCG infection, even up to an interval of eight weeks there was no development of an infection (9).

A study conducted in 2021 showed that the median age of onset of West syndrome was four months. It was shown that prematurity, neonatal asphyxia, neonatal encephalopathy and small for gestational age were among the features that can lead to this syndrome. Some characteristics found in the electroencephalogram were hypsarrhythmias; in the MRI, on the other hand, were periventricular leukomalacia, cystic encephalomalacia and in very few cases a normal MRI. It was also shown that Trina, prednisolone, and adrenocorticotropic hormone treatment are the best alternative for the cessation of spasms (10).

In 2021, a total of 198 patients were studied in Argentina between 2004 and 2017. It is evident that five patients had a spontaneous remission related to an infection, and this was demonstrated through electroencephalographic manifestations and absent clinical manifestations. Of these five children, four were male and one was female. Apparently, these patients had remission of their pathology and normalisation of their EEG following upper respiratory infection; four of them had remission and one of them had sudden exanthema (11).

There is a clinical case report of three cases of three infants with West syndrome, all three infants were breastfed, considering that the mothers follow a strict vegan diet. In one of the three infants West syndrome developed during treatment with vitamin B12 and was maintained until vitamin B12 levels were normal. The fundamental point to mention in this review is the fact that supplementation of the mother can prevent the development of disease in the child. Early initial treatment with vitamin B12 and replacement therapy are of vital importance to prevent severe neurological problems and improve the clinical course of the patient (12).

However, the mothers of patients with West syndrome are also of consideration. A study conducted in 2021 reports that mothers had high levels of depression and anxiety. The scales used for anxiety were the Hamilton Anxiety Scale and for depression the Beck Depression Scale. Maternal anxiety, depression and parental stress were significantly higher in all mothers of children with West syndrome compared to mothers without children with West syndrome (13).

A randomised study conducted in 2021, studying the effectiveness of intravenous methylprednisolone versus oral prednisolone administration, used children aged 2:30 months with West syndrome who were randomised to be administered methylprednisolone at a dose of 30 mg per kilogram per day for three days or oral prednisolone at a dose of 4 mg per kilogram per day for two weeks. The first outcome found was cessation of spasms on day 14. The second outcome was response time in which electro-clinical remission was seen at around two to six weeks (14).

One of the features evident in the treatment of West syndrome has been that cessation of spasms has been seen in 53% when they occur within short periods of evaluation,

ISSN (Online): 2455-3662
EPRA International Journal of Multidisciplinary Research (IJMR) - Peer Reviewed Journal
Volume: 8| Issue: 9| September 2022|| Journal DOI: 10.36713/epra2013 || SJIF Impact Factor 2022: 8.205 || ISI Value: 1.188

although it should be noted that 2/3 of these patients relapse. Electroencephalogram is abnormal in 78% and it is important to mention that good response to standard therapy and a non-structural aetiology are associated with better seizure control (15). A study in 2021 evaluated quality of life at six years in patients who were treated with West syndrome based on hormone therapy. The conclusions drawn from this study were: age of onset, delay in treatment and early spasm control were several factors that did not have a major influence on the patient's quality of life. This means that despite epilepsy control, the boys suffered from an inadequate quality of life (16).

CONCLUSIONS

West syndrome, based on several scientific articles mentioned above, has been shown to have several factors influencing early treatment, adequate seizure control and the different therapeutic modalities encountered. However, it is necessary to mention that all the studies mentioned above evaluate patients from various perspectives; all this is of vital importance to emphasise because the treatment of patients in general is not strictly the same. In some of the studies mentioned above, the use of vigabatrin, methyl prednisolone, adrenocorticotropin-based therapy, prednisolone, among others, is mentioned; however, each individual must be properly evaluated so that the treatment given to the patient is the best solution for the control of West syndrome.

BIBLIOGRAPHY

1. *Bustamante Amador J, García-Segovia R, García Boyano M, Miño-León G. West Syndrome in Children With Congenital Zika Virus Infection. Pediatr Infect Dis J. 2021;40(12):1108-1110. doi:10.1097/INF.0000000000003230*
2. *Goswami M, Sharma S. "West Syndrome-Infantile Spasms": A Pediatric Case Report. Int J Clin Pediatr Dent. 2021;14(2):323-326. doi:10.5005/jp-journals-10005-1922*
3. *Fortini S, Valenzuela GR, Fasulo L, Caraballo R. West syndrome: A study of 26 patients receiving short-term therapy. Epilepsy Behav. 2021;114(Pt A):107235. doi:10.1016/j.yebeh.2020.107235*
4. *Sharma R, Gulati S. Methylprednisolone Pulses in West Syndrome: A New Weapon in the Armory-Is it Needed?. Indian J Pediatr. 2021;88(7):643-644. doi:10.1007/s12098-021-03782-w*
5. *Mytinger JR. Definitions and Diagnostic Criteria for Infantile Spasms and West Syndrome - Historical Perspectives and Practical Considerations. Semin Pediatr Neurol. 2021;38:100893. doi:10.1016/j.spen.2021.100893*
6. *Gibaud M, Barth M, Lefranc J, et al. West Syndrome Is an Exceptional Presentation of Pyridoxine- and Pyridoxal Phosphate-Dependent Epilepsy: Data From a French Cohort and Review of the Literature. Front Pediatr. 2021;9:621200. Published 2021 Mar 5. doi:10.3389/fped.2021.621200*
7. *Yoshitomi S, Hamano SI, Hayashi M, et al. Current medico-psycho-social conditions of patients with West syndrome in Japan. Epileptic Disord. 2021;23(4):579-589. doi:10.1684/epd.2021.1301*
8. *Gilboa I, Israeli G, Brener A, et al. Cardiometabolic outcomes in children and adolescents with West syndrome. BMC Pediatr. 2021;21(1):412. Published 2021 Sep 18. doi:10.1186/s12887-021-02871-1*
9. *Maki Y, Natsume J, Hori I, et al. Risks of ACTH therapy for West syndrome following BCG vaccination. Epilepsy Behav. 2021;118:107924. doi:10.1016/j.yebeh.2021.107924*
10. *Gehlawat VK, Arya V, Bhardwaj H, Vaswani ND, Kaushik JS. Clinical profile of children with West syndrome: A retrospective chart review. J Family Med Prim Care. 2021;10(1):350-353. doi:10.4103/jfmpc.jfmpc_1405_20*
11. *Caraballo R, Fortini S, Espeche A. Spontaneous remission of West syndrome associated with acute infection and fever in five patients. Epilepsy Res. 2021;174:106663. doi:10.1016/j.eplepsyres.2021.106663*
12. *Pavone P, Sullo F, Falsaperla R, et al. Vitamin B12 Deficiency and West Syndrome: An Uncommon but Preventable Cause of Neurological Disorder. Report on Three Cases, One of Them with Late Onset during Vitamin B12 Treatment. Neuropediatrics. 2021;52(4):333-336. doi:10.1055/s-0041-1725013*
13. *Kumar P, Arya V, Khanna A, Kaushik JS. Depression, Anxiety, and Parenting Stress among Mothers of Children with West Syndrome. Indian J Pediatr. 2021;88(12):1247-1249. doi:10.1007/s12098-021-03915-1*
14. *Kapoor D, Sharma S, Garg D, et al. Intravenous Methylprednisolone Versus Oral Prednisolone for West Syndrome: A Randomized Open-Label Trial. Indian J Pediatr. 2021;88(8):778-784. doi:10.1007/s12098-020-03630-3*
15. *Arvinder, Chakrabarty B. Long-Term Outcome in West Syndrome: The Facts and the Scope to Improve It Further. Indian J Pediatr. 2022;89(8):749-750. doi:10.1007/s12098-022-04240-x*
16. *Wanigasinghe J, Arambepola C, Murugupillai R, Kapurubandara R, Jayawickrama A. Quality of life at 6 years in children treated for West syndrome with hormonal therapy. Epilepsy Behav. 2021;119:107974. doi:10.1016/j.yebeh.2021.107974*