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PHARMACOTHERAPEUTIC MANAGEMENT OF INFANTILE EPILEPTIC SPASM SYNDROME (IESS): A CASE STUDY

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ABSTRACT

Infants with Infantile Epileptic Spasm Syndrome, formerly known as West syndrome, suffer from a severe form of epileptic encephalopathy. It is distinguished by Clusters of tonic spasms of the axial and limb muscles, known as epileptic spasms and hypsarrhythmia which is a disordered and chaotic brain wave pattern. Psychomotor delay or arrest may also occur, which can result in long-term disabilities. Numerous factors, such as genetic mutations, brain disorders, and the presence of other medical conditions, can lead to IESS. About 10–40% of patients have cases where no underlying cause can be found. Adrenocorticotropic hormone, vigabatrin, and high dosages of oral steroids are the most effective treatments. Age of onset is usually between 4-6 months, with the majority of cases presenting within the first year of life.

KEYWORDS: West Syndrome, Hypsarrhythmia, Psychomotor delay, Mutation, Adrenocorticotropic hormone.

INTRODUCTION

Infantile Epileptic Spasm Syndrome (IESS), also known as West syndrome, is a rare and severe form of epilepsy that affects infants, typically within the first year of life.[1] This

complex condition is characterized by clusters of sudden, brief muscle contractions, known as epileptic spasms, which can occur in series and are often accompanied by a chaotic and disorganized EEG pattern known as hypsarrhythmia. IESS can result from various causes, including genetic mutations, brain malformations, infections, and metabolic disorders. The predictive role of etiology in seizure remains controversial[2,3], making diagnosis and treatment challenging.

The symptoms of IESS can be subtle, and diagnosis often requires a comprehensive evaluation, including EEG, imaging studies, and genetic testing. Prompt diagnosis and treatment are crucial to optimize outcomes and minimize long-term effects. American Academy of Neurology and Child Neurology Society Quality Measure Set in 2017 suggest that high- dose Prednisolone and ACTH are first-lone treatment for IESS.[4,5] Early identification and intervention can significantly impact the prognosis and quality of life for infants with IESS.

The prognosis for IESS varies depending on the underlying cause, response to treatment, and presence of developmental delays. Some infants may experience significant developmental delays or long-term disabilities, while others may have more favorable outcomes. A pediatric neurologist can provide guidance on diagnosis, treatment, and ongoing care, working closely with the family to develop a comprehensive treatment plan.

Ongoing research aims to improve understanding and treatment of this complex condition, ultimately enhancing the lives of affected infants and their families. With proper management, some infants may experience improved outcomes, while others may continue to face developmental challenges. It is essential for families to work closely with their healthcare team to develop a comprehensive treatment plan and access support services to address the unique needs of their child.

The impact of IESS extends beyond the infant, affecting the entire family. Providing emotional support and connecting families with resources and support groups can help them navigate the challenges associated with this condition. By advancing our understanding of IESS and developing more effective treatments, we can improve the lives of infants and families affected by this complex condition. This case study describes the pharmacotherapeutic management of a 6-month-old infant diagnosed with IESS.

CASE REPORT

A 6-month-old male infant presented with a history of spasms and developmental delay. Electroencephalogram (EEG) revealed hypsarrhythmia, and the patient was diagnosed with IESS. The patient was started on adrenocorticotropic hormone (ACTH) therapy, and vigabatrin was added later due to inadequate response.

PHARMACOTHERAPEUTIC MANAGEMENT

The patient's treatment regimen included

1. ACTH therapy: 150 IU/m²/day for 2 weeks, followed by a taper.
2. Vigabatrin: 100 mg/kg/day, titrated to 150 mg/kg/day.

OUTCOME

The patient showed significant improvement in spasm frequency and EEG findings after 6 weeks of treatment. However, the patient experienced some adverse effects, including irritability and weight gain.

DISCUSSION

This case study highlights the importance of early diagnosis and treatment of IESS. ACTH therapy and vigabatrin are effective treatments for IESS, but careful monitoring of adverse effects is necessary. Most studies show that seizures are controlled more quickly and more often with hormonal therapy than with vigabatrin.[6,7] In another study[8], the adverse events related to corticosteroid administration were mostly tolerable. The most common events were irritability and excitement, which primarily occurred during intravenous hormonal treatment. Weight gain and cushingoid faces were also observed during follow-up.

CONCLUSION

Etiology was identified in approximately half of the IESS cases, with structural etiology found to be the most common. Pharmacotherapeutic management of IESS requires a comprehensive approach, including careful selection of medications, dosing, and monitoring of adverse effects. This case study demonstrates the effectiveness of ACTH therapy and vigabatrin in managing IESS.

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CONFLICTS OF INTEREST

The author would like to inform that there are no conflicts of interest regarding the publication of this case report.

REFERENCE

1. SM Zuberi, E Wirrell, E Yozawitz, JM Wilmshurst, N Specchio, K Riney, et al. ILAE classification and definition of epilepsy syndromes with onset in neonates and infants: position statement by the ILAE Task Force on Nosology and Definitions Epilepsia (2022).
2. R. Riikonen Infantile Spasms: Outcome in Clinical Studies Pediatr Neurol, 2020; 108: 54-64.
3. G Gul Mert, MO Herguner, F Incecik, S Altunbasak, D Sahan, I Unal Risk factors affecting prognosis in infantile spasm Int J Neurosci, 2017; 127: 1012-1018.
4. AD Patel, AT Berg, L Billinghurst, D Fain, E Fecske, T Feyma, et al. Quality improvement in neurology: child neurology quality measure set: Executive summary Neurology, 2018; 90: 67-73.
5. S Li, X Zhong, S Hong, T Li, L. Jiang Prednisolone/prednisone as adrenocorticotropic hormone alternative for infantile spasms: a meta-analysis of randomized controlled trials Dev Med Child Neurol, 2020; 62: 575-580.
6. Glauser TA, Morita DA, Stannard KM. Infantile Spasm (West Syndrome) Medscape, Apr 26, 2010; [Accessed July 17, 2011]. http://emedicine.medscape.com/article/1176431-overview.
7. Hancock E, Osborne J, Milner P. Treatment of infantile spasms. Cochrane Database Syst Rev., 2003; 3: CD001770. doi: 10.1002/14651858.CD001770. [DOI] [PubMed].
8. P Chellamuthu, S Sharma, P Jain, JS Kaushik, A Seth, S. Aneja High dose (4 mg/kg/day) versus usual dose (2 mg/kg/day) oral prednisolone for treatment of infantile spasms: an open-label, randomized controlled trial Epilepsy Res, 2014; 108: 1378-1384.