EUROPEAN ACADEMIC RESEARCH
Vol. III, Issue 10/ January 2016

Impact Factor: 3.4546 (UIF)
DRJI Value: 5.9 (B+)

ISSN 2286-4822
www.euacademic.org

Infantile spasms, West Syndrome: Old strategies and new treatment of Infantile Spasms in Children. An overview on the past, present and future.

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Abstract:

West syndrome described for the first time in 1841 by a doctor of the same name, who began his studies on the disease that caused the death of his son, is still one catastrophic epilepsies in children.

The aim of the study: to note the evolution of disease treatment protocols, their effectiveness in children with West Syndrome.

Method: Jane under study, 15 children diagnosed with West Syndrome Clinic Neuropediatrike University of Siena Italy from 1995 to 2004.

There are used as study material for children tplotesuara clinical records with the relevant examinations, history of illness, their treatment.

Results: there was an early manifestation of disease in 6.7 % of cases, classical performances in 86.6 % of cases and the recent performances of disease in 6.7 % of cases.

Symptomatic were 53.3 %, 40 % Kriptogjenike, and 6.7 % idiopathic.

ACTH is used 2UI/kg/dite average dose for 4 weeks with a result of stopping the crisis within 4.8 days.

Vigabatrin is used as the first choice in 33.3 % of cases, with a response to the crisis ban within 6.5 days in two cases, one of which with tuberous sclerosis.

There are used as other choices as well AED Valproik Ac, TPM, Vit B6, Leveracetam, lamotrigine.

Used treatments show a broad range response to them.

The best dose is as unclear as the drug of choice of mirei.

ACTH and Vigabatrin probably are effective medication treatment with West syndrome in children.

Key words: spasms, West syndrome, therapy, GEFS

Objective

To assess therapeutics regimes used in these children.
To confront these with developing new protocols.

Method

We analyzed 15 children diagnosed with Infantile Spasms Syndrome, hospitalized and followed at the University Clinic Neuropediatrics to Siena Italy from the years 1995 to 2004.

They used clinical records, history of illness, laboratory data, and radiological examinations therapy as a source of information for the study.

Throughout history...

Name the syndrome has been put by Dr. W. J. West, who gave the first description of infantile spasms that were shown to his son in 1841. This detailed clinical description was followed about 100 years later by reporting:

Infantile spasms
Interictal typical display of EEG hypsarrhythmia.
Most patients have some degree of delay in mental development
Mental can development or regression, although the diagnosis may also occur if one of these three elements may be missing or

regression arrest Mental Although the diagnosis Can Be Made (international classification).

Definitions over the years

Livingston et al, 1958 - Minor motor seizures. Chariton, 1975 Myoclonis seizures as myoclonics massive attacks.

International Classification, Gastaut, 1969, ISS - a variant of the generalized epileptic convulsions.

Revision 1981 threw medicines use of both terms.

1985 and 1989: Display of symptoms before age 12 months.

1991 IS includes age groups and may appear small nemoshat and childhood.

2001 Tokyo began the first proposals for standardization, definitions of cases, their appearance and actions to be taken Neisse.

In the U.S.; Infantile spasm epilepsies constitute 2% of childhood epilepsies but 25 % of the display screens in the 1000 jetes. Incidence 0, 25-0.60 for the new-borns. Prevalence of 0.15 to 0.2 per 1000 children. age ≥ 10 yr.

FREQUENCY

International

The age -specific prevalence was 2.0 per 10,000 among children 10 years of age. With SGA children were more likely to develop ISS.

Mortality / Morbidity: ~ 20,000 to 50,000 cases seen each year ISS. Best rates of complete responses reported 40 %. Relapse is between 35-50 %. 3-5 % of these patients die.

90 % of infantile Spasms begin in children younger than 12 muajshd.

The maximum incidence of performances is at age 3-7 months. Boys are affected more often than girls.

B / G - 1,1:1 to 2,8:1 . A family history is not common, 4 % of the cases (Sugai et al 2001.)

A history of familial epilepsies of each type is found in 6-17 % (Chevrie & Aicardi, 1967; Cowan & Hudson, 1991). Cases probability is family to represent a manifestation of severe genetic disorders.

PATHOPHYSIOLOGY:

Infantile Spasms is thought to reflect abnormal interactions between cerebral cortex are structures.

Focal lesions very early in life can affect secondary other brain areas, and this can present hypsarrhythmia anomalous activity stemming from multiple brain areas.

Frequency of occurrence of infantile spasms suggest that an immaturity of the central nervous system may be important in pathogenesies.

CHR hypothesis

NMDA hypothesis

Kyrenine - serotonin hypothesis. (J.Rho 2004)

Infantile Spasms usually occur in groups, often multiple, separated by 5-30 seconds. Spasm often appears before bedtime or during the wake. They can be observed during sleep, although this is rare. Spasms may be flexor, extensor, or a mixture of it flexion it and extension and can be asymmetrical. Sudden spasm starts, fast, tonic contractions of the trunk and gradually relax during muscles 0.5-2 seconds. Contractions may last 5-10 seconds flat. The intensity can range from a Fast contraction head to strong contractions of the body.

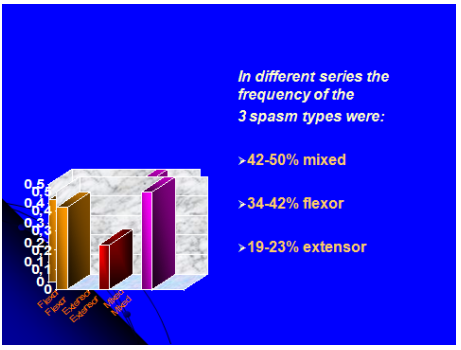
Flexor Spasms: Consist with contractions of muscle flectore short neck, trunk and extremities. They may resemble a movement of embracing and often are accompanied by a cry or weep. The patient relax and attack repeat This situation appear in groups during the day and last game in less than 1 minute der 10-15 minutes or longer in some patients.

Extensor Spasms: Konsisitojne we extensors muscle contractions with sudden neck extension and the extension of the trunk and extremities abduction. Extensor Spasms and asymmetrical unilateral spasms are often associated with symptomatic cases.

Mixed Spasms: Jane most common type, consisting of cervical flexion and extension arms with the legs, the legs with flexion or extension of the arms.

In some studies it has been seen frequency of 3 types of spasms are: 42-50 % 42 % mixed34 - flexor extensor 0.19 to 23 % .

WEST SYNDROME



Medicine is a tradition?

Treatment options for children with West syndrome are divided into 2 groups.

Hypsarrhythmia is seen in 75 % of patients West syndrome.

Hypsarrhythmia whereof diffuse giant wave of high voltage, > 400 microvolts) with a chaotic background of peaks and unruly waves and multifocal and very little synchronization between the two hemispheres. During sleep EEG can identify the wave synchronic Polispikes and can show pseudoperiodics. Slow or persistent outbreaks in background

hypsarrhythmic epileptiforme May be present and may present a focal area or dysfunction . Hypsarrhythmise different variants can appear.

Clinical spasms are associated with a significantly suppression background spasms long lasting. This answer feature called "electrodecremental. EEG answer is assisting in the successful trial of treatments West syndrome.

Usually soon after treatment with adrenocorticotrophic hormone (ACTH) or Vigabatrin, stop spasms and hypsarrhythmia disappear. Hypsarrhythmia rarely persists after age 24 can months can develop slow waves seen in LGS.

Interictal electroencephalogram

Hypsarrhythmia is characteristic EEG which consists of yeast multifocal and pathological wave with a chaotic, with a highest voltage polymorphic delta and theta rhythm. Variants of this image are available, including focal or asymmetric hypsarrhythmia.

Ictal electroencephalogram: In a study with common sight found in 38 % of patients with convulsion was a high voltage, frontal dominant slow wave followed transitional voltage Visible episode called electrodecremental. Electrodecremental episodes were found in 71 % of convulsions.

Medical assistance

The goals of treatment of children with West syndrome are:
Better quality of life without seizures
As fewer side effects and treatment
Smaller number of drugs used
Treatment with anti-epileptic drugs (AEDs) are therapy for children with West syndrome.
As first line (ie, ACTH, prednisone, vigabatrin, pyridoxine [vitamin B - 6].

Second -line treatment (ie , benzodiazepines, valproic acid, lamotrigine, topiramate, zonisamide)

ACTH: the most popular since 1958 : The drug of choice in the U.S. by .88 % was used with a dose of 40 UI / day for 1-2 months f , the choice was not influenced by Etiology. (Neu. May 2004 AAN)

Many studies have been done to identify the lowest dose that is effective in minimizing Relapse and side effects .

Tip of authors

Choose ACTH immediately after a failure to VGB Within 3 week starting dose - 3 UI / kg / day for 3-4 weeks no improvement ?Double every 2 weeks up to a maximum of 12 UI / kg / day. Best forms and doses of ACTH has not yet been determined.

Vigabatrin (VGB)

This has been used since 1990, various comparative outside USStudime been made to better identify effective dosage to individual goes from 18 to 200 mg / kg / day.

Best results were observed in cases with tuberous sclerosis ST, and > 3 months to show that spasms.

Not reach a complete agreement regarding a delay in response to VGB and Relapse percentage. Effects for a long time are still panjohura. The long - term effects of VGB are largely serious side unknown. Effects have been reported (visual field loss & Anomaly of retinal function).

Topiramate (TPM)

In various studies have been made to identify trajtues.Doza regime (2-24 mg / kg / day)

In some studies TPM has seen since injuring effects on TP in several studies as Monotherapy in ISS.

Lamotrigine (LTG)
Some studies have shown small doses of LTG effective in ISS, you do not respond by VGB & ACTH .

Zonisamide (ZNS). I used in Japan since the 1989, in the U.S. since 2000.
Several studies have been done to identify regime definitions and treatment response.

DAS can jet sending his option to treat patients with ISS. Nevojiten further studies .

Valproic Acid
It seems to have prime ISS. These studies are done to identify the best dosage ranges from 100-300 mg / kg / day. Several authors use it as therapy in nderprerejes time that ACTH or steroids.

Pyridoxine
Vit B6 Japan is the first line of treatment theistic, followed by VGB and VPA, ACTH.

Ketogenike role of diet in the treatment of children with West syndrome has not been determined.

Combined therapy: ACTH + VGB, Sod. Valproate + Hydrocortisone are proven sufficient evidence for use.

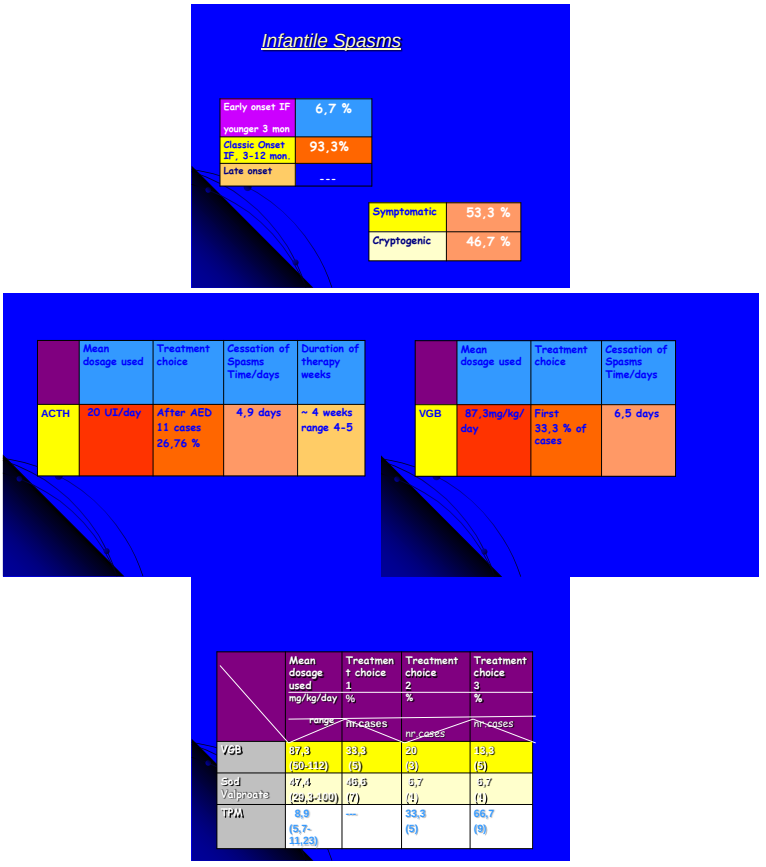
Prognosis:
Prognosis is poor long term and is directly connected with the etiology.

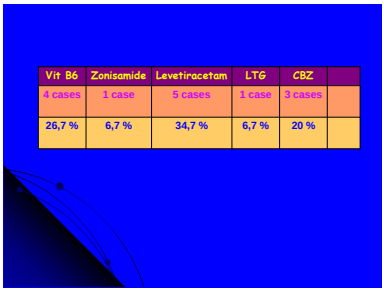
Looks worse prognosis in children with other types of seizures persistent EEG anomalies, the inconsistent response to ACTH, delay the onset of treatment. Children with idiopathic

West syndrome have a better prognosis than those with symptomatic West is mental syndrome. Mental retard severe in 70 % of patients , often with psychiatric problems as autistic or hyperactivity performances . 25-56 % of patients develop other types of epilepsy as LGS.

The tables below show more clinic e medical information of this study, including the age of onset, the type of spasms, type of treatment the duration of treatment and the result of cessation of spasms, of different medications used and the comparative information between them

Google





New developments

Remission spontaneous spasms in West syndrome, viral infections intervention. (exanthema subitum predominates). This link between viral infection and spontaneous remission suggests a process immuno - inflammatory in these patients with West syndrome.

Vit B6 (150-300mg/kg) and Sulthiame. (10 mg / kg) result in stopping the spasms in (30 %)

Sulthiame has a positive effect on the primary therapy of West syndrome ,compared with Vigabatrin. Patients with TS did not respond STM Patients with TS. Chochrane Review, May 2004 AAN.

Probably ACTH is an effective agent in treating short time treatment of infantile spasms IS.

There is an insufficient- evidence that oral corticosteroids are effective in the treatment of Infantile Spasms.

Vigabatrin is effective for treating probability short IS treatment. And the majority of children with tuberous sclerosis. There is insufficient evidence to recommend other treatments (Ac.Valp, Benzodiazpines, pyridoxine, new AED, or other therapies in the treatment of infantile Spasms)

CONCLUSIONS

Used treatments show a wide range of percentages of responses. Results belonging efficiency and security often come from small uncontrolled studies. The best dose is as unclear as the best medicament. No single agent has shown to be as good treatment of IS.

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