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Sorel L & Dusaucy-Bauloye A M. A propos de 21 cas d'hypsarhythmia of Gibbs. Son traitement spectaculaire par l'ACTH (About 21 cases of hypsarhythmia of Gibbs. His spectacular treatment with ACTH). *Acta Neurol. Psychiat. Belg.* 58:130-41, 1958. [Department of Neurology, Catholic University of Louvain, Belgium]

This paper covers a series of cases of West syndrome, or hypsarhythmia, treated with adrenocorticotropic thyroid hormone as described by F.A. Gibbs. [The *SCI*® indicates that this paper has been cited in more than 115 publications.]

Solving Seizures

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The West syndrome is a disease that begins in the child before the age of one year. In its typical and complete form, it manifests a symptomatic triad presenting epileptic seizures in the form of flexion or extension spasms (generally in salvos), a halt in intellectual development (the children developed an intellectual decline which was irremediable), and a characteristic EEG image trace, described for the first time by F.A. Gibbs in Chicago and called "hypsarhythmia."

The primary form of West syndrome appears in a previously healthy brain while the secondary form develops in a brain that has been injured.

The disease had no therapeutic treatment until 1952. At that time, the epileptological considerations of our laboratory were a bit unusual. Our publications and meetings were based on an analytic conception of the epilepsy. We studied seizures, intercritic state, etiology, and EEG trace, and we envisaged no global concept. We looked for the establishment of different epileptic entities essentially according to their age of onset. We spoke about childhood epilepsy, adolescent epilepsy, and epilepsy in adults.

For us, the epilepsies appeared as many different entities. By epileptic entity, we mean a whole array of symptoms, describing a syndrome which is included in an evolutive field. The pathological dysfunctioning is probably the same, but the etiologies can be different. In view of the time it appears and its subacute evolutive field, the West syndrome had its place in this context.

In trying a new therapeutic treatment, we obviously tried it in relation to the different entities and not in relation to the different kinds of seizures.

Corticoids had been tested in other laboratories, but the results relating to seizures were disappointing. We tested them with regard to the different entities, and we thought that the entities with subacute evolution responded. This occurred with the West syndrome and to some degree with the Lennox-Gastaut syndrome.

Another study based on that same concept of epileptic entities established the existence of atypical and incomplete forms of the West syndrome. The main element of the diagnosis remains the age of onset of the first manifestations, their development, and the resistance to the classical antiepileptic treatments.

The interest in this concept is not academic. The diagnosis of the atypical or incomplete forms allowed us to propose the corticoid treatment with the chance to improve the symptomatology as though it concerned the typical triad.

The results of the therapeutics have been differently appraised in the literature because the authors do not discern the range of essential elements in the results of their statistical studies.

The results depend on the form of the primary or secondary West syndrome, on the moment of the beginning of the treatment, and on the way in which the adrenocorticotropic thyroid hormone (ACTH) is used.

In primary West syndrome, if an appropriate treatment has been prescribed in the first eight weeks of the evolution, complete recovery occurs in more than 90 percent of cases. In other children, the treatment must be undertaken because the results will be appreciable even if they are partial.

The ACTH, per os or by injection, is the primary medication. Two months of therapy is a necessary minimum to avoid a recurrence, which is frequent in that kind of disease when treatment is too short.

The results of our work were spread thanks to a meeting organized in 1958 in Chicago by Gibbs,[1] who had been honored with the Golden Brain Award from the University of Chicago.

In Europe, the diffusion of the therapy has been influenced by a 1960 meeting in Marseilles. The discovery of the treatment of the West syndrome provided the basis for a meeting where the results of many research groups were presented.[2]

1. **Gibbs F A,** ed. *Molecules and mental health.* Philadelphia: Lippincott, 1959. 123 p.
2. **Gastaut H, Soulayrol R, Roger J & Pinsard N,** eds. *L' encéphalopathie myoclonique infantile avec hypsarythmie* (*Myoclonic infantile encephalopathy with hypsarhythmia*). Paris: Masson, 1964. 223 p.

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