Reviewer’s report

Title: West syndrome followed by juvenile myoclonic epilepsy: a coincidental occurrence?

Version: 2 Date: 18 February 2013

Reviewer: ville dorothee

Reviewer’s report:

1-Remarks of reviewers were taken in account by authors for revision of the manuscript; we propose some suggestions in order to clarify message of discussion

2-Introduction: definition of idiopathic epilepsies should include data about outcome

3-Discussion

a) The relationship between the “Hairy Elbows Syndrome” and the West syndrome is still unknown since even the molecular analyses, including ARX gene, mainly involved in brain development, were negative in our patient. However, the favourable clinical course, the lack of other seizure types, and the family history of recurrence of different age-dependent epileptic weaken the link between WS and HES [2-4], making this coexistence a coincidental occurrence as previously presumed [11]. You should moderate, because you cannot exclude link between WS and HS only on the base of absence of genetic or chromosomal abnormality identified, and on the base of outcome; more and more genetic abnormalities are identified associated doi mild phenotype.

b) You should introduce this paragraph in order to clarify the message: for example “From our point of view, there is no obvious relationship between such presumed symptomatic spasms and JME, but it is striking to observe coexistence of two age-dependent epileptic syndromes in the same patient”. Recently, the transition between the more common age-dependent epilepsies has been documented and some overlapping pathophysiological processes and common genetic factors have been hypothesized [13]. Furthermore, the transition from WS to CAE has recently been described in two children[7]. Likewise, the clinical course of our patient might be read as a transition from an age-dependent epilepsy (WS) to another one (JME). The linking between WS and the JME proves to be a complex relationship since to our knowledge common genetic alterations have not been reported in the literature.

c) In agreement with this view, it has been recently hypothesized that some types of epilepsy, including WS and JME, may depend on the dysfunction and on a specific susceptibility of a given neural system to epileptogenic factors (system epilepsy) [15]. It is likely that some genes other than those currently known or
non-conventional genetic influences such as epigenetic, epistatic (?) or environmental factors play a role in seizure predisposition. Discussion of common origin of these three apparently independent diseases (presumed symptomatic WS, HS, JME) is one of the challenges of this article but this paragraph is not clear and it’s not sure that reference to “system epilepsy” provide contribution?

**Level of interest:** An article of importance in its field

**Quality of written English:** Acceptable

**Statistical review:** No, the manuscript does not need to be seen by a statistician.