Juvenile myoclonic epilepsy

Sometimes called bilateral myoclonus. 5–10% of cases of epilepsy. An idiopathic generalized epilepsy syndrome with age-related onset consisting of 3 seizure types:

1. myoclonic jerks: predominantly after waking

2. generalized tonic-clonic seizures

3. absence

EEG → polyspike discharges. Strong family history (some studies showing linkage to the HLA region on the short arm of chromosome 6).

Most are responsive to depakene.

The purpose of a study was to investigate personality characteristics and clinical parameters in two well-defined epilepsies: mesial temporal lobe epilepsy related to hippocampal sclerosis (MTLE/HS) and juvenile myoclonic epilepsy (JME) through NEO Revised Personality Inventory (NEO-PI-R) and Neurobehavior Inventory (NBI) standardized instruments.

One hundred patients undergoing corticoamygdalohippocampectomy (CAH), 100 patients with JME, and 100 control subjects answered the personality measures. Clinical parameters such as psychiatric symptoms, seizure frequency, duration of epilepsy, and side of the lesion in MTLE/HS group were investigated. Statistical analysis consisted of the mean and standard deviation (SD) of each variable. Student's t-test or Fisher exact test were used according to the variable studied.

The three groups were within the average range of NEO-PI-R and NBI, although 'tendencies' and differences were demonstrated. The MTLE/HS and control subjects had a similar profile: low scores in Neuroticism and high in Conscientiousness ($r = -0.330$; $p < 0.001$/$r = -0.567$; $p < 0.001$, respectively) in opposition to what occurred in JME, low in Conscientiousness and high in Neuroticism ($r = -0.509$; $p = 0.005$). The NBI 'sense of personal destiny' trait was higher (3.15; $p = 0.003$) in MTLE/HS than in JME and controls. The JME 'law and order' scores were lower than in other groups ($p = 0.024$). A tendency towards specific NBI traits differentiates MTLE/HS (Factor 3) from JME (Factor 1) groups. Psychiatric symptoms and seizure frequency were correlated with worse scores in NBI and, especially, in Neuroticism domain of NEO-PI-R.

Specific personality features were linked to each epileptic disease. These findings highlight the importance of considering unique features linked to epilepsy conditions in daily clinical observation to develop support programmes [1)].

Juvenile myoclonic epilepsy (JME), is commonly considered the archetypical syndrome of the idiopathic generalized epilepsies.

Genetically determined dysfunctions of important cognitive systems like visuomotor coordination and linguistic communication appear now as key mechanisms of seizure generation in JME.

A review suggests a new paradigm to consider JME as a system disorder of the brain analogous to other neurological system disorders [2].

Mutations in the Myoclonin1/EFHC1 gene are an important cause of juvenile myoclonic epilepsy (JME) in Mexican patients [3].

1)
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2)
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