

# Jeavons syndrome



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# HISTORY



In 1977 Jeavons described **eyelid myoclonia and absences** as a separate type of **photosensitive epilepsy** with this quotation:

**“Eyelid myoclonia and absences show a marked jerking of the eyelids immediately after eye closure and there is an associated brief spike and wave activity. The eyelid movement is like rapid blinking and the eyes deviate upwards, in contrast to the very slight flicker of eyelids which may be seen in a typical absence in which the eyes look straight ahead. Brief absences may occur spontaneously and are accompanied by 3 cycles per second spike and wave discharges. The spike and wave discharge seen immediately after eye closure does not occur in the dark. Their presence in the routine EEG is a very reliable warning that abnormality will be evoked by photic stimulation.”**

- **P.M. Jeavons , Consultant in Electroencephalography, Dudley Road Hospital, Birmingham; Honorary Research Fellow, University of Aston and University of Birmingham**
- **Nosological Problems of Myoclonic Epilepsies in Childhood and Adolescence**





## Absence and Myoclonic seizures

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graph TD; A[Absence and Myoclonic seizures] --> B[Epilepsy with myoclonic absences (EMA; Tassinari syndrome)]; A --> C[Facial myoclonic epilepsy with absences]; A --> D[Eyelid myoclonia and absences (ELMA; Jeavons syndrome).];
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Epilepsy with myoclonic absences (EMA; Tassinari syndrome)

Facial myoclonic epilepsy with absences

Eyelid myoclonia and absences (ELMA; Jeavons syndrome).

## Demographic data

- Onset is typically in childhood with a peak at age 6–8 years (range 2–14 years).
- Predominant in girls.
- The prevalence of Jeavons syndrome is around 3% among adult patients with epileptic disorders and 13% among those with Idiopathic generalized epilepsy with absences.



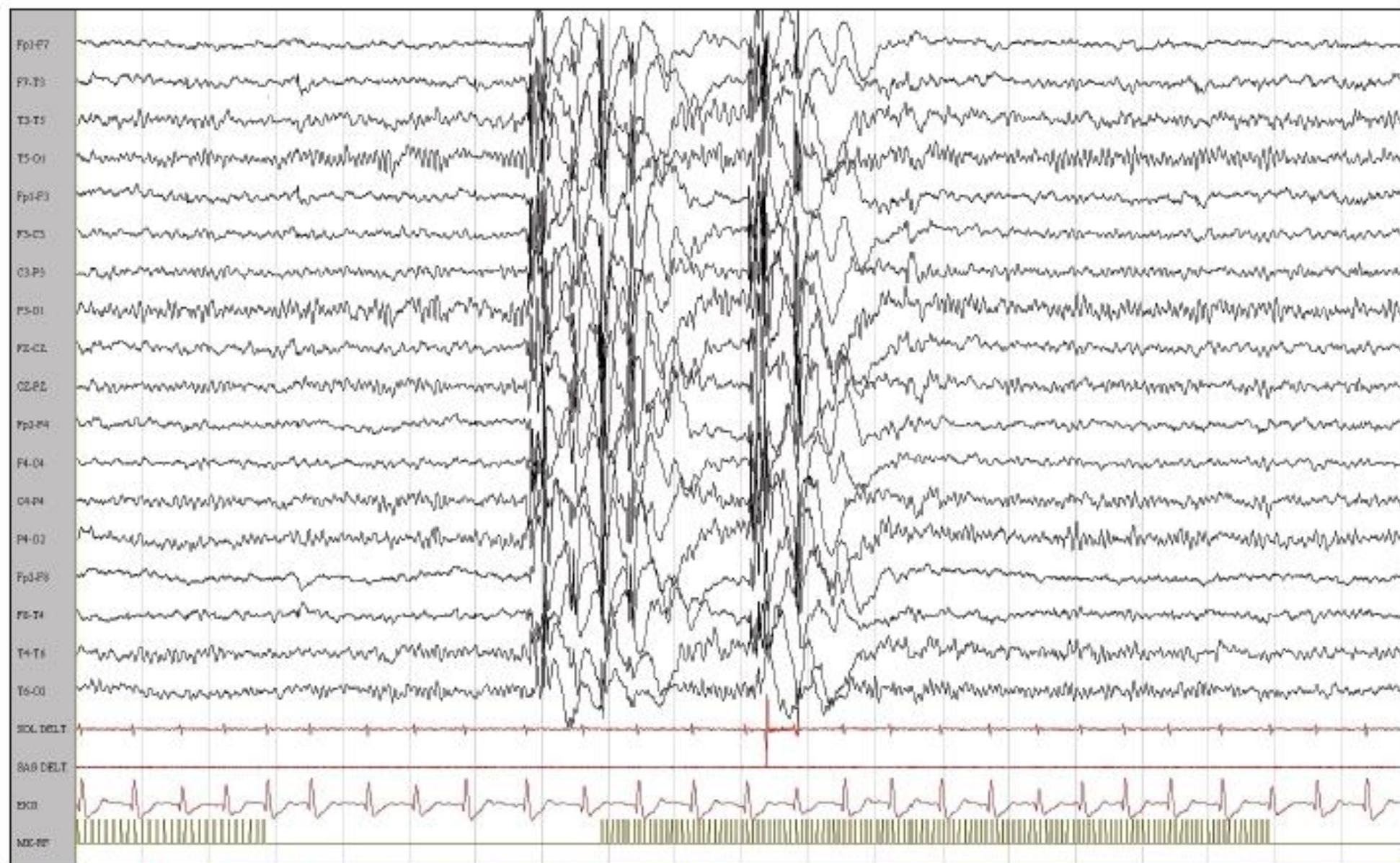
# Definition

Jeavons triad of:

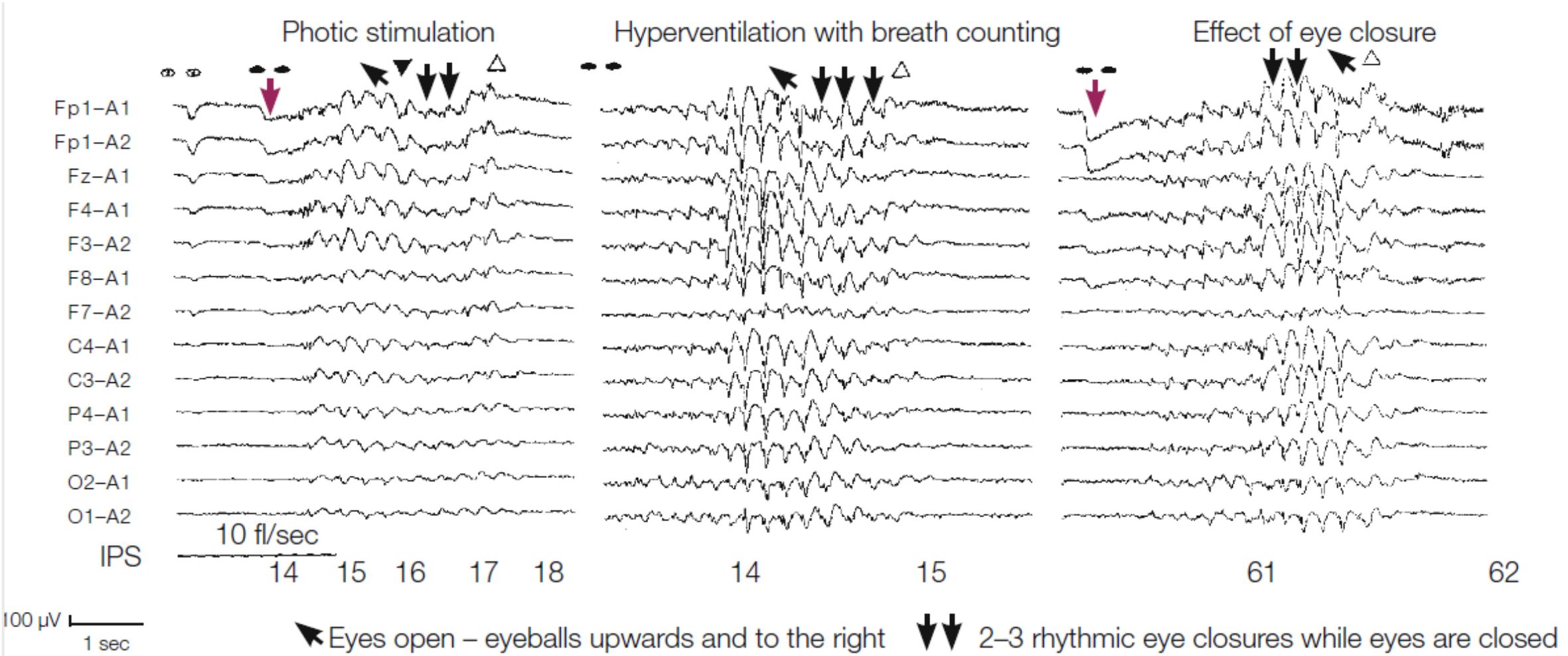
1. eyelid myoclonia with and without absences
2. eye closure-induced seizures, EEG paroxysms
3. photosensitivity

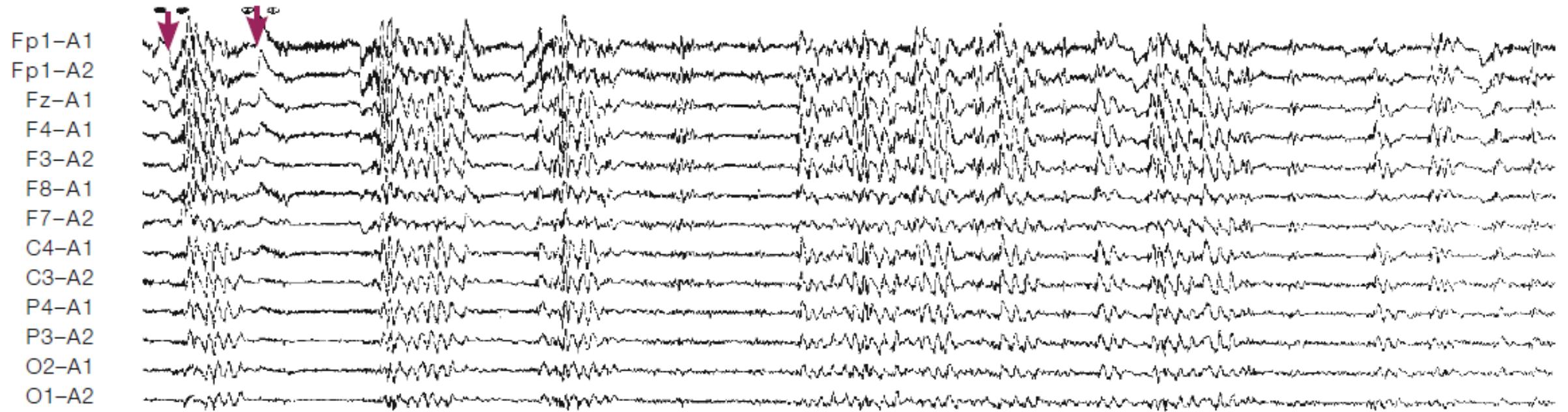
Jeavons syndrome refers to an *idiopathic reflex epilepsy*, which eyelid myoclonia is the defining seizure type.



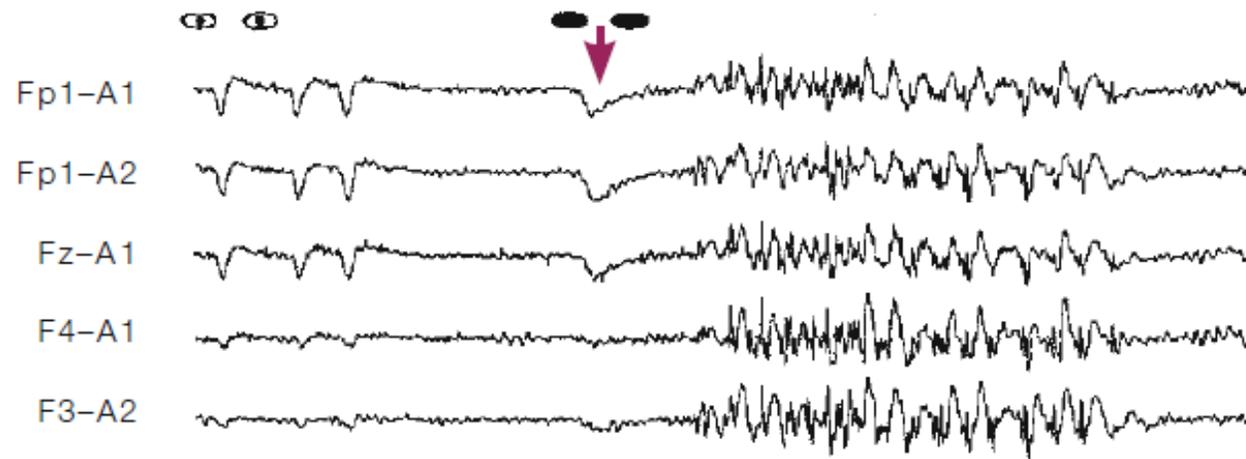


**Fig. 1.** About 2–3 s of generalized multiple spike-and-wave discharges accompanying clinically observed eyelid myoclonus during VEM of a 12-year-old male patient with JS.

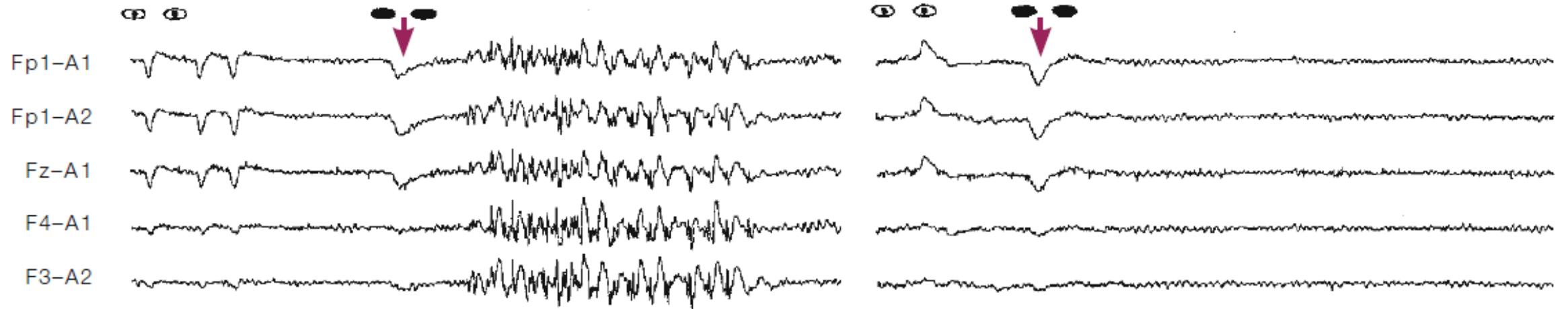


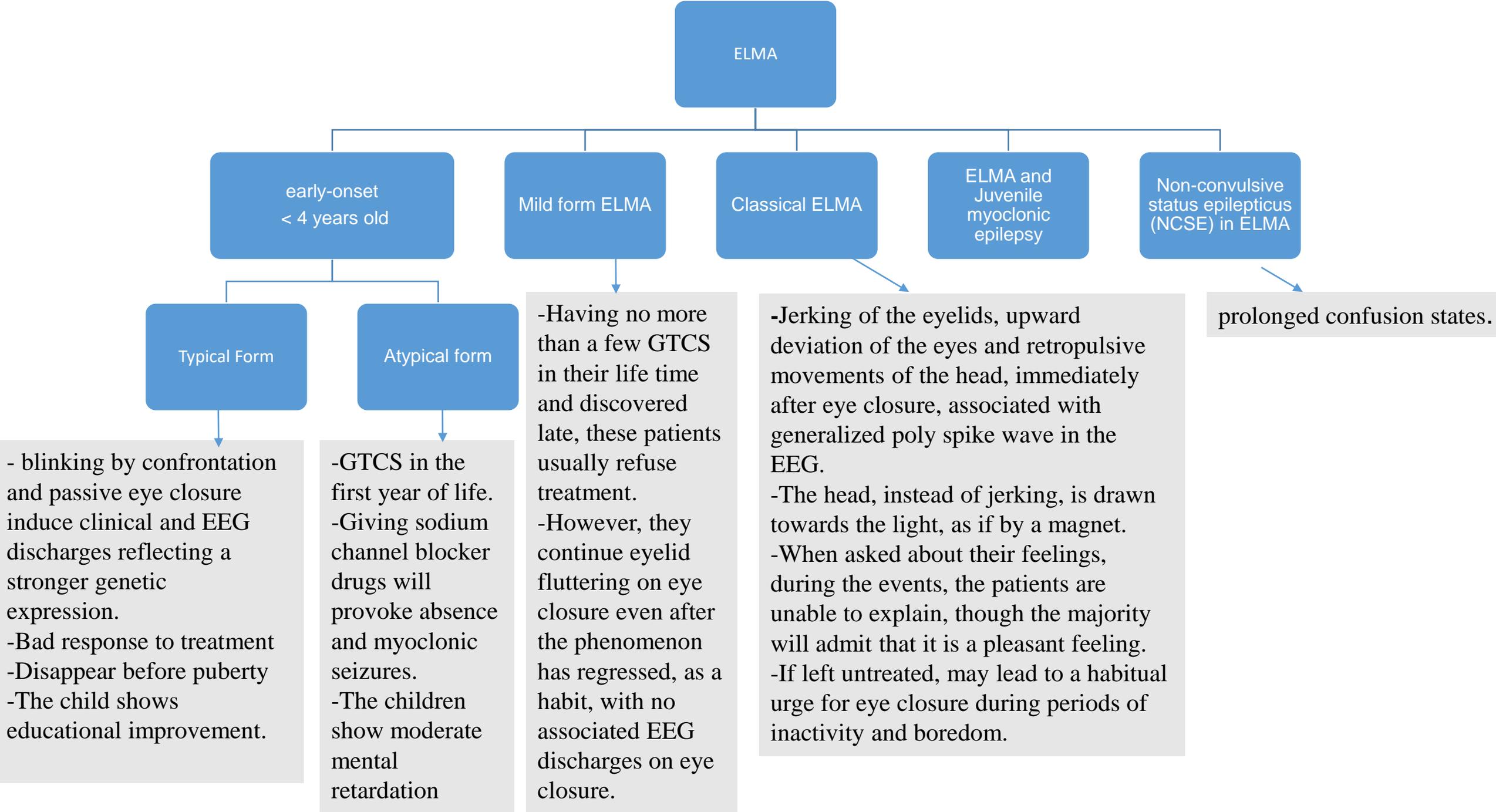


Recording in a lit room



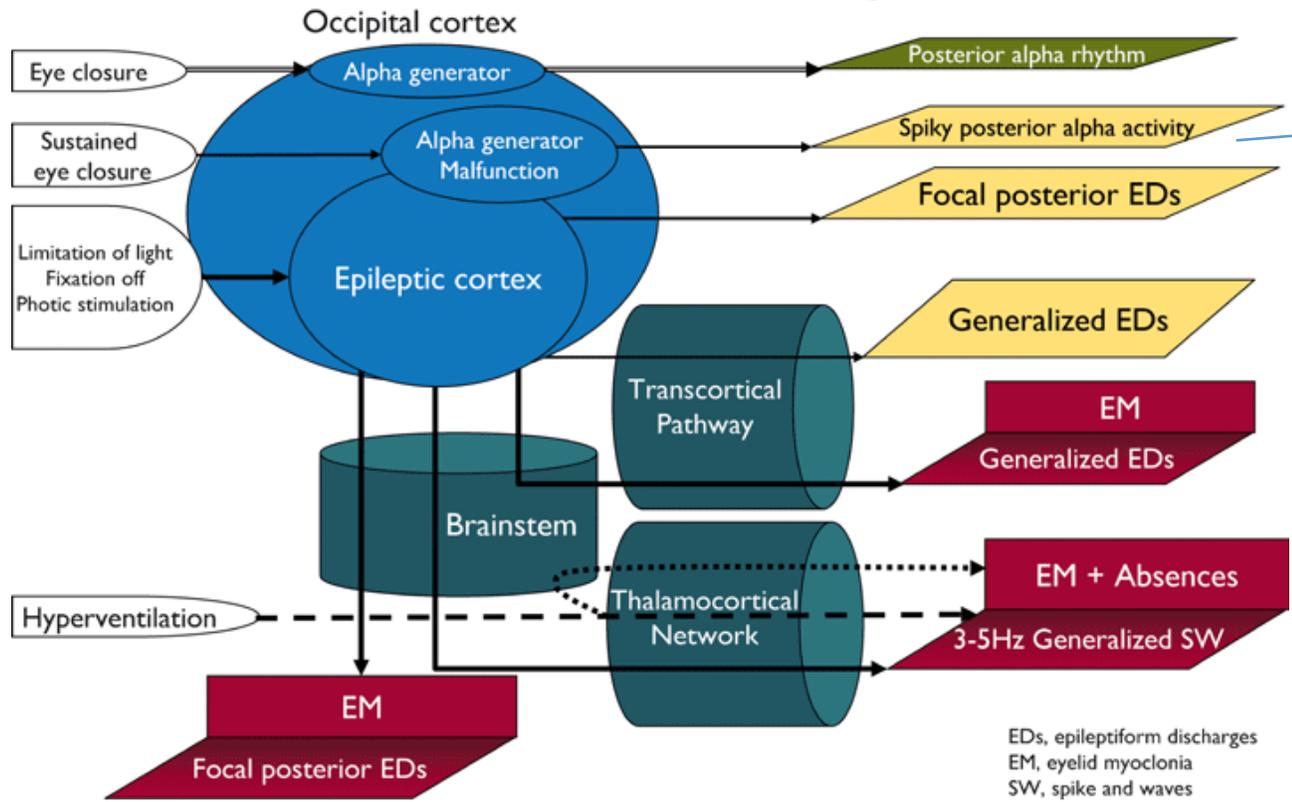
Recording in complete darkness (opaque glasses)







# Jeavons syndrome, Epileptic network, EEG and clinical findings



These spiky posterior alpha activities in JS may support the hypothesis of alpha rhythm generator malfunction in the occipital lobe

## Diagnostic procedures

All tests apart from the EEG are normal.

### Electroencephalography

Video-EEG is the single most important procedure for the diagnosis of eyelid myoclonia with or without absences. It shows frequent high-amplitude 3–6 Hz of mainly polyspikes .

Typically these are

- related to eye closure, i.e. they occur immediately (within 0.5–2 s) on closing the eyes in an illuminated recording room and they are eliminated in total darkness
- brief (1–6 s, commonly 2 or 3 s).



# Differential Diagnosis



**Table 1. Epileptic conditions sharing common features with eyelid myoclonia with absences**

Conditions with eye closure sensitivity

- Eyelid myoclonia with absences
- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Idiopathic generalized epilepsy with tonic-clonic seizures
- Idiopathic occipital lobe epilepsy

Conditions with eyelid myoclonia (with or without absences)

- Eyelid myoclonia with absences
- Childhood absence epilepsy
- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Sunflower syndrome(s)
- Self-induced seizures
- Idiopathic generalized epilepsy with phantom absences
- Myoclonic status in nonprogressive encephalopathies

Conditions with photosensitivity

- Eyelid myoclonia with absences
- Visual reflex seizures
- Sunflower syndrome(s)
- Benign myoclonic epilepsy in infancy
- Childhood absence epilepsy
- Juvenile myoclonic epilepsy
- Idiopathic generalized epilepsy with tonic-clonic seizures
- Juvenile absence epilepsy
- Idiopathic photosensitive occipital lobe epilepsy
- Severe myoclonic epilepsy of infancy (Dravet syndrome)
- Progressive myoclonic epilepsies
- Alcohol or benzodiazepines withdrawal

Eyelid myoclonia is often misdiagnosed as facial tics, sometimes for many years.

# Management

- Valproate alone, or most probably in combination with clonazepam, levetiracetam, lamotrigine or ethosuximide, appears to be the most effective regimen.
- Clonazepam monotherapy is highly efficacious in eyelid myoclonia and myoclonic jerks;
- Levetiracetam may be the most effective, because of its anti myoclonic and anti photosensitive properties.
- Lamotrigine is very effective in absence seizures but may exaggerate myoclonic jerks.
  
- Non-pharmacological treatments used for photosensitive patients (such as wearing special glasses or the newly commercially available blue Z1 lenses)



## Prognosis

- Jeavons syndrome is a lifelong disorder, even if seizures are well controlled with AEDs.
- Men have a better prognosis than women.
- There is a tendency for photosensitivity to disappear in middle age, but eyelid myoclonia persists.
- It is highly resistant to treatment and occurs many times a day, often without apparent absences and even without demonstrable photosensitivity.



# Take Home Message

- As a simple rule of thumb, eyelid myoclonia is highly suggestive of Jeavons syndrome. This becomes more likely when eyelid myoclonia is combined with photosensitivity, and it is pathognomonic of the syndrome when it also occurs after eye closure.
- The diagnosis of Jeavons syndrome is simple because the characteristic eyelid myoclonia, if seen once, will never be forgotten or confused with other conditions. Furthermore, the EEG with the characteristic eye-closure-related discharges and photosensitivity leaves no room for diagnostic error.



# References

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