

# Epileptic spasms without hypsarhythmia in infancy and childhood: tonic spasms as a seizure type

Luciana R De Marchi<sup>1</sup>, Evelyn A Seraphim<sup>1</sup>, Jeana T Corso<sup>1</sup>,  
Pedro VF Naves<sup>1</sup>, Kelly Cristina de Carvalho<sup>1</sup>, Milton David H  
Ramirez<sup>2</sup>, Taissa Ferrari-Marinho<sup>1</sup>, Mirian SB Guaranha<sup>1</sup>,  
Elza Márcia T Yacubian<sup>1</sup>

<sup>1</sup> Department of Neurology, Neurosurgery, Universidade Federal de São Paulo, São Paulo, Brazil

<sup>2</sup> Department of Neuropediatrics, Universidad Militar de Colombia, Bogotá, Colombia

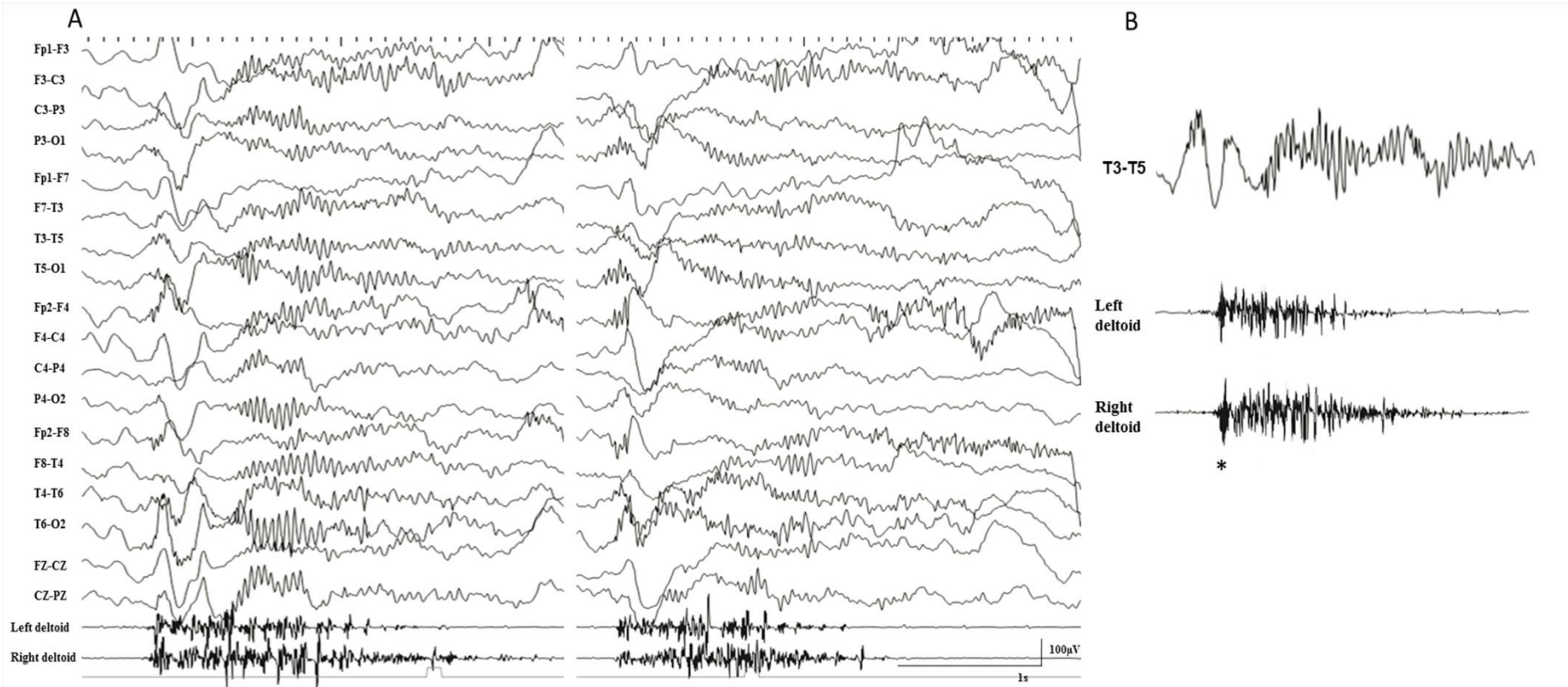
Received December 03, 2014; Accepted February 08, 2015

# Epileptic spasms without hypsarrhythmia and tonic spasms

- Epileptic spasms are still considered as part of the group of unknown seizure types.
- ES without hypsarrhythmia have been described in some series of patients, occurring either in infancy or childhood.
- More prolonged ES were defined as a different seizure type by Fusco and Vigevano (1994), who proposed to separate these from true ES, and suggested the term “tonic spasm seizures”.

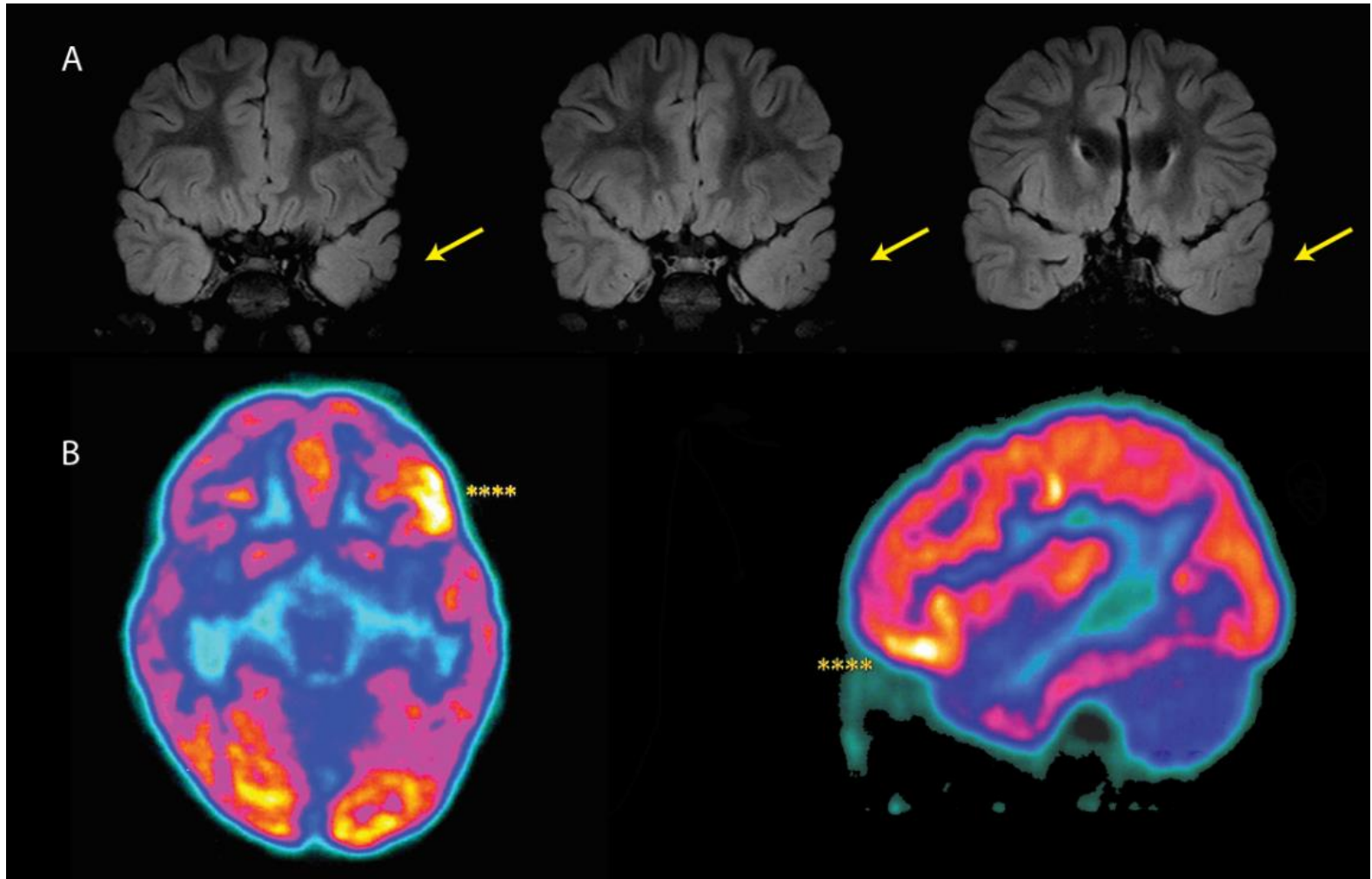
# Case study

- A five-year-old boy who had ES without hypsarrhythmia at eight months of age, effectively treated with oxcarbazepine.
- With the withdrawal of this medication, ES returned, and the patient was refractory to several medications.



**Ictal EEG: diffuse periodic complexes of high-voltage slow waves, superimposed with low-voltage fast activity, followed by an electrodecremental phase with fast activity associated with tonic spasms. (B) Details of complexes: a brief low-voltage fast activity burst, superimposed onto a slow wave, with the characteristic diamond shape (\*) on deltoid EMG.**

(A) MRI showed left temporal lobe atrophy, grey/white matter blurring, and increased T2/FLAIR signal in the left temporal pole (arrows). (B) Ictal PET-CT showed left frontal hypermetabolism (dots).



# Case study

- Vigabatrin was introduced with remission of seizures.
- Video-EEG monitoring three months later revealed normal background activity.

# Epileptic spasms without hypsarrhythmia and tonic spasms

- ES without hypsarrhythmia associated with tonic spasms may be a variant of focal spasms.
- As suggested by Caraballo et al. (2011), this might be considered an epileptic syndrome referred to as “ES without hypsarrhythmia in infancy and childhood.”

# References

- Berg AT, Berkovic SF, Brodie MJ, et al. Revised terminology and concepts for organization of seizures and epilepsies: report of the ILAE Commission on Classification and Terminology, 2005-2009. *Epilepsia* 2010; 51(4): 676-85.
- Caraballo RH, Fejerman N, Bernardina BD, et al. Epileptic spasms in clusters without hypsarrhythmia in infancy. *Epileptic Disord* 2003; 5: 109-13.
- Fusco L, Vigevano F. Tonic spasm seizures: a particular and previously unreported type of seizure. *Epilepsia* 1994; 35(7): S87.
- Ishikawa N, Kobayashi Y, Fujii Y, Tajima G, Kobayashi M. Ictal electroencephalography and electromyography features in symptomatic infantile epileptic encephalopathy with late-onset spasms. *Neuropediatrics* 2014; 45(1): 36-41.
- Kellaway P, Hrachovy RA, Frost JD Jr, Zion T. Precise characterization and quantification of infantile spasms. *Ann Neurol* 1979; 6(3): 214-8.