

Benign spasms of infancy or benign myoclonus of early infancy: polygraph-EEG recordings

Nelly Álvarez Álvarez¹, Rafael Pardo de la Vega¹, María Antonia Gutierrez Trueba², Luis Santovenía González³, Ana González Acero¹, Raquel Blanco-Lago⁴, Antonio Hedrera-Fernandez⁵, Ignacio Málaga Diéguez⁴

¹ Pediatric Neurology, Hospital Universitario de Cabueñas Ringgold standard institution, Gijon, Asturias, Spain

² Neurophysiology Department, Hospital Universitario de Cabueñas Ringgold standard institution, Gijon, Asturias, Spain

³ Neurophysiology Department, Hospital Universitario Central de Asturias Ringgold standard institution, Oviedo, Asturias, Spain

⁴ Pediatric Neurology, Hospital Universitario Central de Asturias Ringgold standard institution, Oviedo, Spain

⁵ Neuropediatría, Hospital Universitario Central de Asturias Ringgold standard institution, Hospital Universitario Central de Asturias Avenida de Roma s/n , Oviedo, Asturias 33011, Spain

Received December 8, 2020; Accepted March 18, 2021

Benign myoclonus of early infancy (BMEI) is a well-defined entity of uncertain etiology, characterized by nonepileptic paroxysmal motor phenomena affecting the muscles of neck, trunk and upper limbs.

It appears in children with normal psychomotor development in the first year of life and has a good long-term prognosis, spontaneously resolving before age 2 years [1-2].

The spectrum of motor phenomena observed in this entity is wide and includes myoclonic jerks, spasms, brief tonic contractions and negative myoclonus. Distinction between BMEI and infantile epileptic disorders can be challenging given the clinical similarities [2-3].

Ictal electroencephalography (EEG) recording is always normal, which makes it the gold standard test for

the differential diagnosis between this entity and the epileptic syndromes in infancy, mainly West syndrome.

Although this entity is widely known, its polygraphic recording is rarely shown in the literature. The EEG recording with polygraphy and video of our two patients helps to the diagnosis of this paroxysmal nonepileptic disorder [4-5].

Case 1: A 2-month-old boy presents repeated brief episodes of elevation of upper limbs without changes in alertness twelve hours after vaccination (diphtheria, tetanus, pertussis) (video sequence 1).

Case 2: A 11-month-old girl presents repeated head drops with no other associated symptoms (video sequence 2).



VIDEOS ONLINE



- Correspondence:
Nelly Álvarez Álvarez
Pediatric Neurology,
Hospital Universitario de
Cabueñas Ringgold standard
institution,
Gijon, Asturias, Spain
<nellyavz612@gmail.com>

Legends for video sequences

Video sequence 1

The video-EEG records episodes compatible with spasms without changes in the EEG tracing.

Video sequence 2

In the video-EEG no ictal modification is observed, only a fast activity produced by motion artifact.

Key words for video research on www.epilepticdisorders.com

Phenomenology: nonepileptic paroxysmal motor phenomena, myoclonic jerks, spasms, brief tonic contractions and negative myoclonus

Localization: neck, trunk and upper limbs

Syndrome: benign spasms of infancy, benign myoclonus of early infancy

Aetiology: unknown

Disclosures.

None of the authors have any conflict of interest to declare.

References

1. Dravet C, Giraud N, Bureau M, Roger J, Gobbi G, Bernardina Dalla B. Benign myoclonus of early infancy or benign non-epileptic infantile spasms. *Neuropediatrics* 1986; 17(1): 33-8.
2. Caraballo RH, Capovilla G, Vigevano F, Beccaria F, Specchio N, Fejerman N. The spectrum of benign myoclonus of early infancy: clinical and neurophysiologic features in 102 patients. *Epilepsia* 2009; 50(5): 1176-83.
3. Ghossein J, Pohl D. Benign spasms of infancy: a mimicker of infantile epileptic disorders. *Epileptic Disord* 2019; 21(6): 585-9.
4. Maydell BV, Berenson F, Rothner AD, Wyllie E, Kotagal P. Benign myoclonus of early infancy: an imitator of West's syndrome. *J Child Neurol* 2001; 16(2): 109-12.
5. Pachatz C, Fusco L, Vigevano F. Benign myoclonus of early infancy. *Epileptic Disord* 1999; 1(1): 57-61.