Original article with video sequences

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Shaking body attacks: a new type of benign non-epileptic attack in infancy

Giuseppe Capovilla

Epilepsy Center, Department of Child Neuropsychiatry, C. Poma Hospital, Mantova, Italy

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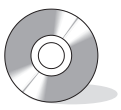
ABSTRACT – Non-epileptic attacks represent a heterogeneous group of clinical entities which frequently pose a challenge for the differential diagnosis of epilepsy. This is particularly the case when motor manifestations are the main clinical features. For the large majority of patients, such motor manifestations have a benign course. A correct diagnosis is important to avoid inappropriate investigations, unnecessary therapy, and parental anxiety.

Here, a previously unreported form of non-epileptic attacks with infantile onset is described which is different from all subtypes of Fejerman syndrome and does not appear to be uncommon. Our series includes 23 patients with an age at onset of the paroxysmal events ranging from 3 to 8 months. The characteristic feature is side-to-side shaking movements of the trunk and limbs. Surprisingly, urinary infection is often a false diagnosis. Home video recording is particularly helpful in recognising the nature of these episodes once their existence is known. *[Published with video sequences]*

Key words: Non-epileptic attacks, infancy, paroxysmal disorders, shuddering, movement disorders, Fejerman syndrome



Correspondence:
G. Capovilla
Epilepsy Center,
Dept. of Child Neuropsychiatry,
"C. Poma" Hospital
46100 Mantova, Italy
<pippo.capovilla@aopoma.it>

More than 30 years ago, Natalio Fejerman reported ten patients presenting with non-epileptic attacks (NEAs) which could be misdiagnosed as infantile spasms of West syndrome (Fejerman, 1976). In the following years, the same and other authors described similar cases in different areas of the world (Vanasse *et al*., 1976; Fejerman and Medina, 1977; Fejerman, 1977; Lombroso and Fejerman, 1977; Giraud, 1982; Gobbi *et al*., 1982; Fejerman, 1984; Holmes and Russman, 1986; Dravet *et al*., 1986; Beltramino, 1987; Galletti *et al*., 1989; Caviedes Altable *et al*., 1992; Martinez Pastor *et al*., 1993; Pachatz *et al*., 1999; Kanazawa, 2000; Maydell *et al*., 2001; Fernandez Alvarez and Aicardi, 2001a, 2001b; Fejerman and Caraballo, 2002; Pachatz *et al*., 2002; Fujikawa *et al*., 2003; Pranzatelli, 2003; Fejerman, 2008), confirming the existence of this benign non-epileptic condition. In these articles, the attacks were described both as true myoclonic and brief tonic, and polygraphic recording of muscle activity confirmed the presence of these different types of attack.

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Another less frequent type of attack was characterized by head drops or loss of tone in the trunk, suggesting negative myoclonus of non-epileptic nature. Furthermore, in a relevant number of cases, the attacks were described as shuddering of the head, shoulders, upper limbs or facial muscles of longer duration, for up to several seconds. A recent report of Caraballo *et al*. (2009) described a large group of infants presenting with all these four types of NEAs, sometimes with more than one type of motor phenomenon in the same patient. Following this publication, Dalla Bernardina (2009) proposed that these infants had Fejerman syndrome.

Here, we describe a previously unreported form of NEA in infancy, distinct from Fejerman syndrome. The infants are clinically different from those with Fejerman syndrome and the differential diagnosis frequently includes conditions other than epilepsy. We propose to name this condition "benign infantile shaking body attacks (BISBA)", since the movements during these episodes are reminiscent of a popular dance of some decades ago.

Patients and methods

We reviewed the clinical charts and EEG data of patients referred to the Epilepsy Center of Mantova between January 1999 and December 2008 for non-epileptic abnormal movements in the first year of life. Exclusion criteria were:

– abnormal neurological examination or neuropsychological development prior to onset;
– occurrence only during sleep;
– occurrence as a reflex reaction to posture or stimuli;
– Non-paroxysmal motor abnormalities or abnormal movements lasting for more than one minute;
– epileptiform interictal or ictal EEG.

All the patients featuring one of the four types of attacks described by Caraballo *et al*. (*i.e*. cases of Fejerman syndrome) were also excluded. In the remaining cases, the following were evaluated: family and personal history, age at onset, the pattern of occurrence (isolated or in clusters), number of episodes per day; time of day, triggering factors, age at disappearance, neuropsychological development, and association with developmental disorders. A minimum follow-up period of 12 months from the disappearance of attacks was a required inclusion criterion.

Results

General data

Twenty-three patients were collected (13 [58%] male and 10 [42%] female). The age at onset ranged from three to eight months, with a mean age of 5.5 months and a median of six months. For five patients there was a family history of epilepsy and febrile convulsions for one patient.

Clinical manifestations

Home video recordings were available for all cases. In addition, ictal video-polygraphic recording was obtained for nine patients. Ictal manifestations were stereotyped in all patients *(table 1)*. The movement was an abrupt and short side-to-side movement of the trunk involving all four limbs *(figure 1; see video sequences*), with asynchronous involvement of

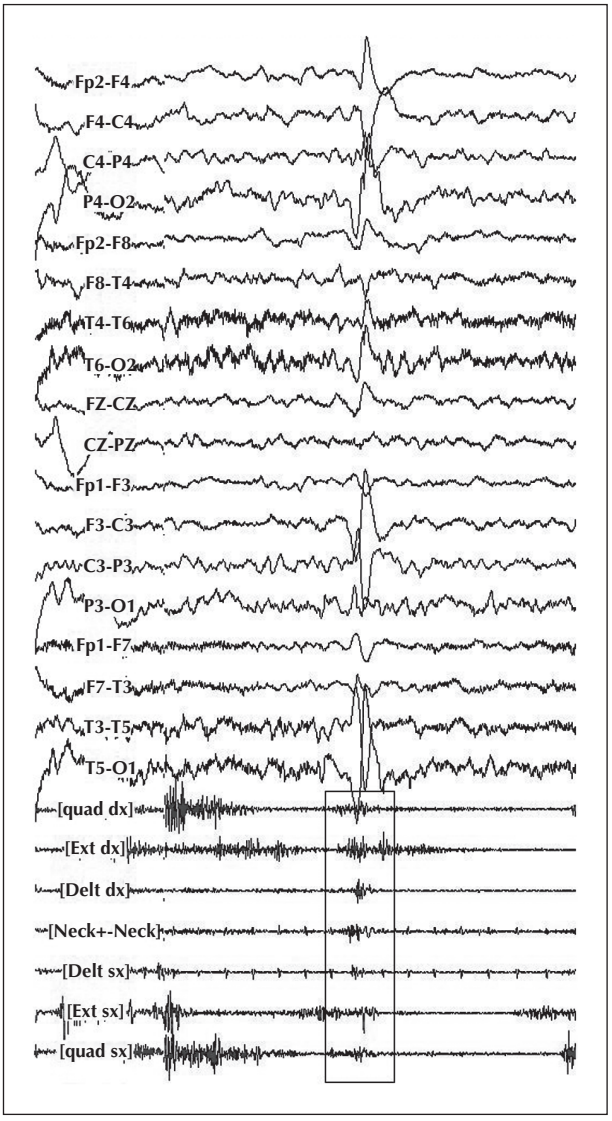

Figure 1. Polygraphic recording of an attack demonstrating asynchronous involvement of all muscles investigated, with variable intensities. The EEG derivations show artefact due to the shaking attacks.

different muscles of both sides of the entire body. The episode resembled the single movement of a popular dance of some decades ago ("the shake"). Movements could be more or less intense and accompanied by crying of the infant. In one patient with independent walking, the attack could lead to a fall to the ground. Episodes usually occurred in the awake state but could occur during drowsiness or also in the first phase of sleep. Episodes were never observed during slow sleep by either myself or parents. In most cases, the episodes occurred several times per day (16/23, 69%), although, in some cases, they were less common, despite an awareness of their short duration. For this reason, it is difficult to be sure about the true frequency of the phenomenon. Triggering factors were sometimes reported for some infants. None of the patients presented with different types of attacks which are included in the so-called spectrum of benign myoclonus of early infancy or Fejerman syndrome.

Table 1. Clinical data of the 23 patients.

Patient	Present age	Family history	Sex	Age at onset (months)	Age at disappearance (months)	duration (months)	Associated with sleep	Isolated	Series	Episodes per day
1	7y	Negative	M	6	7	1	No	Yes	Yes	Multiple
2	9y	FC	M	3	11	7	No	Yes	No	Rare
3	4y7m	E	M	4	8	4	No	No	Yes	Multiple
4	6y	Negative	F	6	12	6	No	Yes	No	Rare
5	8y	Negative	M	6	12	6	No	Yes	No	Multiple
6	6y6m	Negative	F	3	6	3	No	Yes	No	Multiple
7	7y3m	E	M	4	6	2	Yes	Yes	No	Rare
8	11y	E	F	2		5	Yes	Yes	No	Multiple
9	12y	Negative	M	6	10	4	No	Yes	No	Multiple
10	9y	Negative	F	8	9	1	No	Yes	Yes	Multiple
11	7y	Negative	F	8	12	4	No	Yes	No	Multiple
12	6y	Negative	M	8	14	6	No	Yes	Yes	Multiple
13	6y6m	Negative	M	7	13	6	No	Yes	Yes	Multiple
14	3y6m	Negative	M	5	9	4	Yes	No	No	Multiple
15	3y	Negative	M	6	11	5	Yes	Yes	No	Rare
16	7y7m	Negative	F	5	7	2	No	No	Yes	Multiple
17	7y	Negative	F	7	10	3	No	Yes	Yes	Multiple
18	4y	Negative	M	7	11	4	Yes	Yes	No	Multiple
19	5y	Negative	F	5	10	5	No	Yes	No	Multiple
20	3y	E	F	6	15	9	Yes	No	Yes	Rare
21	2y1m	Negative	M	5	12	7	Yes	Yes	No	Sporadic
22	2y3m	E	F	3	10	7	No	No	Yes	Multiple
23	1y11m	Negative	M	7	11	4	No	Yes	Yes	Sporadic

M: male; F: female; y: year; m: month; FC: febrile convulsions; E: epilepsy.

Evolution and prognosis

For all individuals, the episodes tended to reduce in frequency over time and disappeared between six and 15 months of age (mean 10.3; median 11 months), and in most cases (15/23, 65%), before the end of the first year of life. The attacks persisted for a period lasting between one and nine months (mean 4.3; median 4.5 months). Language and cognitive development were normal in all cases.

Differential diagnosis

Unlike the patients described in the literature in the spectrum of benign myoclonus or Fejerman syndrome, the overwhelming majority of the cases described here were not subjected to referral due to a suspicion of epileptic seizures. Indeed, the belief of the parents and, often, of the paediatricians, was that the infant had a probable infection of the urinary tract and that urination could induce a sensation of urethral "burning", responsible of the clinical phenomenon. The consequence was that all these patients had repeated urinalysis, which proved to be normal in all cases. Some patients were investigated based on suspicion of a seizure disorder, due to a positive familial history of epilepsy or febrile convulsions.

Discussion

The differential diagnosis between epileptic and non-epileptic attacks is extremely challenging for epileptologists. In some cases, this can be particularly difficult due to confounding factors and a positive family history for convulsive disorders may create anxiety for the parents. Moreover, as well as the differential diagnosis, the diagnosis *per se* poses a significant challenge. Indeed, in modern diagnostic medicine, the patients or their parents may demand a definitive diagnosis and are not satisfied with generic answers. Video-EEG capture of these events is not always possible but domestic videos may be easily recorded by the parents using both cameras and mobile telephones. From a nosological point of view, the cases presented here might be classified within the spectrum of benign myoclonus of early infancy, however, the peculiar clinical symptoms clearly differentiate these cases from others previously described (Caraballo *et al*., 2009). In particular, the common characteristic feature of patients described with spasms in the literature is the myoclonic or brief tonic jerks with both a symmetric and forward movement. On the other hand, the patients described here demonstrated a side-to-side movement with asynchronous involvement of different muscles of both sides of the entire body, as documented by video and polygraphic recordings. In conclusion, this peculiar clinical picture can be added to the spectrum of non-epileptic attacks in the first year of life. Recognition of such episodes is important both to avoid making an incorrect diagnosis of epileptic seizures and repeating unnecessary examination, such as CT and MRI scans or urinalysis. In the cases described here, diagnosis of infection of the urinary tract could not be easily discarded until a definitive diagnosis was made. □

Legends for video sequences

Video sequence 1
A six-month-old infant (Patient 1), some weeks before starting similar episodes. Note in the awake state, the abrupt side-to-side movement, followed by crying. The episodes persisted for one month.

Video sequence 2
A six-month-old infant (Patient 3). From four months of age, sporadic attacks occurred in the awake state. The video shows different episodes characterized by a side-to-side movement involving the trunk and the four limbs. In the second part of the video, the attacks were clustered in a short series. The polygraphic recording demonstrates asynchronous involvement of muscles of both sides of the body. At the age of eight months, the attacks stopped.

Video sequence 3
A twelve-month-old infant (Patient 20). In this episode, the polygraphic recording clearly demonstrates the asynchronous shaking movement limited to the four limbs. Note the absent of neck involvement. The infant is drowsy and the abrupt occurrence of the attack wakes her.

Video sequence 4
(Patient 20). The patient from *video 3* is shown with another attack in drowsiness. The shaking movement is faster than that in *video 3* and there is also a side-to-side involvement of the trunk. The infant continues to sleep.

Disclosure.
None of the authors has any conflict of interest or financial support to disclose.

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