Original article with video sequences

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Head atonic attacks: a new type of benign non-epileptic attack in infancy strongly mimicking epilepsy

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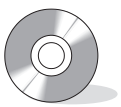
ABSTRACT – Non-epileptic attacks (NEAs) are a heterogeneous group of clinical entities which often complicate the differential diagnosis of epilepsy. NEAs usually have a benign course and are limited to a specific period of life. If motor manifestations are strongly suggestive of an epileptic phenomenon, the risk of misdiagnosis is greater. Here, we describe a novel NEA with infantile onset, characterised by repeated head drops, mimicking epileptic negative myoclonus of the neck. The frequency of the episodes was very high, at hundreds or thousands per day. The episodes appeared in the second semester of the first year of life and spontaneously disappeared within a few months. [*Published with video sequences*]

Key words: NEAs, non-epileptic attacks, infancy, paroxysmal disorders, atonic attacks



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The false diagnosis of epileptic infantile spasms for patients with myoclonic manifestations has been known since the seventies (Fejerman, 1977a; Fejerman, 1977b). Similar cases were subsequently described worldwide (Vanasse *et al*., 1976; Fejerman and Medina, 1977; Fejerman, 1977b; Lombroso and Fejerman, 1977; Giraud, 1982; Gobbi *et al*., 1982; Fejerman, 1984; Holmes and Russman, 1986; Dravet *et al*., 1986; Galletti *et al*., 1989; Caviedes Altable *et al*., 1992; Pachatz *et al*., 1999; Kanazawa, 2000; Maydell *et al*., 2001; Fernandez Alvarez and Aicardi, 2001; Fejerman and Caraballo, 2002; Pachatz *et al*., 2002; Fujikawa *et al*., 2003; Fejerman, 2008), and the existence of this benign non-epileptic condition has been repeatedly confirmed. Polygraphic video-EEG recordings of these non-epileptic attacks (NEAs) has provided evidence of different types of motor manifestation and include: true myoclonic activity, brief tonic activity, loss of tone in the trunk, and shuddering of the head, shoulders, and upper limbs or facial muscles with a long duration of up to several seconds. A recent report by Caraballo *et al*. (2009) described a large group of infants with NEAs,

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presenting with all four types of motor manifestation, with rarely more than one type of motor phenomenon in the same patient. Among these patients, cases of head drops were included, however, the head drops were more prolonged and sustained, and often lateral, completely different from the cases presented here (Caraballo, personal communication). Subsequently, Capovilla (2011) described a large number of patients presenting with a peculiar clinical phenomenon of non-epileptic nature and named this condition "shaking body attacks". More than 20 years ago, Brunquell *et al*. (1990) described a group of 24 paediatric patients presenting with three different kinds of head drop, of both epileptic (17/24) and non-epileptic (7/24) nature. However, the seven cases with non-epileptic attacks presented with different types of epileptic seizures and were often mentally retarded. Here, we describe three neurologically and mentally normal non-epileptic patients, both at onset and during follow-up. The patients presented with a novel NEA with infantile onset, characterised by repeated head drops that could be easily confused with an infantile epileptic condition in the first year of life.

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 2010 for non-epileptic abnormal movements in the first year of life. Exclusion criteria were the presence of abnormal neurological examination or neuropsychological development prior to onset and interictal or ictal EEG epileptiform activity. We selected three patients with similar features, with regards to both electro-clinical manifestations and profile of evolution. In these cases, we evaluated: family and personal history of neurological disease, age at onset, pattern of occurrence (isolated or in clusters), frequency per day, time of day of occurrence, triggering factors, age at disappearance, neuropsychological development, and association with developmental disorders. A minimum follow-up period of 18 months from disappearance of non-epileptic abnormal movements was a required inclusion criterion.

Results

General data

Data was collected for three patients (two female and one male). The age at onset ranged from 6 to 7 months. For one patient, there was a family history of febrile convulsions.

Clinical manifestations

Both home videos and ictal video-EEG polygraphic recordings were obtained for all patients. Ictal manifestations were stereotyped for all infants. The movement was an abrupt loss of muscle tone of the neck, causing a head drop (*figure 1 and video sequences 1, 2, and 3*). Head drops were more or less intense and accompanied by crying of the infant. In all cases, the episodes occurred several times per day, characteristically in cluster, and tended to be present throughout the day. Triggering factors were not reported for any infants (*table 1*). None of the patients presented with different types of attacks included in the so called "spectrum of benign myoclonus of early infancy" or Fejerman syndrome.

Evolution and prognosis

For all the individuals, the episodes tended to reduce in frequency over time and disappeared by the end of the first year of life. The attacks persisted for a period lasting for five to six months. Language and cognitive development were normal in all our cases.

Patient 1
A male patient was born with uneventful pregnancy and normal perinatal period. Growth and psychomotor development was normal. At 6 months and 3 weeks of age, the infant showed head dropping movements. Initially, these movements were observed rarely by the parents, but on some days they became very frequent (20-30 per day). There was no triggering factor and often episodes clustered without any circadian correlation. Episodes were recorded during repeat EEG performed at another epilepsy centre. Artefacts resembling high-voltage, slow waves were considered to be the EEG correlate of head drops and the infant was believed to be epileptic. Levetiracetam was started (20 mg/Kg/die), but episodes persisted. At the age of 9 months, the infant was referred to our epilepsy centre where we recorded the episodes during a video-EEG polygraphic recording (*video sequence 1*) which revealed the non-epileptic nature of the attacks. Levetiracetam was rapidly withdrawn and episodes persisted until the age of 12 months when they completely disappeared. Psychomotor development was normal during follow-up at age 2 years and 10 months.

Patient 2
A female was born with uneventful pregnancy and normal perinatal period. Psychomotor profile was normal. Febrile seizures were reported for the mother. The patient presented with several episodes of head dropping per day at the age of 7 months. In some

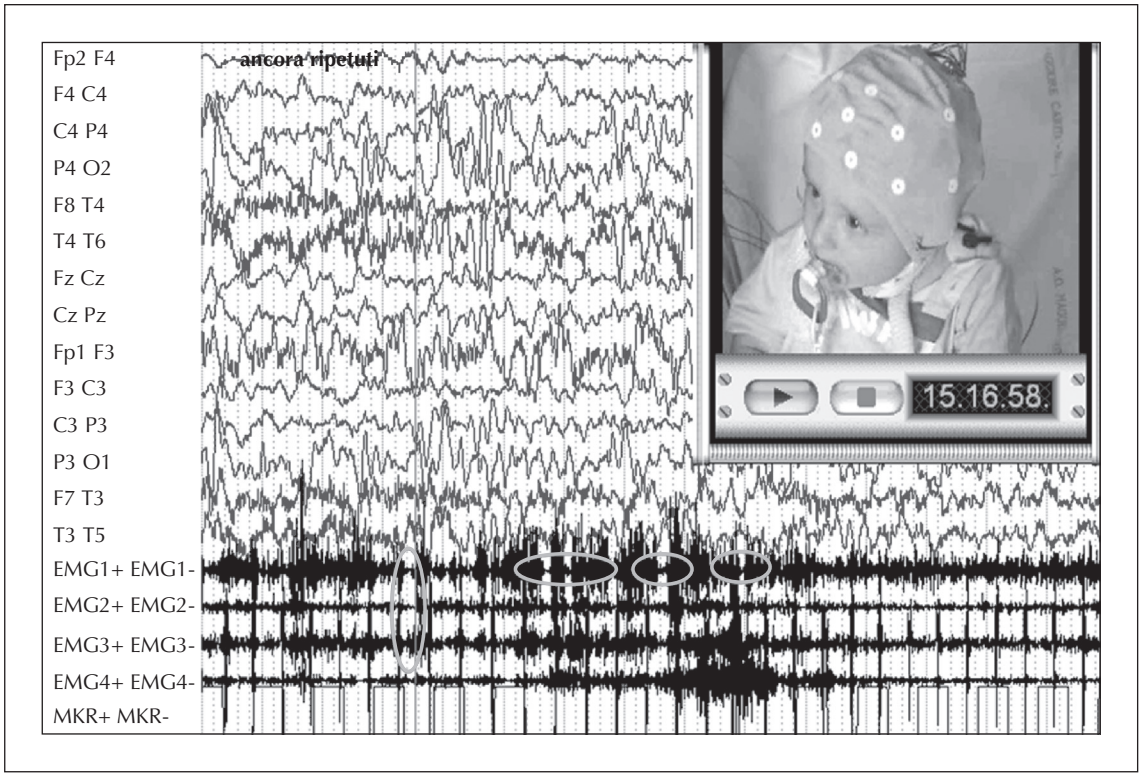

Figure 1. Case 3. Polygraphic recording of repeated head atonic attacks.
The EMG trace of the neck muscles (EMG1) is characterised by a flattening (ellipsis), not evident in the other muscles examined (right deltoid EMG2, left deltoid EMG3, and right radial triceps EMG4). Note the absence of EEG correlate.

Table 1. Clinical data of patients.

Patient	Age at follow-up	Family history	Sex	Age at onset	Age at disappearance	Persistence	Isolated	Series	Frequency	Triggering factors
1	2y 10m	negative	M	6m 3w	12m	5m 1w	yes	yes	daily	no
2	2y 11m	FC	F	7m	12m	5m	yes	yes	daily	no
3	10y	negative	F	6m	11m	5m	yes	yes	daily	no

M: male; F: female; FC: febrile convulsions; y: years; m: months; w: weeks.

instances, the infant presented with many episodes in a few minutes without triggering factors or circadian correlation. Manifestations were recorded in another epilepsy centre during video-EEG monitoring and a diagnosis of epileptic spasms was made. Vigabatrin was prescribed but head drops persisted, therefore a second antiepileptic drug (valproic acid) was introduced, without any clinical amelioration. The parents were referred to our centre for a second opinion when the infant was 10 months old. Several episodes were recorded (*video sequence 2*) and their non-epileptic origin was documented. Valproic acid and vigabatrin were stopped after a few weeks. At the age of 12 months, the episodes spontaneously disappeared. Psychomotor development was normal during follow-up at age 2 years and 11 months.

Patient 3

A female patient was born with uneventful pregnancy and delivery. Growth and psychomotor development were normal. At 6 months of age, the patient had several episodes a day, characterised by shock-like head drops. These were sometimes isolated but more characteristically repetitive, without triggering factors or circadian correlation. The episodes were recorded at another epilepsy centre at the age of 8 months.

The manifestations captured by video-EEG monitoring (*video sequence 3 and figure 1*) were evaluated by a second epileptologist and deemed to be epileptic, and an antiepileptic treatment was proposed. We evaluated the video-EEG recording before initiation of treatment and considered the episodes to be non-epileptic. Treatment was never started. Head atonic attacks persisted until 11 months of age. During follow-up at age 10, psychomotor development was completely normal.

Differential diagnosis

The patients were referred following a diagnosis of epileptic seizures, and for two cases antiepileptic therapy had already been initiated. The high frequency of attacks and occurrence in clusters, resembling epileptic negative myoclonus, was suggestive of a false diagnosis of epilepsy, in particular of West syndrome or infantile spasms. The EEG polygraphic recording can be misleading because the EEG artefact induced by the head drops might be considered as an ictal paroxysmal event. Other types of non-epileptic negative myoclonus typically occur in adult age and are observed in metabolic encephalopathies or vascular accidents of thalamic or brainstem nuclei.

Discussion

The differential diagnosis between epileptic and non-epileptic attacks is extremely challenging for epileptologists. Moreover, the problem is not only the differential diagnosis but also the diagnosis *per se*. The cases described here represent a new type of NEA in infancy and this entity should be carefully considered for the differential diagnosis of West syndrome. In fact, this type of NEA can be easily misdiagnosed as an epileptic condition due to a number of different contributing factors: firstly, the lack of description of similar cases with adequate video-EEG documentation in the literature; secondly, EEG artefact caused by head drops can be misinterpreted as an EEG abnormality or as a short epileptic seizure with electrodecremental activity; and finally, epileptologists are often more concerned about misdiagnosing patients as non-epileptic rather than epileptic. If the spontaneous disappearance of attacks occurs when the infant still assumes AED therapy, AED treatment may be considered to be successful, thus reinforcing a wrong diagnosis of epilepsy. An additional negative consequence is that the infant may continue to assume an unnecessary AED treatment probably for many years, considering that West syndrome and infantile spasms are recognised as epileptic syndromes with a poor prognosis and high risk of recurrence of epileptic seizures. In conclusion, recognition of NEA is extremely important in clinical practice due to the high risk of misdiagnosis of epilepsy. Video-EEG monitoring is critically important in the paediatric population with "refractory" paroxysmal events to prevent misdiagnosis and potentially unwarranted exposure to drugs that might impair growth and development. □

Disclosures.
None of the authors has any conflict of interest or financial support to disclose in connection with the published text.

Legends for video sequences

Video sequence 1

Nine-month-old infant. A sudden, shock-like head drop is evident. The head returns to initial posture very rapidly.

Video sequence 2

This 10-month-old infant presents with three episodes of head atonia. The intensity of the attacks is variable and the last one is very mild. Note that the compensatory movement of the infant can be of variable intensity.

Video sequence 3

Eight-month-old infant. The video sequence shows head atonic attacks both isolated and repeated.

Key words for video research on www.epilepticdisorders.com
Syndrome: non epileptic paroxysmal disorder
Etiology: not applicable
Phenomenology: atonic seizure (drop attack); myoclonus (negative)
Localization: not applicable

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