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CASE REPORT

Ictal video-electroencephalogram of breath-holding attack

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Breath-holding attacks (BHAs) are non-epileptic paroxysmal events that affect 0.1% to 4.6% of infants from 6 months to 5 years of age.[1] Their frequency peaks at about 2 years of age and reduces by 5 years. The detailed description of the event is the key point for the diagnosis: the child typically emits a short loud cry, followed by breath holding, cyanosis, rigidity or limpness and transient loss of consciousness for a few seconds. Then, an inspiration marks the resolution of the spell.[1] The BHA are considered rare before 6 months of age, although they have been reported in the neonatal age.[2] BHA might be a frightening experience for parents, but its course is known to be benign.[1] Some patients have severe and frequent BHA up to five times per day, so their management can be complicated, especially when occurring under 6 months of age. Electroencephalogram (EEG) is used in specific cases to exclude tonic epileptic seizures.[3]

Case History

Since the age of 1 month and half an infant presented, usually during crying, episodes of apnea with cyanosis, and hypertonia, sometimes followed by hypotonia, pallor, loss of consciousness and finally sleep. For this reason, at 2 months of life, he was admitted to the hospital where physical, neurologic and cardiologic examination, brain ultrasound, standard EEG, blood cell count, serum ferritin level and electrocardiogram (ECG) showed no abnormalities. The infant was discharged with a diagnosis of BHA and a cardiorespiratory monitor was suggested.

His medical history was remarkable for obstructive apnea due to gastroesophageal reflux disease and mild congenital laryngomalacia. His mother's history was unremarkable, except for exposure to a stressful event during pregnancy since, at 20 weeks of gestational age, fetal ultrasound showed shortness of long bones, suggesting a possible congenital disease. This event increased the anxious state of the mother already greatly upset by COVID-19 pandemic issues. However, the suspicion of congenital disease was ruled out.

After discharge from the hospital, these episodes increased in intensity and frequency up to five times per day becoming so alarming that the patient was admitted to the Paediatric Intensive Care Unit (ICU) and Ambu bag ventilation was used during the episodes. The mother was frightened, fearing for the infant's life, so her capacity to console him was reduced. A long-term video EEG (vEEG) (Video S1) was performed, recording a typical event and demonstrating the non-epileptic nature of the paroxysmal event. After vEEG, resuscitation procedures with Ambu bag were stopped during the episodes and reassurance and psychological support was offered to the parents. In the following days, episodes decreased to 2 per day. Iron supplementation and piracetam 40 mg/kg divided in two daily doses was prescribed and in the following weeks the situation further improved. At 8 months of age he presented a normal development, BHA are still present although with minor manifestations, that the family has learned to manage.

Key points

1 We report the complete video electroencephalogram (vEEG) features of a breath-holding attack event, describing the pathophysiological sequence of events.
2 vEEG could be a useful instrument in cases with uncertain and frequent events to reduce hospitalisation and other unnecessary tests.

Video EEG

Background EEG was normal: the awake EEG activity was represented by continuous theta activity with normal voltage and symmetrical distribution and sleep non-REM pattern was characterised by theta-delta activity with physiological spindles, without slow nor epileptiform abnormalities.

Following an emotional trigger, the infant cried, and tachycardia was registered (180 bpm from basal 140 bpm) for a few seconds (Video S1). He remained for 30 s in a forced expiration status with crying facial grimace, cyanosis and opisthotonic posturing. Cerebral activity at EEG, 13 s after the breath holding, showed diffuse slow-wave in the delta band with no epileptic discharges, and polygraphy marked severe oxygen desaturation with SpO_2 going down to 53% (Fig. 1). Then, relative bradycardia (100–110 bpm) was recorded

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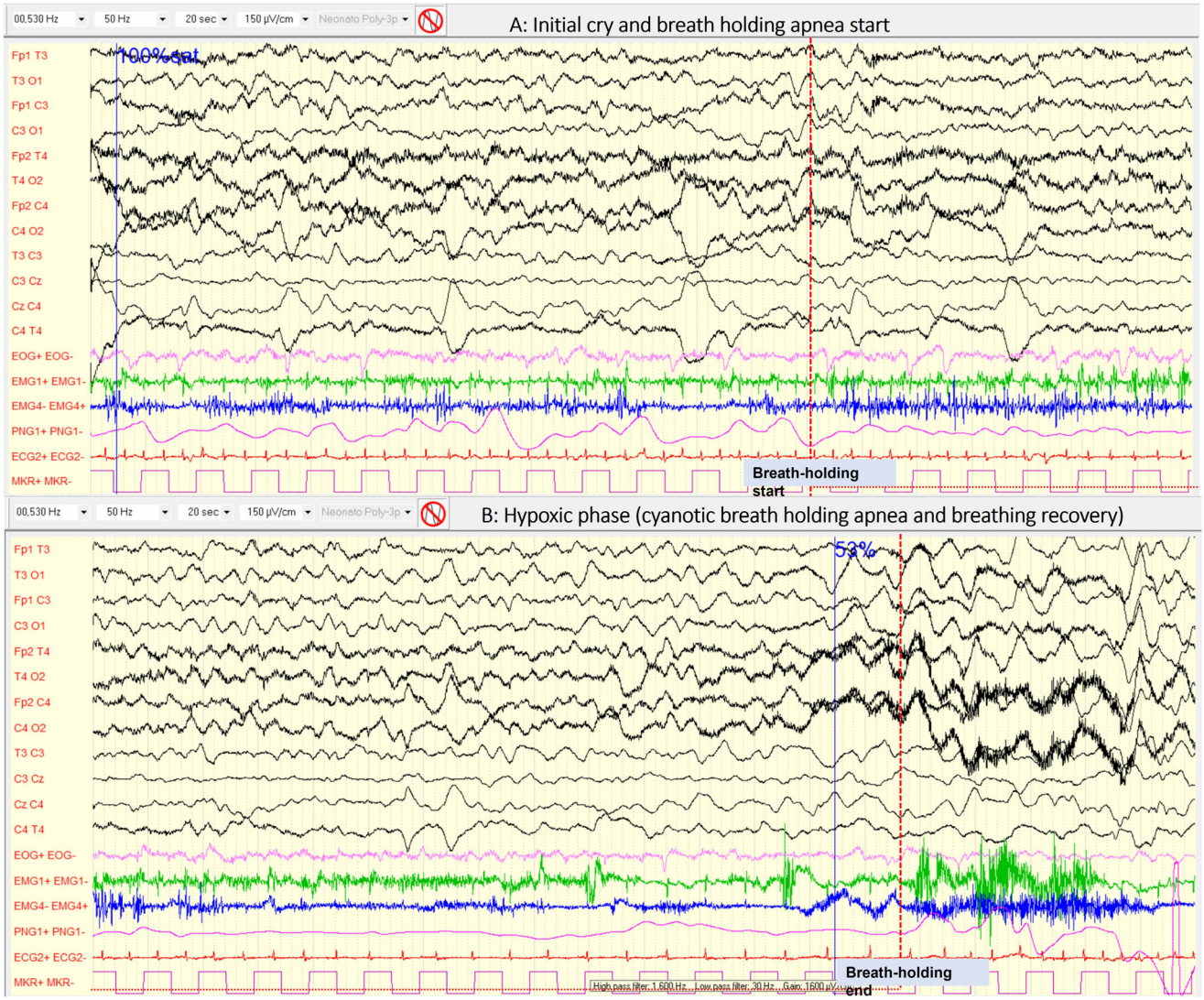

Fig. 1 The first part of a BHA episode in 3-month-old boy. The upper trace (a) shows the EEG during the prodromal phase, when the cry is bursting after an emotional trigger accompanied by a relative tachycardia compatible with activation of the sympathetic system. In correspondence with the vertical red dotted line breath holding starts and lasts 30 s. The lower trace (b) shows the second part of BHA when oxygen desaturation appears together with increasing electrocortical slowing and finally relative bradycardia and breath recovery. Fp1, T3, C3, 01, Cz, Fp2, T4, C4, 02, EEG electrodes; EOG, electrooculography, EMG1, left deltoid muscle surface electromyography; EMG4, right deltoid muscle surface electromyography; PNG, respiratory movements; ECG, electrocardiography.

before an inspiration act and hyperpnea followed for 3 s. A 'syncopal' phase lasting 10 s occurred: the infant became pale, floppy, with loss of consciousness and no motor reaction in response to external stimuli; in this phase, the EEG presented slightly increased delta activity while oxygen saturation was still low (Fig. 2). Relative tachycardia followed and progressive normalisation of EEG correlated with clinical recovery and voluntary movements. Finally, the baby fell asleep, and a typical normal sleep EEG pattern was recorded.

Discussion

BHA is commonly reported above the age of 6 months, but it should be considered also below this age, as this case and few other published cases demonstrate.[2] EEG is usually not needed for the diagnosis, but it could be useful when the semiology is unclear.[3,4]

In a study by Low *et al.*,[3] on 129 children with BHA aged 7 months–11 years, a normal interictal background EEG activity during wake and sleep was usually found. In their study, they registered only one patient during a BHA episode: a 13-month-old girl who during the attack at EEG showed slow activity 'such as occurs in anoxia and with stupor from various causes'.[3] No polygraphy or video was performed at that time. In addition to this, descriptions of ictal EEG are very rare. As we show during the BHAs EEG initially shows a slowing of the background activity and, as bradycardia deepens, further slowing of the cerebral activity manifests. As the consciousness was regained, the background EEG rapidly returned to normal.[4]

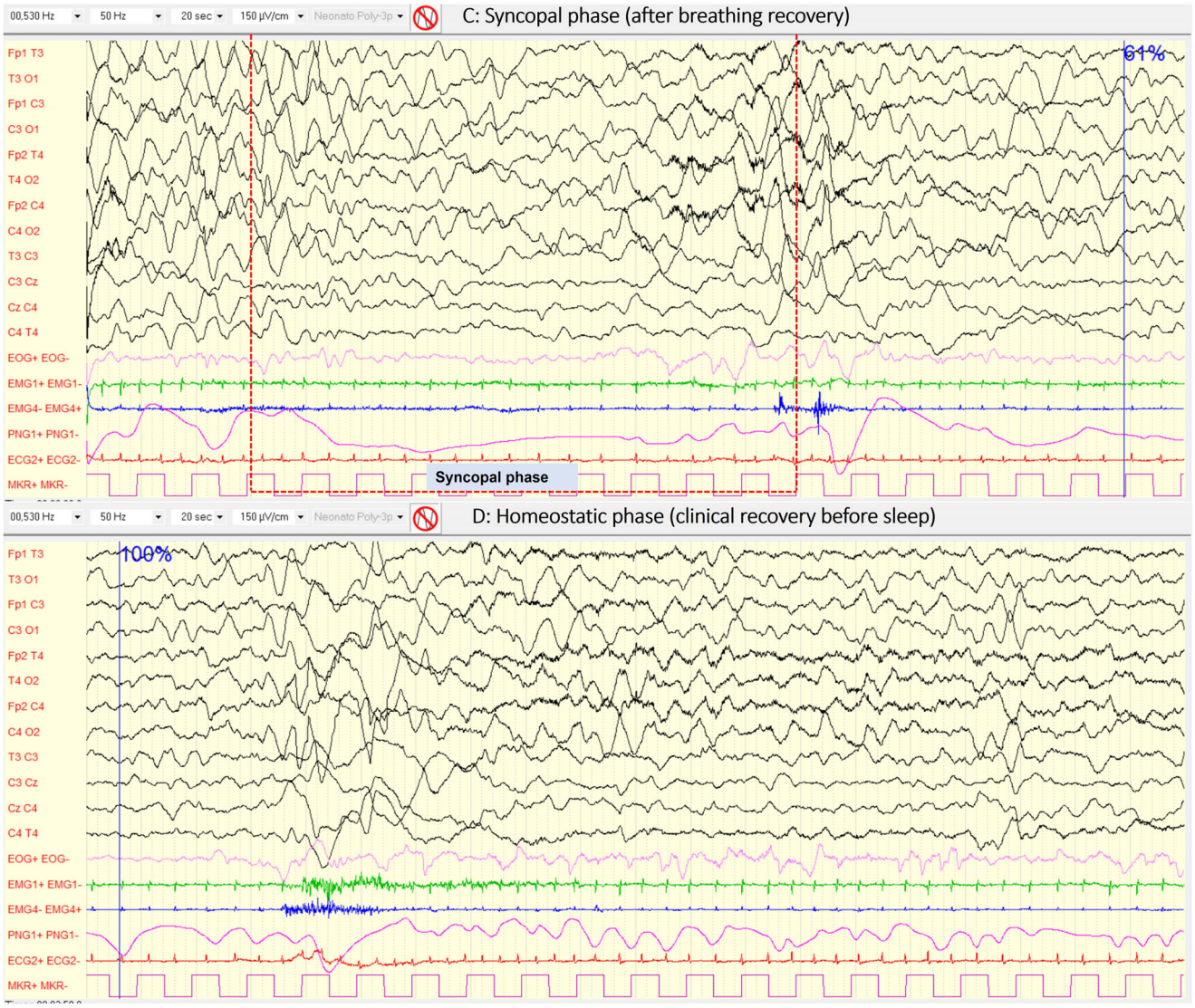

Fig. 2 The second part of the BHA episode in a 3-month-old boy. In the upper trace (c) after inspiration few seconds of hyperpnea occurred, followed by a syncopal phase (during the 10 s among the vertical red dotted lines) where pallor has been observed together with relative bradycardia and transitory hypopnea, followed by relative heart rate instability, respiratory, clinical and oxygen saturation progressive recovery. In the lower trace (d) the homeostatic phase is reached with oxygen saturation normalisation. Fp1, T3, C3, 01, Cz, Fp2, T4, C4, 02, EEG electrodes; EOG, electrooculography; EMG1, left deltoid muscle surface electromyography; EMG4, right deltoid muscle surface electromyography; PNG, respiratory movements; ECG, electrocardiography.

We provide an interesting video EEG with polygraphy providing insights on the pathophysiology of BHA. At first, the breath-holding is associated with cyanosis and oxygen desaturation causing EEG slowing (brain manifestation of hypoxia). Further electrocortical slowing then appears together with pallor, bradycardia and syncope, suggesting that hypotension (during the pale skin phase) probably had an additive role in the worsening of EEG slowing. The heart rate up-down variations observed at the ECG seems to reflect a sympathetic activation shifting toward a parasympathetic activation before reaching a homeostatic phase of clinical recovery. These findings resemble the phenomenon of vaso-vagal syncope, since the contemporaneous recording of both EEG and ECG during the episodes was consistent with these cardiovascular episodes.[5] Interestingly, patients with BHA in adult life present vaso-vagal syncope more frequently than the control population.[6] Moreover, this recording shows that the distinction between cyanotic and pallid BHA is not exclusive, as a pallor phase may follow a cyanotic phase.

BHA are known to have a benign prognosis with a reduction in the frequency of episodes and a spontaneous remission before adolescence, with no neurological consequence.[6] However, an increased predisposition for syncopal attacks and concentration problems were also reported.[6] In a single article, not confirmed by other studies, some authors reported a slight increase in the plasma level of S-100B protein (related in other studies to brain injury) in a group of BHA patients in comparison with a control

group,[7] without evidence of a clinical meaning of this finding. Future studies would be required to clarify this point.

In line with other studies,[8,9] we acknowledge the possibility that psycho-educational interventions could improve the parents' mental wellness.[10]

Our main message is that a vEEG monitoring with polygraphy (oxygen saturation, respiratory effort, ECG) to record the BHA episode should be early considered in cases with frequent or severe events and uncertain diagnosis to obtain objective evidence of BHA for physicians and families. This diagnostic approach in those peculiar cases may facilitate clinical management, early reassuring families and providing the appropriate interventions, possibly avoiding repetitive hospitalisation, unnecessary investigations or admission to ICU (causing a high cost), as in our patients before reaching a sure diagnosis thanks to the vEEG monitoring.

Informed consent was obtained from the subject legal representatives involved in the study.

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Supporting Information

Additional Supporting Information may be found in the online version of this article at the publisher's web-site:

Video S1: At the beginning the baby is crying: cerebral activity at EEG shows diffuse theta-delta activity, normal voltage; tachycardia is recorded (140–190 b/m). At 0:03 he stops crying and for 30 s maintains forced expiration, facial grimace, moves arm and legs. At 0:26 cyanosis appears, followed by opisthotonus posture and behavioural arrest (0:37): cerebral activity at EEG shows diffuse slow waves in the delta band, and polygraphy shows relative bradycardia (100 b/m) and oxygen saturation (SpO_2) decrease till SpO_2 53%. At 1:09 progressive clinical recovery occurs with respiration and spontaneous movements regain, EEG normalisation, relative tachycardia and SpO_2 100%. In the end the baby falls asleep.