Epileptic Disord 2008; 10 (1): 53-5

Ictal hiccup during absence seizure in a child

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Received September 4, 2007; Accepted January 17, 2008

ABSTRACT – Absence seizures are one of the features of idiopathic generalised epilepsy (IGE) and occur as component of many different syndromes. They are commonly associated with various clinical features such as mild clonic components, change in postural tone, automatisms and autonomic phenomena. Childhood absence epilepsy (CAE) is the prototype IGE with typical absence seizures. We report a child who had prominent ictal hiccups during an absence seizure, and discuss the possible mechanisms. *[Published with video sequences]*

Key words: absence seizure, ictal hiccup, 3Hz spike and wave complexes

Absence seizures, both simple and complex, are a feature of childhood absence epilepsy (CAE) and other epilepsy syndromes (Binnie *et al.* 2003). Ictally, mild clonic components, change in postural tone, automatisms and autonomic phenomena are reported, with many children having a mixture of these during an absence attack. The clonic components usually take the form of a mild, rhythmic contraction of the eyebrows, eyelids, corners of the mouth, fingers, arms or shoulders. Both *de novo* and perseverative automatisms are common, and correlate with longer seizure duration (Penry *et al.* 1975). However, ictal hiccups (clonic contraction of diaphragm) has not been reported in absence seizures. We report a child who had prominent ictal hiccups during one of her absence seizures.

Case report

An otherwise healthy nine-year-old girl presented with a five-month history of 15-20 episodes each day of brief staring associated with lip smacking and eyelid blinking, without any aura or post-ictal confusion. There was no history of any other seizure types. She was not on any medication. Her video-EEG showed normal background activity. She had five of her habitual, brief, vacant episodes during the recording, three during hyperventilation, each lasting for about eight seconds. During the vacant episodes, her EEG showed generalised 3Hz spike and wave discharges, and in some there was also a leading spike over the frontal region with variable lateralisation. Most of the spikes were single or double. During the fourth episode, which occurred after 150 seconds of hyperventilation, she became unresponsive to command and had hiccups which were time-locked with 3Hz spike and wave discharges. She had a single hiccup at the beginning of the discharges, followed by five continuous hiccups in two seconds and then one towards the end of the discharges. She did not have hiccups during the resting recording or the other four absence attacks.

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doi: 10.1684/epd.2008.0164

She showed minor manual automatisms and eyelid flickering during the absence episodes. Immediately after the attacks, she became fully alert and responsive. There was one brief, inter-ictal, 3Hz generalised spike and wave discharge lasting for two seconds, with bi-frontal emphasis. Photic stimulation was unremarkable *(see video sequence)*.

Discussion

To our knowledge, ictal hiccups has not been reported during absence seizures. Hiccups are intense, synchronous contractions of the diaphragmatic and inspiratory intercostal muscles, lasting about 500ms. This is followed, 30ms later, by glottal closure causing the characteristic "hiccup" sound. The pathophysiology of hiccup genesis is not known for certain. It is probably due to the irritation of one or more components of the so-called hiccup reflex arc. Some postulate a brainstem hiccup centre adjacent to the respiratory centre in the medulla, while a spinal centre in C3 to C6 segments has also been suggested. Pathology in supratentorial areas such as the hypothalamus, temporal lobe and reticular activating system are all capable of stimulating hiccups (K Marsot-Dupuch *et al.* 1995). In adults, brain stem disorders are the most common, central cause *(figure 1)*.

In absence seizures, the pathophysiology is believed to affect the T-type calcium channels in thalamic relay neurons, resulting in paroxysmal activation of the cortex leading to 3Hz spike and wave discharges in the EEG (Chang and Lowenstein, 2003). The well-recognised, additional motor phenomena in absence seizures vary from seizure to seizure and are thought to reflect selective cortical activation, which in this case would most probably be the temporal lobe or the hypothalamic component of the hiccup reflex arc. Motor phenomena such as eye movement, eyelid flutter, or yawning have an efferent pathway involving the brain stem, and hiccups could result from a similar mechanism. Alternatively, this could possibly reflect a direct effect on the brain stem. A *de novo* automatism is less likely as an explanation because hiccups have not been described as an automatism in this or any other seizure type, particularly temporal lobe seizures (Luders and Noachtar, 2000). An adult patient with temporal lobe pathology in whom hiccups persisted even when seizures

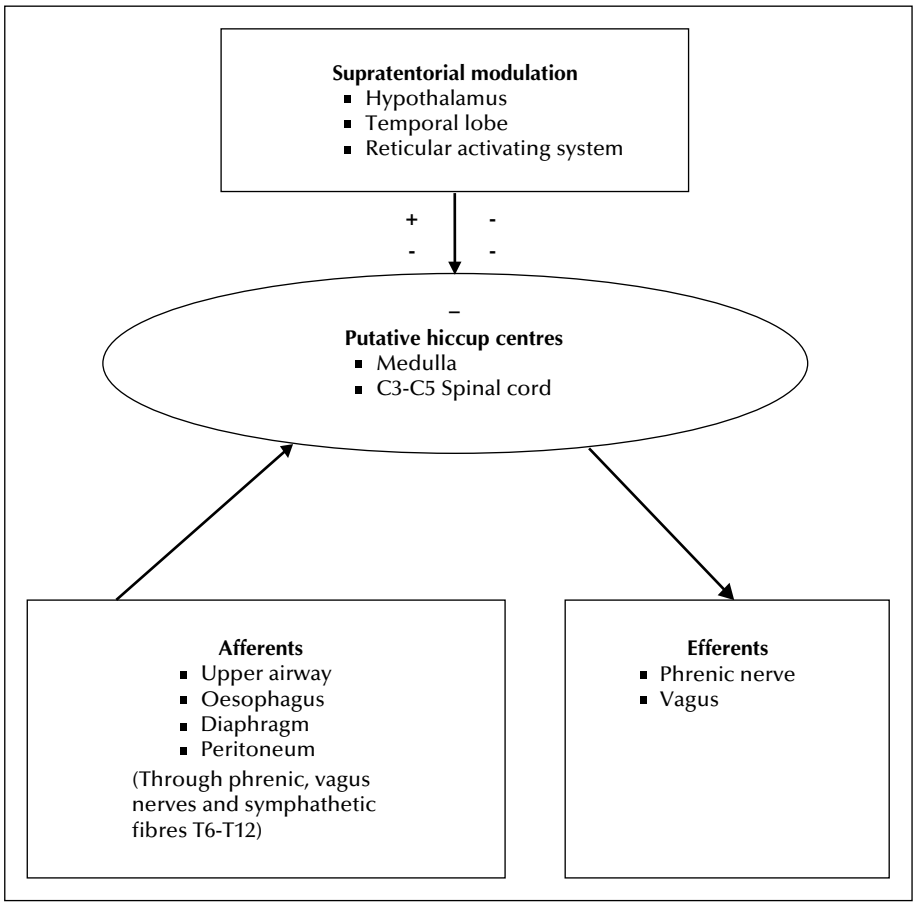

Figure 1. Physiology of hypothetical hiccup reflex arc.

were controlled by anti-epileptic therapy, has been reported (Jansen *et al.* 1990), suggesting a temporal lobe influence in generating hiccups. Hiccups have also been described in one child with left temporal lobe epilepsy, but it is not clear whether it occurred before, during or after the seizure. In this child, it was considered to be an autonomic component of the partial seizure (Fogarasi *et al.* 2006). Similarly, the hiccups in the child reported may represent an autonomic component of absence seizures.

A definitive epilepsy syndrome classification in this child is difficult at the present time as we need follow up to understand the evolution of the seizure disorder. However, it is clear that she has a form of primary generalised epilepsy with absence seizures. Her absences are well controlled at present with oral sodium valproate therapy. We think that she probably has childhood absence epilepsy syndrome. The absence of early morning myoclonic jerks, GTCS and photosensitivity on the EEG makes juvenile myoclonic epilepsy unlikely, but it cannot be entirely ruled out at this time. It is also unlikely that she has myoclonic absence epilepsy as she has no learning difficulties; the clonic movements observed were not sustained, proximal arm muscles were not involved and her EEG also showed normal background rhythms (Panayiotopoulos, 2002). We suggest that in our patient, the abnormal thalamo-cortical excitation during the absence seizure caused ictal hiccups. □

Legend for video sequence

Ictal hiccup during an absence seizure.

Acknowledgements. We are grateful to Zoe Unwin, EEG technologist, Royal Hallamshire Hospital for her technical assistance and Rebecca Sarah Penn-Critten, for her secretarial assistance.This work is not supported by any grant. This material has not been presented in any other local, regional or international meetings.

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