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EPISODIC MIGRAINE (S NAHAS, SECTION EDITOR)

Exploding Head Syndrome: a Review

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Abstract
Purpose of Review To explain our current understanding of exploding head syndrome (EHS), an unusual and underreported sensory parasomnia.
Recent Findings Based on findings from recent studies of EHS, the prevalence is higher than previously suggested by the literature, which historically has consisted mostly of case reports. The typical presentation also has been better illustrated by recent case series, and diagnostic criteria have been defined. Its pathophysiology is still unclear.
Summary EHS is underrecognized and its symptoms are alarming, but a review of our current state of knowledge will allow physicians to make a diagnosis of this benign condition with greater confidence.

Keywords Exploding head syndrome · EHS · Parasomnia · Sensory sleep start

Introduction

Exploding head syndrome (EHS) was given its provocative name by J.M.S. Pearce in 1988 when he described 10 patients who had complained of experiencing an abrupt sensation of noise like an explosion that awakened them from sleep [1]. Many now credit Silas Weir Mitchell with the first clinical description of the peculiar phenomenon in 1890, when he wrote of a "sensory shock" in which patients experience a loud sound "like that of a pistol-shot" during sleep [2]. It has been suggested more recently by Goadsby and Sharpless that the syndrome be renamed "episodic cranial sensory shock" to more accurately describe the experience of the patient and to credit Dr. Mitchell's original description [3]. There has even been speculation that René Descartes may have had EHS, and that the credit for the first written description of the syndrome should really go to his biographer, Adrien Baillet, who in 1691 wrote of Descartes' unusual dream of a "sharp and shattering noise" waking him from sleep [4].

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Clinical Presentation

The American Academy of Sleep Medicine classifies EHS as a sensory parasomnia and lists diagnostic criteria of an experience of sudden loud noise or sense of explosion occurring at the wake-sleep transition or during sleep, followed by immediate arousal often with a sense of fright, and without significant pain [5•]. A variety of other symptoms have been reported to occur, the most common being tachycardia, brief muscle jerks or twitches, and visual phenomena, often described as a flash of light [6••]. About 5% of patients have a sensation of momentary respiratory arrest and requiring a deliberate effort to breathe again [7•]. Several cases of EHS occurring with sleep paralysis have been reported [8, 9], as well as a case of a woman with EHS and sleep-related orgasms [10]. A female to male ratio of 1.5:1 has been reported with median age of onset of 54 [11••].

Though EHS classically is described as painless, an association with headache has been noted in some patients. EHS has been reported occurring simultaneously with idiopathic stabbing headache attacks [12]. Some patients with a pre-existing diagnosis of migraine have reported EHS attacks occurring prior to a migraine exacerbation [8, 13]. EHS may also occur in patients with a predisposition to headaches, but not in direct temporal association to a headache attack. A review of all reported cases in the literature up to 2014 concluded that the most common co-morbid condition occurring with EHS is a primary headache disorder, with migraine without aura being the most common type. Migraine with aura, primary stabbing headache, chronic tension-type headache, primary

exertional headache, and primary headache associated with sexual activity were also reported [11••].

Prevalence is difficult to determine, as the disorder is likely both underreported and underrecognized. It is reported that only 11% who screen positive for EHS have actually discussed their symptoms with a healthcare provider [6••], and some patients have been met with disbelief when they reported their symptoms to their doctors [7•]. The Munich Parasomnia Screening (MUPS) questionnaire developed in 2007 is a self-rating instrument used to assess prevalence and frequency of parasomnias. The developers found that 13.8% of psychiatric patients (total $n = 65$), 10.0% of patients with a known sleep disorder ($n = 50$), and 10.7% of healthy controls ($n = 65$) screened positive for EHS. A random subsample ($n = 36$) underwent clinical interview, and 11.1% were diagnosed with EHS [14]. A recent study of 211 undergraduates found a lifetime prevalence of 18% and a recurrent prevalence of 16.6%. The same study found similar prevalence in men and women, which differs from the observations of many authors that EHS is more common in women [9]. As most of the cases reported in the literature have been of middle- or older-aged patients, it is possible that prevalence in women may increase with age.

Differential Diagnosis

The primary concern that brings many patients to the office with a complaint of EHS is worry about bleeding or tumor [7•]. Many patients are so alarmed by their symptoms that they may have a hard time determining if there is pain or not, which may necessitate work-up for a low-grade subarachnoid hemorrhage [15]. Migraine, cluster headache, hypnic headache, and chronic paroxysmal hemicrania may all cause awakening from sleep and should be considered in the differential [16]. Hypnic headache in particular is more common in women in their 60s–80s and occurs only during sleep [17]. Pain should never be the primary complaint in EHS, and if present, should be brief and not severe. Nightmare disorder and flashbacks of post-traumatic stress disorder may also be included in the differential, as these may include perceptions of an explosion with associated fear, but the attack of EHS should have no other content [18]. The brief, often stereotyped presentation of EHS may also raise concern for seizure, though there should be no post-ictal state.

Pathophysiology

The etiology of EHS is unknown. When Pearce first wrote of EHS, he speculated that the perception of sound may be due to a brief disinhibition of the cochlea or its central connections [1], or that it may be a phenomenon similar to nocturnal myoclonus [7•]. The current theory discussed by multiple authors is that EHS is a sensory variant of a hypnic jerk [15, 19, 20]. A delay in areas of the reticular formation in switching off during transition to sleep causes a paroxysm of neuronal activity, which may cause myoclonus or the perception of a loud sound or flash of light [21]. In one case report, a patient who presented with EHS was found to have sarcoidosis, and MRI disclosed a nonenhancing pontomesencephalic lesion around the periaqueductal gray reaching into the tegmentum. The patient's symptoms improved with clonazepam, suggesting that there may be impaired GABAergic transmission from the PAG to the serotonergic neurons in the dorsal raphe nucleus in patients with EHS. In rats, GABAergic neurons in the brainstem decrease the firing rate of serotonergic neurons in the dorsal raphe nucleus at the time of sleep initiation, lending some credence to this hypothesis [20]. EHS also has been reported with withdrawal from both benzodiazepines and SSRIs, further supporting the plausibility of this mechanism [22]. Anecdotal evidence of response to nifedipine [12] and topiramate [23] suggests there may be transient calcium channel dysfunction playing a role in EHS as well.

Several studies have utilized polysomnography to assess biophysiologic changes during EHS events. In one study of 9 patients with EHS, 5 of them reported events during recording, all reportedly arising from sleep. Surprisingly, all recorded events occurred while the patients were awake and relaxed. It is also notable that several of these patients had very long sleep latencies and poor sleep quality overall [24]. Other studies have captured events arising at the transition from wake to sleep [13, 23], as well as events arising from stage 2 non-REM sleep [13, 19, 25]. Though patients commonly report EHS as waking them from sleep, it appears that more commonly it occurs near the wake-sleep transition. This supports the theory that EHS is due to abnormal activity of neurons in the brainstem involved in sleep initiation.

The role of stress must also be considered in EHS. Many patients have reported their symptoms beginning in the setting of increased stress [7•, 24]. Case reports have described EHS beginning after the death of a patient's son [25] and with increased stress at work [22]. One patient developed psychogenic nonepileptic spells shortly after presenting with EHS symptoms and was thought to have a dissociative disorder triggered by stress [19]. Given the poor sleep quality observed in some polysomnography studies, it is possible that emotional stress and anxiety may be causing disrupted sleep which in turn may trigger EHS symptoms in some people [24].

Treatment

For many patients, no treatment is needed for EHS. This is a benign condition, and some patients may improve with reassurance alone [1, 24]. Given the association with stress, relaxation techniques may be an effective treatment option. Some, however, may be so distressed by their symptoms that they actively try to avoid sleep [18]. Clinically significant distress and functional impairment is reported by 2.8% of patients [9], and reassurance alone may not be enough. There have been no

clinical trials investigating treatment options for EHS, but should drug therapy be warranted, there is anecdotal evidence for several medications. Benzodiazepines, including clonazepam [20] and clobazam [25], have been reportedly effective. The tricyclic antidepressants clomipramine [24] and amitriptyline [11••] have also been used successfully in some cases, though some authors report failure of amitriptyline [12, 20]. Nifedipine [12], flunarizine [26], and topiramate [23] have all been effective in case reports. Medications reported to be unsuccessful include propranolol, valproic acid, oxycodone, and gabapentin [12]. As the symptoms of EHS are extremely brief and benign, aggressive treatment likely is not indicated in most patients, and side effects weighed against potential benefit must always be taken into consideration.

Conclusions

EHS is traditionally classified as a headache disorder, though it is not actually a painful condition for most. It may be regarded as a sensory parasomnia that typically occurs at the wake-sleep transition. EHS is often considered to be a rare disorder, but recent studies suggest it is underreported. Though a benign condition, it may cause significant distress, especially when it goes undiagnosed. Recognizing symptoms of EHS and providing reassurance are the foundation of managing this disorder, but further research is needed to better understand its pathophysiology and to elucidate the best treatment options when reassurance alone is insufficient.

Compliance with Ethical Standards

Conflict of Interest Claire Ceriani declares no conflict of interest. Dr. Nahas reports personal fees from Allergan, Amgen, electroCore, Ely Lilly, Avanir, and Supernus, all outside of the submitted work.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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