Hot water epilepsy:
case report and literature review

Efterpi Pavlidou, Anastasia Gkampeta, Evangelia Dosadi, Evangelos Pavlou

Journal of Pediatric Sciences 2013;5:e197

How to cite this article:
Hot water epilepsy: case report and literature review

Efterpi Pavlidou, Anastasia Gkampeta, Evangelia Dosiadi, Evangelos Pavlou
Second Pediatric Department, AHEPA Hospital, Aristotle University of Thessaloniki, Greece

Abstract: Hot water epilepsy is a rare type of seizures, which belongs to reflex epilepsies and occurs while bathing. It is geographically distributed, mainly to India and Turkey. The differences among countries are attributed to bathing habits and genetic susceptibilities. It can present as an episode of altered or loss of consciousness accompanied by staring gaze, pallor, unresponsiveness and postictal somnolence. Pathophysiology of this clinical entity is complex and unknown. It seems that a default of the thermoregulatory system exists and leads to the occurrence of seizures due to rapid rise of temperature. Blood brain barrier plays its unique role in the genesis of hot water epilepsy. Herein, we report a new caucasian case of a two years old child, that presented with impaired consciousness during bathing with hot water. We perform an extensive literature review and discuss the questions that arise regarding the pathophysiology, etiology and management of hot water epilepsy.

Keywords: Hot water epilepsy, bathing epilepsy, children, pathophysiology, management

Corresponding author: Efterpi Pavlidou, M.D., Ph.D, Pediatrician, Consultant in Pediatric Neurology, 2nd Pediatric Department, AHEPA Hospital, Aristotle University of Thessaloniki, Greece
Telephone: 00306932232175
Fax: 00302310994803
e-mail: efterpi.pavlidou@gmail.com

Introduction

Hot water epilepsy (HWE) belongs to reflex epilepsies and is characterized by seizures while bathing with hot water [1]. It was described for the first time in 1945 and subsequently there have been many reports of its prevalence in both children and adults [2].

It is considered to be a geographically distributed epileptic syndrome since it mainly occurs in India and Turkey [3,4]. The differences among countries may depend on bathing habits and mainly on genetic susceptibilities [5,6]. Almost all cases of HWE are seen in healthy children, more frequent among males than females [4,7]. Seizures usually occur when hot water (40°C-50°C) is poured over the head.

However, up to ten per cent of patients have crisis during body immersion in hot water [8]. Additionally, subsequent spontaneous seizures have been reported in 16–62% of patients, showing a common underlying genetic predisposition [9].

Case report

A previously healthy two-years-old boy presented with multiple episodes of loss of consciousness with staring, pallor and unresponsiveness during warm bathing. The episodes were initiated four months ago, they lasted few seconds and had always postictal somnolence. The crises gradually had longer duration that lasted several minutes. Due to those episodes, the child was hospitalized and an EEG and a brain MRI
were performed. Both were normal. He was referred to a psychiatrist, who diagnosed a neuropsychiatric disorder and advised family counseling. Since the episodes continued, the child started anti-epileptic treatment with sodium valproate (25 mg/Kg of body weight), but he did not get benefit and he was referred to our department for further investigation.

The child was born after an uncomplicated pregnancy from an unconsanguineous marriage. Family history was negative for epilepsy and febrile seizures. Psychomotor development was normal. Height, weight and head circumference were consistent with age. Neurological examination was normal. Routine blood tests, ophthalmological examination, inter-ictal EEG, cardiac ultrasound and a holter-ECG were also normal. Due to technical difficulties we could not proceed to a video-EEG, while provoking the episode. However, we triggered the episode by bathing the child with hot water and we recorded it on a video. The boy had an episode of altered consciousness with unresponsiveness, pallor, hypotonia and gaze lasting three to four minutes at the end of the bath following by postictal somnolence. We suggested to the parents to wash the child with cool water (temperature below 37°C) and we increased the dose of sodium valproate to 30mg/kg, since the therapeutic levels were low. However, the child continued to have seizures while bathing. The decision to alter the medication to oxcarbamazepine was taken. During his latter hospitalization the child had a co-existent febrile infection of the lower respiratory system, without triggering of the seizures. Eighteen months after the initial crisis, the child has no precipitating seizure and his psychomotor development is normal.

**Discussion**

Reflex epilepsies are seizures provoked exclusively by an external stimulus and account to six per cent of all epilepsies [10]. HWE belongs to reflex epilepsies and seizures are precipitated by bathing in hot water. It seems that they require a specific thermal cutaneous stimulus [11]. Affected infants have often been described as becoming lethargic, with a staring gaze, pallor, blue lips even apnea, hypotonia, loss of consciousness, followed by prolonged postictal somnolence or limpness [12,13]. The majority of the seizures, almost 70%, are focal with or without secondary generalization [14]. Adults or older children usually describe an aura prior to the crisis with a sense of fear, vertigo, activity arrest, confused speech, visual and auditory hallucinations or complex automatisms. Seizures last from seconds to several minutes and can appear at any time during bathing [8,15]. Subsequent spontaneous non-reflex seizures have been reported to occur few years later in 16–38% of patients [16], while history of epilepsy among the family members has been reported only in 7–22.6% of cases.

According to a large retrospective study of Yalcin et al. in 25 cases with HWE, age at onset of the first seizure ranged from six months to 37 years old. The majority of the patients had their first seizure within the first decade of life, with male predominance. Most of the seizures were complex partial seizures with secondarily generalization. Spontaneous seizures occurred subsequently one to six years after reflex ones [17].

Incorpora et al. in their study, showed the complexity of this kind of epilepsy [18]. They reported the case of two five-years-old monozygotic twins that presented with episodes of hot water reflex seizures from their first days of life followed, since the age of 20 months, with clinical episodes of cerebral dysfunction including alternating hemiplegia of childhood. These attacks occurred only during immersion in a hot bath or when hot water was sprinkled over the body. Clinically, the attacks were characterized by irritability, a fixed stare with head deviation and upper-limb hypertonia of the one side. The episodes lasted from few seconds to three to four minutes, and usually ceased with a cry while the mother was drying the child [18]. The occurrence of hot water seizures in twins and with so early clinical presentation shows mainly the strong genetic predisposition of this entity.

Only in one study, the mean triggering temperature of water was calculated to about 41.4°C [19]. Precise bath water temperature measurements are few, however the experience up to now suggests that even ‘ideal’ or ‘normal’ water temperature is sufficient to be a precipitating factor of paroxysmal events in different entities [20]. Therefore, one can argue that water bathing itself is responsible for the occurrence of seizures, rather than true hot water with a temperature above the ‘normal’. Moreover, we do not know the exact ‘ideal’ or ‘normal’ water temperature for each person.

Another study questioned whether the triggering factor for the seizures while bathing, had to do with the light reflected from the surface of the bath or
even the sound of ‘splashing’. However it seems that this is not really the case, since eye pads and head-
phones that were used, did not prevent bath-induced
episodes [21].

The pathophysiologic mechanism of HWE is not only
unknown but it is rather foggy too. These seizures
may be a response to hyperthermia, which is
provoked by high temperature-dependent stimuli
during bathing [22-24]. However, seizures have been
reported during a bath even when hot water is not
poured over the head. It seems that a default of the
thermoregulatory system occurs, due to the rapid rise
of temperature, and leads to the production of
seizures [25,26].

In addition, Satishchandra et al. (1998) suggested that
patients with HWE probably have a genetically
determined aberrant thermoregulatory system and,
because of this, they are sensitive to rapid rises in
temperature. They also reported that in these patients,
it takes longer for body temperature to return to
baseline after bathing compared with normal healthy
controls [16]. Although, in experimental animal
models, the role of the temporal lobe or amygdala
was impressive, there was no evidence of the
involvement of these structures in the human study
[25]. In HWE, as in the majority of reflex epilepsies,
an underlying lesion of the cerebral cortex exists
[27]. Case reports have proved cortical dysplasia to
be the responsible underlying pathology of HWE
[7,28,29].

Channelopathies have also been implicated in the
pathophysiologic mechanisms of triggering HWE.
For instance, SCN1A mutations, as in Dravet
syndrome, underlie induction of epileptic seizures in
HWE. This thought justifies the Japanese, who
perform screening test for Dravet syndrome to
patients with hot-water precipitated seizures as a
predictive factor. A mutant chloride channel, that is
also implicated in hyperekplexia, and a calcium-
channel blocker ‘flunarazine’, which is connected to
the genesis of alternating hemiplegia of childhood,
may also have a relative link to HWE [12].

In molecular basis, blood-brain-barrier (BBB) seems
to play a very important role in the pathogenesis of
HWE. BBB is responsible not only for brain
homeostasis, but also for keeping out inflammatory
triggers. It is generally assumed that in chronic
epilepsy, it may be impaired. What is not clear is
whether seizures contribute to BBB failure or
whether loss of selective permeability and glucose
transport may be implicated in epileptogenesis [30].

Possibly rapid increase of temperature at the scalp
can cause hyperactivity in neurons and activation of
the sympathetic nervous system, which in turn can
lead to an increase in vascular resistance and arterial
blood pressure and therefore induce seizure. This
mechanism, like a ‘domino effect’, can represent a
possible pathogenetic pathway for HWE [22,31,32].

In the differential diagnosis of bath-induced
paroxysmal disorders apart from HWE are also other
conditions, such as alternating hemiplegia of
childhood, hyperekplexia and infantile syncope.
Although alternating hemiplegia of childhood is rare,
A third of affected individuals have a history of
paroxysmal episodes precipitated by bathing [12,20].
Additionally, seizures could be triggered by hot water
bathing, in patients with severe myoclonic epilepsy in
infancy, therefore it should be in our differential
diagnosis especially if it is the first time that this type
of seizures occurs [33]. Differential diagnosis should
also be done, in some cases, from febrile convulsions,
vasovagal syncope, breath-holding spells or even
aquagenic urticaria [3]. However, it is more than
obvious that at least in the majority of cases thorough
history and a good clinical examination can lead us
safely to the correct diagnosis. In some uncertain
cases, provocation of an episode can resolve the
puzzle. Ictal EEG, whenever possible, could also help
to confirm the diagnosis. Although children with
Dravet syndrome could have seizures provoked by
water immersion, the clinical presentation, evolution
and development are completely different from HWE
[34].

Interictal EEGs are usually normal, whereas ictal
ones mainly show focal epileptic activities and
paroxysmal discharges characterized by secondary
generalization [9]. The fact that complex partial
seizures are the most common clinical presentation
and ictal or interictal recordings show an epileptic
focus in the temporal lobe suggest the presence of a
structural lesion in the temporal lobe [35]. Interictal
EEG can be normal in the majority of the cases,
while only 15–20% can reveal diffuse abnormalities
[36]. Lateralized or localized spike-wave discharges
in the anterior temporal regions have been reported in
few isolated cases. Ictal recording has technical
limitations and is difficult to obtain; however, several
reports have demonstrated ictal EEGs by provoking
an episode. They demonstrated left temporal
rhythmic delta activity, sharp and slow waves in the
left hemisphere, bilateral spikes, and temporal
activity [9,37,38,39].
HWE is known as a benign and self-limited reflex epilepsy. Only by avoiding hot water or long showers, it may be sufficient for a patient to become seizure-free. However, approximately one-third of patients with HWE continue to have seizures even during regular baths. In these patients, carbamazepine, sodium valproate and levetiracetam might be preferred [9]. Some authors suggest the use of intermittent oral benzodiazepines, by using five to ten milligrams of oral clobazam one to two hours prior to bathing [8,40]. In our case, the boy continued to have seizures although the water’s temperature was low. This was the main reason that we insisted on anti-epileptic treatment and since sodium valproate proved insufficient, oxcarbamazepine was tried and helped us to achieve seizure control.

Conclusions
Precautions against precipitating factors are generally enough in order to avoid a reflex epilepsy. Antiepileptic therapy is needed as an additive treatment in a small proportion of cases and the final prognosis of these patients is also favourable [41]. Since seizures show a tendency to decrease spontaneously, withdrawal of medication, if it had been given, should be carefully undertaken only after several months [14]. To play the role of ‘devil’s advocate’, since such seizures have not been reported in saunas or furnaces, a complex tactile stimuli, rather than rapid rise of temperature per se, might have the most relevant part on seizure precipitation [41]. Question marks arise, when one thinks that HWE can be initiated in adulthood or even during pregnancy and not reveal itself years earlier although the triggering factor (i.e. hot water bathing) is still present. Another question is why different types of epilepsy (e.g. hot water epilepsy, febrile seizures, Dravet syndrome and severe myoclonic epilepsy of infancy) share an overlapping pathology; probably due to a common genetic background. Undoubtedly, further research on genetics and pathophysiology may reveal the generating mechanisms and treatment alternatives not only for HWE but also for other reflex epilepsies. A complicated combination of elements such as genetic predisposition, geographical distribution and cultural features is the key to understand this rare but fascinating form of epilepsy.

References
13. Franzoni, E., Gentile, V., Grosso, S., Brunetto, D., Cordelli, D.M., & Balestri,


32. Fukuda, M., Morimoto, T., Nagao, H. & Kida, K. Clinical study of epilepsy with severe febrile seizures and seizures