

Hot water epilepsy under 3 years of age: A short term follow-up study

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Abstract

Aim: Hot water epilepsy (HWE) is a type of reflex epilepsy that occurs as a result of immersing or pouring hot water over the head. Clinical and laboratory findings and short-term neurodevelopmental outcomes of patients were discussed in this report.

Materials and Methods: Eight patients under the age of three and followed-up with a diagnosis of HWE between 2017 and 2020 were enrolled in the study. Patient files were evaluated retrospectively.

Results: All our patients except one were boys. The patients' mean age at admission was 16.6 months (min: 10, max: 36), and the mean follow-up period was 13.5 months (min: 6, max: 25). Two patients also experienced non-reflex motor seizures not related to bathing, and these received daily oral antiepileptic therapy. Six patients had focal onset secondary generalized seizures, while one had focal seizures and one has hypomotor seizures accompanied by loss of consciousness. neurodevelopmental milestones were compatible with the patient's age in all followed-up cases. Clobazam administration before bathing is currently an effective and easily applicable method for this condition.

Conclusion: Hot water epilepsy is mostly benign. Clobazam before bathing is an effective option. Daily oral antiepileptic therapy should be administered to patients with seizures accompanying HWE.

Keywords: Childhood; clobazam; hot water epilepsy

INTRODUCTION

Reflex epilepsy is a type of epilepsy involving seizures that occur with any sensory stimulus. It occurs when the cortical and subcortical brain areas over-react to a physiological stimulus. Photosensitive reflex seizures are the most common form. Music, reading, and eating are rarely seen as triggers. Hot water epilepsy (HWE) is another type of reflex epilepsy that occurs as a result of immersing or pouring hot water over the head. It was first described by Allen in 1945 (1). While common in Asia, there are also reports from many other parts of the world (2).

The prevalence of HWE in India varies between 3.6 and 6.9% (3). In Turkey, HWE is present in 0.6% of epileptic patients (4). This study evaluated the demographic data, laboratory results and treatments of HWE patients under three years of age. To the best of our knowledge, this report is the largest case series of HWE in children under three.

MATERIALS and METHODS

Following approval by the local ethics committee, eight patients under the age of three and followed-up with a diagnosis of HWE between 2017 and 2020 were enrolled in the study. Patient files were evaluated retrospectively. Neuroimaging studies, electroencephalography (EEG) and laboratory results were obtained from hospital files.

Since videos taken by the patients' families at home could not be recorded onto the patient files, the views and comments of the clinician who watched the videos were employed instead. Due to the difficulties involved in bathing patients in the laboratory environment, ictal EEG could not be performed in any case. At the time of diagnosis all patients underwent sleep-deprived EEG with 21 electrode electroencephalography according to the international 10-20 system using a Nihon Kohden electroencephalograph. Patients who were followed-up for less than six months or lost to follow-up were excluded from the study.

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RESULTS

All our patients except one were boys. The patients' mean age at admission was 16.6 months (min: 10, max: 36), and the mean follow-up period was 13.5 months (min: 6, max: 25). All neurological and systemic examinations were normal. All patients have no history of epilepsy in their family except one. Routine laboratory tests including complete blood count, serum biochemistry, thyroid function test and vitamin B12 were normal.

Seizures are classified by the ILAE 2017 operational classification of seizure types (5). Two patients also experienced non-reflex motor seizures not related to bathing, and these received daily oral antiepileptic therapy. Six patients had focal to bilateral tonic-clonic seizures, while one had focal onset seizures without awareness and one has hypomotor seizures accompanied by impaired awareness and behavioral arrest. During the

first hospital visits, four patients were misdiagnosed with breath-holding spells and one with syncope. The correct diagnosis was made with the help of home video recordings and clinical findings. In all cases, the temperature of the bathwater was reduced before medical treatment, but no benefit was obtained. Five patients are currently benefiting from clobazam (0.5 mg/kg/dose) before bathing. One patient uses carbamazepine (20 mg/kg/day) and another uses levetiracetam (20 mg/kg/day), these two not only have HWE but also seizures not related to bathing previously. While patient no.1 had only one non-provoked seizure, patient no.2 had two focal onset seizures with impaired awareness. All patients underwent 12-channel electrocardiography for cardiac arrhythmias, but no pathology was detected.

The patients' demographic and clinical features are summarized in Table 1.

Table 1. Patients' demographic and clinical characteristics

Case	Age, month	Seizure onset, month	Sex	Family History	Neuro developmental state	Initial diagnosis	Seizure type	EEG	MRI	Seizure out of bath	Treatment	Follow up, month
1	20	11	F	Non	Normal	HWE	Focal onset BTC	N	N	Present	LEV+ CLBZ before bath	9
2	43	18	M	Non	Normal	HWE	Focal onset BTC (previously pathologic)	N	N	Present	CBMZ+ CLBZ (before bath)	25
3	24	14	M	Non	Normal	HWE	Hypomotor	N	N	Non	CLBZ (before bath)	10
4	16	10	M	Non	Normal	BHS	Focal onset BTC	N	N	Non	CLBZ (before bath)	6
5	48	36	M	Non	Normal	BHS	Focal onset BTC	N	N	Non	CLBZ (before bath)	12
6	24	17	M	Present	Normal	BHS	Focal	N	N	Non	CLBZ (before bath)	7
7	36	15	M	Non	Normal	Syncope	Focal onset BTC	N	N (arachnoid cyst)	Non	CLBZ (before bath)	21
8	30	12	M	Non	Normal	HWE	Focal onset BTC	N	N	Non	CLBZ (before bath)	18

CLBZ: Clobazam, CBMZ: Carbamazepine, BTC: Bilateral tonic clonic, HWE: Hot water epilepsy, LEV: Levetiracetam

DISCUSSION

The cause of HWE is still unclear. Convulsions have been observed when rats are placed into hot water. It has therefore been suggested that repetitive exposure to high temperature causes neuronal excitability (6). However, it is still unclear whether the temperature is the only cause. Although the water temperature in all our cases was reduced from 40-50°C to 30°C, the seizures still could not be controlled. Kowacs et al. proposed two distinct entities, hot water epilepsy and bathing epilepsy, depending on the temperature of the bathing water (7). HWE cases are thought to involve a central thermoregulation defect. Patients with Dravet syndrome also experience recurrent seizures due to hot water immersion and fever associated with central thermoregulation disorder. Two of our patients also had seizures without bathing. No other finding indicative of Dravet syndrome was detected in our patients, and further genetic screening was not therefore required. SLC1A1 mutation has also been reported to be capable of affecting glutamate uptake and kinetics and of causing HWE (7). Consistent with our patients, focal and focal onset secondary generalized seizures have been mostly observed in the literature (9). No cases with generalized onset seizures exist in our study.

Previous reports have frequently described focal onset seizures without and with awareness. HWE in children exhibits heterogeneous symptomatology, such as slowing of movements, blank stares, and perioral cyanosis. Although patients had no lateralizing neurological deficit, 3 Tesla magnetic resonance imaging (MRI) of the brain was performed to detect any cerebral focal lesions, since focal onset seizures were present in all cases. MRI was normal in all cases, except for one patient with an arachnoid cyst in the posterior fossa, which was not considered to be epileptogenic. Gross et al. reported a pediatric case with a parietal lobe lesion and presenting with HWE (10). It may therefore be concluded that structural or functional disorders of the parietal lobe may be responsible for the pathophysiology in HWE. The temporal lobe was also implicated in a report in which single-photon emission computed tomography was applied to a small number of patients (11). In none of our cases, pathology was detected in the temporal or parietal lobe.

Electroencephalographic evaluation was performed on all patients, and no epileptic activity or disorganization was detected in any case. EEG performed at another center was reported to be pathological in patient No. 2, but repeat examinations were normal. This patient also had focal onset seizures not related to hot water. Case reports in the literature have almost always reported no pathology on EEG (12). The frequency of EEG abnormality in HWE studies in Turkey is higher than elsewhere in the world. Gokcil et al reported pathological EEG in seven out of eight patients, and Bebek et al. in 13 out of 21, while all 10 children in Erdem et al.'s study were pathological (5,13,14). Interestingly, in Erdem et al.'s study, despite the patients' EEG pathologies, one patient had only recurrent febrile seizures other than HWE.

Erdem et al. also reported that pathology was detected on the EEGs of all patients, and that one patient even exhibited 3-3.5 Hz generalized spike and slow-wave activity. Paradoxically, however, that patient experienced absence seizures only with hot water. To the best of our knowledge, no such case has been described in the previous literature. Sirsi et al. reported generalized spike and wave activity in a case with SYN1 mutation, and autism spectrum disorder, cognitive impairment and drug-resistant reflex epilepsy, similar to this patient (14). Our patients had normal neurological and systematic examinations, had no dysmorphic findings, and had no history of febrile convulsions, this can be a reason why most EEGs are normal in our series.

Vasovagal syncope, breath-holding spells and arrhythmias should be considered at differential diagnosis of children with HWE. Iron deficiency anemia due to nutritional problems in developing countries may cause particular diagnostic confusion by precipitating breath holding spells. A detailed history may be the best guide. Video recordings of the event taken with mobile phones are also very valuable when bathing children at home.

The home video recordings of all but one of our cases were evaluated by a pediatric neurologist, and this supported information obtained from histories. Although HWE often begins in infancy, the age at diagnosis in India and Turkey is higher than in other countries. This may be due to clinicians being less knowledgeable about the disease.

The prognosis of HWE is often favorable. The duration of follow-up in our cases varied between 6 and 25 months. Even if patients had non-reflex seizures accompanying HWE, neurodevelopmental milestones were compatible with the patient's age in all followed-up cases. It may therefore be concluded that non-reflex seizures accompanying to HWE are not a poor prognostic indicator.

Therapeutic approaches may differ between centers. Reducing the bath-water temperature is often beneficial in adulthood (14). Phenytoin and phenobarbital were mostly used in the 20th century (13). However, benzodiazepine administered before bathing is currently an effective and easily applicable method in patients who fail to benefit from modifications to the physical environment in the bathroom (like decreasing water temperature) (16,17). "Benzodiazepine before baths" is frequently used in patients without non-reflex seizures accompanying HWE, since it increases drug compliance and causes fewer drug-related side effects. In our case series, clobazam therapy was initiated one hour before bathing, except for two cases with non-reflex seizures accompanying HWE, and all seizures stopped. The other two cases (cases 1 and 2) are currently seizure-free with daily levetiracetam and carbamazepine therapies, respectively. In these patients, bathing habits also were changed. Their daily antiepileptic treatment was effective in non-bathing seizures, but benzodiazepines before baths were used for bathing seizures and became more effective.

Some patients turn hot water attacks into a ritual event and self-induced seizures being observed (4). Behavioral therapeutic options should be considered in these patients. "Self-abortion" is also an effective method in adult patients (18).

CONCLUSION

Hot water epilepsy can also occur in early childhood and can be misdiagnosed as a non-epileptic phenomenon. The condition is mostly benign. Clobazam before bathing is an effective option. Daily oral antiepileptic therapy should be administered to patients who have also non-reflex seizures accompanying HWE.

Competing Interests: The authors declare that they have no competing interest.

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REFERENCES

1. Allen IM. Observations on cases of reflex epilepsy. *N Z Med J* 1945;44:135-42.
2. Kurata S. Epilepsy precipitated by bathing: A follow-up study. *BrainDev* 1979;11:400-5.
3. Satishchandra P, Shivaramakrishna A, Kaliaperumal VG, Schoenberg BS Hot water epilepsy: a variant of reflex epilepsy in Southern India. *Epilepsia* 1988;29:52-6
4. Bebek N, Gurses C, Gokyigit A et al. Hot water epilepsy: clinical and electrophysiologic findings based on 21 cases. *Epilepsia* 2001;42:1180-4.
5. Fischer RS, Robert S, French, JA. et al. Operational classification of seizure types by the International League Against Epilepsy: Position Paper of the ILAE Commission for Classification and Terminology. *Epilepsia*, 2017;58:522-30.
6. Klauenberg BJ, Sparber SB. A kindling-like effect induced by repeated exposure to heated water in rats. *Epilepsia*. 1984;25:292-301.
7. Kowacs PA, Marchioro IJ, Silva EB Jr et al. "Hot-water epilepsy", "warm-water epilepsy", or bathing epilepsy? Report of three cases and considerations regarding an old theme. *Arq Neuropsiquiatr* 2005;63:399-401.
8. Karan KR, Satishchandra P, Sinha S, Anand A. Rare SLC1A1 variants in hot water epilepsy. *Hum Genet* 2017;136:693-703.
9. Satishchandra P. Hot-water epilepsy. *Epilepsia* 2003;44:29-32.
10. Grosso S, Farnetani MA, Francione S, et al. Hot water epilepsy and focal malformation of the parietal cortex development. *Brain Dev* 2004;26:490-3.
11. Patel M, Satishchandra P, Aravinda H et al. Hot water epilepsy: Phenotype and single photon emission computed tomography observations. *Ann Indian Acad Neurol* 2014;17:470-2.
12. Kaplan Y, Kurt SG, Karaer H et al. Intra-familial incidence and characteristics of hot water epilepsy. *Can J Neurol Sci* 2009;36:575-81.
13. Erdem E, Topcu M, Renda Y, et al. Hot water epilepsy. *Clin Electroencephalogr* 1992;23:152-8.
14. Zeki G, Ilker IH, Hidir UU, Zeki O. Hot water epilepsy: seizure type, water temperature, EEG findings and treatment published correction appears in *Neurologist* 2010;16:210.
15. Sirsi D, Armstrong D, Munoz-Bibiloni J et al. SYN1 Gene Mutation in a Child with Focal Epilepsy and Reflex Bathing Seizures *J Pediatr Epilepsy* 2017;6:119-24.
16. Incecik F, Herguner MO, Elkay M, Altunbasak S. Hot water epilepsy - a report of three cases. *Indian Pediatr* 2004;41:731-3.
17. Dhanaraj M, Jayavelu A. Prophylactic use of clobazam in hot water epilepsy. *J Assoc Physicians India* 2003;51:43-4.
18. Savitha MR, Krishnamurthy B, Ashok DA, Ramachandra NB. Self abortion of attacks in patients with Hot Water Epilepsy. *Indian Pediatr* 2007;44:295-8.