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Hot water epilepsy with alone and spontaneous seizures in childhood

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ABSTRACT

Objectives: Hot water epilepsy (HWE) is a type of epilepsy that primarily affects children. This study was aimed to evaluate the clinical, electroencephalogram (EEG), neuroimaging findings, and treatment options in children with HWE.
Methods: The medical records of 24 patients who had HWE were evaluated retrospectively.
Results: There were 2767 patients diagnosed with epilepsy during the seven-year period, and 0.86 % of the patients had HWE. The median age of the patients was three (range 1.2–7 years), with a male predominance (male/female ratio: 7.1). Six patients (25 %) had HWE with spontaneous seizures (HWESS) and 18 patients (75 %) had HWE alone (HWEA). 11 patients had focal onset seizures, 13 patients had generalized onset seizures. EEG abnormalities were found in 7 patients (29.2 %). Three patients (12.5 %) had nonspecific MRI findings. Developmental abnormalities (autism spectrum disorder, learning disability and speech disturbance.) were detected in 8 patients (33.3 %). Only one patient's (4.2 %) seizure could be controlled by changing bathroom habits. Twenty-three patients (95.8 %) were given antiepileptic drugs. 18 of 24 patients had come for follow-up visits for two years, nine of them used monotherapy and seizures did not recur. The treatment response was 55.5 %. Oxcarbazepine (8 patients, 33.3 %) and valproic acid (7 patients, 29.2 %) were the most chosen two drugs for HWE. The genetic tests performed were not accepted relevant to the patients' clinical conditions and epilepsy.
Conclusion: The frequency of the HWE was not as high in the literature. Male predominance, EEG abnormalities may be seen. Changing bath room habits did not improve the treatment as a first line management, all the patients except one used antiepileptic drug treatment. Until now, there has been no study in Turkey showing the frequency of HWE exclusively in children.

1. Introduction

Reflex epilepsy is mostly triggered by visual stimuli. Hot water epilepsy (HWE) is a form of reflex epilepsy provoked by bathing with hot water and pouring it overhead (Epilepsydiagnosis.org., 2021). It is the second most common type of reflex epilepsy, accounting for 6.9 % of all epilepsies (Satishchandra and Sinha, 2013; Sharma and Sankhyan, 2021). Although the exact pathogenesis of HWE remains unclear, some hypotheses have been suggested. It may be related to damage to the thermoregulation center in the hypothalamus. Studies have identified a focus associated with local hypometabolism in the temporo-occipital region without structural lesions (Satishchandra and Sinha, 2013; Pejaver et al., 2015). This type of reflex epilepsy distinguishes between bathing epilepsy and hot water epilepsy (HWE). BE is caused by a complex combination of lukewarm water and other environmental sensory stimuli. HWE have a genetic predisposition (Turkey and India), and immersion in hot water (40–50°C) causes seizures. (Kowacs et al., 2005, Accogli et al., 2021). Seizure may occur as an up-rolling of the eyes, staring, suddenly becoming floppy afterwards with generalized seizures, and stiffening of all four limbs followed by jerky movements (Bharathi et al., 2021). It may progress to spontaneous seizures with variable outcomes. Using lukewarm water with some antiepileptic drugs is among the treatment options (Pejaver et al.,2015; Meghana et al., 2012). The objective of this study is to evaluate the clinical, electroencephalogram (EEG), neuroimaging findings, and therapeutic outcomes among children with HWE.

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2. Material and methods

2.1. Study population

Patients under 18 years of age were included in the study. There were 2767 patients diagnosed with epilepsy during the seven-year period, and 24 patients had HWE. The medical records of 24 patients with HWE were evaluated retrospectively at the pediatric neurology outpatient clinic.

2.2. Seizure classification

Seizures were classified according to the new terminology of the International League Against Epilepsy (ILAE) (Scheffer et al., 2017). ILAE 2017 classification of seizure types expanded version and common descriptors of behaviors during and after seizures were used. (Fisher et al., 2017).

Behavior arrest was accepted as focal onset, nonmotor seizure; cyanosis, vomiting, abdominal pain, and headache were grouped as focal, nonmotor onset autonomıc seizures. Simple partial seizures were accepted as focal onset aware seizures,Tonic clonic seizures were accepted as generalized onset motor seizures. Atonic seizures were accepted as generalized onset motor sezures.

2.3. Data collection

The seizure type was determined based on the history taken from the parents and the video footage, if any. Turkish people often bath by pouring hot water (40–50 °C) over their heads repeatedly.This study included participants whose seizures were triggered by pouring hot water over head. Patients who had seizures with HWE were classified as HWE alone (HWEA). Patients who had spontaneous seizures besides HWE were classified as having HWE with spontaneous seizure (HWESS). Patients' gender, age, development, history and family history of febrile seizures and epilepsy, genetic testing, electroencephalography (EEG), magnetic resonance imaging (MRI), and treatment options were evaluated. Change in bathroom habits were advised of all patients. Advised bathroom habits were using lukewarm water, initially washing the body parts other than head first, washing head at last, and getting shower not pouring the water with a stoup over head.

2.4. Ethical conduct of research

The local ethics committee approved the study.

3. Results

3.1. Sample description

During the seven-year period, 2767 patients were diagnosed with epilepsy. A total of 24 patients with HWE were included in this study. The frequency of the HWE was 0.86 % in the present study. Six patients (25 %) were HWESS, and 18 patients (75 %) were HWEA. Of these, 21 (87.5 %) were male and three (12.5 %) were female (male/female ratio 7.1). The median age of the patients was three (range 1.2–7 years), with a mean age of 4.31±3.54. The mean age of patients with HWESS was 7.08 ± 4.82, and the mean age of patients with HWEA was 3.38 ± 2.56.

3.2. Seizure characteristics

According to the ILAE seizure classification, there were focal onset seizures in 11 patients (45.8 %); cyanosis in 5 patients (20.8 %), behavioral arrest in 3 patients (12.5 %), vomiting in 2 patients (8.4 %), and abdominal pain and headache in 1 patient (4.1 %). 13 (54.2 %) of the patients had generalized-onset motor seizures; tonic clonic seizures in 11 patients (45.8 %) atonic seizures in 2 patients (8.4 %). Clinical findings are shown in Table 1.

3.3. Factors associated with HWE

Seven patients had genetic testing. One only had chromosome analyisis (CA), the other six patients had chromosomal microarray (CMA) besides CA. One had CA, CMA, fragment analysis of FMR1 gene. Patient 3 had 9p13.1p12 and 22q11.23q12 deletion (1.7 megabase, 55 marker); Patient 10 had normal CMA and Fragile X pattern; Patient 12 had Xq28 deletion(5.8 kilobase, haploinsufficiency score 3); Patient 16 had normal CMA, Patient 17 had 5p.13.2 deletion(426 kilobase, 25 marker), 6q26 duplication (224 kilobase, 31 marker); Patient 23 had normal CMA; Patient 4 had normal CMA. These seven patients had normal CA. The results of CMA were not relevant to the clinical features of the patients.These results were not considered significant. The patients did not have a family history of HWE. Patient 7,9,12,16,17,19 had family history of epilepsy (25 %); Patient 4 had family history of febrile seizures. Table 2 compares the clinical characteristics of the patients with genetic testing in the current study to pediatric, and adult patients with HWE and BE in the literature. EEG abnormalities were detected in 7 patients (29.2 %), including sharp wave complexes in 5 patients and spike wave discharges in 2 patients (Fig. 1). While brain MRI was normal in 21 patients (87.5 %), three patients (12.5 %) had nonspecific findings (arachnoid cyst in two patients, corpus callosum cyst in one patient) (Fig. 1). Three of the patients had febrile convulsions (Patients 1, 9, and 17) before HWE; one had a family history of febrile seizures (Patient 4). Patient 17 previously presented with febrile seizures. Then hot water spells started. Afterwards he had spontaneous seizures. Four patients (Patients 19, 20, 21, and 23) experienced spontaneous seizures prior to HWE, Patient 21, also had post-traumatic epilepsy. In patient 12, hot water seizures were followed by spontaneous seizures.

Neurological examination of all patients were normal except patient 17. He had nystagmus.

Neurological development was normal in 16 patients (66.7 %), while developmental delay was present in eight (33.3 %) patients. Three (37.5 %) of them with abnormal development had HWESS, and five (62.5 %) had HWEA ($p = 0.36$). Autism spectrum disorder (ASD) was seen in four patients (16.7 %), speech disturbance in two patients (8.3 %), and learning problems in two patients (8.3 %). EEG was abnormal in 3 (18.8 %) of 16 patients with normal development and 4 (50 %) of 8 patients with developmental delay ($p = 0.16$). Epileptic activity was detected in 4 (19 %) of 21 male patients and 2 (66.7 %) of 3 female patients ($p = 0.075$). EEG was epileptic in 3 (50 %) of 6 patients with HWESS and 4 (22.2 %) of 18 patients with HWEA ($p = 0.3$). EEG abnormalities were detected in 29.2 % of the patients (sharp wave complexes in 5 patients and spike wave discharges in 2 patients). Sharp wave discharges (Patient 19), and spike activity over the C3 electrode (Patient 22) were observed in the central and parietal regions There was no significant relationship between neuromotor development and HWESS, family history of epilepsy ($p = 0.31$, $p = 0.76$, respectively).

3.4. Treatment

Since the patients had seizures every time they had poured hot water over head, it was first recommended to change their bathroom habits. Since the frequency and severity of the seizures in all but one patient did not alter with the regulation of bathroom habits, antiepileptic treatment was started. Twenty-three patients (95.8 %) were treated with antiepileptic drugs. Oxcarbazepine (8 patients, 33.3 %) and valproic acid (7 patients, 29.2 %) were the most chosen drugs for HWE**.** The prophylactic clobazam was used for three patients (12.5 %). Clobazam was less preferred because there was a shortage in its supply in our country. Six (25 %) out of 24 patients did not come for follow-up visits (patients 1,5, 7, 14, 15, 21). Four of these patients had used monotherapy. Seizures recurred in two of them. 18 of 24 (75 %) patients had come for follow-up visits for two years. Five (20.8 %) patients had HWESS. The other 13

Table 1
Patient Characteristics.

Patient	Age(Years/ Gender	Charectristics of Seizure/ Classification	EEG	MRI	1st Drug	2nd Drug	Development	Types of Seizure with HWE
1	1.5/M	Cyanosis/Focal onset	Normal	Normal	PB	None	Normal	HWEA
2	2/M	Vomiting/Focal onset	Normal	Normal	PCLBZ	None	Normal	HWEA
3	7/M	Headache,abdominal pain/ Focal onset	Normal	Normal	OXC	VPA	Learning Disability	HWEA
4	4/M	Vomiting/Focal	Normal	Normal	OXC	None	Normal	HWEA
5	2/M	Tonic clonic convulsion/ Generalized onset	Normal	Normal	OXC	None	Normal	HWEA
6	1/M	Cyanosis/Focal	Normal	Normal	PB	None	Normal	HWEA
7	1/M	Tonic clonic convulsion/ Generalized	Normal	Normal	OXC	None	Normal	HWEA
8	5/M	Tonic clonic convulsion/ Generalized	Normal	Arachnoid cyst	OXC	VPA	Normal	HWEA
9	1/M	Tonic clonic convulsion/ Generalized onset	Normal	Normal	VPA	LEV	Normal	HWEA
10	3/M	Tonic clonic convulsion/ Generalized onset	Generalized Sharp-wave complexes	Normal	VPA	None	Autism	HWEA
11	4/M	Cyanosis/Focal onset	Bilateral temporal Sharp-wave complexes	Normal	OXC	VPA	Normal	HWEA
12	1/F	Tonic clonic convulsion/ Generalized onset	Generalized Sharp-wave complexes	Normal	PB	VPA	Speech disturbance	HWESS
13	1/M	Cyanosis/Focal onset	Normal	Normal	VPA	OXC	Normal	HWEA
14	1/M	Tonic clonic convulsion/ Generalized onset	Normal	Normal	OXC	LEV	Autism	HWEA
15	8/M	Behavioral arrest/Focal onset	Normal	Normal	CBZ	LEV	Normal	HWEA
16	7/M	Tonic clonic convulsion/ Generalized onset	Generalized Sharp-wave complexes	Normal	VPA	None	Normal	HWEA
17	1,5/M	Behavioral arrest/Focal onset	Normal	Normal	VPA	LEV	Normal	HWESS
18	1,5/M	Tonic clonic convulsion/ Generalized onset	Normal	Normal	OXC	None	Speech disturbance	HWEA
19	7/M	Atonic seizure/Generalized onset	Left centroparietalSharp-wave complexes	Normal	CBZ	None	Autism	HWESS
20	11/M	Tonic clonic convulsion/ Generalized onset	Normal	Normal	CBZ	None	Normal	HWESS
21	12/M	Tonic clonic convulsion/ Generalized	Normal	Arachnoid cyst	VPA	None	Normal	HWESS
22	8/F	Cyanoisis/Focal onset	Left central Spike -wave activity	Normal	VPA	None	Normal	HWEA
23	10/M	Behavioral arrest/Focal onset	Generalized Spike -wave activity	Normal	LTG	LEV	Autism	HWESS
24	3/F	Atonic Seizures/ Generalized onset	Normal	Callosal Cyst	Bathroom habits	None	Learning disability	HWEA

HWEA: HWE alone HWESS: HWE with spontaneous seizure, M: Male, F: Female, CBZ: Carbamazepine, LEV: Levetıracetam, LTG: Lamotrigine, VPA: Valproic acid, OXC: Oxcarbazepine, PB: Phenobarbital, PCLBZ: Prophylactic clobazam,

(54.2 %) patients had HWEA. Of the 18 patients who continued with follow-up visits, 9 of them (50 %) overwhelmed the fits with monotherapy. One patient was treated with bathroom habits, and seizures did not recur. Seizures recurred only in 5 patients with HWEA and 3 patients with HWESS. Considering the response to treatment in the patients who were followed up, the treatment response in our study was 55.5 %. Seizure recurrence was not related to the type of HWE ($p = 0.6$).

Valproic acid was used in four patients (16.7 %), levetiracetam was used in five patients (20.8 %), and oxcarbazepine was used in one patient (4.2 %) as the second choice drug. The findings of the patients with HWESS and HWEA are shown in Table 3.

4. Discussion

There is differential diagnosis between HWE and BE. Kowacs et al. (2005) stated that the absence of similar spells in other hot environments, such as saunas or furnaces, suggests the presence of complex tactile stimuli. Temperature may not be the most important factor for triggering seizures. Their reports of two of the three cases support this view, as bathing at a warm but not hot temperature also caused seizures. This finding supports the use of the term "bathing epilepsy" instead of "hot-water epilepsy" to describe patients with this disorder.

HWE is a type of epilepsy that primarily affects children (Satishchandra and Sinha, 2013). HWE is precipitated by the stimulus of bathing by pouring hot water over the head. Some have seizures even during body washing. Children and males are affected more frequently than adults and females. Because of their bathroom habits, it is more common in India and Turkey (Satishchandra and Sinha, 2013; Meghana et al.,2012; Gürses et al., 2019). Turkish people frequently bath by pouring hot water (above 39 °C) over their heads. Hot water epilepsy may be seen frequently for this reason.

HWE accounts for 6.9 % of all epilepsy cases (Satishchandra and Sinha, 2013; Sharma and Sankhyan, 2021). In Turkey, Bebek et al. (2001) reported HWE in 0.6 % of epileptic patients, both children and adults. We found a frequency of 0.86 % in the present study. Çıraklı et al. (2020) and Gürbüz et al. (2021) did not mention the frequency among children. While HWE is mostly reported in the form of case reports in Turkey, no study indicating its frequency in children with hot water epilepsy has been conducted so far.

There is a male predominance; Meghana et al. (2012) observed a male predominance of 4:1, whereas Bharathi et al. (2021) reported a male-to-female ratio of 3:1 with the participants, who were 36 drug-naive patients with HWE. We also found male predominance (male/female ratio: 7.1).

Table 2
Review of the Literature with current study.

Reference		Gender	Age Onset	Epillepsy Type	Seizure	Genetic Tests	Interictal EEG	Treatment
Türay and Eröz (2021)		M	21 months	HWE	Cyanosis after bathing with hot water	POGZ gene frame shift variation (Sutton syndrome)	Sharp wave discharge in the right posterior area	CBZ, LEV, PCLBZ
Belcastro et al. (2009)		M	15 years	HWE	Generalized tonic clonic seizures, sense of fear, dazed look, irrelevant speech	Gs-alpha gene (GNAS 1α) (Mc Cune Albrıght Syndrome)	Normal	Not available
Santos-Silva et al. (2015)		M	5 years	HWESS	Infantile spasm, Eye deviation, and flexion of the neck tonic movement of upper and lower limbs with hot water	GPR56 Mutation 811 C>T (R271X)	High amplitude frequent bursts of multifocal spikes	POBZP (Prophylactic oral benzodiazepine)
Gürbüz et al.	Case 1	F	11 months	HWE	Focal onset Bilateral tonic clonic	Not available	Normal	LEV,PCLBZ
	Case 2	M	18 months	HWE	Focal Onset Bilateral tonic Clonic	Not Available	Normal	CBZ, PCLBZ
	Case 3	M	14 months	HWE	Hypomotor accompanied by impaired awareness	Not available	Normal	PCLBZ
	Case 4	M	12 months	HWE	Focal onset Bilateral tonıc clonıc	Not available	Normal	PCLBZ
Patient 3		M	7 years of age	HWEA	Headache, abdominal pain, Focal onset, autonomic, nonmotor onset	9p13.1p12 deletion, 22q11.23q12 deletion (benign, not relevant to clinical findings	Normal	VPA
Patient 10		M	3 Years of age	HWEA	Tonic clonic generalized onset	Normal	Generalized sharp wave complexes	VPA
Patient 12		F	1 years	HWESS	Tonic clonic generalized onset	Xq28 deletion (Clin Gene HI:3), not relevant to clinical findings	Generalized sharp wave complexes	VPA
Patient 16		M	7 years	HWEA	Tonic clonic generalized onset	Normal	Generalized sharp wave complexes	VPA
Patient 17		M	1.5 years	HWESS	Behavioral arrest Focal onset, nonmotor	5p13.2 deletion, 6q26 duplication, not relevant to clinical findings	Normal	VPA, LEV
Patient 23		M	10 years	HWESS	Behavioral Arrest, Focal onset, nonmotor	Normal	Generalized Spike Wave Activity	LTG, LEV
Patient 24		F	3 years	HWEA	Atonic Seizures, Generalized onset	Normal	Normal	Changing bath habits
Mosquera-Gorostidi et al. (2019)	Case 1	M	10 months	Bathing Epilepsy	Pallor, cyanosis, limpness, staring gate	Not available	Normal	VPA
	Case 2	F	13 months	Bathing Epilepsy	Perioral cyanosis, head drop, staring gaze	Not available	Normal	Changing bath habits
Kuang et al. (2024)		F	4 months 20 days	Bathing Epilepsy	Eyes gazing to right with clonıc movements of right face and lips, bradycardia impaired consciousness	SMC1A gene (c.298+2 T>C)	Mild slow back ground, focal interictal epileptic activity	Drug resistant epilepsy, LEV,VPA, TPM
Braun et al.		M	19 months old	Bathing Epilepsy	Unresponsive, eyes deviating to left, shallow irregular breathing	2q22.3q23.2	Normal	LEV, CLBZ
Kowacs et al.	Case 1	M	28 years old (Age onset: 14 years)	Bathing Epilepsy	Headache, cephalic parasthesia, and negative motor phenomenon on his left upper limb	Not available	Right rolandic focus, left temporal spikes	CBZ
	Case 2	F	30 years of age (Age onset: 26 years)	HWE and Bathing Epilepsy	Brief complex visual hallucinatory phenomenon, speech arrest, vomiting, generalize seizure	Not available	Not available	CBZ, phenobarbitone
	Case 3	F	20 years	Bathing Epilepsy	Partial complex, fugacious with the left hand and lips closing.	Not available	Normal	Phenytoin, CBZ, Clonazepam
Altun et al. (2017)		M	24 years	Bathing Epilepsy	Frequent difficulty breathing, paleness, teeth squeezing, purple discoloration of the lips, oral automatisms, unconsciousness	Not available	Normal	VPA
Accogli et al.	Case 1	M	18 years (Age onset: 5 years)	Bathing Epilepsy	Impaired awareness, pallor, cyanosis, oral automatism, hypotonia	SYN1 mutation c.1264 C>Tp (Arg 422)	Right temporal and left anterior temporal epileptic findings	CLBZ, VPA

(*continued on next page*)

Table 2 (*continued*)

Reference		Gender	Age Onset	Epillepsy Type	Seizure	Genetic Tests	Interictal EEG	Treatment
	Case 2	M	47 years of age	Bathing epilepsy	Focal impaired awareness, orolingual automatism, salivation and spitting, right hand tapping, left ward head and gaze deviation	SYN1 mutation c.1266delAp. (Gb423Serfs*244)	Normal	CBZ, LTG, VPA
Nguyen et al. (2015) (Proband of the study)		M	29 years of age (Age onset: 4 years)	Bathing Epilepsy	Complex partial seizure after bath, chilling aura alteration of consciousness, oral and manual automatism	SYN1 Q555X	Normal	PB, CBZ,CLB, PHT, LTG

GNAS 1α: Gs-alpha gene, GPR: G protein-coupled receptor proteolytic site, HI: Haplotype insufficiency, HWE:Hot water epilepsy, HWESS: HWE with spontaneous seizures, HWEA: HWE alone, M: Male, F: Female, CBZ: Carbamazepine, LEV: Levetıracetam, LTG: Lamotrigine, VPA: Valproic acid, PB: Phenobarbital, PCLBZ: Prophylactic clobazam, PHT: Phenytoin, PGOZ: pogo transposable element-derived protein with zinc finger domain POBZP: Prophylactic oral benzodiazepine

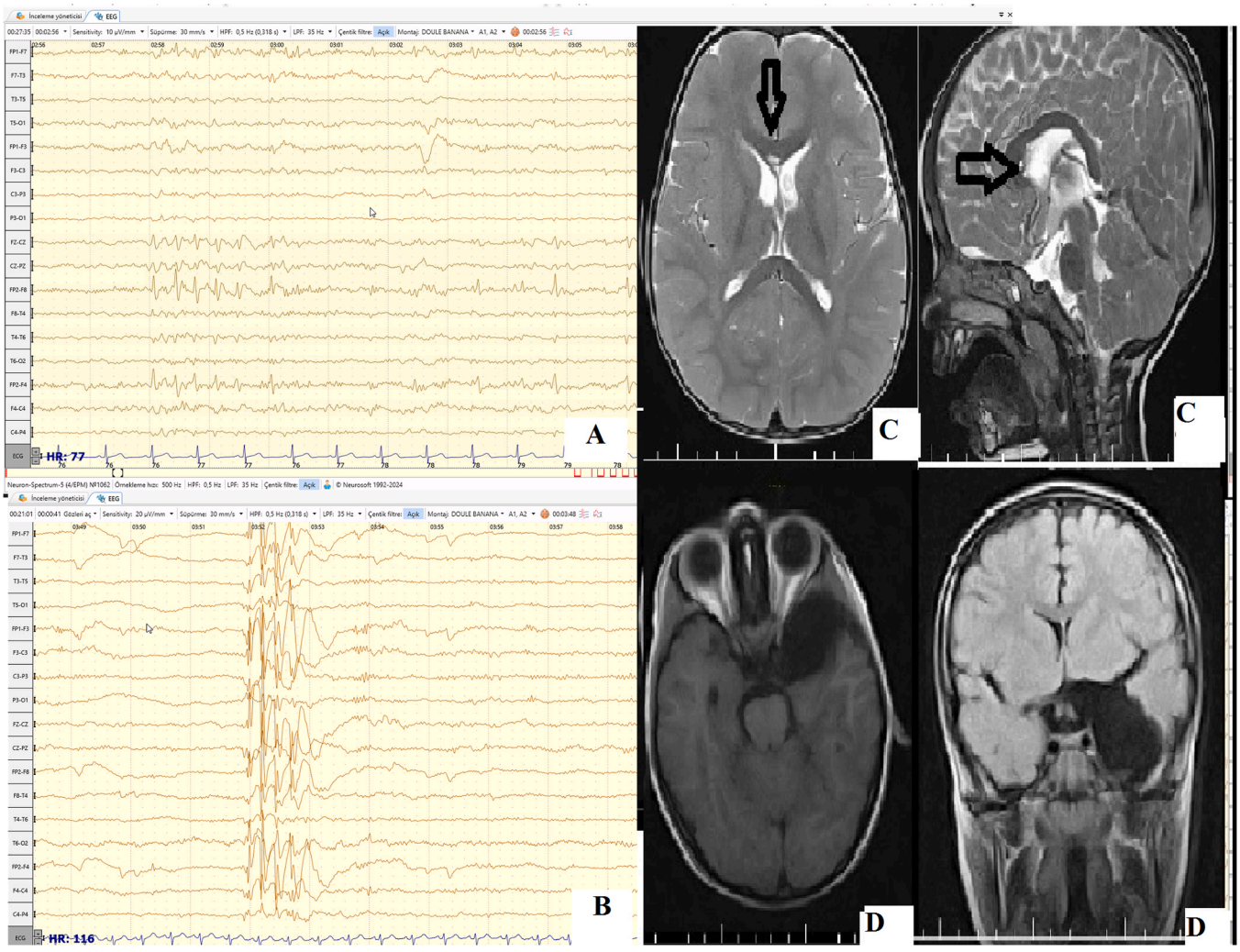

Fig. 1. A. Patient 23, biphasic sharp wave complexes over right frontal and central electrodes (10μV/mm, 30 mm/sec, 0.318 sec). B. Patient 19, sharp wave discharges (20μV/mm, 30 mm/sec, 0.318 sec) C. Corpus callosum cystD. Temporal arachnoid cyst C. EEG findings Patient 23 spikes multiple spike wave discharges.

The disease's onset age, which is in the first decade, varies in the literature (Satishchandra and Sinha, 2013; Meghana et al., 2012). Meghana et al. (2012) reported a mean age of onset of HWE 18.7 ± 10.2 years. Gürses et al.(2019) reported case series with a mean age of 11.4 years. Bharathi et al. (Bharathi., et al., 2021) found that the age at onset of HWE was mostly between 1 and 5 years of age. Çıraklı et al. (2020) reported a median age of 30 months with HWE and Gürbüz et al. (2021) reported a mean age of 16.6 months. We found that the disease onset was in the first decade with a mean age of 4.31±3.54.

Family history of HWE, epilepsy, febrile seizures, and head trauma are the risk factors for HWE (Gürses et al.,2019). The studies showed that 11 %–27 % of HWE patients had a history of febrile seizures before the development of this type of reflex epilepsy (Satishchandra and Sinha, 2013).Our study revealed similar findings with the literature.

In the ILAE 2017 classification, reflex epilepsy is categorized under electroclinical syndrome of variable age onset. According to the new ILAE 2017 seizure classification, complex partial seizure is defined as focal onset awareness or impaired awareness, and complex partial

Table 3
The findings of the patients with HWESS and alone.

	Hot Water Epilepsy			
Characteristics	HWESS		HWEA	
	n	%	n	%
Gender				
Male	5	20.8	16	66.7
Female	1	4.2	2	8.3
Clinical Findings				
Cyanosis	-	-	5	20.8
Vomiting	-	-	2	8.3
Tonic Clonic Seizure	3	12.5	8	33.3
Atonic Seizure	1	4.2	1	4.2
Headache Abdominal Pain	0	-	1	4.2
Behavioral Arrest	2	8.3	1	4.2
Seizure Type				
Generalized onset	3	12.5	10	41.7
Focal onset	3	12.5	8	33.3
EEG				
Normal	3	12.5	14	53.8
Abnormal	3	12.5	4	16.7
MRI Findings				
Normal	5	20.8	16	66.7
Anormal	1	4.2	2	8.4
Development				
Normal	3	12.5	13	54.2
Abnormal	3	12.5	5	20.8
FS				
Yes	1	4.2	2	8.3
No	5	20.8	16	66.7
FS history in family				
Yes	-	-	1	4.2
No	6	25	17	70.8

EEG: Electroencephalogram, FC: Febrile seizure, HWEA: HWE alone HWESS: HWE with spontaneous seizure, MRI: Magnetic resonance imaging

seizure with secondary generalization is defined as focal to bilateral tonic-clonic seizures (Scheffer et al., 2017). According to Gürbüz et al. (2021), focal-onset-impaired awareness seizures were the most common type of HWE; generalized-onset seizures may also be seen. Seizures are characterized by a sense of fear, auditory or visual hallucinations, meaningless looks, and tonic-clonic seizures (Scheffer et al., 2017; Gürses et al., 2019). Gürses et al. (2019) described three patients with focal-onset-impaired awareness seizures. We found that generalized -onset seizures were most common and mostly seen with tonic clonic convulsions. Meghana et al. (2012) detected that the frequency of patients with HWEA was 64 %, while the frequency of patients with HWESS was 36 %. We also found that HWEA was seen mostly.

Interictal scalp EEG is usually normal but may show localized or lateralized spike discharges in the anterior temporal regions and in the front temporal region on either or both sides. Most focal abnormalities, predominantly in the temporal region, are reported on the EEG (Satishchandra and Sinha, 2013; Meghana et al.,2012). In their study, epileptic activity was detected on the EEG in 21 % of the patients. We found epileptic activity on the EEG at a similar rate. We found epileptic activity in the central, centroparietal, and temporal regions. EEG abnormalities were more common in patients with HWESS.

In the majority of cases, there is no structural abnormality on neuroimaging studies. MRI imaging may demonstrate parietal cortex lesions triggering seizures or focal hypometabolism of the temporoocipital region without structural lesions (Satishchandra and Sinha, 2013; Gürses et al., 2019). Our study revealed nonspecific brain abnormalities.

Patients with HWE may have developmental delays. Ratnapriya et al. (2009a), (2009b) described two patients with a locus for autosomal dominant reflex epilepsy precipitated by hot water that had normal development. Bebek et al. (2001) reported two HWE patients with intellectual disabilities.

In this study, we detected developmental abnormalities, such as autism, learning disability, and speech disturbance. Developmental abnormalities may be a consequence of genetic abnormalities. Since our study was a retrospective study, the genetic tests performed included CA,and CMA in a few patients. The genetic results found in the CMA were not accepted relevant to the patients' clinical conditions and epilepsy. Prospective studies, including genetic studies, may provide us with new information about both HWE and syndromes.

Treatment choices may change according to the patient and specialist. If HWE is not treated, it might progress to spontaneous seizures with variable outcomes (Satishchandra and Sinha, 2013; Meghana et al.,2012; Okudan and Özkara, 2018). Decreasing the hot water temperature is an effective management strategy. Treatment options include carbamazepine, lamotrigine, phenytoin, phenobarbital, sodium valproate, oxcarbazepine, levetiracetam, and prophylactic clobazam taken orally prior to bathing. Phenytoin and carbamazepine may be used as treatment options when a patient has spontaneous seizures in addition to hot water-induced seizures (Satishchandra and Sinha, 2013; Meghana et al.,2012; Okudan and Özkara, 2018). Satischandra and Shinha, (2013) and Ekici et al. (2017) reported the prophylaxis usage of intermittent clobazam (5–10 mg orally) 1.5–2 hours before bathing.

In this study, only one patient's seizure could be controlled by lowering the water temperature and changing bathroom habits. The rest of the patients were given antiepileptic drugs. Oxcarbazepine was the most used drug.

5. Conclusion

We found that spontaneous seizures and developmental abnormalities accompanied HWE. EEG and neuroimaging can often found to be normal But in the current study the patients had epileptic abnormalities on interictal EEGs. Most of the patients who had monotherapy were HWEA. Although all patients were advised to change their bathroom habits first, this was ineffective as the first treatment option.

Until now, there has been no study in Turkey showing the frequency of HWE exclusively in children.

The frequency of HWE was not as high as in the literature. It may be due to the fact that the patients may mistake the symptoms of HWE or that the doctors who referred the patients to the pediatric neurology department can mistakenly identify HWE as a non-epileptic occurrence.

HWE may not be directly related to developmental delay. Further genetic testing may contribute to understanding HWE with developmental delay and syndromes.

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CRediT authorship contribution statement

Arzu Ekici: Supervision, Methodology, Conceptualization. **Sevgi Yimenicioglu:** Writing – review & editing, Writing – original draft, Data curation, Conceptualization.

Declaration of Competing Interest

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