

## REFLEX-EPILEPSY INDUCED BY TAP WATER BATH. A Case Report and Review of the Literature.

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### ABSTRACT

Reflex epilepsy is a condition in which seizures can be provoked habitually by exclusively external stimulus or, less commonly, internal mental processes. It may coexist with spontaneously occurring seizures. Reflex seizures are epileptic rather than psychogenic in nature, presenting often as either focal-onset or primary generalized seizures, with epileptiform EEG changes. Bathing epilepsy or as commonly named hot water epilepsy (HWE), is a form of reflex or sensory epilepsy in which seizures are precipitated by the contact of hot or tepid water over the head and body. We report a case of an 18-month-old Sudanese toddler boy, presented with recurrent abnormal funny turns, noticed by his parents, since he was 6-month old. These events occur while having a bath by pouring tap water over his head. The diagnosis of epilepsy was supported by interictal electroencephalography (EEG). This rare type of epilepsy has a geographical distribution. To our knowledge, this might be the first case report from African and Arab Nations.

**Key words:** *Benign reflex epilepsy, boy, hot/warm water epilepsy, ictal/ interictal EEG, physiopathology, Sudanese toddler, water-immersion epilepsy.*

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### Case Report

An 18 month-old Sudanese boy, presented to our clinic with abnormal funny turns noticed by his parents, following a bath, on different occasions. His condition was recurrent, not associated with any other specific provoking stimuli. Shortly after a bath he had abnormal eye gaze. He used to become floppy and unconscious and collapsed without tonic, clonic or myoclonic movements. He did not respond to any vocal or tactile stimuli during the attack which usually last about one minute and ended with yawning. The child was a product of a full term uneventful pregnancy and labour. He had no history of neurological illness or trauma and he had normal developmental milestones. No family history of similar conditions. Physical examination including detailed neurological assessment was unremarkable.



**Fig. 1** The child interacting happily with staff before the bath.



**Fig.2** The mother bathing the child.

The authors witnessed the event following an experiment in the hospital, after having a formal consent of both parents. The child was bathed by pouring tap water over his head. See the sequence of the event as demonstrated by the attached photos. Figures: 1, 2, and 3(a & b).

Ictal EEG was technically difficult; however interictal EEG was performed while the child succumbed to sleep by chloral hydrate. The record showed normal sleep changes of stage I and II superimposed on a normal background brain slowing. Bursts of high amplitude intermixed regular and irregular slow and sharp waves observed frequently, they were sometimes preceded and followed by electrodecremental activities. Occasionally they cluster around the parieto-posterior temporal regions bilaterally. During the awake period no epileptogenic focus discharge was observed Figures 4, 5 and 6(a & b). These EEG features were suggestive of a generalized active seizure

- discharge probably of bilateral parieto-posterior-temporal initiation. MRI brain scan was normal.



a)



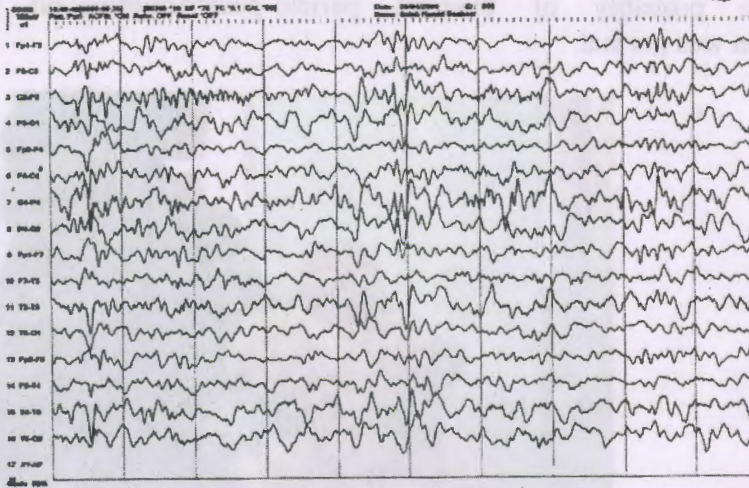
b)

**FIG. 3 (a) & (b) The child in the ictal phase**

The child was started on sodium valproate. Three months later, at first follow-up visit, his convulsions had decreased but did not completely stop during regular bath. We built up the dose, at a second follow-up visit, now he is seizure-free even with a bath.

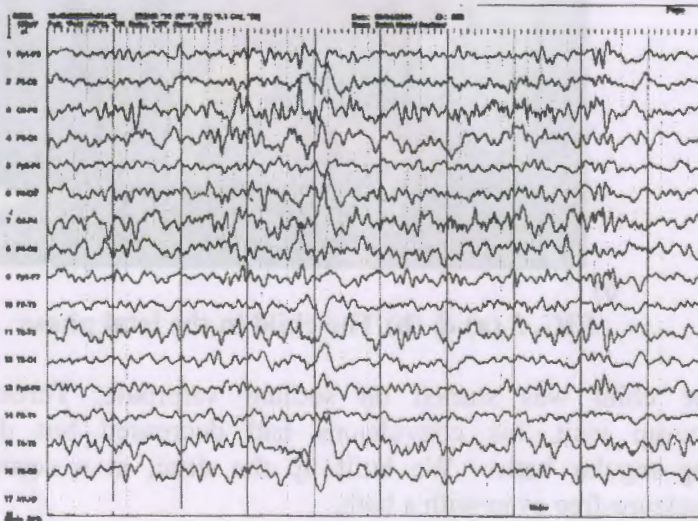
#### **Discussion**

The immediate factors that provoke any given seizure are complex and the occurrence of seizures in someone with epilepsy is rarely predictable. In contrast, reflex seizures are predictable in response to a specific stimulus.



**Fig.4 EEG record showing normal sleep changes of stage I and II superimposed on a normal background brain slowing.**

Hot water epilepsy, also known as bathing epilepsy or water-immersion epilepsy is a rare form of epilepsy, refers to a specific type of reflex epilepsy precipitated by the stimulus of bathing in hot water. This is a benign form of reflex epilepsy, first described in 1945<sup>1</sup>. Its incidence could be underestimated because of confusion with febrile convulsions, vagal fits, or aquagenic urticaria<sup>2</sup>. *Beaumanoir A:*



**Fig. 5 Burst of high amplitude intermixed regular and irregular slow and sharp waves.**

Hot water epilepsy is considered to be a geographically specific epileptic syndrome, it accounts for 6.9% of all epilepsies in certain communities. It is most prevalent in India and Turkey than in Europe, Japan or North America. The induction of epileptic seizures by immersion in hot water has only rarely been reported in western countries<sup>2</sup>. Different bathing habits and genetic factors may be responsible for the high incidence of HWE in these regions. The exact pathogenesis of HWE is not known<sup>3</sup>.



**Fig. 6. Intermixed slow and sharp waves preceded and followed by electrodecremental activities.**

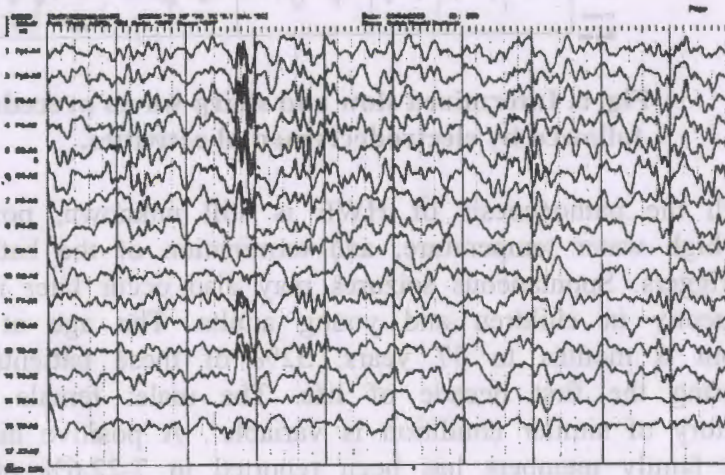
Although the pathogenesis of HWE is still unknown, pouring water over the head, high water temperature, and termination of the bath were the most common triggers. Spontaneous seizures may also occur later in life<sup>1</sup>. It occurs most frequently in children and young adults. The age at onset of HWE ranged from 6 months to 37 years, 52% of these patients had their first seizure during the first decade of life. The male: female ratio was (3:1). Family history of similar condition is variable<sup>1</sup>. A positive history of epilepsy among the family members has been reported in 7-22.6% of cases. Children are of normal psychomotor development and they continue to develop normally after seizure. None had any neurological deficits<sup>2</sup>.

The pattern of seizures is of complex partial with or without secondary generalization. Onset include dazed look, sense of fear, irrelevant speech, visual and auditory hallucinations with complex manifestations<sup>4</sup>. One third of all reported cases have primary generalized tonic-clonic seizures. Usually these seizures last 1-3 minutes and manifests either at the beginning or at the end of the bath<sup>5</sup>.

*Auvin, S.*<sup>6</sup> and his colleagues reported that seizures episodes consisted of a loss of consciousness absence followed by left predominant hypotonia with right fronto-temporal high voltage slow waves on the ictal-EEG. Temporal lobe has been thought to take part in the epileptogenesis. Interictal EEGs obtained in 22 patients showed epileptiform abnormalities over temporal regions in 41%. Brain MRIs in 15 patients and CTs in 2 were normal except for cortical atrophy in 1 and mesial temporal sclerosis in 1. Treatment with antiepileptic drugs (AEDs), mainly carbamazepine, in 16 patients prevented seizure recurrence; avoidance of the precipitating factor prevented seizures in the remainder<sup>4</sup>. A classification proposed by ILAE task force in 2001 include hot-water epilepsy, under reflex epilepsy<sup>3</sup>.



(A)



(B)

**Fig. 7: Slow wave foci on the parieto-posterior temporal regions (a) evolved to secondary generalization (b).**

### Pathophysiology

The trigger mechanism of this unique type of epilepsy as a type is hyperthermia. Aberrant thermoregulation in the genetically susceptible population with possible co-existing environmental influence could be the probable mechanism responsible for this epilepsy. Complex tactile stimuli might play the most relevant role on seizure triggering, as well as water temperature with an additive effect over cutaneous stimulation. Partial and generalized tonic-clonic reflex seizures related to hot water bathing have been described as temperature-related stimulation. Seizures was reported to be precipitated by very hot water head baths (40–50°C). *Auvin, S.*<sup>7</sup> et al, reported a water reflex epilepsy in a 6 year-old girl, occurring at lower temperature than the core temperature. Still the mechanism of HWE is unclear. A hot-air stimulus to the scalp failed to induce attacks in susceptible patients.

Nechay, A. et al,<sup>8</sup> reviewed the paroxysmal disorders of infancy and of the newborn in which the normal process of bathing may be an important trigger, particularly focused on infant bathing in normal water temperature (37 degrees C, range 36-38 degrees C) rather than in hot water that is above body temperature. Four principal diagnostic categories emerged: bathing

epilepsy, alternating hemiplegia of childhood, hyperekplexia and paroxysmal extreme pain disorder. Bathing or water immersion epilepsy was the best studied and is arguably distinct from hot water epilepsy. The paroxysmal episodes previously attributed to aquagenic urticaria may have been examples of bathing epilepsy with a genetic component. Despite suggestions in the literature to the contrary, no convincing reports of bath-induced infantile syncope have been found. The underlying mechanisms of bath-induced paroxysmal disorders in infancy remain poorly understood, but all have autonomic manifestations and some if not all may be channelopathies<sup>8</sup>.

Animal models of genetic reflex epilepsy have been described. These include the baboon, *Papio papio*, in which generalized convulsions can be induced by visual stimulation. Audiogenic seizures characterize genetic reflex epilepsies in predisposed strains of mice, rats, and birds. Rat model and human experimentation had proven that HWE is a type of hyperthermic seizure with possible kindling on repeated stimulation in animals<sup>9</sup>. Genome-wide linkage analysis of three out of five families showing concordance for the disease haplotype to chromosome 10q21. Sequence analysis of a group of functional candidate genes, the ion channels *KCNMA1*, *VDAC2* and solute carriers *SLC25A16*, *SLC29A3* revealed no potentially pathogenic mutation<sup>10</sup>. An association with McCune-Albright syndrome have been reported; this is a rare disease due to post-zygotic and somatic mutations of the *Gs-alpha* gene that results in cellular mosaicism. The main features occur in the bony skeleton, the skin, the endocrine system and in atypical presentations, in other non-endocrine tissues. It is unknown whether or not an expression of the *GNAS1* product in the brain does exist. Although the association of McCune-Albright with hot water epilepsy may be merely casual, it brings up the possibility that the salient phenotypic variability of MAS might also include epilepsy<sup>11</sup>.

### **Electroencephalography**

EEG recordings demonstrate focal activity in the temporal or frontal lobe. Interictal scalp EEG is usually normal; 15 to 20% might show diffuse abnormalities. Ictal EEG recordings has technical limitations and difficult to obtain. Ullal, G. R. et al., studied kindling and mossy fibre sprouting in the rat hippocampus following hot water induced hyperthermic seizures, induced epileptic seizures in 36 male Wistar albino rats by means of hot water sprays at 48 h time intervals. Seizure discharges (EEG) were recorded from ventral hippocampus in six of these rats<sup>9</sup>. Yalcin, A. D. et al., on studying hot water epilepsy clinical and EEG features of 25 cases, reported interictal EEGs epileptogenic abnormalities located over the temporal regions in nine patients<sup>12</sup>. For detailed characterization of clinical seizure type or in cases requiring differential diagnosis or pre-surgical monitoring, the EEG-video monitoring must be used<sup>12</sup>.

### **Imaging studies**

MRI examination should be considered for this group of patients, as in the evaluation of other focal epilepsies, guided by the electro-clinical features of the seizures. Since some types of reflex seizures can occur in the context of symptomatic, localization-related epilepsy, perform brain MRI as in the evaluation of other focal epilepsies<sup>13</sup>.

The high rate of epileptogenic abnormalities localized in the temporal region and the complex partial seizures observed in most reported patients

indicate the considerable role of the temporal lobe mesial focus in hot water epilepsy. However, there were no demonstrable structural changes on MRI and CT scans except in a few case reports, such as hippocampal sclerosis, dysplasia, and a huge cystic lesion<sup>13</sup>.

### Treatment

Therapy of reflex seizures involves limiting exposure to the provoking stimulus, as well as standard epilepsy medications. These seizures usually respond to valproate treatment<sup>3</sup>. Carbamazepine and oxcarbazepine are also useful permitting the disappearance of seizures and an improvement of executive disorders. However, antiepileptic medications are selected on the basis of electro-clinical seizure type and when choosing antiepileptic medications, considerations are similar to those in other types of epilepsy. Electroclinical seizure type, prior treatment history, patient age, comorbidities and medication adverse effects are primary considerations. Parents of infants with seizure susceptibility should be counseled to avoid pouring hot water over their infants' heads during bathing.

### Prognosis:

Hot water epilepsy is a benign form of epilepsy<sup>1,2</sup>. Seizure course and psychomotor development are usually favourable because of a good response to pharmacological treatment and the tendency toward spontaneous remission of the seizures<sup>3</sup>.

Avoiding lukewarm water is indicated and Understanding that HWE is a hyperthermic seizure changes the use of regular AEDs management for these seizures to intermittent prophylaxis with benzodiazepines<sup>14</sup>.

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