

An Unusual Cause of Syncope in Children: Hot Water Epilepsy

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Abstract

The Syncope is defined as a sudden transient loss of consciousness with inability to maintain postural tone. Neuro cardiogenic or vasovagal syncope is the commonest cause of syncope in children. Approximately 30–50% of children are likely to have had a benign fainting episode before 18 years of age. However, syncope may be the 1st manifestation of an underlying life-threatening cardiac anomaly like prolonged QT syndrome or cardiomyopathy. Syncope in a young child is unusual and always warrants a thorough investigation. We present an interesting case of a one-year old Omani girl referred to us for evaluation of syncope.

Keywords: Syncope • children • Hot water epilepsy

Introduction

A one-year-old Omani female infant was referred to the cardiology clinic with history of 3 episodes of sudden loss of consciousness and floppiness with the last episode followed by generalized abnormal movements. The attacks had occurred over past one month, always while bathing the child and particularly after giving a head bath. The water was warm as per parents. The child would suddenly become floppy, hypotonic and lose consciousness. The last episode was followed by generalized twitching movements lasting about 2 minutes. Within minutes of removing from the bath, the child would return to normal. Her growth and development were appropriate for age. Her perinatal history was normal. There was no history of sudden death or epilepsy in the family.

On assessment, she was conscious and oriented. Her vital signs were normal. Her heart sounds were normal and there were no added sounds. There were no pyramidal tract signs or localizing signs. Routine blood tests including a full blood count, blood glucose and biochemistry were normal. An Electrocardiogram (ECG) done showed no abnormalities with a corrected QT interval of 424 msec and the baseline echocardiogram was normal. Her evaluation was completed by referring her to neurology clinic where an inter-ictal sleep electroencephalogram done was normal. A brain MRI done was also within normal limits. A working diagnosis of Hot Water Epilepsy (HWE) or reflex epilepsy was made. The parents were explained the diagnosis and were reassured that it was not a serious or life-threatening condition. She was not given any anti-convulsant medications and the reasoning was explained to them. They were advised to bathe her with lukewarm water. On follow-up visits, there were no seizure recurrences.

This convinced the parents about the diagnosis. They were counselled about the appropriate water temperature for bathing the child and about avoiding pouring water directly on the child's head to avoid seizure triggering.

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They were also educated about the need for caution to be taken in the future especially related to taking showers and swimming.

Discussion

Syncope is a common presentation in children and the causes may vary from benign to life threatening conditions. Prolonged syncope may cause seizures secondary to hypoxia and some seizure types like atonic seizures may mimic syncope. Certain arrhythmias may be precipitated by activities like bathing, swimming and exercise especially in those with prolonged QT syndrome [1,2].

Reflex epilepsies, as a group, are rare and constitute a group where some, or all, epileptic seizures happen due to a specific trigger. Triggers may be external, like flashing lights or internal, like emotional or behavioral stress. As a group they constitute about 6% of all epilepsies [3,4]. HWE is an uncommon form of reflex epilepsy, usually brought about by a hot water bath. It is postulated to be mediated through specific thermal cutaneous stimuli [3,4]. Seizures may be partial or generalized through a possible common afferent pathway [5,6]. Sporadic cases have been reported world over with a large series from South India where head bathing of infants is part of a ritual [7,8].

Some severe myoclonic epilepsies of infancy are known to be triggered by hot water. However, HWE is a distinct entity. Seizures have been reported after taking a cold water bath, a tub bath or even when showering [8,9]. The exact etiology of HWE remains elusive and there have been reports of seizures even with below the head bathing. In the series from India, ritual head immersion in water at around 45°C was seen to produce seizures. A dysfunctional thermo-regulatory center sensitive to sudden changes in local temperatures during bathing especially in colder months could precipitate HWE. Febrile epilepsy was not seen in these patients [8]. An animal model to simulate the events occurring during HWE was studied to unravel the possible underlying neurophysiological basis [9]. Occurrence of cases within families lead to the possibility of an underlying genetic predisposition to HWE and a candidate locus 10q21.3-q22.3 was identified [10].

The diagnosis is usually clinical based on the events described by the patient as usually brain imaging studies and electroencephalograms during the inter-ictal period do not reveal abnormalities. Due to some reports of HWE like seizures even while bathing with normal or cold water, some groups have recommended to rename this entity as bathing epilepsy [11]. The possibility of an underlying channelopathy or dysfunctional autonomic regulation has been proposed as an underlying etiology [12]. Initial clustering of cases in certain geographic locations led some researchers to believe this to be an area specific syndrome in a genetically predisposed population triggered by external environmental stimuli [13]. Studies have demonstrated seizure

discharges on EEG after hot water immersion as a confirmation of this entity; however, the same is not essential for diagnosis of HWE [6].

HWE is more common in males and is seen throughout childhood. Treatment usually is restricted to modifications of the water temperature and bathing technique with avoidance of pouring water directly over the head or showering neck downwards. Anti-epileptic medications are usually not prescribed [14]. Anti-epileptic drugs like clobazam taken 1-2 h before bath have been shown to have good results with minimal side effects. [5,15]. in event of occurrence of other types of seizures or seizures occurring with daily regular baths despite above precautions would warrant the use of anti-epileptics [15]. As the condition tends to improve with time usually, a trial of gradual downgrading and stopping the medications may be warranted in some cases after a seizure-free interval of more than a year or so. Good control of HWE can usually be achieved with behavioural modifications of bathing techniques without use of medications [3].

Conclusion

The case report was an example of how non cardiac conditions may come to cardiology clinic as disorders mimicking as syncope. Paediatricians and paediatric cardiologists need to keep an open mind for non-cardiac causes of syncope at the same time ruling out serious cardiac conditions.

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The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Data Availability

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