Head up tilt test in the diagnosis of neurocardiogenic syncope in childhood and adolescence

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Background: Neurocardiogenic syncope (NCS) is a common paroxysmal disorder that is often misdiagnosed as a seizure disorder. Head up tilt test (HUTT) has been used to confirm this diagnosis. There is no data available of its use in children / adolescents from India. Aim: To study the usefulness of the HUTT in children and adolescents with suspected NCS. Settings and Design: This was a part retrospective and later prospective study set in a tertiary child neurology outpatient department (OPD). Material and Methods: Patients with a strong clinical suspicion of syncope were recruited for the study. Clinical and treatment details were either retrieved from the chart or prospectively recorded in later patients. The HUTT was then carried out at baseline and after provocation and the results correlated with the clinical diagnosis. Results: Eighteen children with a mean age of 10.8 years were studied. Eight had precipitating factors. Thirteen had premonitory symptoms. Pallor, temperature change, diaphoresis, headache, tonic / clonic movements, post-ictal confusion and peri-ictal headache were symptoms noticed. Sixteen had a positive HUTT. Seven were on long-term anti-epileptic drugs (AEDs). Two had epileptiform abnormalities on their electroencephalogram (EEG). Conclusion: The diagnosis of syncope is often confused with epilepsy. Head up tilt test has a high sensitivity in the diagnosis of NCS in children / adolescents. It is fairly safe and easy to perform.

Key Words: Syncope, Head up tilt test, childhood

Introduction

Syncope is a common paroxysmal disorder in children and adolescents. It is characterized by a sudden, brief loss of consciousness and postural tone followed by spontaneous recovery.1 It needs to be differentiated from other paroxysmal disorders like seizures, vertigo, panic attacks and hysteria. Syncope can result from a variety of cardiac conditions e.g. aortic stenosis, bradyarrhythmias and from neuropsychiatric disorders. However, in childhood and adolescence, neurally-mediated mechanisms are responsible for most cases of syncope, which is now called neurocardiogenic syncope (NCS).

For several years, the diagnosis of NCS has been achieved only after excluding other similar disorders by history, examination and investigations such as two-dimensional echocardiography (2D-Echo), ambulatory electrocardiographic monitoring and electroencephalography (EEG).2 Though adequate in the majority this approach tends to lead to a misdiagnosis. This is especially true with the convulsive form of NCS where epilepsy is often diagnosed.3

The head up tilt test (HUTT) has been used in the evaluation of NCS since the last 15 years.4 It has also been used in children since the early nineties.5,6 We describe our experience in Indian children and adolescents.

Material and Methods

We reviewed all our patients who had been subjected to the HUTT over 2 years (2000-2001) to confirm or exclude the diagnosis of NCS. The last few patients were enrolled prospectively after undertaking the study. Hence our study was retrospective and part prospective. We excluded those who did not undergo tilt testing. The details of clinical history and examination were retrieved from the hospital’s outpatient medical records. Specific clinical details sought included the presence of prodromal symptoms, number of episodes, circumstances surrounding each episode and eyewitness observations of the actual episode. Inquiry was also made for the presence of associated migraine-like symptoms as well as similar symptoms in family members. Whether the event/events had been diagnosed as seizures/epilepsy and whether any anti-epileptic drugs (AEDs) were being taken was also noted. CNS and CVS examination details were specifically looked for.

A 12-lead electrocardiogram (ECG) was taken in all patients.

The HUTT was undertaken in a cardiology office setup after informed consent. After a variable period of fasting and a pretest pe-
Neurocardiogenic syncope is the commonest type of syncope in children/adolescents with a prevalence rate of 15% between 8-18 years. Cardiac causes are only rarely seen. In one study of 35 patients no cardiogenic syncope was seen. Because of its paroxysmal nature, it is often misdiagnosed as a seizure disorder. History in a case of NCS can be typical with episodes being momentary, occurring in an upright position in the wakeful state and usually (but not always) precipitated by acute illness, noxious stimuli, emotional upsets, use of certain medications or acts such as micturition, though not always. However, several difficulties may be encountered in reaching the diagnosis. It may not always be possible to get an accurate history as these episodes often occur in school or outside homes and may not be witnessed or witnesses may not be available for interview. Convulsive syncope, where clonic movements are seen sometimes during the episode, adds to the diagnostic conundrum. It is commonly believed that disorientation after the event and unconsciousness for more than 5 minutes favors the diagnosis of a seizure. However, presence of post-episode confusional state was seen in 50% of our patients with syncope. The clinical examination is unremarkable as it is expectedly normal and there are no biochemical markers for this disorder. It is of note that 1/3 of the patients had active migraine. This association is well described.

Thus, the diagnosis rests on the exclusion of other disorders on the basis of examination and investigations. EEGs or ambulatory ECG monitoring for paroxysmal disorders like seizures and cardiogenic syncope may be false negative. On the other hand, the presence of EEG abnormalities does not necessary mean the presence of seizure disorder as up to 5% of normal children are known to have epileptiform abnormalities. These problems often lead to a misdiagnosis of epilepsy. In our series 1/3 of the patients were on AEDs, some for prolonged periods.

The HUTT provides a simple confirmatory test and has
been in use since the mid-1980s. The methodology of HUTT, however, is not yet standardized. Various tilt angles are in use and have different sensitivities/specificities. Agents such as isoproterenol infusion, or sublingual glyceryl trinitrate are used as provocating agents to increase the sensitivity of the test. However, this is not recommended in all protocols.

Testing in children has been gaining acceptance in recent years. However, there is hardly any published Indian literature available.

In our series, the majority with clinically definite NCS had a positive test. The one negative test was in a young child where the provocative test with nitroglycerine was not carried out. The inability to perform this in very young children may constitute a limitation of the test. The precipitation of patient-specific syncope-like symptoms during the test is crucial to the specificity of the test. Our positive rate of 94.1% is higher than the 44-76% rates reported. This is possibly due to the following reasons: 1) we performed the test close to the time of the symptoms, which increases the positive yield, 2) we used a large tilt angle (80 degrees), which has been associated with a higher positive rate, and 3) we used glyceryl trinitrate as a provocative agent thereby increasing the positivity rate as well. False positive tests are of concern and have been variably estimated at between 6-17%. Higher tilt angles and increased duration of the HUTT seem to increase sensitivity at the expense of specificity. In teenage subjects in a recent study, using different tilt angles, found unacceptably high false positives, as high as 60%, casting doubts on the veracity of the tilt test. Other reports however do not confirm this.

As we were using intermittent BP monitoring every 2-3 minutes it is theoretically possible that we may have missed a brief drop in BP between measurements. This is unlikely to have occurred as at the time of any reported symptoms two simultaneous BP readings were taken. Symptom reproduction is necessary along with hypotension and/or bradycardia for the HUTT to be accepted as positive. Hence even if we did miss an asymptomatic brief drop in BP, the test would not have been classified as positive and would not have changed our results.

Another event missed by the routine HUTT is the recently described cerebral syncope. Here, symptoms of syncope are reproduced by the tilt test in the absence of systemic hemodynamic changes. This often leads to a misdiagnosis of a psychogenic disorder. Simultaneous transcranial doppler ultrasonography and near infrared spectroscopy have however demonstrated paradoxical cerebral vasoconstriction and transient cerebral hypoxia induced by the tilt test resulting in transient loss of consciousness. We did not encounter any such patient in our series.

Our study confirmed the safety of this test. No adverse events were noted in any patient.

Our study, therefore, confirms that HUTT is useful to help reach a diagnosis of NCS. There are concerns about false positivity and one needs to interpret the results in light of the clinical scenario. A positive HUTT can be used to reassure the parents about the usually benign nature of the diagnosis. It can also be used to minimize, if not eliminate, the unnecessary use of anti-epileptic drugs.

References


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