Clinical score to predict the short-term outcome of pediatric status epilepticus

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ABSTRACT

INTRODUCTION

Unlike in adults, clinical scoring system to predict outcome and treatment response in pediatric status epilepticus is lacking. We used a pediatric modification of adult scoring system, Status epilepticus in pediatric patients severity score (STEPSS) to determine if it can accurately predict short term outcome.

MATERIAL AND METHODS

In this prospective study, children aged 1 to 18 years with seizure duration of ≥ 5 minutes or actively convulsing at emergency room were enrolled. STEPSS score was calculated at admission and outcome such as mortality, Pediatric overall performance category (POPC) at discharge and treatment response were noted. The diagnostic utility of STEPSS score to predict unfavorable outcome was calculated.

RESULTS

One hundred and twenty-five children with mean age 5.1 years were enrolled. Seven children died and 29 had unfavorable outcome. The predictive accuracy of STEPSS at cutoff >3 for mortality had sensitivity of 85.3, specificity of 60.2 and negative predictive value of 98.6 with area under curve 0.751 [95% CI: 0.593-0.908]). Likewise, it predicted unfavorable outcome with sensitivity of 51.7, specificity of 60.4 and negative predictive value of 80.6.

CONCLUSION

The STEPSS, a simple bedside clinic score was found to be valuable in predicting outcome and treatment response in children with status epilepticus.

KEYWORDS

Status epilepticus, STEPSS, Outcome

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INTRODUCTION

Status epilepticus (SE) is one of the commonly encountered neurological emergency in pediatric population with mortality around 3-12 %.[1-3] The outcome of pediatric status epilepticus is determined by various factors including age of child, presence of any neurological deficit at presentation and response to drug therapy.[4-8] Various clinical scoring system has been designed to determine outcome in status epilepticus in adults.[9-11] Status Epilepticus Severity Score (STESS) is one these widely used scoring system in adults. However, such scoring system is still lacking in pediatric population. Determination of such clinical parameters will not only help practicing clinicians to determine severity outcome but also decide which category of patients would require more aggressive therapy at presentation.

In this study, we decided to use a modified version of STESS known as Status Epilepticus in Pediatrics patients Severity Score (STEPSS) to predict short term outcome in status epilepticus in children.

MATERIAL AND METHODS

This was a prospective cohort study conducted in a tertiary level hospital in Western Nepal from April 2024 to October 2025. Children from 1-16 years presenting with seizure for at least 5 minutes duration or presenting at emergency department in active convulsing state were enrolled in our study. Ethical clearance for the study was taken from Institution review committee. (Reg. No: UCMS/IRC/061/24) Informed and written consent were taken for all parents before enrolling them into our study.

STESS score and its pediatric modification STEPSS

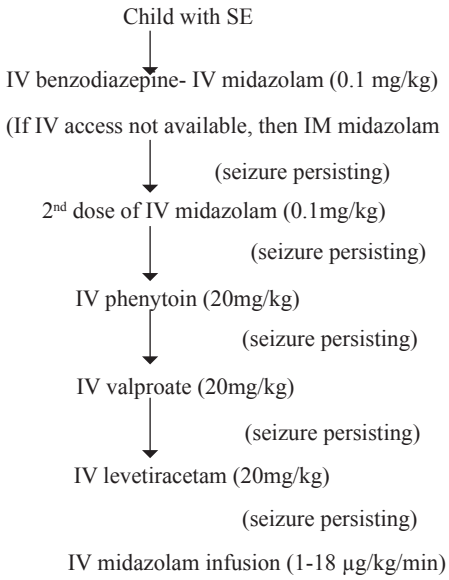
STESS is a simple bedside clinical score which is widely used in adult patients with SE. It comprises of 4 clinical variables- age of the patient, level of consciousness, past history of seizure and type of seizure. STESS score ranges from 0-6 with lower score indicating less severe form of presentation while higher score predicting poor outcome. In our study, age less than 2 years at presentation was given score of 2 points. This pediatric modification of STESS score was done based on review of available literature on prognostic factors of SE in children. Studies show that age less than 2 years have increased risk of mortality and therefore this age cutoff of 2 years was taken in STEPSS scoring system.[12] Rest of the clinical variables and the scoring system was similar to adults (table 2).

Case definition

SE was defined as seizure lasting for more than 5 minutes or if child is actively convulsing at presentation to pediatric emergency department.

Procedure

All children who presented with SE to our hospital were treated as per the standard hospital protocol. All base line clinical parameters were recorded as well as all baseline investigations as per treatment protocol were sent. The etiology of SE was determined according to information gathered from relevant history, clinical findings and investigations done.



Flow diagram: Management protocol for pediatric status epilepticus at UCMS

The anti-seizure medication (ASM) was considered effective if there was cessation of SE within 10 minutes of the initial dose of medication and there was sustained absence of seizure for 30 minutes. A patient was labelled as Benzodiazepine (BZD) responsive if SE responded with first or second dose of BZD. Established SE was defined as SE which responded to second line ASM after BZD, usually phenytoin. Refractory SE was defined as SE that persisted despite adequate doses of two appropriate ASM (BZD and phenytoin) and responded only to third line ASM or midazolam infusion. Super-refractory SE was defined as SE that persisted 24 hours or more despite onset of anesthesia or withdrawal of anesthesia.

Outcome

The short-term outcome of patient was assessed using the Pediatric Overall Performance Category (POPC) scale at the time of discharge (table 1). POPC scale scores of 1 and 2 were considered as favorable outcome whereas scores of 3 or more was labelled as unfavorable outcome. In children with pre-morbid developmental delay or disability, a return to baseline functional status was considered as favorable outcome whereas a decline from before was considered as unfavorable. The primary outcome was to determine the sensitivity of STEPSS to predict an unfavorable outcome and mortality. We also studied other diagnostic parameters such as specificity, negative predictive value (NPV) and positive predictive values (PPV). Likewise, we also analyzed the utility of STEPSS to predict treatment outcome as refractory and super refractory status epilepticus.

Table 1. POPC Scale

Score	Category	Description
1	Normal	Normal, at age-appropriate level, school-age child attending regular school classroom
2	Mild disability	Conscious, alert, and able to interact at age-appropriate level, school-age child attending regular school classroom, but grade perhaps not appropriate for age, possibility of mild neurological deficit
3	Moderate disability	Conscious, sufficient cerebral function for age-appropriate independent activities of daily life, school-age child attending special education classroom and/or learning deficit present
4	Severe disability	Conscious, dependent on others for daily support because of impaired brain function
5	Coma or vegetative stage	Any degree of coma without the presence of brain death criteria, unaware, even in awake in appearance, without interaction with environment, cerebral unresponsiveness and no evidence of cortex function (not aroused by verbal stimuli), possibility of some reflexive response, spontaneous eye-opening, and sleep-wake cycle
6	Brain death	Apnea, areflexia, and/or electroencephalographic silence

Statistical analysis

With the precision of 10% on either side and with confidence interval of 95%, 30 patients with unfavorable outcome were required; expecting 25% proportion of unfavorable patients, a total of 120 patients were needed for this study. Data was analyzed through SPSS 25. Data were presented as means, standard deviations and proportions with percentages. The predictive accuracy of STEPSS score for death, unfavorable outcome and development of refractory and super refractory status epilepticus was calculated using sensitivity, specificity, positive predictive value and negative predictive value at various cut off points. The receiver operating characteristic (ROC) curve was drawn and optimal cut-off point was determined using Youden's index (sensitivity+specificity-1).

RESULTS

Demographic and clinico-etiological profile

A total of 125 children with mean age of 5.1 years (standard deviation of 4.2 years) were enrolled in the study. Out of 125 children, 84(67.20%) were males. The approximate median duration of seizure prior to presentation to hospital was 15 minutes (Inter-quartile range: 10-25 min). Around 52% of the children had received treatment prior coming to the hospital. Only 23.2% of the children were known case of epilepsy while remaining had first episode of seizure presenting as status epilepticus.

The most common etiology was acute symptomatic which was present in 54 (43.20%) children followed by febrile status epilepticus which was present in 33 (26.4%) children. Remote symptomatic SE was present in 12(9.60%) patients. In 15.2% children, no definite cause was identified and labelled as idiopathic. Pre-existing co-morbidities were present in 14 (11.20%) children, of which cerebral palsy was present in 6 (4.8%) patients, developmental delay/ intellectual disability in 5 (4%) patients, hearing impairment in 2 (1.6%) patients and autism spectrum disorder in 1 (0.8%) patient.

STEPSS scores in the study population

109 (87.20%) children were either alert or somnolent/ confused at presentation to hospital while remaining 16 (12.80%) were stuporous or comatose. Generalized tonic clonic seizure was the most common seizure type present in 103 (82.40%) children followed by focal seizure which was present in 21 (16.80%) children. Only 1 child had non convulsive status epilepticus confirmed by continuous EEG recording. (table 2)

Table 2. Components of STEPSS score in study population (N = 125)

Features		Scores	N (%)
Consciousness	Alert or somnolent/confused	0	109 (87.2)
	Stuporous or comatose	1	16 (12.8)
Seizure type	Simple partial, complex partial, Absence, myoclonic	0	21(16.8)
	Generalized tonic clonic	1	103(82.4)
	Non convulsive SE	2	1 (0.8)
Age	≥ 2 years	0	78 (62.4)
	< 2 years	1	47 (37.6)
History of previous seizure	Yes	0	29 (23.2)
	No or unknown	1	96 (76.8)

Treatment response and outcome

Out of enrolled children presenting with SE, 35 (28%) were benzodiazepine (BZD) responsive while 76 (60.80%) children had established SE, i.e., unresponsive to second dose of BZD and required phenytoin. 10 (8%) children had refractory SE, i.e., unresponsive to BZD and phenytoin while 4 (3.20%) had super refractory SE. Seven (5.60%) children died. Overall, 96 (76.80%) children had a favorable outcome while 29 (23.2%) children had unfavorable outcome.

Predictive accuracy of STEPSS

The predictive accuracy of STEPSS at cut- off >3 for unfavorable outcome (POPC score ≥ 3) had sensitivity of 51.7, specificity of 60.4 and negative predictive value of 80.6 with Youden's index of 12.1. This predictive accuracy of STEPSS at cut- off >3 was more than another cut-off score (table 3). On analyzing with ROC curve, the area under curve was 0.48 (95% CI: 0.35-0.61) (fig 1) Hence STEPSS

score of more than 3 can be used to predict unfavorable outcome in case of pediatric SE. Table 3 summarizes the predictive values for various STEPSS scores in determining unfavorable outcome.

Table 3. Predictive accuracy of various STEPSS Scores in determining unfavorable outcome

STEPSS Score	Sensitivity	Specificity	PPV	NPV	Youden's Index
>0	89.7	2.1	21.5	40	-8.2
>1	65.5	18.8	19.6	64	-15.7
>2	24.1	70.8	20	75.6	-5.1
>3	51.7	60.4	28.3	80.6	12.1
>4	10.3	97.9	60	78	8.2

Likewise, the ROC curve (area under curve 0.751[95% CI: 0.593-0.908]) suggested that STEPSS score was very useful in predicting mortality in children with SE (Fig 2). STEPSS score of >3 had sensitivity of 85.3, specificity of 60.2 and negative predictive value of 98.6 in predicting mortality outcome. (table 4). Also, STEPSS had excellent predictive accuracy in determining refractory and super-refractory SE with area under ROC curve of 0.666(95% CI: 0.492-0.841) and 0.674(95% CI: 0.458-0.889) respectively (Fig 3and 4). Then predictive values of STEPSS score >3 in determining which child with SE will have refractory and super-refractory status is summarized in table 4.

Table 4. STEPSS score (>3) Predicting the accuracy of Mortality, Refractory and Super refractory status epilepticus

Outcome	Sensitivity	Specificity	PPV	NPV	Youden's Index
Mortality	85.3	60.2	11.3	98.6	45.5
Refractory	70	60	13.2	95.8	30
Super Refractory	75	58.7	5.7	98.6	33.7

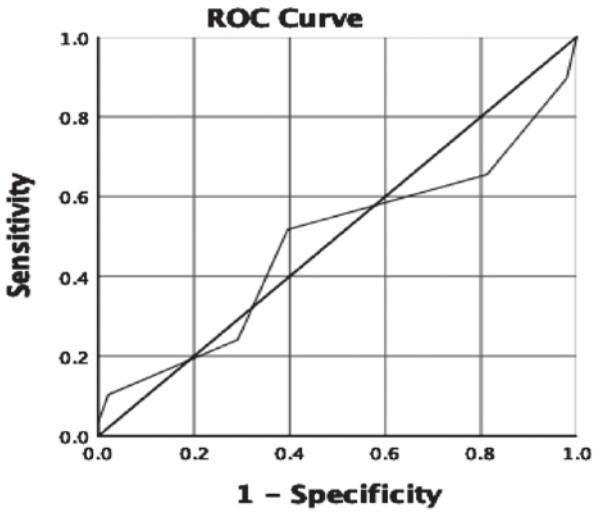


Figure 1. Receiver Operating Characteristic (ROC) curve of STEPSS score for unfavorable outcome [the area under curve was 0.48 (95% CI: 0.35-0.61)]

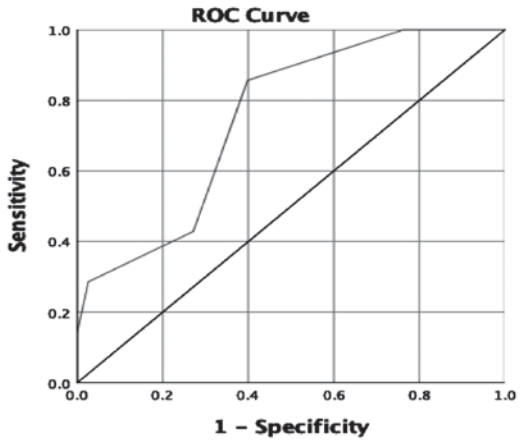


Figure 2. Receiver Operating Characteristic (ROC) curve of STEPSS score for mortality outcome [the area under curve was 0.751 (95% CI: 0.593-0.908)]

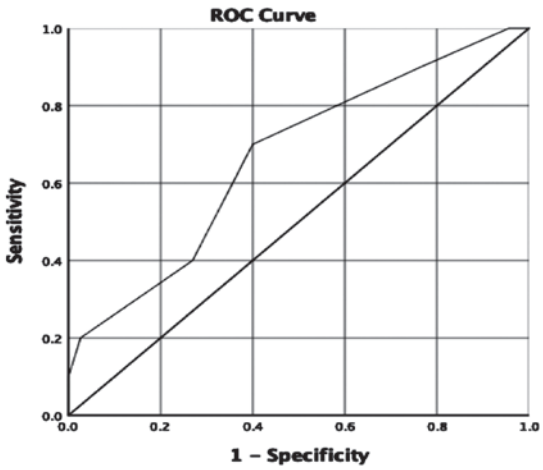


Figure 3. Receiver Operating Characteristic (ROC) curve of STEPSS score for refractory status epilepticus [the area under curve was 0.666 (95% CI: 0.492-0.841)]

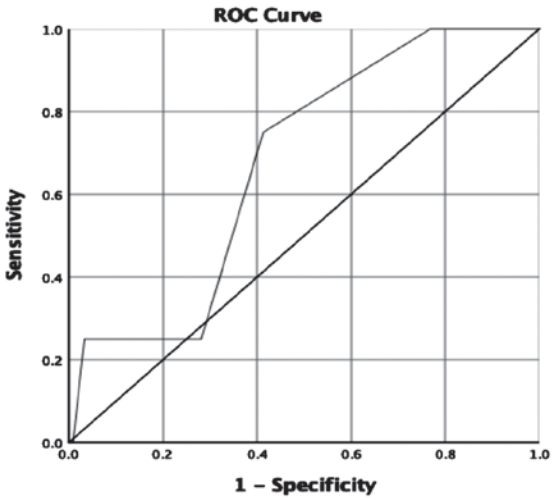


Figure 4. Receiver Operating Characteristic (ROC) curve of STEPSS score for super-refractory status epilepticus [the area under curve was 0.674 (95% CI: 0.458-0.889)]

DISCUSSION

In this study, we evaluated pediatric epilepticus severity score to predict short term outcome in pediatric SE. Our study concluded that STEPSS score was fairly accurate in predicting mortality, unfavorable outcome at the time of discharge and occurrence of refractory and super-refractory SE.

The demographic and clinic-etiological profile of the study population was similar to other studies in pediatric status epilepticus especially in developing countries.[13,14] Symptomatic etiology was the most common cause of pediatric SE as reported by other studies too. Mortality rate in our study was around 5.6% which is similar to study by Sidharth et al.[13] The findings are contrary to those observed in studies in adults where mortality rate was found to be as high as 16-25%.[11] The reason of high mortality in adults can be explained with the fact that adults usually present more severe form of symptomatic cases such as stroke which has a very mortality and morbidity unlike children where relatively benign causes such as febrile SE is more common. Another reason might be the duration of seizure taken as threshold to define SE. Our study used only 5 minutes duration to label as SE while most adult studies define 30 min operational threshold to label SE.

The STESS was developed in 2006 as a simple clinical bedside tool to predict the outcome and treatment response of status epilepticus in adults.[15] Most of the studies done in adults have depicted STESS ≥3 have excellent sensitivities, specificities and predictive values to determine functional outcome, mortality and treatment response.[16-23]. In 2018, Wang et al concluded that the diagnostic value of STESS for patient survival had sensitivity of 81% and specificity of 53% with area under curve (AUC) of 0.81.[24] There have been very few similar studies of epilepticus severity score in pediatrics. A study by Sidharth et al is one of first few studies studying the predictive accuracy of pediatric modification of STESS score to determine outcome in terms of mortality, functional outcome at discharge and treatment response.[13] Our study used the same pediatric modification as STEPSS score to determine the short-term outcome in SE. in our study, STEPSS score >3 showed 85.3% sensitivity, specificity of 60% and negative predictive value of 98.6%. This was similar to the study by Sidharth et al. Also, our study showed STEPSS score of >3 to have excellent sensitivity of >70% and NPV of >96% in determining poor response to treatment. In all reported studies, the STESS has been reported to have excellent negative predictive value.[15,16-23].

There are other scoring systems too that can be used in pediatric SE to determine prognosis and outcome. One such scoring system is Epidemiology-based mortality score (EMSE)[17] and the other is Encephalitis Nonconvulsive Status Epilepticus Diazepam Resistance Imaging Tracheal Intubation (END-IT)[25]. In contrary to STEPSS, both these scoring systems use parameters such as etiology which is not always known at the time of presentation and requires further investigation. This is one of the major advantages of STESS and its pediatric modification STEPSS as both of them used only those clinical parameters that can be readily recorded at the time of presentation and therefore timely guide treatment and prognostication.

One of major strengths of our study is its prospective design and adequate sample size and unfavorable cases. Also, we followed a standard treatment algorithm for management of all SE cases and adequate follow up was ensured to determine short term outcome especially the functional outcome and treatment responses. The main limitation of our study is that it was limited to hospital study only. So, the study has to be evaluated in other pediatric population in different settings so that the findings of the study have better validity. Another drawback was unable to study the long-term outcome as SE is likely to have chronic morbidity. Lastly non-availability of continuous EEG monitoring at emergency department at presentation had limited our ability to diagnose more cases of non-convulsive status epilepticus. This would likely affect the STEPSS scoring.

CONCLUSION

We used a modified pediatric version of epilepticus severity score (STEPSS) in our study and evaluated in a prospective cohort of children with SE. The STEPSS was found to be useful in predicting short term outcome like death and functional outcome as well as treatment response. Further studies in larger group of children are needed to confirm the predictive accuracy of STEPSS in children with status epilepticus.

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CONFLICT OF INTEREST

None

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None

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