

Spectrum of Nonepileptic Paroxysmal Events in Children from Southern India

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Abstract

Background Nonepileptic paroxysmal events (NEPEs) present with episodes similar to epileptic seizures but without abnormal electrical discharge on electroencephalogram (EEG). NEPEs are commonly misdiagnosed as epilepsy. Epilepsy is diagnosed on the basis of a detailed history and examination. Emphasis during history to rule out the possibility of NEPE is important. The wrong diagnosis of epilepsy can lead to physical, psychological, and financial harm to the child and the family. Hence, this study was planned.

Objective The objective of the study is to evaluate clinical profile, frequency, and spectrum of NEPE in children.

Materials and Methods This is a prospective observational study. Patients with NEPE between January 2014 and August 2016 aged < 18 years were enrolled. NEPEs were diagnosed on the basis of history, home video, and EEG recordings. Patients were divided into different categories according to age, specific type of disorder, and system responsible. Patients were followed for their NEPE frequency and outcome.

Results A total of 3,660 children presented with paroxysmal events; of them 8% were diagnosed with NEPE. Patients diagnosed with NEPE were classified into three age groups on the basis of their age of onset of symptom; of the total 285 patients, there were 2 neonates (0.7%), 160 infants (56%), and 123 children and adolescents (43.1%). Fifty-eight percent patients were boys. The most common diagnoses were breath-holding spells 113 (39%), followed by syncope 38 (13.3%) and psychogenic nonepileptic seizures 37 (12.9%). About 9 and 5% of patients had concomitant epilepsy and developmental delay, respectively.

Conclusions NEPEs account for 8% of paroxysmal events. Most common NEPEs were breath-holding spells among infants and syncope and “psychogenic nonepileptic seizures” in children and adolescents.

Keywords

- ▶ breath-holding spells
- ▶ nonepileptic paroxysmal events
- ▶ syncope

Introduction

Nonepileptic paroxysmal events (NEPEs) present with episodes similar to epileptic seizures but without abnormal electrical discharge on electroencephalogram (EEG). Twenty percent of patients seen at epilepsy referral centers are found to have NEPE.¹ The situation is again complicated by the co-occurrence of NEPE, mainly psychogenic nonepileptic seizures (PNES) in 30% of patients with genuine epileptic seizure.²

The incidence of epilepsy in children aged < 18 years ranges from 50 to 100/100,000. The incidence is highest in the first year of life^{3,4}; although frequencies and types of NEPE have been studied in adult populations, the data in children and adolescents are limited.⁵ However, the prevalence of epilepsy outnumbers that of NEPE.

Distinction between epileptic and nonepileptic events helps to prevent physical, psychological, and financial harm to the child and the family. Detailed history from a good

eyewitness and detailed clinical examination help in differentiating NEPE from epilepsy. The EEG is helpful in classifying and differentiating epileptic from nonepileptic events; however, it lacks sensitivity and specificity.^{6,7}

NEPE can be classified into four broad categories—syncope and other generalized paroxysms, movement disorders and other abnormal movements, sleep disorders, and oculomotor abnormalities. NEPE can also be classified based on the system involved—cardiac, vascular, neurological, respiratory, etc. In children, physiologic and organic disorders constitute a major chunk of NEPEs in addition to PNES.¹ In our study, we have evaluated clinical profile, frequency, and spectrum of NEPE in pediatric patients encountered during a 30-month period.

Materials and Methods

This was a prospective observational study from January 2014 to August 2016. Children aged < 18 years diagnosed with NEPE were included in the study. All the children with paroxysmal events were analyzed; NEPEs were diagnosed clinically by history; and video recording of the event was reviewed when available. EEG was sought when in doubt and was proved to be negative before confirming the diagnosis of NEPE. Demographic and clinical details of children with NEPE were noted, and all individuals were analyzed on the basis of phenomenological and clinical features of the paroxysmal motor event and a home video. Ethical clearance was obtained from the institutional ethical committee.

Results

In the study period of 30 months, 3,660 children presented to our department with paroxysmal episodes, and of them, 285 (8%) children were confirmed as NEPE. Of the 285 participants, 120 (42%) were girls and 165 (58%) were boys. Home video recording helped in recognition of type of NEPE in 35% of children. Only two children presented to us in neonatal life: one with hyperekplexia and the other with benign neonatal sleep myoclonus. The most common age group presenting with NEPE was infants (160 infants) of whom 71 were females and 89 males. Twelve children (7 males and 5 females) were aged between 1 and 5 years and rest 111 were aged between 5 and 18 years (67 males and 44 females) at presentation.

The events are further subclassified into four broad categories—syncope and other generalized paroxysms, movement disorders and other abnormal movements, sleep disorders, and oculomotor abnormalities. The frequency and clinical features of further subclassified events are elaborated in ►Table 1. The events were classified according to the system involved and are described in detail in ►Table 2.

Of 285 children with paroxysmal events, 111 children presented with movement disorders and other abnormal movements. The mean age of presentation was 77.31 months (range: 1–185 months) with 66 boys and 45 girls. There were two neonates. Of the 39 infants who presented with movement disorders and other abnormal movements, gratification disorder (12/39, mean age at presentation was 8.9 months),

and tremors (12/39) were the common diagnoses. There were seven children aged < 5 years, and five of them had presented with stereotypic movements. There were 63 children who presented with movement disorders and other abnormal movements between 5 and 12 years of age and 36 of them presented with PNES (mean age: 123 months).

A total of 166 children presented with syncope and other generalized paroxysms with mean age of 42 months and range of 3 to 190 months. Breath-holding spell (113 children) was the most common diagnosis among syncope and other generalized paroxysms. Of 113 children with breath-holding spell, 111 children presented in infancy (mean age of 9 months) and the rest 2 presented within 24 months. Syncope was the next common under syncope and other generalized paroxysms with 38 children (mean age of presentation 124.5 months) and all except one presented between 5 and 18 years of age.

A total of seven children (mean age: 15.5 months) presented with sleep disorder and six of them with sleep myoclonus (3 boys and 3 girls with mean age of 8 months) and one child with sleep transition disorders. In oculomotor abnormalities, one female infant presented with spasms nutans.

Discussion

The “NEPEs” are common in childhood and present with a wide clinical spectrum.⁸ In the present study, 285 (8%) children were NEPE among 3,660 children presenting with paroxysmal events. In a study by Kotagal et al, NEPE accounted for 23% of children admitted for video-EEG monitoring.¹ In Kim et al’s study, 143 (12.9%) patients had NEPE among 1,108 children reviewed with long-term video-EEG.⁹ Kutluay et al noted that 94 (23%) of 416 pediatric patients were found to have NEPE.¹⁰ The above findings are comparable to our study. However, Bye et al encountered NEPE in 285 (43%) of 666 patients,¹¹ and Chen et al noted NEPE in 31 (38.3%) of 81 infants and toddlers.¹² This high percentage of NEPE in the last two studies may reflect a referral bias.

Twenty-five patients (9%) had concomitant epilepsy and 13 patients (5%) had developmental delay in our study. Kotagal et al noted concomitant diagnoses of epilepsy in 19 patients (28%), developmental delay in 21 patients (34%), and both in 17 patients (25%).¹ Kutluay et al’s study puts the association of NEPE with epilepsy at 25%.¹⁰ High association of epileptic and NEPE in the above-mentioned studies is because the study was performed in patients who were referred for suspected epilepsy, whereas our study assessed NEPE among patients presenting with paroxysmal events.

Only two children presented to us in neonatal life: one with hyperekplexia and the other with benign neonatal sleep myoclonus. Very less number of patients (2 out of 285) being presented with NEPE in neonatal age group can be because the study population comprised of children attending pediatric neurology department. Furthermore, in a study by Chen et al who monitored 81 children aged ≤2 years in their pediatric epilepsy monitoring unit, 31 (38.3%) had at least one NEPE event during the study period and only 2 patients were neonates.¹²

Table 1 Frequency and clinical characteristics of nonepileptic paroxysmal events

Morphology of event	Female (120/285; 42%)			Male (165/285; 58%)			
	5–18 years	1–5 years	Infants	5–18 years	1–5 years	Infants	Neonates
Movement disorders and other abnormal movements (111/285; 38.9%)							
Benign myoclonus of early infancy (4)	0	0	2	0	0	2	0
Benign neonatal sleep myoclonus (1)	0	0	0	0	0	0	1
Cataplexy (1)	0	0	1	0	0	0	0
Gratification disorder (12)	0	0	10	0	0	2	0
Hyperekplexia (4)	0	0	0	0	0	3	1
Paroxysmal dyskinesias (1)	0	0	0	1	0	0	0
Psychogenic seizures (37)	11	0	0	25	1	0	0
Psychological disorders including factitious disorder imposed on another, malingering (17)	9	0	0	8	0	0	0
Shuddering attacks (7)	0	0	0	0	0	7	0
Stereotypies (6)	0	1	0	1	4	0	0
Tics (8)	2	0	0	6	0	0	0
Tremor (13)	0	1	8	0	0	4	0
Oculomotor abnormalities (1/285; 0.3%)							
Spasmus nutans (1)	0	0	1	0	0	0	0
Sleep disorders (7/285; 2.4)							
Sleep myoclonus (6)	0	0	3	0	0	3	0
Sleep transition disorders (1)	0	1	0	0	0	0	0
Syncope and other generalized paroxysms (166/285; 58.24%)							
Benign paroxysmal vertigo (2)	1	0	0	0	1	0	0
Breath-holding spells (113)	0	2	45	0	0	66	0
Cyclical vomiting syndrome (5)	2	0	0	3	0	0	0
Hyperventilation spells (3)	3	0	0	0	0	0	0
Reflex anoxic seizures (5)	1	0	1	1	0	2	0
Syncope (38)	15	0	0	22	1	0	0
Total (285)	44	5	71	67	7	89	2

Majority of our patients were infants, constituting 56% (160 children) of our total patients with NEPE. Majority of these infants were diagnosed with breath-holding spell (111 children), gratification disorder (12 children), tremors (12 children), shuddering attack (7 children), and benign sleep myoclonus (6 children). In the study by Kim et al, common NEPEs observed in infants were tonic posturing (28.6%), breath-holding spell (14.3%), shuddering (14.3%), nonepileptic myoclonus (8.6%), sleep myoclonus (8.6%), and gastroesophageal reflux (5.7%).⁹

In children aged < 5 years, the common diagnosis was stereotypic movements. Kim et al noted staring (72.7%), tonic posturing (94.1%), sleep myoclonus (72.7%), and sleep disorder (69.2%) in children younger than 6 years.⁹ Among the 111 children (39%) presenting between 5 and 18 years of age, 37 children (33.3%) presented with PNES and 37 children

(33.3%) with syncope attack. Among the children older than 6 years of age, PNES was noted in 13/15 patients (86.7%) by Kim et al.⁹ PNES constituted almost 72% among patients aged 12 years and above in a study by Kutluay et al.¹⁰ The limitation of the study was that video-EEG recording was not undertaken because of the lack of available facility.

Conclusions

NEPEs are common in children, especially in infants. Neonates, infants, and young children present with various types of NEPE than older children. The home video recording is of immense value in the diagnosis of NEPE. Most common NEPEs were breath-holding spells among infants and syncope and “psychogenic nonepileptic seizures” in children and adolescents.

Table 2 Nonepileptic paroxysmal events showing based on the system involved

Morphology of event	Female (42%)			Male (58%)			
	5–18 years	1–5 years	Infants	5–18 years	1–5 years	Infants	Neonates
Cardiac							
Reflex anoxic seizures	1	0	1	1	0	2	0
Channelopathies							
Cyclical vomiting	2	0	0	3	0	0	0
Neurological							
Cataplexy	0	0	1	0	0	0	0
Hyperekplexia	0	0	0	0	0	3	1
Paroxysmal dyskinesias	0	0	0	1	0	0	0
Tics	2	0	0	6	0	0	0
Tremor	0	1	8	0	0	4	0
Psychological							
Gratification disorder	0	0	10	0	0	2	0
Psychogenic pseudo seizures	11	0	0	25	1	0	0
Stereotypies	0	1	0	1	4	0	0
Respiratory systems							
Breath-holding spells	0	2	45	0	0	66	0
Sleep related							
Sleep myoclonus	0	0	3	0	0	3	0
Sleep-wake transition disorder	0	1	0	0	0	0	0
Unclassifiable or more than one system							
Benign myoclonus of early infancy	0	0	2	0	0	2	0
Benign neonatal sleep myoclonus	0	0	0	0	0	0	1
Benign paroxysmal vertigo	1	0	0	0	1	0	0
Hyperventilation spells	3	0	0	0	0	0	0
Shuddering attacks	0	0	0	0	0	7	0
Somatoform disorder	9	0	0	8	0	0	0
Spasmus nutans	0	0	1	0	0	0	0
Syncope	15	0	0	22	1	0	0
Total (285)	44	5	71	67	7	89	2

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None.

Conflict of Interest

None declared.

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