Children with psychogenic non-epileptic seizures (PNES): A detailed semiologic analysis and modified new classification

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Abstract

\textbf{Purpose}: To analyze children with psychogenic non epileptic seizures and propose a modified new classification. \textbf{Methods}: This retrospective analysis included 56 children aged <18 years (M:F = 26:30; mean age: 12.3 ± 4.0 years) diagnosed PNES on video-EEG monitoring. The semiological characteristics like pattern of bodily movements, emotional signs, stereotypy, ictal vocalization, responsiveness, delay in diagnosis etc. were recorded. We analyzed our data as per previous adult classifications and proposed a modified classification. \textbf{Results}: There were 190 recorded attacks (range: 1–9, median: 3) recorded. The age at onset of PNES was 8.9 ± 4.1 years (range: 0.4–15.8 years; median: 9 years), age at diagnosis: 11.9 ± 4.1 years (range: 2–17; median: 12.0 years), delay in diagnosis: 3.2 ± 3.7 years (range: 0–15; median: 2.0 years). Anxiety disorder was seen in 9 (16.1%), stress in 6 (10.7%) children. Flexion/extension bodily movements were seen in 40 (70.1%), negative emotional signs in 17 (30.4%) and tremors in 14 (25%) cases. Thirty-three (58.9%) patients diagnosed as having true seizures initially and were on anti-epileptic drugs (AEDs), 14 patients (25.0%) initially diagnosed of PNES which remained unchanged after VEEG, nine patients (16.1%) had both PNES and true seizures. Twenty-six (46.4%) of our patients into the existing classifications. We then classified our patients into categories of a modified new classification: Hypermotor: 13 (23.2%), partial motor: 8 (14.3%), affective/emotional behaviour phenomena: 2 (3.6%), dialeptic: 8 (14.3%), ‘aura’: 3 (5.4%), mixed: 22 (39.3%). \textbf{Conclusion}: Incorrect diagnosis of epilepsy leads to unnecessary drug treatment. A detailed analysis of semiology and classification helps in early diagnosis of PNES. A modified systematic classification of PNES is proposed which would help in better standardization of PNES.

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Keywords: Childhood epilepsy; Classification; Psychogenic non-epileptic seizures; PNES

1. Introduction

Psychogenic non-epileptic seizures may be defined as paroxysmal involuntary events of altered motor, sensory or behavioural phenomena which resemble epileptic seizures but are not associated with the epileptiform discharges on electroencephalography (EEG) \cite{1,2}. The prevalence of PNES varies from 5\% to 33\% in the outpatient epilepsy clinics and 10–58\% in patients admitted for the evaluation of the refractory seizures \cite{3}. The...
prevalence of PNES in children is 3.5–20% of children undergoing video EEG (VEEG) monitoring [4,5]. Several studies have shown that the childhood PNES are fairly common but the literature about the PNES in children is limited [4,6]. The diagnosis of PNES is often challenging in children especially if the child also has concurrent epilepsy [7]. Most of them experience their first PNES between 10 and 19 years of age but the diagnosis of PNES is established at a later date [8,9]. The diagnostic delay in the patients leads to unnecessary anti-epileptic drugs (AEDs) intake [10]. After the availability of VEEG, the diagnosis and differentiation of PNES from true seizures has become easier. The prognosis of PNES seems to be better in children and adolescents than in adults [11] provided an early diagnosis and intervention has been made. Studies have been done in an attempt to classify the PNES according their semiology in adults [12,13] but there are very few studies available in the paediatric age group [6,14]. Hence, a systematic and uniform classification of childhood PNES is required for an early diagnosis and wider usage for easy comparison.

In the present study, we analysed a large cohort of children with PNES diagnosis based on VEEG observations. The aim is to study in detail the different semiology patterns of psychogenic seizures in the paediatric age group and to classify them in a new modified classification.

2. Patients and methods

This observational study was carried out in the Departments of Neurology, Child and Adolescent Psychiatry and Psychiatry in a tertiary university hospital in South India from August 2005 to August 2012. Out of 1281 VEEGs recorded during this time, 139 (10.9%) patients were diagnosed with PNES. Fifty-six out of 139 (40.3%) patients were children (<18 years of age), who were included in this study. All the patients were admitted in the epilepsy monitoring unit (EMU) either to characterize the undiagnosed event (epileptic vs. PNES) or to diagnose the type of epilepsy/epileptic syndrome or for pre-surgical evaluation. Written informed consents were taken from their parents/legal guardians before the VEEG recording.

The Video EEG recordings were done for an average of 2 days (range: 1–3 days). It was carried out in Galileo EB Neuro system (Italy) as per the standard protocol and criteria. Standard 10–20 system of electrode placement was used for scalp ictal EEG recordings during the attack. Patient’s response during and after the attacks, response to induction/suggestion techniques (if any) were also recorded. Hyperventilation and photic stimulation were used wherever required. The recorded clinical attacks were confirmed as typical and habitual by the accompanying relatives and/or parents.

The video and EEG data of the 56 patients were retrieved from the server maintaining the records and were analyzed by two epileptologists independently who have experience in diagnosing PNES (SS and PSC). The differences in the analysis of PNES attacks were sorted out after discussion. The criteria used to diagnose the PNES [13] were (a) at least one typical attack should have been recorded on VEEG (b) no EEG changes should be noticed during the event (c) no post ‘ictal’ slowing on EEG (d) no evidence of any neurological condition responsible for the events. Co-existent epilepsy was not an exclusion criterion.

The parameters noted in the VEEG record were presence/absence of dizziness or ‘aura’, duration of the event, preictal pseudosleep, pattern of limb/bodily movements, head movements, jerking, periods of rest during the attack, emotional state, breathing patterns, suggestion techniques, response to verbal commands during the attack, vocalization, post ‘ictal’ state etc.

The mean, median, standard deviations, frequency were calculated for various variables using the R statistical software.

3. Results

3.1. Patients

A total of 56 children aged <18 years (M:F = 26:30; mean age: 12.3 ± 4.0 years, range: 2–17 years) diagnosed to have PNES on VEEG were included in the study. There were 190 attacks (range: 1–9, median: 3) recorded which were archived for the detailed analysis. The mean age at onset of PNES was 8.9 ± 4.1 years (range: 0.4–15.8; median: 9 years). The mean age at diagnosis was 11.9 ± 4.1 years (range: 2–17; median: 12.0 years). The mean duration of the illness before the diagnosis of PNES was 3.2 ± 3.7 years (range: 0–15; median: 2.0 years).

The reasons and/or accompaniments of the PNES in 42 children were anxiety disorder – 9 (16.1%); family history of epilepsy – 9 (16.1%); stress related to studies and parental control – 6 (10.7%); non-specific somatization symptoms like abdominal pain, headache – 6 (10.7%) each; depression – 6 (10.7%); psychiatric co-morbidity among close family members – 2 (3.6%). No obvious causes were identified in the rest.

There was coexistent epilepsy observed in 9 (16.1%) patients. The seizure types were: complex partial – 5 (8.9%), generalized tonic-clonic – 3 (5.4%) and simple partial – 1 (1.8%) seizures. Routine EEG showed interictal epileptiform discharges in all of them (n = 9/9; 100%). On analysis, it was found that prior to VEEG recording, 33 (58.9%) patients were initially misdiagnosed as epilepsy and they were on AEDs; in 14 patients (25.0%), the initial diagnosis of PNES was made which remained unchanged after VEEG, and nine patients
(16.1%) had both PNES and epilepsy. A total of 44 patients (78.6%) were on AEDs (mean duration: 2.9 ± 2.0; range: 0–17 years) at the time of VEEG recording. The common AEDs in use were: carbamazepine (n = 21; 47.7%), clobazam (n = 16; 36.4%), sodium valproate (n = 9; 20.5%), phenytoin and oxcarbazepine (n = 8 each, 18.2%) (Table 1). MR imaging of brain was carried out in 32 patients (57.1%) and was abnormal in 11 (34.4%) of them. In a subgroup of patients with only PNES (n = 14; 25%), all had undergone MRI which was normal in 12 (85.7%) patients; unidentified bright objects and non-specific signal changes in white matter were seen in 1 (7.1%) patient each. Among patients with both PNES and epilepsy (n = 9; 16.1%), MRI was done in all and it was found to be abnormal in 6/9 (66.7%), (MTS = 3, non-MTS = 3), and normal in 3/9 (33.3%).

3.2. Description of events (Table 2)

Two (3.6%) children experienced some ‘uneasiness’ or ‘aura’ before the event. The attack began abruptly in 43 (76.8%) children and 9 (16.1%) patients were observed to be apparently in sleep with no evidence of sleep record on EEG suggestive of preictal pseudosleep phenomena.

Out of phase asynchronous body movements were observed in 11 (19.6%), pelvic thrusting in 5 (8.9%) (forward: 3 (5.4%), backward: 2 (3.6%)) and generalized violent thrashing/grabbing movements were noted in 6 (10.7%) patients. Flexion/extension movements were seen in upper limbs in 15 (26.8%) and in lower limbs in 16 (28.6%) children. Flexion/extension movements of the head was seen in 9 (16.1) and side to side body turning in 12 (21.4%) patients. Tremors were observed in 14 (25.0%), while no motor activity (whole body ‘flaccidity’) during the attack was observed in 12 (21.4%) patients.

Two (3.6%) children were noted to have vocalization during the start of the attack while 6 (10.7%) patients had vocalization starting in the middle of the event which persisted during the whole attack. Emotional signs were seen in 17/56 (30.4%) patients. Moaning was most commonly seen in 6 (10.7%) while gasping/hiccups was seen in 4 (7.1%) without any signs and symptoms of respiratory distress, screaming and grunting were less commonly observed in 2 (3.6%) cases while 5 (8.9%) children had a cry/weep during the attack. Urinary incontinence, tongue bite and coughing was seen in 1 (1.8%) child each. Hyperventilation was seen in 7 (12.5%) children.

The eyes were closed in 25 (44.6%) children and 10/25 (40.0%) of them opened their eyes readily on verbal commands. In 31 (55.4%) patients, eyes remained opened during the whole attack. Brief intervals of pause/rest during the on-going attack were observed in 19 (33.9%) children. Stereotypy in attacks was seen in 41 (73.2%) patients.

In 17 (30.4%) children, suggestion techniques were used to induce the attack while in rest, 39 (69.6%), spontaneous attacks occurred. Thirty-one (55.4%) children followed the simple motor commands given by the EEG assistant during the event like naming, raising hand, opening and closing mouth, which along with semiological characteristics helped in diagnosing the PNES.

3.3. Classification

Based upon the classification proposed by Seneyviratne et al. [13] which is based on adult PNES, we classified our cohort. Three (5.4%) patients were classified...
into rhythmic motor, 5 (8.9%) as complex motor, 2 (3.6%) in hypermotor, 8 (14.3%) in dialeptic and 3 (5.4%) in ‘non-epileptic aura’ category. Mixed semiologies of different patterns of PNES were observed in 9 (16.1%) children. We could not classify the rest of our cohort (n = 26, 46.4%) according to this classification (Table 3).

Based on our analysis of childhood PNES and considering the limitations of existing classification of the PNES, we classified our cohort of childhood PNES into five major categories (Table 4) (I) Abnormal motor comprising of (IA) hypermotor (n = 13, (23.2%)), (IB) partial motor (n = 8, (14.3%)) (II), affective/emotional behaviour phenomena (n = 2, (3.6%)) (III), dialeptic (n = 8, (14.3%)) (IV) ‘aura’ (n = 3, (5.4%)) and (V) mixed pattern (n = 22, (39.3%)). The above classification of PNES is similar to the classification proposed by an earlier study[15].

We further described the semiologies in each category. Hypermotor type included asynchronous out of limb movements, thrashing/grabbing/violent/kicking movements, whole body rigidity and flaccidity; partial motor type included flexion/extension or side to side movements of the head/neck/limbs; affective/emotional behaviour phenomena included grimacing, weeping, grunting, moaning, screaming; dialeptic type included coma like state, fall or no response to external stimuli and ‘aura’ included subjective feeling without any external manifestations, dizziness before the attack and pressing the alarm button himself. The mixed type comprised of combination of the two or more of above subtype in a patient.

### Table 3
Comparison with the classification proposed by Seneviratne et al. [13].

<table>
<thead>
<tr>
<th>Categories proposed by Seneviratne et al. [13]</th>
<th>Their cohort (%)</th>
<th>Present cohort N = 56 (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rhythmic motor</td>
<td>46.7</td>
<td>3 (5.4)</td>
</tr>
<tr>
<td>Hyper motor</td>
<td>3.3</td>
<td>2 (3.6)</td>
</tr>
<tr>
<td>Complex motor</td>
<td>10.0</td>
<td>5 (8.9)</td>
</tr>
<tr>
<td>Dialeptic</td>
<td>11.2</td>
<td>8 (14.3)</td>
</tr>
<tr>
<td>‘Aura’</td>
<td>23.6</td>
<td>3 (5.4)</td>
</tr>
<tr>
<td>Mixed</td>
<td>5.2</td>
<td>Unclassified = 26 (46.4)</td>
</tr>
</tbody>
</table>

### Table 4
Proposed new classification of childhood PNES.

<table>
<thead>
<tr>
<th>Type</th>
<th>Semiological characteristics</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I Abnormal motor</td>
<td>Movement of the whole body (head, neck, limbs and trunk), pelvic thrusting, out of phase limb movements</td>
<td>13 (23.2)</td>
</tr>
<tr>
<td>A Hyper motor</td>
<td>Thrashing/grabbing/violent/kicking/punching movements. Whole body rigidity, whole body jerky movements, opisthotonic movements</td>
<td>8 (14.3)</td>
</tr>
<tr>
<td>B Partial</td>
<td>Head and neck: side to side, flexion/extension movements; limbs: flexion/extension, abduction/adduction movements, jerking, facio-pharyngio-respiratory: coughing, gagging, hyperventilation</td>
<td>2 (3.6)</td>
</tr>
<tr>
<td>II Affective/emotional behavior phenomena</td>
<td>Weeping, grimacing, screaming, moaning, grunting</td>
<td>8 (14.3)</td>
</tr>
<tr>
<td>III Dialeptic</td>
<td>Coma like state, no response to external stimuli, fall, flaccidity</td>
<td>3 (5.4)</td>
</tr>
<tr>
<td>IV ‘Aura’</td>
<td>Subjective feeling during the attack without any external manifestations, dizziness, ‘pressing alarm himself phenomena’</td>
<td>22 (39.3)</td>
</tr>
<tr>
<td>V Mixed</td>
<td>Hyper motor + affective/emotional behavior phenomena</td>
<td>10 (17.9)</td>
</tr>
<tr>
<td>A</td>
<td>Hyper motor + affective/emotional behavior phenomena</td>
<td>3 (5.4)</td>
</tr>
<tr>
<td>B</td>
<td>Hyper motor + dialeptic</td>
<td>1 (1.8)</td>
</tr>
<tr>
<td>C</td>
<td>Hyper motor + non epileptic aura</td>
<td>5 (8.9)</td>
</tr>
<tr>
<td>D</td>
<td>Partial motor + affective/emotional behavior phenomena</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>E</td>
<td>Partial motor + dialeptic</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>F</td>
<td>Partial motor + non epileptic aura</td>
<td>3 (5.4)</td>
</tr>
<tr>
<td>G</td>
<td>Affective/emotional behavior phenomena + dialeptic</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>H</td>
<td>Affective/emotional behavior phenomena + non epileptic aura</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>I</td>
<td>Dialeptic + non epileptic aura</td>
<td>0 (0.0)</td>
</tr>
</tbody>
</table>

3.4. EEG observations (Fig. 1)

As per the definition of PNES, none of the EEG record in the present cohort was associated with epileptiform EEG discharges except in the children having concurrent epilepsy who had inter-ictal EEG changes. We observed different EEG patterns in different types of PNES which were mainly due to the movement and muscle artifacts. These EEG changes started and ended abruptly with the event. After analysing EEG record using different filter settings and montages, it was seen that in the hypermotor PNES (IA), the record was obliterated by very high amplitude movement and muscle artifacts, while in partial type (IB) the EEG was also studded with muscle and movement artifacts. During rhythmic or semi-rhythmic movement of body parts, similar artifacts were observed. In affective/emotional behaviour phenomena, there were either no EEG artifacts or very subtle movements.
changes, or subtle muscle artifacts mainly involving facial, jaw or pharyngeal muscles. During dialeptic and ‘aura’, the EEG remained largely unchanged.

4. Discussion

The aim of this study was to study the semiological pattern of psychogenic non-epileptic seizures (PNES) in children and to propose a modified classification for the systematic categorization of childhood PNES. We found that like adult psychogenic seizures, the semiology of PNES in children is highly varied but show similar patterns. We could classify all our patients into the proposed modified classification.

4.1. Patients

The mean age at onset of PNES in children in our study was 8.9 ± 4.1 years which is lower than the reported (>10 years) in literature [6,16] while the mean age at diagnosis was 11.9 ± 4.1 years which is comparable to previous studies [17,18]. The mean duration of diagnostic delay in our study was 3.2 ± 3.7 years which is lesser than reported in the adults [8]. Early referral to a specialized centre may be responsible for this difference. There was a slight female predominance (53.6%) noted in our studies which is less than 67–74% reported in previous studies on adults [12,13,19] but support some authors describing a decrease in this tendency in the paediatric age groups [6,20,21]. Studies have shown that psychiatric co-morbidities play as common stressors in childhood PNES [22]. We found that anxiety disorder was a common stressor which is comparable to previous studies [22,23] but unlike some studies [24,25], severe psychopathologies like major depression were not observed in this study. Concurrent epilepsy in childhood PNES have been reported in 15–74% cases [6,7,20,21], our study showed similar results within this range (16%) but on a lower side. In the present study, about 60% of the children were unnecessarily taking AEDs which is well within the range of 35–75% shown in the previous studies [20,26].

4.2. Events

Pre-ictal pseudosleep was seen in 16% of our cohort which is much less than the reported (55.5%) [27]. Tremors were observed in 25% of cases which is similar to previous studies [12] and supports the evidence that tremors represent common motor phenomena in both paediatric and adult PNES [12,14]. We observed pelvic thrusting in about 8.9% of patients, similar to previous study [28] and suggest that this motor phenomena is not commonly seen in children. Forward and backward
pelvic thrusting were seen in three and two patients, respectively in this study which is contrary to previous study on adult PNES [29] suggesting that forward pelvic thrusting is more indicative of PNES.

Thrashing and grabbing movements are noted variably from 3.3% to 18% of adults with PNES [29]. This was observed in 10% of patients in our cohort. There was involvement of both upper and lower limbs in almost equal number of the patients, we observed side to side body turning and whole body flaccidity in equally number of patients (21%) which have not been reported in the previous studies [13,14]. Negative emotional signs like weeping, moaning, screaming have been reported in childhood PNES [30,31], which we also observed in 30% of cases, making these an important marker for paediatric PNES [32]. Stereotypical attacks were observed in about 73% of our patients which is similar to previous studies [6] and contradict the common belief that the stereotypical attacks are organic seizures.

4.3. Rationale for modification in the classification

We used the classification proposed by Seneviratne et al. [13] as it is extensive and has been studied previously to classify the paediatric PNES attacks by Szabo et al. [14]. We believe that the comparison of our cohort with the previous PNES classifications [12,19] is not required as all of them are based on adult PNES studies. As mentioned before, we could not classify about half of our patients into the categories proposed by Seneviratne et al. [13]. On classifying the patients of our study into the categories, we observed that a large number of the patients fell into the mixed category, because they had attacks of varying combinations of the other five categories mentioned above. Previous studies have also shown a comparatively large number of patients classifying into mixed category which may be due to insufficient recognition of the main semiologic patterns. The hypermotor category in the classification proposed by Seneviratne et al. [13] includes those patients who presented with violent, kicking, thrashing, punching type of movements which are bilateral, asynchronous and asymmetric. There is a considerable overlap between the hypermotor category and the complex motor category of Seneviratne et al. [13]. In the present classification, the hypermotor category includes those patients who had movements involving the whole body which is different from the partial motor category which includes patients having movements involving a part of the body. There is no clear mention about the state of consciousness of the patients during the attack among different categories. Several studies [33,34] found that a large number of the patients present with emotional signs like moaning, weeping, screaming, grunting which have not been included in the previous classifications [14]. The number of patients who had mixed semiologies was more in the present as well as in the classification proposed by Seneviratne et al. [13]. So, an attempt was made to subdivide the mixed category for better standardization in this study. The authors have also reported a similar classification of semiological characteristics in the adult PNES and hence this modified classification system in children will be uniform [35]. We were able to classify all our 56 patients into the five categories of the modified classification.

The major limitation of the present study was its retrospective design. Inaccurate diagnosis of epilepsy leads to exposure to side effects of AEDs, high drug costs and various psycho-social stigmas associated with epilepsy which may have additional impact on development of the children and failure to provide specific psychosocial interventions needed for PNES. Hence, early recognition of PNES and a focused classification is very important for the proper management. We invite a general consensus on the classification of childhood PNES attacks. This proposed classification is based on a large cohort but further multicentric studies are required for improved sub-categorization.

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References


