Seizure 59 (2018) 67–71

Contents lists available at ScienceDirect

Seizure

journal homepage: www.elsevier.com/locate/yseiz

Psychogenic nonepileptic seizures in adults with epilepsy and intellectual disability: A neglected area

Jans S. van Ool[a,*], Alexandra I. Haenen[a], Francesca M. Snoeijen-Schouwenaars[a], Albert P. Aldenkamp[b,c], Jos G.M. Hendriksen[c,d], H. Jurgen Schelhaas[e], In Y. Tan[a], Richard H.C. Lazeron[e], Nynke M.G. Bodde[b]

[a] Department of Residential Care, Epilepsy Centre Kempenhaeghe, P.O. Box 61, 5590 AB Heeze, The Netherlands
[b] Department of Behavioural Sciences, Epilepsy Centre Kempenhaeghe, P.O. Box 61, 5590 AB Heeze, The Netherlands
[c] Department of Neurology, Maastricht University Medical Centre, P.O. Box 5800, 6202 AZ Maastricht, The Netherlands
[d] Centre of Neurological Learning Disabilities Kempenhaeghe, P.O. Box 61, 5590 AB Heeze, The Netherlands
[e] Department of Neurology, Epilepsy Centre Kempenhaeghe, P.O. Box 61, 5590 AB Heeze, The Netherlands

ARTICLE INFO

Article history:
Received 2 February 2018
Received in revised form 30 April 2018
Accepted 4 May 2018
Available online xxx

Keywords:
PNES
Functional seizures
Developmental disability
Behaviour
Differential diagnosis

ABSTRACT

Purpose: To describe the main characteristics of psychogenic nonepileptic seizures (PNES) in adults with epilepsy and intellectual disability (ID), and to analyse the differences regarding psychosocial functioning, epilepsy severity and ID between patients with PNES and a control group without PNES.
Methods: Medical records of adults with ID and epilepsy living at an epilepsy care facility (N = 240) were screened for PNES and evaluated by a neurologist. A control group consisting of patients with epilepsy and ID, without PNES, was matched according to age, sex and level of ID. Characteristics of PNES and epilepsy were provided by the subject's nursing staff or retrieved from patient charts, psychosocial data were collected by standardised questionnaires and level of ID was individually assessed using psychometric instruments.
Results: The point prevalence of PNES was 7.1%. The patients with PNES (n = 15) were most often female and had a mild or moderate level of ID. Compared to controls, they showed more depressive symptoms, experienced more negative life events and had more often an ID discrepancy (ID profile with one domain particularly more impaired than another). Stress-related triggers were recognised in a large majority by the nursing staff.
Conclusion: PNES appears to be a relatively rare diagnostic entity among inpatients with both epilepsy and ID. However, the complexity of diagnosing PNES in this population, and the similarities in stress-related triggers for PNES in patients with and without ID, suggest that PNES may be underdiagnosed in the ID population. Diagnostic challenges of PNES and, as subcategory, reinforced behavioural patterns are discussed.

1. Introduction

Psychogenic non-epileptic seizures (PNES) are defined as sudden and involuntary paroxysmal events that resemble epileptic seizures, but are not induced by an organic cause. In addition, there is positive evidence or it is strongly suspected that the events are related to a psychogenic cause. These events can involve changes in behaviour, motor activity, sensation, cognitive processing, or autonomic function [1,2]. The term PNES can be misleading, as one not only needs to exclude epilepsy, but also other organic causes that can lead to a similar semiology.

The diagnosis of PNES consists of a two-phase process, of which the patient needs to be informed as soon as possible. First, organic causes, including especially epilepsy, have to be excluded as a cause of the seizures. Epilepsy may coincide with PNES, however, and it is necessary to determine whether or not the paroxysmal event can be attributed to epilepsy. The gold standard for excluding epilepsy is video-EEG monitoring of a characteristic seizure that does not show the electrographic discharges seen during an epileptic seizure [2,3]. In the second phase, psychological aetiologies that cause the paroxysmal events must be assessed.

* Corresponding author.
E-mail addresses: oolj@kempenhaeghe.nl (J.S. van Ool), haenena@kempenhaeghe.nl (A.I. Haenen), schouwenaarsf@kempenhaeghe.nl (F.M. Snoeijen-Schouwenaars), aldenkampb@kempenhaeghe.nl (A.P. Aldenkamp), hendriksenj@kempenhaeghe.nl (J.G.M. Hendriksen), schelhaasj@kempenhaeghe.nl (H. J. Schelhaas), tanf@kempenhaeghe.nl (I.Y. Tan), lazeronr@kempenhaeghe.nl (R.H.C. Lazeron), bodden@kempenhaeghe.nl (N.M.G. Bodde).

https://doi.org/10.1016/j.seizure.2018.05.002
1059-1311/

The highest level of certainty, "documented PNES", is reached when a non-epileptic seizure with semiology typical to PNES is captured on video-EEG, along with a patient history of psychosocial characteristics consistent with PNES [2]. As this certainty level cannot always be reached, for example because of limited access to video EEG, the recognition of PNES with a lower level of certainty (i.e., "possible", "probable", or "clinically established") also becomes relevant.

PNES is considered to be a multifactorial biopsychosocial disorder [4]; many psychosocial and biological factors have been described that contribute to its development or prolongation (e.g., [1,5]. Studies have shown that the majority of patients with PNES are female (75%) and report previous trauma (up to 70%); also, a history of co-morbid psychiatric or psychosocial problems is common [2].

PNES are also recognised among patients with ID (e.g., [6,7,4]. A below average intelligence quotient (i.e. IQ <85) might be a risk factor for PNES [8], although it remains unclear whether this study also included patients with ID (IQ <70). There is limited evidence regarding the presentation and incidence of PNES in this subpopulation, as patients with ID are often excluded from studies. Duncan and Oto [9] compared patients with PNES with and without ID and concluded that a diagnosis of epilepsy, the use of anti-epileptic drugs, episodes of psychogenic nonepileptic status, and situational or emotional triggers were more prevalent among those with ID. Sexual abuse seemed to be more frequent among those without ID. Another theory suggested that PNES in people with ID manifests less profoundly as an emotional conflict, but more as a reinforced behavioural pattern, which can be considered a subcategory of PNES. By exhibiting this reinforced behavioural pattern a secondary gain is reached, such as receiving attention or avoiding demands or unpleasant situations [10]. By producing seizure-like events that are paradoxically reinforced by caregivers, these patients may have unconsciously and unintentionally learned how to control the environment. This idea was elaborated upon in a study by Magudda et al. [11], who described characteristics of a patient group with mild ID. Remarkably, this subgroup developed PNES after a decrease in epileptic seizure frequency. All of these patients had early-onset epilepsy, for which caregivers probably provided much attention. The authors hypothesise that the decrease in epileptic seizure frequency or cessation of epilepsy might have led to a loss of this advantage, after which the epileptic seizures had been substituted by PNE Baslet et al. [6] identified a subgroup of PNES patients who presented with neurological impairments and ID, but showed less severe psychiatric impairment. Psychopathology, including depression, anxiety and somatic distress, was often present, however.

The aims of the present study are twofold: (1) to describe (clinical) characteristics of PNES in adults with ID and epilepsy, and (2) to compare epilepsy severity and psychological and behavioural characteristics between those with PNES and a matched control group without PNES, all with epilepsy and ID.

2. Methods

2.1. Participants

Electronic charts of adult patients living at the residential care facility of Kempenhaeghe, a tertiary epilepsy centre in The Netherlands, were screened for evidence of non-epileptic events between January 2014 and December 2016. Only those who met the following criteria were included: impaired intellectual functioning (IQ < 70), age $\geq$18 years, and diagnosis of PNES following evaluation by a neurologist and, when necessary, other medical specialists. Those with PNES must have had more than one seizure-like event in the past two years, which had to include a hypothesised behavioural or psychosocial component. Seizures with an organic cause were excluded. In this article we consider the reinforced behavioural pattern as a subcategory of PNES. Our screening of 240 eligible patients yielded 17 patients with PNES (7.1%). As two patients did not provide consent for the study, a total of 15 subjects with PNES were included in the final analyses.

A control group consisting of 15 patients with epilepsy and ID, without PNES, was matched according to age, sex, and level of ID. For each PNES subject, all matching patients were identified and one of the possible matched was randomly automated selected.

2.2. Instruments and procedure

This cross-sectional, observational study is part of the TRIANGLE study (The Relation between epilepsy, ID, And Neuropsychiatric comorbidities in a Group of patients in Long-term care for Epilepsy). TRIANGLE is approved by the local ethical committee of Kempenhaeghe (No. 15.01). All subjects or legal representatives (if appropriate) provided consent for the study.

All information regarding PNES was collected through a questionnaire completed by the subject's nursing staff (see Appendix A). This questionnaire was created by a research team including a health care psychologist, psychotherapist and neurologist. Both objective (e.g., frequency, time and location, and injuries as a result of PNES) as well as subjective characteristics (e.g., suspected triggers and impact on daily life) were addressed.

The level of ID was diagnosed according to DSM-5 in terms of mild, moderate, severe or profound [12]. Each ID domain, i.e., conceptual, social and practical, was assessed separately using an abbreviated version of the Wechsler Adult Intelligence Scale – fourth edition [26] and the Vineland-II subscales Socialization and Daily Living Skills [14]. A significant difference between domains was considered to be an ID domain discrepancy (for more information regarding this method, see [13]).

The severity of epilepsy was determined using the Epilepsy Impact Scale Kempenhaeghe (EPIEK; [15], which is based on five aspects: seizure frequency, number of anti-epileptic drugs, use of emergency anti-epileptic drugs, use of protective measures for epilepsy, and adjustments in the subject's daily schedule after a seizure. The relevant information was retrieved from the subject's medical records. The EPIEK yields an epilepsy severity score ranging from 0 to 10, a higher score indicating a more severe form of epilepsy.

For the assessment of depressive symptoms, anxiety symptoms, aggressive/destructive behaviour, and life events, three standardised questionnaires were administered among the subject's nursing staff. Depressive and anxiety symptoms were assessed using the Anxiety, Depression, And Mood Scale (ADAMS) [16,17] and aggressive/destructive behaviour was assessed using the Behavior Problems Inventory (BPI) [18,19], higher scores reflecting more severe symptoms or behaviour. Both the ADAMS and BPI have been validated among people with ID [17,18]. The number of life events in the past year was calculated using the Checklist Life Events (CLE) [20,21].

2.3. Analyses

First, clinical characteristics of PNES are described. The correlation between frequency of PNES and epileptic seizures in the past year was examined using Spearman's rank correlation analysis. As neither variable met the criteria for a normal distribution, a log-transformation was performed prior to the analysis. Second, differences between subjects with PNES and the control group are analysed with statistical analyses appropriate for case-control studies, i.e., paired T-test or Wilcoxon signed rank test for continuous variables and McNemar's test for dichotomous