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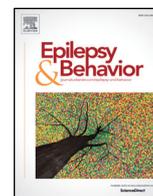
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Challenging behavior in adults with epilepsy and intellectual disability: An analysis of epilepsy characteristics

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ABSTRACT

Purpose: The study aimed to describe the frequency and severity of self-injurious, stereotyped, and aggressive/destructive behavior in adults with both epilepsy and intellectual disability (ID) who reside at a tertiary epilepsy center and to investigate the associations between challenging behavior and epilepsy and ID characteristics.

Method: The frequency and severity of self-injurious, (motoric) stereotyped, and aggressive/destructive behavior among 189 patients was assessed using the Behavior Problem Inventory. Comparisons were made with an adult reference population with ID, based on gender, to determine whether the behavior was clinically deviant. Epilepsy characteristics, including age at onset, epilepsy type, seizure types, seizure frequency, and use of antiepileptic drugs (AEDs), were retrieved from patient files. The level of ID was classified using the Diagnostic and Statistical Manual of Mental Disorders – Fifth Edition (DSM-5) and an ID domain discrepancy was allocated if there was a substantial difference between two domains of adaptive behavior within a subject.

Results: Self-injurious behavior was present in 35% of subjects, stereotyped behavior in 60%, and aggressive/destructive behavior in 63%. The behavior exceeded clinical norms in 7%, 18%, and 12%, respectively. Aggression was the behavior evaluated most often as being problematic, despite its reported frequency being the lowest. When adjusting for level of ID and use of psychotropic medication, logistic regression analyses showed that self-injurious behavior was significantly associated with a lower number of AEDs (odds ratio (OR) = 0.4); that stereotyped behavior was significantly associated with a higher number of seizure types (OR = 1.4) and a lower number of AEDs (OR = 0.4); and that aggression was significantly associated with the presence of an ID domain discrepancy (OR = 3.1).

Conclusion: Challenging behavior is a serious issue among adults with epilepsy and ID. Although some of the epilepsy and ID characteristics seemed to contribute independently to these types of challenging behavior, the effects of epilepsy-related characteristics are modest when compared with ID.

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1. Introduction

Challenging behavior is a serious concern among people with epilepsy and intellectual disability (ID) [1]. It is defined by Emerson [2] as “culturally abnormal behavior(s) of such an intensity, frequency or duration that the physical safety of the person or others is likely to be placed in serious jeopardy, or behavior likely to seriously limit use of, or result in the person being denied access to, ordinary community facilities”. Various types of challenging behavior are encountered in the daily care for this population, such as aggression, self-injury, noncompliance,

hyperactivity, and stereotyped mannerisms. These behaviors can result in a fear of harm or actual injury to the person or to others and might have adverse consequences for the individual's development and opportunities for community integration [3].

The prevalence of challenging behavior among people with ID was studied in multiple large population studies, which resulted in point prevalence rates between 10 and 22.5% [4–7]. The prevalence in those with both epilepsy and ID is less well-documented. Two systematic review studies on challenging behavior in this population concluded that people with epilepsy and ID did not clearly exhibit more challenging behavior when compared with those without epilepsy [8, 9] although the results were inconclusive. More specifically, having epilepsy was not associated with aggression, behavioral disturbances, social impairments,

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or challenging behavior in people with ID [10–13]. McGrother et al. [14], however, found higher rates of being uncooperative, seeking attention, and disturbing others at night in people with epilepsy and ID compared with those without epilepsy, after adjusting for gender, age, and level of intellectual understanding. Studies comparing ID populations with and without epilepsy might oversimplify the association between epilepsy and challenging behavior, considering epilepsy is a very heterogeneous disorder with variability in localization, syndromes, etiology, seizure type and frequency, and treatment strategies often including polypharmacy in people with ID.

The literature on the impact of specific epilepsy-related characteristics on challenging behavior in people with ID is scarce. Espie et al. [15] explored associations between epilepsy factors as well as nonepilepsy concerns and challenging behavior and psychiatric symptoms. They concluded that psychiatric symptoms were most strongly related to epilepsy characteristics, such as seizure frequency and severity, whereas behavioral outcomes were most strongly predicted by nonepilepsy concerns, including sensory, intellectual, and motor impairments, as well as adverse effects of drugs [15]. Other studies on behavioral changes associated with antiepileptic drugs (AEDs) show that effects vary among different AEDs, with positive as well as negative effects, although high-quality evidence in people with ID is lacking [1].

The aim of the study was to describe the frequency and severity of self-injurious, stereotyped, and aggressive/destructive behavior in adults with both epilepsy and ID who reside at a tertiary epilepsy center and to investigate the associations between challenging behavior and epilepsy and ID characteristics.

2. Method

2.1. Study design and participants

This study had a cross-sectional design and was part of the TRIANGLE study (The Relation between epilepsy, ID, And Neuropsychiatric comorbidities in a Group of patients in Long-term care for Epilepsy), which was conducted within the tertiary care facility of Kempenhaeghe, the Netherlands. The TRIANGLE study is approved by the medical-ethical committee of Kempenhaeghe (No. 15.01), and the medical-ethical committee of Erasmus University Medical Center concluded that the rules laid down in the Medical Research Involving Human Subjects Act do not apply to this study (MEC-2016-408). The inclusion criteria were as follows: 1) age \geq 18 years, 2) diagnosis of epilepsy according to the clinical definition by Fisher et al. [16], 3) diagnosis of ID or current adaptive functioning at the level of ID as evaluated by the individual's psychologist, and 4) currently living at the residential care facilities of Kempenhaeghe for at least 1 year. The consent was provided by individuals themselves if they were capacitated, by their legal guardian in case individuals did not have the capacity, or by both the individual and their legal guardian if the individual was capacitated but also had a legal guardian.

2.2. Instruments and procedure

Data on challenging behavior, epilepsy characteristics, and ID were collected using the multiple methods listed below.

2.2.1. Challenging behavior

To assess challenging behavior, the Dutch version of the Behavioral Problem Inventory-01 (BPI) [3] was completed by a professional caregiver who had been familiar with the subject for at least 1 year. The BPI consists of three subscales: self-injurious behavior (SIB; 15 items), stereotyped behavior (25 items), and aggressive/destructive behavior (11 items). Self-injurious behavior was defined as behavior that may cause damage to the person's body and that occurred repeatedly in an unvarying manner (e.g., head-hitting); stereotyped behavior included peculiar or inappropriate voluntary acts that occurred repetitively and

habitually (e.g., rocking); and aggressive/destructive behavior referred to deliberate, abusive attacks against others or objects (e.g., hitting others). For each item, the caregiver was asked to evaluate the frequency and severity of the particular behavior in a subject during the past two months. Frequency was rated on a five-point scale (never, monthly, weekly, daily, or hourly) and – if the item occurred at least monthly – the severity was rated on a three-point scale (slight problem, moderate problem, or severe problem). The sum of items yields a continuous (nonstandardized) frequency and severity score per subscale, with a higher score representing more frequent or severe challenging behavior. In addition, whether the scores were clinically deviant was examined by comparing the subject's score with clinical norms of an international population with ID (USA, UK, the Netherlands, and Romania) [17]. A score was considered clinically deviant if it exceeded the mean score plus 1.5 standard deviation (>93 rd percentile) of the adult group with the corresponding gender.

The BPI is found to have good psychometric properties [3, 18]. Reliability analyses in this study showed Cronbach's alpha values ranging from 0.684–0.858 (internal consistency) and split-half reliability values of 0.557–0.864. The BPI was also found to have good factor and criterion validity [3].

2.2.2. Epilepsy characteristics

Epilepsy characteristics, including age at onset, epilepsy type and etiology, seizure type, number of seizures (including nocturnal seizures) in the past year, and the use of AEDs, were retrieved from the subject's medical records. With respect to all aspects of epilepsy, the patients are regularly followed up by a neurologist specialized in epilepsy. Seizures were recorded by the direct support staff and relied therefore on direct or secondary observations. Nonepileptic events, such as psychogenic nonepileptic seizures, were excluded. Non-EEG seizure-detection systems were used to detect nocturnal seizures if applicable. The epilepsy type was classified according to the most recent classification system by the International League Against Epilepsy (ILAE) [19].

2.2.3. ID

Regarding the ID, there were two variables of interest: overall level of ID and ID domain discrepancy. The level of ID was based on the three domains of adaptive deficits as described in the Diagnostic and Statistical Manual of Mental Disorders – Fifth Edition (DSM-5) [20]: the conceptual, social, and practical domains. Each domain was assessed separately. The conceptual domain was assessed using a psychological test in combination with an expert opinion by the subject's psychologist. The psychological test applied was either a 4-subtest version of the Wechsler Adult Intelligence Scale – Fourth Edition (WAIS-IV) [21], in case of expected mild to (high-) moderate level of ID ($n = 79$), or the Peabody Picture Vocabulary Test – Third Edition (PPVT-III) [22], in case of (low-) moderate to severe level of ID ($n = 57$). The remaining 53 subjects were classified by expert opinion of the subject's psychologist. The WAIS-IV short form was validated among people with neurological disorders and impaired intellectual functioning [23]. The PPVT-III is a measure of receptive vocabulary and is considered a valid screening tool for global cognitive functioning [24, 25]. The social and practical domains were assessed using the corresponding Daily Living Skills and Socialization subscales of the Vineland-II Expanded Interview Form [26] (Dutch translation by Dijkxhoorn and Verhaar [27]), which were completed on all subjects.

The results for each domain were converted into a classification of mild, moderate, severe, or profound deficits. Internationally used cutoff points, described by the Diagnostic and Statistical Manual of Mental Disorders – Fourth Edition (DSM-IV), the International Statistical Classification of Diseases – tenth edition (ICD-10), and Vineland II, were applied, all using cutoff points of 70–50/55 for mild deficits, 50/55–35/40 for moderate deficits, 35/40–20/25 for severe deficits, and below 20/25 for profound deficits [26, 28, 29]. The lower-end values were applied. An ID profile was considered to be discrepant when there was a

substantial intra-individual difference between two DSM-5 domains, indicating that one domain is considerably more or less deficient than the other(s). An ID domain discrepancy was attributed if one of the following criteria was met: (1) a difference between the Vineland II subscales Daily Living Skills and Socialization with a significance level of .01 [25] or (2) a difference of at least one complete classification level between the conceptual domain and the social or practical domain (if the conceptual domain was determined by the PPVT-III or expert opinion).

2.3. Analyses

Descriptive statistics were calculated for each subscale of the BPI as well as the most reported items and the number of subjects with clinically deviant challenging behavior. Associations between subscales were examined using Kendall's tau-b rank correlation analyses, as the subscale variables violated the normality assumption and had a high number of tied ranks (e.g., both subscales with scores of 0). In order to examine the extent to which the frequency scores were predictive of the severity scores, separate regression analyses were performed. Each score was standardized to adjust for unequal item numbers so that slopes could be compared between subscales.

As the frequency and severity subscales were highly correlated, subsequent analyses were performed using only the frequency variables, as this is the more objective measure. Multiple binary logistic regression analyses were performed to examine direct associations between predictors and clinically deviant behavior, as well as multivariate logistic regression including all predictors and controlling for the use of psychotropic medication. Categorical variables were only included if they had sufficient statistical power. Hence, the level of ID was divided into mild/moderate and severe/profound, and the type of epilepsy included only focal epilepsy or a combined type with both generalized and focal epilepsy, as the number of subjects with generalized epilepsy only was small ($n = 20$). Results were considered significant if $p < .05$. All analyses were conducted in IBM SPSS Statistics version 24.

3. Results

3.1. Sample characteristics

A total of 240 patients were invited for the study, 189 of whom provided consent (inclusion rate: 78.8%). Participants were significantly younger than nonparticipants (mean difference = 6.04 years, $p = .015$) and were using psychotropic medication more often (41.8% versus 14.0%, respectively, $p < .001$). Individuals who participated did not differ from nonparticipants with respect to level of ID or gender.

The sample comprised 58.7% males, and mean age was 47.9 years ($SD = 15.6$; range: 18.3–85.9 years). The majority of subjects resided in residential facilities of Kempenhaeghe (76.2%); the others lived in community settings. The epilepsy originated in childhood in more than half of the subjects (54.4%). Most subjects experienced focal epilepsy or a combination of focal and generalized epilepsy (41.3% and 44.4%, respectively), with a seizure frequency of at least once a week (55.5%). Those who were seizure-free in the past year were currently using AEDs. Nearly all subjects were using AEDs (99.5%), and 41.3% were prescribed psychotropic medication on a daily basis. The level of ID was mild in 20.1%, moderate in 30.7%, severe in 29.1%, and profound in 20.1%, and an ID domain discrepancy was present in 32.8%. Clinical characteristics are depicted in Table 1.

3.2. Frequency and severity of challenging behavior

Self-injurious behavior was reported in 34.9% of subjects, stereotyped behavior in 59.8%, and aggressive/destructive behavior in 63%; 87.8% of subjects exhibited one or more of these challenging behaviors. The frequency of the behavior was clinically deviant compared with the

Table 1
Clinical characteristics of the study sample.

Characteristics	Values
Age at onset of epilepsy (years)	Mdn = 2.0, IQR = 0–5.5, range: 0–53
Infancy (<1 yr)	32.8%
Childhood (1–12 yr)	54.0%
Adolescence (12–18 yr)	10.1%
Adulthood (18+ yr)	3.2%
Epilepsy type ^a	
Generalized only	10.6%
Localized only	41.3%
Both generalized and localized	44.4%
Unknown	3.7%
Number of seizure types (semiology) ^a	Mean = 2.8, SD = 1.9, range: 0–8
Seizure frequency (last year)	Mdn = 70.0, IQR = 11.5–153.0, range: 0–1206
Seizure-free	12.7%
Yearly	12.2%
Monthly	19.6%
Weekly	43.9%
Daily	11.6%
Etiology of epilepsy ^a	
Structural	28.6%
Genetic	20.1%
Infectious	6.3%
Metabolic	1.1%
Unknown	43.9%
Daily use of antiepileptic drugs	Mean = 3.1, SD = 1.1, range: 0–6
Daily use of psychotropic drugs	41.8%
Psychiatric classification (DSM-IV)	20.6%

^a Based on ILAE 2017 criteria [18]. Mdn = median, IQR = interquartile range, SD = standard deviation.

norms for adults with ID, adjusted for gender, in 7.1%, 17.5%, and 11.6% of subjects, respectively. In total, 55 of 189 subjects (29.1%) had at least one type of clinically deviant behavior. For descriptive statistics and Kendall's tau-b correlation coefficients between BPI subscales, see Fig. 1 and Table 2. The self-injurious behaviors that were reported most often were self-scratching (13.8%), self-biting (9.5%), and head-hitting (9.5%). With respect to stereotyped behavior, grimacing (29.6%), yelling/screaming (25.4%), rocking/repetitive body movements (18.0%), and gazing at hands or objects (18.0%) were reported most frequently. Being verbally abusive with others (32.8%), hitting others (26.9%), and pushing others (26.9%) were the aggressive/destructive behaviors which occurred most often.

Item-level analyses showed that self-injurious and aggressive/destructive behaviors were most often displayed on a monthly basis (in 37.8% and 52.9% of cases, respectively) whereas stereotyped behaviors occurred most often daily (in 51.3% of cases). A different trend was identified for the severity of behaviors. Self-injurious and stereotyped behaviors were most often evaluated as being a slight problem (39.5% and 37.1% of cases, respectively) whereas aggressive/destructive behaviors were mostly perceived as a moderate problem (40.3% of cases). The behavior was considered a severe problem in 27.2% of cases for self-injurious behavior, in 25.1% of cases for stereotyped behavior, and in 27.8% of cases for aggressive/destructive behavior. As some subjects showed multiple types of behavior, the total number of subjects who exhibited at least one behavior that was considered a severe problem was 56 (29.6%).

Kendall's tau-b correlation analyses yielded very high positive correlation coefficients between the frequency and severity of each behavioral type ($\tau_b = 0.876$ – 0.972 , all p values $< .001$). Separate linear regression analyses with standardized scores confirmed these associations and showed that frequency scores explained 82.4% of variance in severity of self-injurious behavior, 80.0% of variance in severity of stereotyped behavior, and 62.7% of variance in severity of aggressive/destructive behavior. The regression lines are depicted in Fig. 2. The slope value was highest for aggressive/destructive behavior ($B = 1.08$, $SE = 0.08$), compared with self-injurious behavior ($B = 0.73$, $SE = 0.04$), and lowest for stereotyped behavior ($B = 0.50$, $SE = 0.02$), indicating that aggressive/destructive behavior is perceived as problematic more quickly than self-injurious or stereotyped behavior.

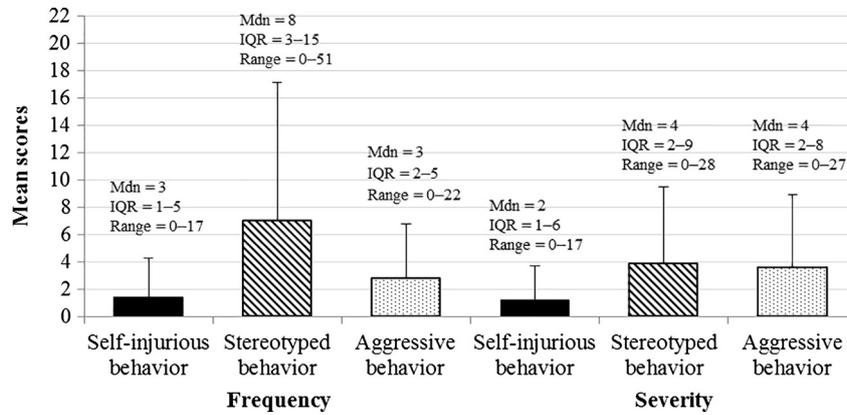


Fig. 1. Means and 1 standard deviation error bars per BPI subscale. Note: Medians (Mdn) and interquartile ranges (IQR) are derived from positively scored items only.

3.3. Associations between epilepsy, ID, and challenging behavior

Results of the bivariate and multivariate logistic regression predicting clinically deviant challenging behavior are presented in Table 3. Bivariate logistic regression analyses showed that a more severe level of ID and a lower number of AEDs increased the likelihood of having clinically deviant self-injurious behavior (OR = 4.16, *p* = .033 and OR = 0.54, *p* = .018, respectively). After adjusting for level of ID, use of psychotropic medication, and the other predictors in a multivariate analysis, the effect of a lower number of AEDs remained significant (OR = 0.39, *p* = .004), but none of the other epilepsy or ID characteristics was significantly associated with self-injurious behavior.

With respect to stereotyped behavior, bivariate analyses indicated direct significant associations between clinically deviant stereotyped behavior and a higher number of seizure types (OR = 1.56, *p* < .001), a higher seizure frequency (OR = 1.02, *p* = .015), a lower number of AEDs (OR = 0.70, *p* = .048), and a more severe level of ID (OR = 14.76, *p* < .001). Although not statistically significant, there was also a clinically meaningful association between stereotyped behavior and epilepsy type, with the likelihood of having stereotyped behavior being twice as high when having a combined epilepsy type (OR = 2.04, *p* = .069). Taking the level of ID, use of psychotropic medication, and the other predictors into account, the likelihood of stereotyped behavior was significantly increased by the number of seizure types (OR = 1.44, *p* = .030), but significantly decreased by the number of prescribed AEDs (OR = 0.44, *p* < .001). The effects of seizure frequency and a combined epilepsy type were no longer present in the multivariate analysis.

Clinically deviant aggressive/destructive behavior was significantly related to a higher number of seizure types (OR = 1.42, *p* = .006) and a more severe level of ID (OR = 5.52, *p* = .003) in bivariate analyses.

After adjusting for the use of psychotropic medication and the other predictors in a multivariate analysis, none of the epilepsy characteristics was associated with aggressive/destructive behavior. However, the presence of an ID domain discrepancy did significantly increase the likelihood of having deviant aggressive/destructive behavior (OR = 3.08, *p* = .043) as well as a more severe level of ID (OR = 9.13, *p* = .002).

Post hoc exploratory analyses were performed by means of a chi-square or Fisher’s exact test in order to assess the use of specific AEDs in relation to clinically deviant behavior. Because of polypharmacy and the concomitant use of, on average, three AEDs, we could not determine the exclusive influence of each individual AED. With respect to self-injurious behavior, valproic acid was used less often in the subgroup with than in the subgroup without clinically deviant behavior (21.4% versus 52.4%, *p* = .028). In addition, none of the subjects in the subgroup with clinically deviant self-injurious behavior used topiramate or lacosamide, compared with 16.0% and 8.6% in the subgroup without (not significant). There were no significant differences in AED use between subjects with clinically deviant aggressive/destructive or stereotyped behavior and those without.

4. Discussion

The present study demonstrates that challenging behavior is a serious cause for concern among people with epilepsy and ID who reside at a tertiary epilepsy center. A total of 29.6% of the sample exhibited self-injurious, stereotyped, or aggressive/destructive behavior that is perceived as a severe problem by daily caregivers. Aggressive/destructive behavior was perceived as problematic more quickly compared with self-injurious or stereotyped behavior. Moreover, 29.1% of the sample exhibited clinically deviant behavior based

Table 2
Kendall’s tau-b correlation coefficients between BPI subscales.

	Self-injurious behavior		Stereotyped behavior		Aggressive behavior	
	Frequency	Severity	Frequency	Severity	Frequency	Severity
Self-injurious behavior						
Frequency	–	.972**	.347**	.361**	.125*	.098
Severity		–	.346**	.365**	.128*	.101
Stereotyped behavior						
Frequency			–	.901**	.053	.025
Severity				–	.106	.078
Aggressive behavior						
Frequency					–	.876**
Severity						–

** *p* < .001.
* *p* < .05.

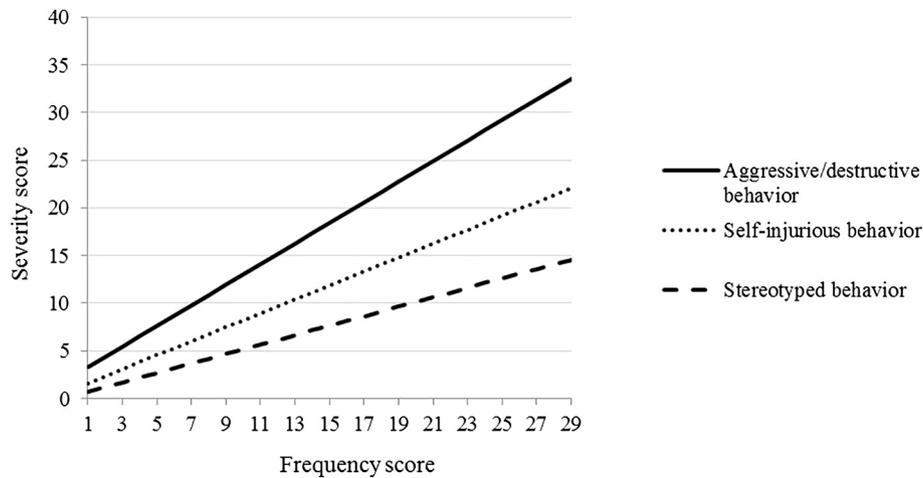


Fig. 2. Regression trend lines predicting standardized severity scores from standardized frequency scores per subscale.

on ID population norms of frequency scores, with (motoric) stereotyped behavior being encountered most frequently compared with ID population norms.

The results showed that some of the epilepsy and ID characteristics were related to challenging behavior. The likelihood of clinically deviant self-injurious and stereotyped behavior (but not aggressive/destructive behavior) tended to be smaller if a higher number of AEDs were prescribed on a daily basis. One might speculate that this might be due to the mood-stabilizing properties of certain AEDs [30]; for example, the use of valproic acid was significantly lower in subjects with self-

injurious behavior. Conversely, there are some AEDs that may have negative effects on behavior [1] although this was not demonstrated in our study, probably due to polypharmacy in nearly all subjects and/or small sample size. In clinical practice, it is important to continue to evaluate efficacy and potential adverse effects regularly. Further research is necessary to determine behavioral effects of (combinations of) AEDs in people with ID, especially because the diagnosis of ID is often considered an exclusion criterion in medication trials. Another explanation for the negative association between the number of AEDs and self-injurious and stereotyped behavior might relate to the treatment effect

Table 3
Logistic regression analyses predicting clinically deviant challenging behavior (frequency).

	Bivariate analyses			Multivariate analyses		
	B (SE)	OR	95% CI	B (SE)	OR	95% CI
<i>Self-injurious behavior</i>						
Age at onset	−0.09 (0.08)	0.91	0.79–1.06	−0.06 (0.08)	0.95	0.81–1.10
Generalized and focal epilepsy	0.80 (0.58)	2.23	0.72–6.94	0.40 (0.64)	1.45	0.43–5.23
Seizure frequency	0.01 (0.02)	1.01	0.98–1.04	−0.01 (0.02)	1.00	0.95–1.04
Number of seizure types	0.27 (0.15)	1.31 ^a	0.98–1.75	0.35 (0.22)	1.42	0.92–2.18
Number of AEDs	−0.62 (0.26)	0.54 [*]	0.32–0.90	−0.94 (0.32)	0.39 ^{**}	0.21–0.74
ID discrepancy	−0.62 (0.67)	0.54	0.14–2.00	−0.16 (0.79)	0.84	0.18–3.99
Level of ID (severe/profound)	1.43 (0.67)	4.16 [*]	1.12–15.42	1.02 (0.79)	2.78	0.59–13.02
				$\chi^2 = 17.83^*$, Nagelkerke $R^2 = 0.223$		
<i>Stereotyped behavior</i>						
Age at onset	−0.05 (0.04)	0.95	0.88–1.03	0.01 (0.05)	1.01	0.92–1.11
Generalized and focal epilepsy	0.72 (0.39)	2.04 ^a	0.95–4.41	0.20 (0.49)	1.22	0.47–3.15
Seizure frequency	0.02 (0.01)	1.02 [*]	1.01–1.04	0.01 (0.02)	1.01	0.98–1.04
Number of seizure types	0.45 (0.11)	1.56 ^{***}	1.25–1.95	0.37 (0.17)	1.44 [*]	1.04–2.00
Number of AEDs	−0.36 (0.18)	0.70 [*]	0.49–1.00	−0.82 (0.24)	0.44 ^{***}	0.27–0.71
ID discrepancy	−0.92 (0.48)	0.40 ^a	0.16–1.02	−0.29 (0.60)	0.75	0.23–2.44
Level of ID (severe/profound)	2.69 (0.63)	14.76 ^{***}	4.32–50.46	2.30 (0.71)	9.56 ^{**}	2.47–40.20
				$\chi^2 = 47.55^{***}$, Nagelkerke $R^2 = 0.376$		
<i>Aggressive behavior</i>						
Age at onset	−0.02 (0.04)	0.98	0.90–1.06	0.02 (0.05)	1.02	0.92–1.12
Generalized and focal epilepsy	−0.03 (0.46)	0.97	0.40–2.37	−0.52 (0.54)	0.60	0.21–1.71
Seizure frequency	0.02 (0.01)	1.02	0.99–1.04	−0.00 (0.02)	1.00	0.97–1.03
Number of seizure types	0.35 (0.13)	1.42 ^{**}	1.11–1.81	0.21 (0.18)	1.23	0.87–1.74
Number of AEDs	0.17 (0.21)	1.18	0.78–1.78	0.02 (0.26)	1.02	0.62–1.68
ID discrepancy	0.61 (0.46)	1.84	0.75–4.54	1.12 (0.56)	3.08 [*]	1.04–9.15
Level of ID (severe/profound)	1.71 (0.57)	5.52 ^{**}	1.79–17.01	2.21 (0.71)	9.13 ^{**}	2.29–36.4
				$\chi^2 = 22.07^{**}$, Nagelkerke $R^2 = 0.219$		

B = beta, SE = standard error, OR = odds ratio, CI = confidence interval, AEDs = antiepileptic drugs, ID = intellectual disability.

^a $p < .1$.
^{*} $p < .05$.
^{**} $p < .01$.
^{***} $p < .001$.

leading to seizure improvement and hence improvements in behavior [e.g., [31]]. More (longitudinal) research focusing on treatment effects of AEDs on behavior in people with ID is recommended.

Furthermore, clinically deviant stereotyped behavior was present significantly more often in those with mixed seizure types, suggesting a severe form of epilepsy that can be difficult to treat. Stereotyped behaviors are controlled behaviors which are thought to have a potential calming effect on an over-aroused nervous system [32, 33]. Hence, it may be that they are (subconsciously) exhibited as a coping strategy for the burden of severe epilepsy, particularly when a person already has limited coping resources because of a more severe level of ID. It can also be hypothesized that the patients with a severe form of epilepsy have an underlying complex or brain impairment that might separately lead to behavioral abnormalities.

Clinically deviant aggression was not significantly associated with an aspect of epilepsy but was linked to ID characteristics. A more severe level of ID and an ID domain discrepancy (i.e., one domain of adaptive functioning being considerably more or less developed than another) significantly increased the likelihood of aggressive/destructive behavior. One could hypothesize that there is a risk the person with an ID domain discrepancy might be overestimated by others or by the person himself, resulting in an aggressive response to (over)demanding situations or internal frustration. Future research is necessary to clarify this mechanism.

We were able to take into account multiple epilepsy characteristics that make up the phenotype of epilepsy and indicate its severity. The etiology of epilepsy (and/or ID) and specific epilepsy syndromes were, however, not included in our study. It might have been that, for some patients, behavioral abnormalities resulted from the same underlying cause that is involved in the epilepsy itself, for example, a severe brain impairment [34]. Information regarding etiology was missing for a large percentage of the subjects; in those with a known cause of epilepsy, the etiologies were heterogeneous and sometimes rare and therefore resulted in small subsample sizes with insufficient statistical power. The representativeness of our sample is limited to patients with often severe epilepsy, majority of whom live in a residential care setting. Results should therefore be validated in other samples. A review of studies on deinstitutionalization and post-deinstitutionalization of adults with ID concluded that studies show no association with emotional well-being, including challenging behavior [35].

To conclude, both epilepsy and ID characteristics may contribute independently to certain types of challenging behavior although the effects of epilepsy-related characteristics are modest when compared with ID. This study does not confirm the hypothesis that severe epilepsy strongly contributes to challenging behavior, something which has been repeatedly suggested in previous research and clinical practice. We did not find evidence for effects of seizure frequency, age at epilepsy onset, or epilepsy type on challenging behavior. The results of this study can help to anticipate challenging behavior and contribute to the search for targeted behavioral or other interventions for people with epilepsy and ID.

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Conflict of interest

None of the authors has any conflict of interest to disclose.

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