

Impact of Disruptive Seizures, Sleep, and Behaviors on Activities of Daily Living and Communication in Individuals Aged 16 Years and Older with Developmental and Epileptic Encephalopathies

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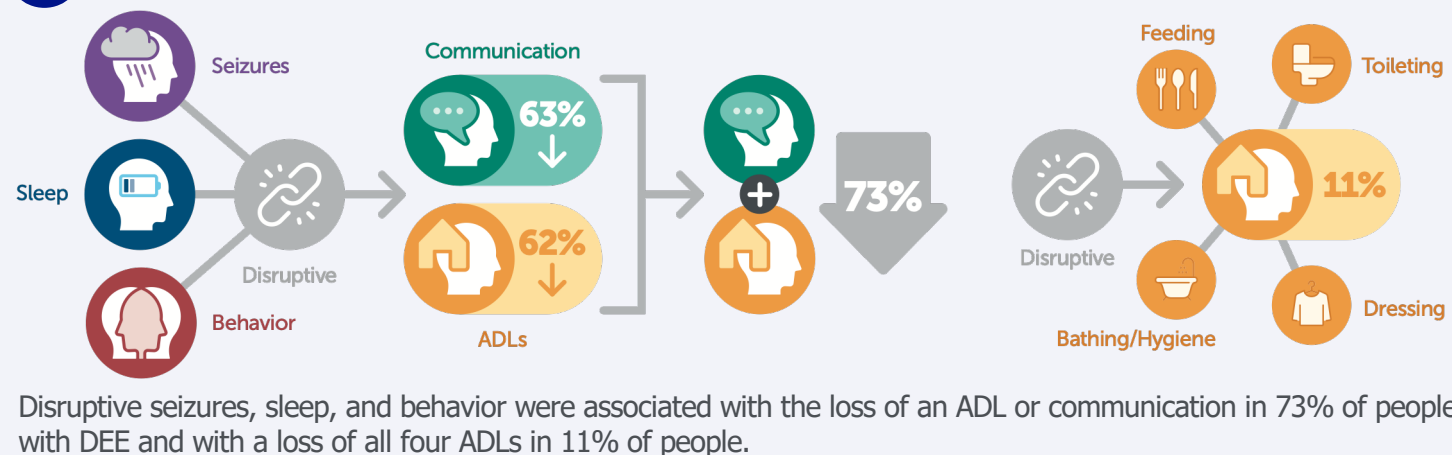
QUESTIONS

- How do caregivers of a person with developmental and epileptic encephalopathy (DEE) ≥16 years old define normal and disruptive seizures, sleep, and behavior?
- What is the relationship between disruptive seizures, sleep, and behavior and loss of ability to perform activities of daily living (ADLs) and communicate?

INVESTIGATION

- An internet-based anonymous survey was distributed via patient advocacy websites, social media, and patient community events
 - Respondents were primary caregivers to a person with DEE
 - Survey questions were designed to:
 - Define, for each individual, normal and disruptive seizures, sleep, and behavior
 - Determine the typical abilities of each individual to perform ADLs and communicate
 - Establish patterns of relationships between disruptive symptoms and typical abilities to perform ADLs and communicate
- Here, responses for people with DEE ≥16 years old were explored

RESULTS



CONCLUSIONS

- Disruptive seizures, sleep, and/or behavior associated with temporary loss of abilities to perform ADLs and to communicate were commonly reported for people with DEE ≥16 years old
- Disruptive patterns and maintenance of typical abilities may be important outcomes of treatment evaluations to caregivers of people with DEE ≥16 years old



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Background

- Developmental and epileptic encephalopathies (DEEs) are characterized by severe and frequent seizures and developmental delays¹⁻³
- People who live with DEE primarily depend on caregivers to perform activities of daily living (ADLs) and have difficulty with communication^{4,5}
 - ADLs include feeding, bathing and hygiene, dressing, and toileting⁶⁻⁸
- Available literature on adults with DEE and quality of life, including the ability to communicate and to independently perform ADLs, is lacking
 - Literature on the impact of disruptive seizures, sleep, and/or behavior on communication and ADLs in people with DEE overall is also minimal
 - These two distinct, yet interrelated, gaps in the existing literature limit the available evidence for guiding the clinical management of adults with DEE

Objective

- To characterize “normal” (typical daily experience during the current phase of the DEE journey) and “disruptive” (deviations from the typical experience) seizures, sleep, and behavior
 - The perceived effect of these symptoms on daily life in individuals ≥16 years old diagnosed with DEE was explored

Methods

- Respondents to this internet-based anonymous survey were primary caregivers to a person diagnosed with DEE
 - Current analyses were conducted on responses from caregivers to individuals with DEE ≥16 years old
- The survey focused on characterizing normal and disruptive seizures, sleep, and behavior
 - Differences between normal and disruptive patterns were explored, and caregiver-perceived effects on communication and ability to perform ADLs independently were evaluated descriptively
 - Disruptive behavior was defined in the survey as any of the following behaviors: hitting, biting, kicking, shouting, hair pulling, swearing, harsh language, throwing objects, refusal to cooperate, destruction of property, threatening with physical harm, invading a person's personal space, and anger

Results

Table 1. Demographics and Baseline Characteristics

	People With DEE ≥16 Years Old N=166
Age, y, median (range)	
Diagnosis	10.5 (0–64.0)
Time of Survey	22.0 (16.0–67.0)
Primary Diagnosis^a, n (%)	
LGS	35 (21.1)
DS	21 (12.7)
STXBP1	17 (10.2)
Other	93 (56.0)
Secondary Diagnosis^b, n (%)	
LGS	31/37 (83.8)
Other	6/37 (17.2)
Country of Residence^c, n (%)	
US	137 (82.5)
UK	6 (3.6)
Canada	6 (3.6)
Australia	5 (3.0)
Other ^d	12 (7.2)
Sibling Living in the Same Home, n (%)	78 (47.0)

^aAt least 10% of people with DEE ≥16 years old.

^bAt least two people with DEE ≥16 years old.

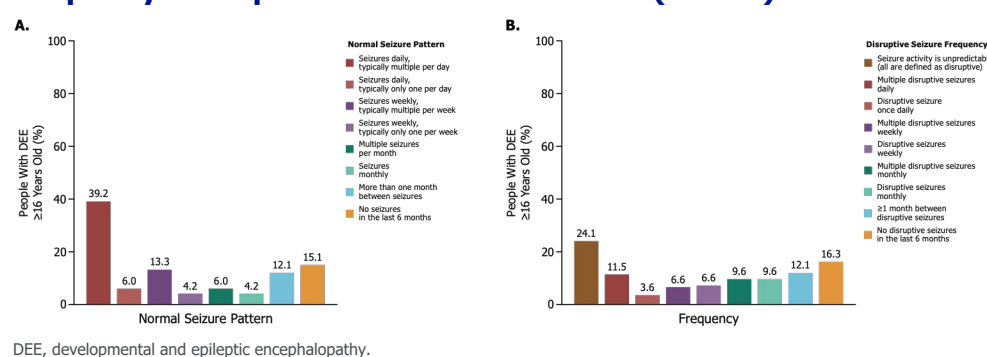
^cAt least 3% of people with DEE ≥16 years old.

^dOther countries of residence were Belgium (n=3, 1.8%), Netherlands, Italy, Spain (n=2 each, 1.2%), Portugal, Ukraine, and Sweden (n=1 each, 0.6%).

There were 140 people with DEE ≥18 years old at the time of the survey. DEE, developmental and epileptic encephalopathy; DS, Dravet syndrome; LGS, Lennox-Gastaut syndrome.

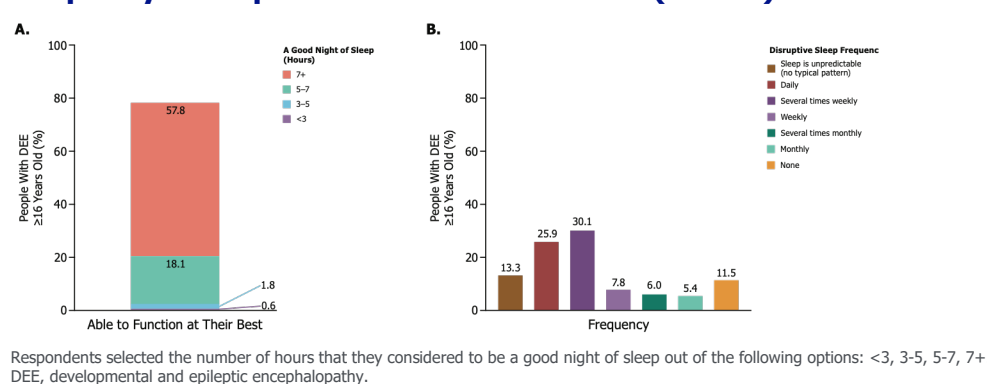
- Nearly half (n=75, 45.2%) of people with DEE ≥16 years old had daily seizures as a part of their normal pattern (**Figure 1A**)
- Forty (24.1%) people with DEE had unpredictable (ie, always disruptive) seizure activity (**Figure 1B**)
 - Daily disruptive seizures were reported for 25 (15.1%) people with DEE
- Seizure clusters were reported for 111 (66.9%) people with DEE ≥16 years old

Figure 1. A. Normal Seizure Patterns and B. Disruptive Seizure Frequency in People With DEE ≥16 Years Old (N=166)



- A good night of sleep for the person with DEE was defined as <7 hours by 59 (35.5%) caregivers
 - As the number of hours required for a good night of sleep increased, the number of people with DEE who were able to function at their best with a good night of sleep also increased (**Figure 2A**)
- Twenty-two (13.3%) people with DEE had unpredictable (always disruptive) sleep (**Figure 2B**)
 - Daily sleep disruptions were reported for 43 (25.9%) people with DEE

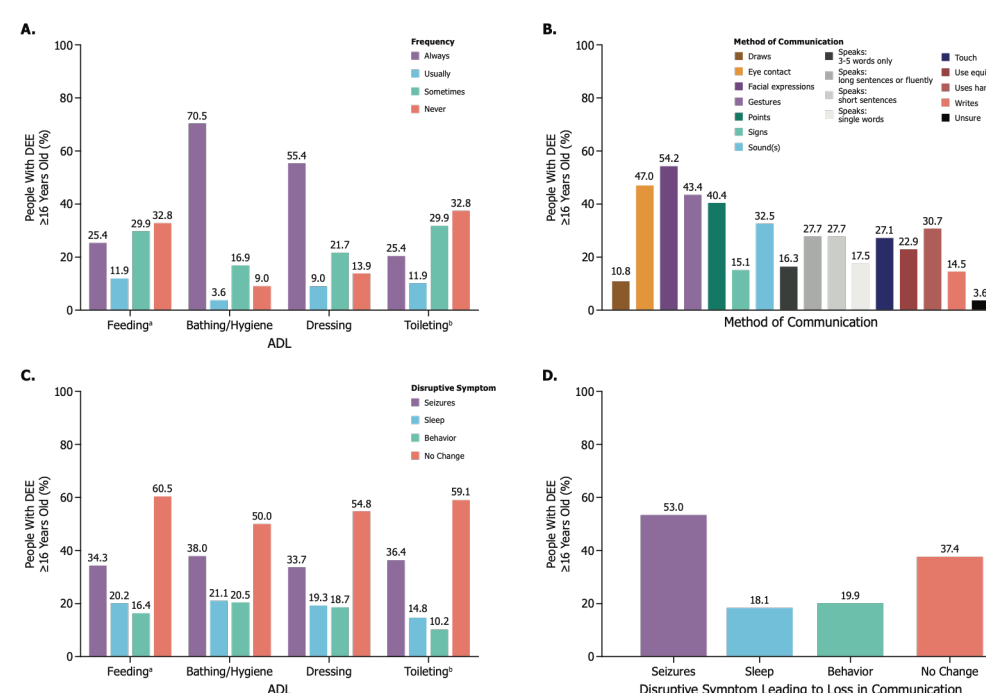
Figure 2. A. Normal Patterns of Sleep and B. Disruptive Sleep Frequency in People With DEE ≥16 Years Old (N=166)



Respondents selected the number of hours that they considered to be a good night of sleep out of the following options: <3, 3-5, 5-7, 7-9. DEE, developmental and epileptic encephalopathy.

- Over half (n=99, 59.6%) of people with DEE displayed at least monthly disruptive behavior
 - Daily, weekly, and multiple disruptive behavior were reported for 44 (26.5%), 33 (19.9%), and 22 (13.3%) people with DEE, respectively
 - Thirteen (7.8%) people with DEE had greater than one month between disruptive behavior episodes
 - Disruptive behavior was not reported for 54 (32.5%) of people with DEE
- Of 134 people with DEE who were not fed by gastrostomy tube (G tube), assistance was always required for feeding by 34 (25.4%) and never required by 44 (32.8%) people (**Figure 3A**)
 - Many (n=117, 70.5%) people with DEE always required assistance for bathing and hygiene, and over half (n=92, 55.4%) always required assistance for dressing
 - Of 88 people who did not use incontinence products at all times, 18 (20.5%) always required assistance with toileting
- The most commonly reported (≥30% of people with DEE ≥16 years old) methods of communication were facial expressions (n=90, 54.2%), eye contact (n=78, 47.0%), gestures (n=72, 43.4%), pointing (n=67, 40.4%), sounds (n=54, 32.5%), and using hands (n=51, 30.7%; **Figure 3B**)
- Disruptive seizures, sleep, and/or behavior were reportedly associated with temporary reduction in abilities to perform ≥1 ADL and/or communicate for 121 (72.9%) people with DEE ≥16 years old
- A temporary loss in ability to perform an ADL independently was reported for 103 (62.1%) people with DEE ≥16 years old
- Disruptive seizures were most commonly perceived as the reason for reduced independence across all ADLs (33.7%–38.0%; **Figure 3C**)
- Disruptive symptoms were also associated with temporary loss of communication methods for 104 (62.7%) people, most commonly due to disruptive seizures (n=88, 53.0%; **Figure 3D**)

Figure 3. A. Normal Patterns of Assistance Required for ADLs, B. Normal Methods of Communication Reported for People With DEE ≥16 Years Old, C. Loss of Ability to Independently Perform ADLs Due to Disruptive Symptoms, and D. Loss of Ability to Communicate Due to Disruptive Symptoms



^aDoes not include responses for those who reported use of a G tube (n=88 included).

^bDoes not include responses for those who reported that incontinence products were used all of the time (n=134 included).

Caregivers were able to choose more than one mode of communication.

ADL, activity of daily living; DEE, developmental and epileptic encephalopathy; G tube, gastrostomy tube.

CONCLUSIONS

- Disruptive seizures, sleep, and/or behavior were reported by most caregivers to an individual with DEE ≥16 years old
- Disruptive symptoms were often associated with a temporary loss of abilities
 - Approximately 73% of people with DEE ≥16 years old had reduced abilities to perform an ADL and/or communicate
 - Disruptive seizures were the most common symptom responsible for loss of any ADL and communication
 - Disruptive sleep and disruptive behavior reportedly had an effect on feeding and bathing/hygiene in approximately 20% of people with DEE
- Disruptive seizure, sleep, and behavior patterns may be important outcomes to caregivers when evaluating treatments for individuals with DEE
 - Additional analyses differentiating normal and disruptive patterns and their relationship with ADLs and communication by DEE diagnosis may provide additional insight into important outcomes within each DEE community

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Disclosures

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