



The “Cinderella Syndrome”: A narrative study of social curfews and lifestyle restrictions in juvenile myoclonic epilepsy

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ABSTRACT

Several factors are thought to contribute to inadequate seizure control in patients with juvenile myoclonic epilepsy (JME), including drug resistance, neuropsychiatric comorbidity, and poor lifestyle choices. Recent evidence supports the existence of frontal lobe microstructural deficits and behavioral changes that may contribute to poor seizure control in a minority of patients. Counseling patients on the importance of adequate sleep hygiene and alcohol restriction is an important part of the management strategy for patients with JME. However, information is lacking on how these lifestyle restrictions impact on patients with JME. We conducted a qualitative descriptive analysis of the social impact of JME on 12 patients, from their own perspective. We identified four prominent themes: the importance of alcohol use as a social “norm”, how JME affected relationships, decision making (risk versus consequences), and knowledge imparting control. Given that these restrictions were interpreted by patients as social “curfews”, we suggest that the term “Cinderella Syndrome” encapsulates the perceived imperative to be home before midnight. Our findings underscore the importance for clinicians to recognize that in counseling patients with JME about lifestyle adjustments, there may be a significant social consequence unique to this patient group.

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1. Introduction

Juvenile myoclonic epilepsy (JME) is a primarily generalized epilepsy that manifests typically in adolescence or young adulthood [1–4]. For the majority of patients with JME, there is an excellent response to medication therapy, with seizures in 85% of patients successfully controlled with appropriate antiepileptic medication and lifestyle adjustments [5–8]. However, several factors contribute to poor seizure control in the remaining 15% of patients, including drug resistance, psychiatric comorbidity, and what has been termed as an “abnormal” lifestyle [5]. Indeed, a specific behavioral profile was highlighted in the seminal article on JME [9]. Recent research has highlighted deficits in frontal lobe function [10,11] and social cognition [12] in patients with JME. Several lifestyle factors are known to provoke seizures in JME, in particular sleep deprivation and excess alcohol [5]. Sleep deprivation is reported as a trigger in up to 89% of JME-related seizures, and a pathognomonic feature is “chronodependency” of myoclonic jerks [13]. As is the case with epilepsy in general, a diagnosis of JME is known to carry with it considerable psychological burden, in part on account of its unpredictable nature, associated stigma, and its

association with low self-esteem, anxiety, and depression [14]. The teenage years are known to be a critical period for the development of self-esteem and identity formation. The emergence of risk taking behavior [15,16] contributes to the teenage years being a particularly challenging period in which to develop a chronic illness. For many teenagers, social interaction may involve late nights out or a sleep-over with friends. For patients with JME, however, a lifestyle which involves sleep deprivation and fatigue, coupled with excessive alcohol intake, is a powerful precipitant of seizures [16]. Avoidance of sleep deprivation, therefore, is a major lifestyle adjustment necessary to reduce the risk of seizures. A bit like Cinderella, who must leave the Ball and be home by midnight, an adolescent's enjoyment may be restricted by such a ‘curfew’.

While several studies have confirmed that adolescents with epilepsy have a poorer quality of life compared to other chronic conditions [14, 17–19], quality-of-life studies in patients with JME are few [20,21]. Knowledge regarding the impact of lifestyle restrictions specifically as it affects adolescents is limited. It has been emphasized that JME provides a good example of where listening to patients can provide novel insights [22]. We therefore wished to explore the potential social impact of living with a diagnosis of JME from a young adult perspective, using a qualitative descriptive approach. Specific objectives of this study included an exploration of the patient's view of having a diagnosis of JME in terms of its impact on relationships, ascertainment of a patient's

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awareness of known seizure triggers, and the potential impact of possible restrictions on their lifestyle choices.

2. Methods

2.1. Patient population and study design

We used a qualitative descriptive research design, using purposeful sampling of a cohort of patients with JME attending a specialist epilepsy clinic at University Hospital Galway, Ireland. The research project was approved by the Research Ethics Committee of Galway University Hospitals. Patients were recruited between January and April 2013 and invited to participate in a semistructured interview conducted by one of the authors (TL). Inclusion criteria included patients aged between 18 and 27 years, with an established diagnosis of JME (an age-related primarily generalized epilepsy associated with myoclonic jerks on awakening, with characteristic electroencephalographic (EEG) abnormalities [3,4]). All patients were diagnosed and being treated by neurologists with subspecialty expertise in epilepsy and who contributed to this study (TC, MJH). As part of routine clinical care, all patients attending the epilepsy clinic received regular counseling regarding the importance of lifestyle adjustments as part of seizure control. Patients are provided with written material and directed to a website (www.epilepsy.ie) for further information.

Patients were excluded if the diagnosis was unclear or they had significant neurological, psychiatric, or medical comorbidity or significant learning disability. Patients gave written informed consent to participate in the recorded interview.

2.2. Data analysis and outcomes

Following a pilot study to establish the reliability of the interview process, data were gathered using a semistructured interview (Table 1), and emergent themes were identified. Audio recording was transcribed verbatim onto a database and anonymized. Initial data were generated into ideas for formal coding into meaningful thematic groups, according to the method of Braun and Clarke [23]. Thereafter the candidate themes were refined by rereading all the coded data extracts for each theme to ensure that the candidate themes sufficiently captured the coded extracts and refine the thematic map. Following completion of thematic analysis, a final report was generated for each patient.

Table 1
Semistructured questionnaire.

| |
|---|
| Question 1 |
| "How did you feel when you were first diagnosed with epilepsy?" |
| Question 2 |
| "Did your diagnosis of epilepsy have any effect on your relationships with friends or making new friends." (Prompt: "In what way?"). |
| Question 3 |
| "What about relationships with your family?" (Prompt: "In what way?"). |
| Question 4 |
| "Since you were diagnosed, do you think epilepsy has affected your day to day life (e.g., social life, family life, sports, work)?" (Prompt: "In what way?"). |
| Question 5 |
| "Lack of sleep and alcohol use is thought to be major triggers in provoking seizures?" "Has this ever happened to you?" |
| Question 6 |
| "Do you feel avoiding these trigger factors restricted you in every-day living?" (Prompt: "In what way?") |
| Question 7 |
| "Are/were there any social occasions or outings that may have been restricted due to your epilepsy (i.e., graduation parties, concerts, sleepovers/camping)?" |
| Question 8 |
| "Is there anything that you would like to add in relation to living with epilepsy?" |

3. Results

3.1. Demographics

Eighteen patients were identified who fulfilled the inclusion criteria, twelve of whom participated in the study. All patients had a clear history of myoclonic jerks on awakening, as well as generalized tonic-clonic seizures. Eleven of the twelve patients had at least one interictal EEG with epileptiform discharges typical of JME. One patient had a normal EEG, recorded while on treatment. Six patients declined to participate. Age of the participants ranged from eighteen to twenty-seven years, and age at diagnosis ranged from twelve to twenty years. Seven participants were female. Four patients were attending college; one had completed high school. Five patients were in full time employment, and two were unemployed at the time of interview. All patients admitted to regular alcohol consumption to varying degrees. Six patients were driving. All patients reported good adherence to anticonvulsant medication. Ten patients were taking monotherapy: sodium valproate (5), levetiracetam (2), lamotrigine (2), and topiramate (1). One female patient was taking a combination of clonazepam and lacosamide, and one patient was taking a combination of sodium valproate and lacosamide. All but one of the participants were seizure-free in the six months prior to interview; one patient had a convulsion secondary to postpartum sleep deprivation.

3.2. Thematic analysis

Four main themes emerged from the qualitative analysis which we have categorized as follows: (i) alcohol – the social "norm", (ii) relationships with others, (iii) choice versus consequences, and (iv) knowledge equals control. Table 2 includes some representative quotations from participants, relating to each theme.

3.2.1. Alcohol – the social "norm"

Alcohol consumption emerged as a strong theme, having a significant social impact in this patient group. Thirty-three percent of the participants reported this issue early in the interview prior to direct questioning regarding their views about alcohol in relation to precipitating factors and seizures. Abstaining from alcohol appeared to be more problematic for participants attending college. Two subthemes relating to alcohol consumption emerged. Firstly, participants described their social environment revolving heavily around alcohol consumption: "Everything in Ireland is built around alcohol". Secondly, patients referred to the impact of abstaining from drinking alcohol in this social environment: "I used to think why should this [abstaining from alcohol] restrict... Why should I let it stop me from doing the things I want to do". Participants also reported difficulties in socializing when they abstained from alcohol: "It's not the same... [in a night club]... when you're stone cold sober" (Table 2). Feelings of isolation and being different were noted following abstinence from alcohol.

3.2.2. Relationships with others

Relationships featured strongly throughout the interviews, with early diagnosis of JME prior to high school felt to be advantageous when forming new friendships. Overall, most participants reported having positive experiences relating to relationships with family, partners, and friends. Notably, many participants reported the importance of a small close-knit circle of friends: "I think overall generally the family did come a bit closer together. I know everyone is looking out over me". Eighty-five percent of participants who were in a relationship (6 of 7 participants) felt particularly supported when socializing with their partners. One participant equated feelings of being safe in their presence (Table 2). The majority of negative comments expressed by participants related to feelings of over protection: "... My whole family, like I have four sisters and two brothers... so they're all giving out to me when I go out". One participant experienced negative perceptions of her

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