Assessing the impact of caring for a child with Dravet syndrome: Results of a caregiver survey

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Objective: The objective of this study was to describe and quantify the impact of caring for a child with Dravet syndrome (DS) on caregivers.

Methods: We surveyed DS caregivers at a single institution with a large population of patients with DS. Survey domains included time spent/difficulty performing caregiving tasks (Oberst Caregiving Burden Scale, OCBS); caregiver health-related quality of life (EuroQoL 5D-5L, EQ-5D-5L); and work/activity impairment (Work Productivity and Activity Impairment questionnaire, WPAI). Modified National Health Interview Survey (NHIS) questions were included to assess logistical challenges associated with coordinating medical care.

Results: Thirty-four primary caregivers responded, and 30/34 respondents completed the survey. From OCBS, providing transportation, personal care, and additional household tasks required the greatest caregiver time commitment: arranging for child care, communication, and managing behavioral problems presented the greatest difficulty. EuroQoL 5D-5L domains with the greatest impact on caregivers (0 = none, 5 = unable/extreme) were anxiety/depression (70% of respondents ≥ slight problems, 34% ≥ moderate) and discomfort/pain (57% respondents ≥ slight problems, 23% ≥ moderate). The mean EQ-5D general health visual analogue scale (VAS) score (0 = death; 100 = perfect health) was 67 (range, 11–94). Respondents who scored ≤ 65 were two- to fourfold more likely to report ≥ moderate time spent and difficulty managing child behavior problems and assisting with walking, suggesting that children with DS with high degrees of motor or neurodevelopmental problems have an especially high impact on caregiver health. On the WPAI, 26% of caregivers missed > 1 day of work in the previous week, with 43% reporting substantial impact (≥ 6, scale = 1–10) on work productivity; 65% reported switching jobs, quitting jobs, or losing a job due to caregiving responsibilities. National Health Interview Survey responses indicated logistical burdens beyond the home; 50% of caregivers made ≥ 10 outpatient visits in the past year with their child with DS.

Conclusions: Caring for patients with DS exerts physical, emotional, and time burdens on caregivers. Supportive services for DS families are identified to highlight an unmet need for DS treatments.

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Abbreviations: DS, Dravet syndrome; EQ-5D, EuroQoL 5D-5L quality-of-life survey; IQR, interquartile range; NHIS, National Health Interview Survey; OCBS, Oberst Caregiving Burden Scale; QoL, quality of life; SD, standard deviation; US, United States; VAS, visual analogue scale; WPAI, Work Productivity and Activity Impairment questionnaire.

1. Introduction

Dravet Syndrome (DS) is a rare, devastating, genetic epilepsy that begins in the first year of life and is marked over the course of the disease by frequent seizures and multiple seizure types, including status epilepticus [1]. Dravet Syndrome is usually determined by clinical diagnosis and is often associated with an SCN1A gene mutation, which may be a loss-of-function mutation. More than 80% of people with DS are found to have a mutation in SCN1A [2], the gene that encodes the pore-forming subunit of the type I voltage-gated sodium channel (NaV 1.1), found in the brain and the heart [3–5]. No drug is currently approved in the United States (US) for DS. It is a highly pharmacotherapy-resistant and refractory
epilepsy syndrome. Attaining substantial seizure reduction (i.e., ≥ 75% reduction) or seizure-freedom in affected children, even with combination therapy, is rare, thereby placing a heavy impact of disease on children with DS and on the caregivers they are dependent upon. Contributing to this impact is the lack of medical literature regarding treatment of the associated characteristics of DS, such as concomitant developmental and behavioral issues. While much focus in the literature has been understandably placed on finding new treatments for these patients, only recently has the literature begun to highlight the broader impact of DS on caregivers and the family as a whole.

In a cohort of Canadian caregivers, Nolan et al. found that persistent severe seizures in patients with DS, along with cognitive, developmental, behavioral, and sleep issues, resulted in high stress for caregivers and little ability to find relief [6]. Stress was characterized in this parental cohort by deterioration of relationships with others (including spouses), as well as fear, uncertainty, and sleep problems [6]. Skuluzak et al. identified grief as an additional stressor on parents and emphasized the need for support in managing the stress of caring for a child with DS [7]. More recent studies have identified additional stress factors on caregivers, including sleep deprivation, reduced mental health, deterioration of social relationships, financial burden [8], and a substantial incidence of depression, in addition to family-related factors such as concern regarding the emotional impact of having a child with DS may be having on siblings [9].

To further identify and quantify the factors impacting the well-being of caregivers of children with DS, we conducted a prospective, single-center survey study within Children's Hospital Colorado, a hospital with a large population of patients with DS.

2. Methods

We assessed the burden of DS on a cohort of caregivers of patients with DS actively being seen at Children's Hospital Colorado. An electronic survey (Appendix) was administered to caregivers, who were eligible if they provided care for a friend or a family member with DS. The survey was administered through REDCap (https://www.project-redcap.org/), a metadata-driven methodology and workflow process for providing translational research informatics support [10]. The study design was reviewed and approved by the Colorado Multiple Institutional Review Board, and all survey participants provided informed consent.

An e-mail inviting DS caregivers to participate in the survey was sent if an e-mail address was available in the patient's electronic health record. Fifteen eligible participants did not have an e-mail on file; a coordinator contacted the caregivers by telephone to see if they were interested in participating and providing their e-mail. Eight agreed; of the remaining 7, 4 could not be reached, 2 were not English-speaking, and 1 declined. Consent was obtained before participants could begin the survey questions. The survey was administered one time for each caregiver and asked for information pertaining to the previous year. Survey areas of assessment included time spent and difficulty performing caregiver tasks, caregiver health-related quality of life (QoL), and caregiver work-related productivity/activity impairment.

To understand the time spent and difficulty performing tasks associated with caregiving, we modified a version of the Oberst Caregiving Burden Scale (OCBS) [11,12], a 15-item instrument that rates caregiving tasks based on time spent (1 = none, 4.5 = a large amount) and difficulty of task (1 = not difficult, 4.5 = very difficult).

Caregiver QoL was assessed using the EuroQol 5D-5L (EQ-5D) health-related QoL survey [13]. The EQ-5D survey is a standardized measure of health status and assesses 5 different domains on a 5-point scale (0 = death; 100 = perfect health). Visual analogue scale scores were used to stratify OCBS and Work Productivity and Activity Impairment questionnaire (WPAI) results to assess if there were differences in these measures for VAS scores <65 or ≥ 65.

Caregiver productivity and activity impairment was assessed using a subset of the WPAI questionnaire [15], which was tailored to focus on the impact of DS caregiving on employment. Work Productivity and Activity Impairment items generate percentages (0%–100%) that quantify absenteeism (percentage of time missed from work), presenteeism (percentage of restriction while at work), overall work restriction (percentage of total restriction due to either absenteeism or presenteeism), and overall activity limitation (percentage of limitation in daily activities) due to caregiving responsibilities, with higher values indicating greater limitation. Only caregivers currently employed (full-time, part-time, or self-employed) were asked to respond about work productivity, but all caregivers were asked about activity limitation.

Finally, to assess logistical challenges required of the caregiver to coordinate healthcare visits of the patient with DS selected and modified questions from the National Health Interview Survey (NHIS) were included in the survey [16]. Items included questions such as “During the past 12 months, how many times has your child gone to a hospital emergency room? (This includes emergency room visits that resulted in a hospital admission)”; see Appendix.

Survey responses for the different OCBS, EQ-5D, WPAI, and NHIS subdomains were totaled and presented as percentages, means (± SD), medians, and ranges as appropriate.

3. Results

The survey was open for 3 weeks, from November 30, 2016 to December 20, 2016; it was e-mailed to 60 primary caregivers of patients with DS routinely seen at the Children's Hospital Colorado who were asked to participate; 34 (57%) agreed. Of those who participated, 30 (88%) fully completed the survey. The 34 participants were primary caregivers for 34 patients with DS; 91.2% of the patients had confirmed sodium channel α-subunit gene mutation in SCN1A. Patients ranged in age from 2 to 22 years (mean ± SD, 11.7 ± 5.8 years). Additional demographic information on caregivers or patients with DS was not collected.

Responses to the OCBS are provided in Fig. 1 and show that a substantial number of caregiver domains were impacted from both a difficulty and time perspective. The top five domains in terms of proportion of caregivers reporting moderate or greater time burden were: providing transportation (93% of respondents), personal (patient with DS) care (87%), additional household tasks (83%), communication (80%), and symptom observation (77%). The top five domains with regard to proportion of caregivers reporting moderate or greater difficulty included: arranging for care (73%), communication (70%), coordinating resources (67%), managing behavior problems (67%), and personal (patient with DS) care (63%).

From the EQ-5D, the domains with the greatest impact on caregivers were anxiety/depression (70% of respondents ≥ slight problems, 33% ≥ moderate) and discomfort/pain (57% of respondents ≥ slight problems, 23% ≥ moderate). The EQol Index score, a summary measure of the five EQ-5D health domains adjusted by population-based preference weights, was 0.78 ± 0.17, suggesting an overall reduction in QoL as compared with perfect health (1 = perfect health). The EQ-5D general health assessment mean VAS score of 67 (range 11–94; Table 1) also provided evidence of substantial reduction from what would be expected in a normative population, where 63% of respondents scored between 80 and 100, and 71 was the mean score reported for those with one major health condition [17]. Forty percent of respondents scored <65 on the VAS scale of general health, where 0 = death and 100 = perfect health, as compared with 22% in a normative population [17].

Work Productivity and Activity Impairment responses (Table 2) indicated a substantial effect of DS on caregivers’ missed work and
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