

# Assessing the Impact of Caring for a Child with Dravet Syndrome: Results of a Caregiver Survey

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## INTRODUCTION

- ▶ Dravet syndrome (DS) is a rare, devastating epileptic encephalopathy characterized by:
  - Frequent seizures and multiple seizure types, including status epilepticus
  - Neurodevelopmental problems
  - Increased rates of sudden unexpected death in epilepsy
- ▶ Studies have shown DS places a heavy burden of disease on the children affected (Brunklau et al, 2011); however, recent literature describes the additional burden on the caregivers they are dependent upon
  - Physical health, mental health, social function, financial resources, sleep, and fatigue have been identified as areas of greatest impact (Jensen 2017)<sup>2</sup>
  - Villas et al (2017)<sup>3</sup> reported that nearly two-thirds of caregivers (66%) had suffered from depression, but only 26% had received some form of family therapy
- ▶ We conducted a prospective, single-center survey study within Children's Hospital Colorado to further assess factors impacting the well-being of caregivers of children with DS

## METHODS

- ▶ An electronic survey to describe and quantify the impact of caring for a child with DS on caregivers' health-related quality of life (QoL) and productivity was administered to caregiver participants, if they provided care for a friend or a family member with DS
  - The survey was administered through Research Electronic Data Capture (REDCap; <https://www.project-redcap.org/>),<sup>4</sup> a metadata-driven methodology and workflow process for providing translational research informatics support
- ▶ The study was reviewed and approved by the Colorado Multiple IRB; all participants provided informed consent
- ▶ Survey domains included time spent/difficulty performing caregiving tasks, health-related QoL, and work-related productivity using:
  - Modified Oberst Caregiving Burden Scale (OCBS)
    - 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)
  - EuroQoL 5D-5L (EQ-5D) health-related QoL survey
    - 5-domain survey that rates QoL on a 5-point scale (0=no problems; 4=severe problems; or 0=none, 4=extreme)
    - A general health assessment measured on a visual analog scale (VAS; 0-100 scale where 0=death and 100=perfect health) was also included
    - An EQoL Index was computed using responses across all 5 EQ-5D domains, adjusted by population-based preference weights
- A subset of the Work Productivity and Activity Impairment (WPAI) questionnaire, which was tailored to focus on the impact of DS caregiving on employment

- Survey 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 about work productivity, but all caregivers were asked about activity limitation
  - EQ-5D VAS scores were used to stratify OCBS and WPAI results to assess if there were differences in these measures for VAS scores <65 or ≥65
  - Select questions from the National Health Interview Survey (NHIS) were also included to better understand the logistical challenges faced by caregivers

## RESULTS

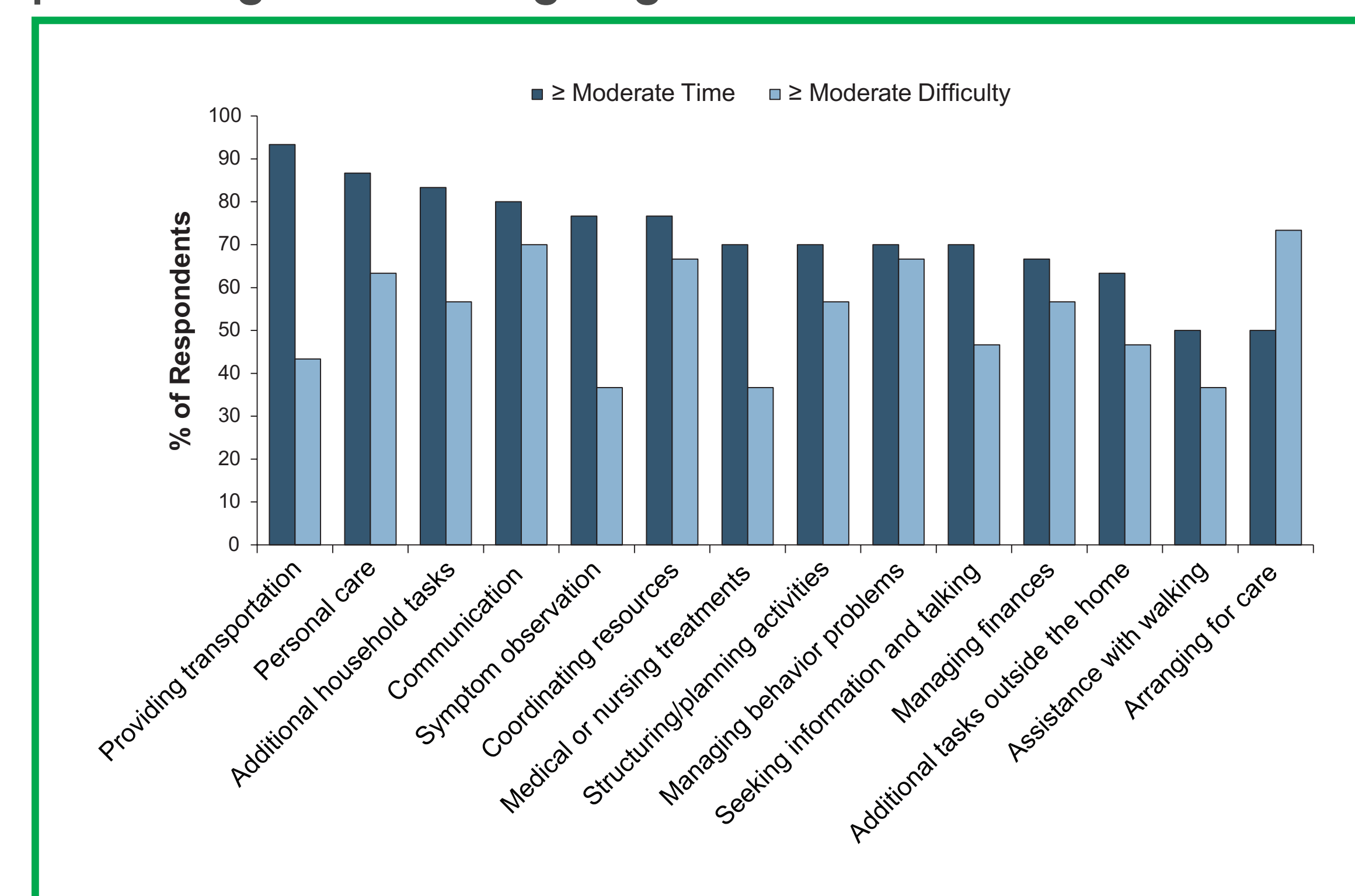
### Demographics

- ▶ Thirty-four of 60 invited primary caregivers (57%) responded to the survey
  - 30 of 34 responders (88%) fully completed the survey
  - DS caregivers' children ranged in age from 2 to 22 years (mean±SD, 11.7±5.8 years)

### OCBS

- ▶ Responses to the OCBS are provided in **Figure 1**
- ▶ The top 5 domains with a moderate or greater time burden included:
  - Providing transportation (93% of respondents)
  - Personal [DS patient] care (87%)
  - Additional household tasks (83%)
  - Communication (80%)
  - Symptom observation (77%)
- ▶ The top 5 domains with a moderate or greater difficulty included:
  - Arranging for care (73%)
  - Communication (70%)
  - Coordinating resources (67%)
  - Managing behavior problems (67%)
  - Personal care (63%)

**Figure 1. OCBS results: percent of DS caregivers experiencing moderate to severe time and difficulty limitations when performing routine caregiving tasks.**



DS, Dravet syndrome; OCBS, Oberst Caregiving Burden Scale.

### EQ-5D (Table 1)

- ▶ Domains with the greatest impact on caregivers were anxiety/depression and discomfort/pain
- ▶ The EQoL Index score was 0.78±0.17, suggesting an overall reduction in QoL as compared to perfect health (1.0)
- ▶ The mean EQ-5D general health VAS score of 67 (range 11-94) 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<sup>5</sup>
  - 40% of responders scored ≤67
  - DS Caregivers were nearly twice as likely to score <65 on the VAS and <70 on the EQ-5D compared to a normative US population<sup>5</sup>

**Table 1. Impact of DS Caregiving on EQ-5D Tasks**

Survey Variable	n	Min	Max	Score=0	Score ≥Slight	Score ≥Moderate
Mobility/walking about (0-4) <sup>a</sup>	30	0	3	80%	20%	13%
Self-care/washing & dressing self (0-4) <sup>a</sup>	30	0	1	93%	7%	0%
Usual activities (0-4) <sup>a</sup>	30	0	4	73%	27%	13%
Pain/discomfort/(0-4) <sup>b</sup>	30	0	4	43%	57%	23%
Anxiety/depression (0-4) <sup>c</sup>	30	0	4	30%	70%	33%
				Mean	±SD	
EQoL Index <sup>d</sup>	30	0.31	1	0.78	0.17	
General Health (0-100; VAS) <sup>e</sup>	30	11	94	67	21	

<sup>a</sup>Scores: 0=I have no problems; 1=I have a slight problem; 2=I have a moderate problem; 3=I have severe problems; 4=I am unable.  
<sup>b</sup>0=I have no pain; 1=I have slight pain; 2=I have moderate pain; 3=I have severe pain; 4=I have extreme pain.  
<sup>c</sup>0=I am not anxious or depressed; 1=I am slightly anxious or depressed; 2=I am moderately anxious or depressed; 3=I am severely anxious or depressed; 4=I am extremely anxious or depressed.  
<sup>d</sup>US time-trade-off mapping using the EQ-5D-3L crosswalk (0=death; 1=perfect health).  
<sup>e</sup>0=death; 100=perfect health.  
 DS, Dravet syndrome; EQ-5D, EuroQoL 5D-5L health-related quality-of-life survey; EQoL, EQ-5D summary index; VAS, visual analog scale.

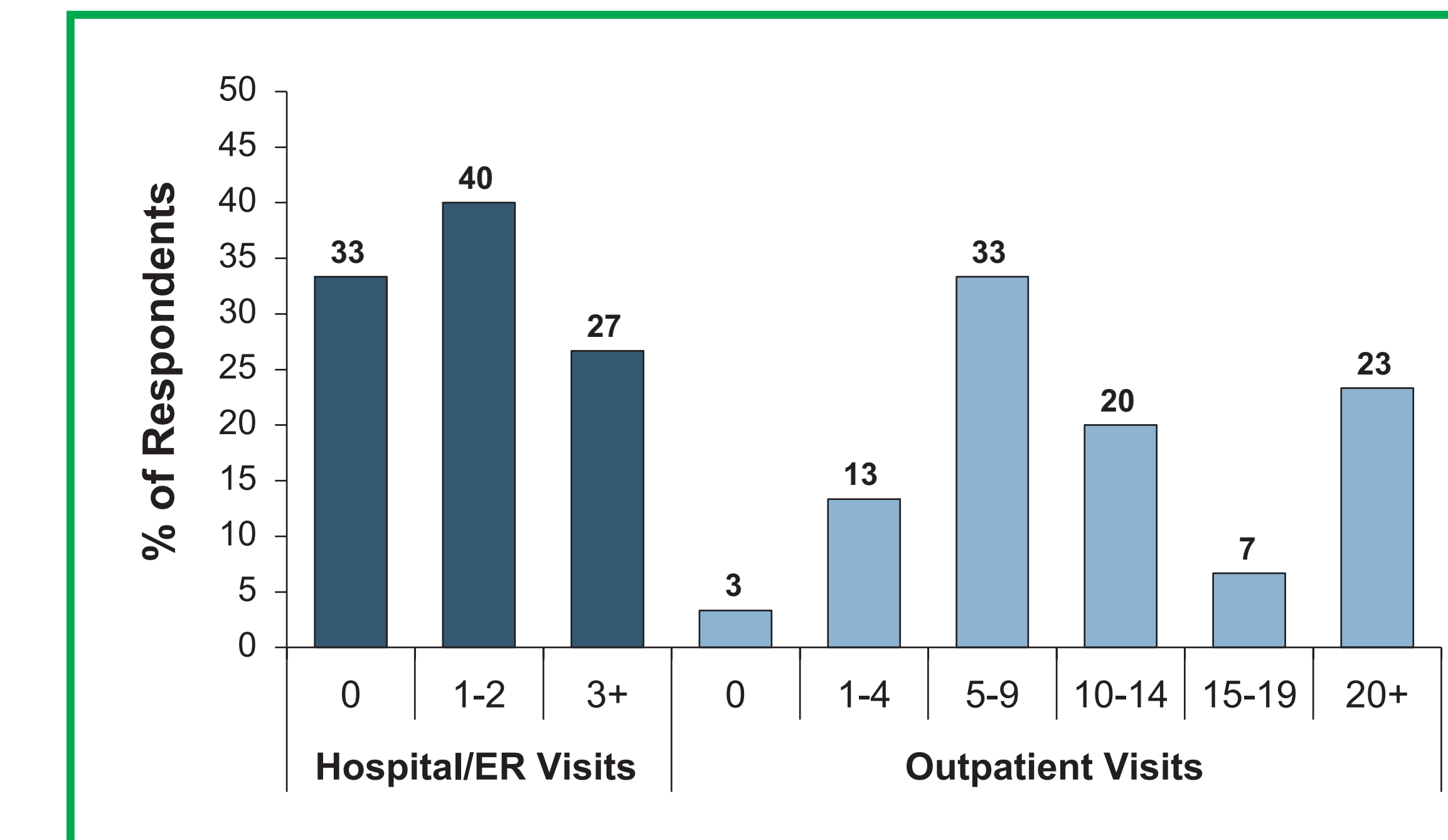
### WPAI

- ▶ Responses indicated substantial effect of DS caregiving on caregivers' missed work and leisure time, work productivity, and regular activities
  - 45% (15/33) either quit, retired early, or lost their jobs
  - 18% (6/33) found it necessary to switch jobs
  - Of caregivers who were not employed (n=9), a majority of respondents (7/9, 78%) felt that caregiving was an impediment to employment

### NHIS

- ▶ The majority of caregivers were faced with coordinating multiple hospital/ER (67% of caregivers) and outpatient visits (97% of caregivers) annually (**Figure 2**)
  - Outpatient visits were an intensely utilized medical service, with 50% of DS caregivers making ≥10 visits annually for their child with DS and 23% making ≥20 outpatient visits annually

**Figure 2. NHIS results: yearly hospital/ER/outpatient visits coordinated by caregivers for children with DS.**



DS, Dravet syndrome; ER, emergency room; NHIS, National Health Interview Survey.

### Stratifications of Survey Results by VAS Score

- ▶ An EQ-5D general health VAS score <65 was used to stratify results from other survey domains
- ▶ VAS <65 vs VAS ≥65 equated to:
  - Approximately 27 mean hours fewer per week for leisure activities, a nearly 2-fold higher impact on work productivity, and a 1.5-fold greater impact on leisure activities (**Table 2**)
  - An approximately 2-fold greater proportion of caregivers reporting moderate or greater difficulty with watching and reporting symptoms, providing transportation, household tasks, managing behavior, and seeking information (**Table 3**)
  - A 2- to 4-fold greater proportion of caregivers reporting moderate or greater time and difficulty dealing with assistance with walking
- ▶ Certain comorbidities such as greater behavioral problems or decreased motor ability may be contributing to an even greater negative impact on caregivers as measured by EQ-5D general health VAS

**Table 2. Influence of DS Caregiving on WPAI Work Productivity and Leisure Time, by EQ-5D VAS Category**

	EQ-5D VAS ≥65 (n=18)		EQ-5D VAS <65 (n=12)	
	Mean (SD)	Median (IQR)	Mean (SD)	Median (IQR)
Weekly time missed from work (hours)	7.4 (15.2)	0.5 (0.0, 2.8)	6.9 (12.3)	0 (0.0, 8.0)
Weekly time missed from leisure (hours)	31.0 (53.9)	7.0 (1.8, 23.0)	57.8 (59.2)	40.0 (7.3, 84.0)
Effect caregiving had on work productivity <sup>a</sup>	39.1 (25.6)	52.0 (13.8, 58.0)	76.9 (19.8)	75.0 (68.0, 90.0)
Effect caregiving had on leisure time <sup>a</sup>	55.1 (24.0)	55.5 (39.5, 69.3)	82.6 (12.4)	81 (77.3, 91.8)

<sup>a</sup>Data are scores; scale: 0=no impact to 100=completely prevented productivity or leisure time.  
 DS, Dravet syndrome; EQ-5D, EuroQoL 5D-5L health-related quality-of-life survey; IQR, interquartile range; SD, standard deviation; VAS, visual analog scale; WPAI, Work Productivity and Activity Impairment.

**Table 3. Influence of DS Caregiving on OCBS Domains, by EQ-5D VAS Category<sup>a</sup>**

	EQ-5D VAS ≥65 (n=18)	EQ-5D VAS <65 (n=12)
Moderate or More Time Spent		
– Assistance with walking	33.3%	75.0%
Moderate or More Difficulty		
– Assistance with walking	16.7%	66.7%
– Watching/reporting symptoms	27.8%	50.0%
– Providing transportation	27.8%	66.7%
– Household tasks	38.9%	83.3%
– Managing behavior	44.4%	100.0%
– Seeking information	33.3%	66.7%

<sup>a</sup>Table shows OCBS domains with an approximately 2-fold or greater difference between VAS groups.

DS, Dravet syndrome; EQ-5D, EuroQoL 5D-5L health-related quality-of-life survey; OCBS, Oberst Caregiving Burden Scale; VAS, visual analog scale.

## CONCLUSIONS

- ▶ In this single-center cohort, DS caregivers surveyed faced substantial physical, emotional, and time burdens, including elevated levels of anxiety/depression and pain
- ▶ Caregivers had a mean EQ-5D general health VAS score of 67, lower than that reported for those in the general population with one major health problem (71)
- ▶ These data corroborate recent findings reported in the literature and further add to our knowledge about the direct impact of DS on caregivers' emotional and financial well-being
- ▶ Our findings identify potential areas for supportive services for DS families and highlight an unmet need for more comprehensive treatments for DS

## STUDY LIMITATIONS

- ▶ Single-center study with small sample size (34 participants) may limit the generalizability of our data
- ▶ A survey depends heavily on the impressions and recollections of participants, thus making our data subject to various response and recall biases
- ▶ The consistency of our findings with observations from recently published studies by Villas et al<sup>3</sup> and Jensen et al,<sup>2</sup> however, suggest that our results may have some validity regarding the larger population of DS caregivers

## REFERENCES

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## DISCLOSURE

JDC: Research support, Zogenix; Consultant, Amgen.

MDW: Research support, Zogenix.

CHK: No conflicts to report.

GRV: No conflicts to report.

KK: Research support, Zogenix and Colorado Department of Public Health and Environment.

AG: Employee, Zogenix; Stock ownership, Zogenix.

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