Interim Findings from the Sibling Voices Survey: Impact of Severe Childhood Epilepsy on Siblings 17 Years of Age or Under

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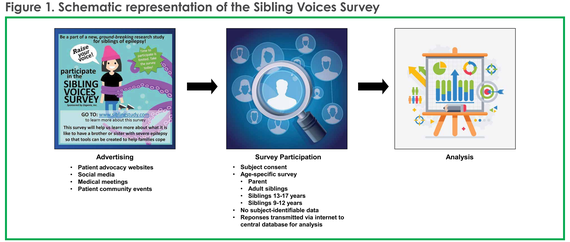
INTRODUCTION

- Dravet syndrome, Lennox-Gastaut syndrome, and other epileptic encephalopathies are severe epilepsy syndromes with onsets in childhood that are typically resistant to treatment and accompanied by neurodevelopmental impairment
- These patients require constant care and monitoring which can significantly impact the broader family as a whole
- Recent data has begun to quantify the impact of these conditions on the primary caregiver, but little information exists about the impact on the entire family unit, and specifically on siblings
- Siblings are often directly and/or indirectly involved in the care of their affected brother/sister
- These siblings have had minimal recognition by health care providers and the medical community
- The effects on a sibling living in a family with a child with severe epilepsy has not been well studied
- The Sibling Voices Survey was developed to evaluate the psychosocial impact on siblings who live with a brother/sister with severe childhood epilepsy

METHODS

- Internet-based survey (www.siblingstudy.com; **Figure 1**)
  - Survey questionnaires were developed in consultation with the Dravet syndrome and Lennox-Gastaut syndrome patient communities and healthcare professionals
    - Questions were further refined following beta testing in a small group of subjects that included parents and siblings from the patient communities
  - 4 separate surveys were developed and used in the study:
    - Parents
    - Siblings 9-12 years
    - Siblings 13-17 years
    - Adult siblings 18 years and older
  - Examples of questions in the surveys for siblings 9-12 years and 13-17 years are shown in **Figure 2**
  - Subjects are recruited through social media advertising with patient advocacy groups
  - No subject-identifiable information (eg, name, town, state, or country of residence) is solicited
  - Protocol was reviewed and approved by the Western IRB (Puyallup, WA) on July 14, 2017
  - The survey was launched on July 19, 2017 and is still open for participation

Figure 1. Schematic representation of the Sibling Voices Survey



- Analysis
  - This interim analysis of the survey responses focused on 2 sibling groups: 9-12 years and 13-17 years
  - Areas of focus for this analysis were:
    - Identifying which aspects of their affected brother's/sister's condition were most difficult/challenging for siblings
    - Identifying the most common emotions reported by siblings
  - To identify the most concerning aspects or emotions most frequently reported by siblings, the number of responses in the 2 top categories ("*A lot*" and "*A medium amount*") were summed for each question and these sums were ranked

Figure 2A. Example of survey questions for siblings 9-12 years used to identify difficult aspects

1. What things about your brother's or sister's [diagnosis] or seizures are the hardest for you to deal with? Please pick the answer that you think best describes how you feel about each statement.
   1a) Feeling worried or scared when they have a seizure
   a. Not at all
   b. A little
   c. A medium amount
   d. A lot

   1b) Not getting as much time and attention from my mom and dad
   a. Not at all
   b. A little
   c. A medium amount
   d. A lot

   1c) Having activities changed because my sibling gets sick or has a seizure
   a. Not at all
   b. A little
   c. A medium amount
   d. A lot

   1d) My parents being stressed or unhappy
   a. Not at all
   b. A little
   c. A medium amount
   d. A lot

   1e) Having to help out more at home with more responsibilities and chores
   a. Not at all
   b. A little
   c. A medium amount
   d. A lot

   1f) My brother's or sister's odd or weird behaviors
   a. Not at all
   b. A little
   c. A medium amount
   d. A lot

   1g) Let us know if there is anything we forgot to ask _____

Figure 2B. Example of survey questions for siblings 13-17 years used to identify emotions

For the next set of questions (19 through 24), please pick the best answer for each statement or question.

19. I have bad dreams or nightmares
    a. Never
    b. Very occasionally
    c. Sometimes
    d. Frequently
    If d, do you think your brother's or sister's epilepsy is the trigger for your bad dreams? YES____ NO _____

20. I feel jumpy or get easily startled (easily scared)
    a. Never
    b. Very occasionally
    c. Sometimes
    d. Frequently

21. Have there been extended periods of time when you felt down and unhappy?
    a. Not at all
    b. A little
    c. Sometimes
    d. A lot

22. Have there been extended periods of time when you felt irritable (easily frustrated)?
    a. Not at all
    b. A little
    c. Sometimes
    d. A lot

23. Have there been extended periods of time when you did not enjoy things like you usually do?
    a. Not at all
    b. A little
    c. Sometimes
    d. A lot

24. Have there been extended periods of time when you felt too tired to do things that you normally do?
    a. Not at all
    b. A little
    c. Sometimes
    d. A lot

RESULTS

- From July 19, 2017 to August 10, 2017 a total of 183 survey submissions were received
- 12 surveys were completed by siblings 9-12 years
- 15 surveys were completed by siblings 13-17 years
- These siblings represented 23 of the 137 families who had participated in the survey
- Demographics of these 2 groups of siblings are presented in **Table 1**

Table 1. Demographic characteristics of sibling respondents and patients

Sibling Survey Respondents	Sibling Respondent Group		Total
	9-12 years	13-17 years	
n	12	15	27
Male:Female	5:7	10:5	15:12
Age, years (mean±SD)	10.1±1.1	13.8±0.9	12.1±2.1
School grade (mean±SD)	5.1±1.3	8.9±1.0	7.2±2.2
Family size (mean±SD)	5.1±1.1	5.5±1.9	5.2±1.5
Number of children in family (mean±SD)	3.1±1.1	3.6±1.8	3.3±1.5

Patients	9-12 years	13-17 years	Total
n[a]	11	13	23
Age, years (mean±SD)	7.5±2.6	7.6±4.3	7.8±3.6
Male:Female	4:7	5:8	8:15
Epilepsy diagnosis, n			
Dravet syndrome[a]	8	6	13
Lennox-Gastaut syndrome	0	1	1
Epilepsy	1	5	6
Severe epilepsy	2	0	2
Unknown epilepsy	0	1	1

[a]The number of patients does not equal the number of siblings in each group because 4 families had 2 siblings participate in the survey. In each case, the patient had been diagnosed with Dravet syndrome. One family pair of siblings was grouped in with 9-12 years, 2 family pairs of siblings were grouped in with 13-17 years, and the final family sibling pair had one sibling in each age group.

Note: All patient demographic information was submitted by the siblings.

- Differences emerged with regard to the aspects that most impacted the 2 sibling groups (**Figure 3**):
  - The sibling group 9-12 years identified both "*Feeling worried or scared when their sibling had a seizure*" and "*Not getting as much time and attention from my mom and dad*" as the most difficult aspects to deal with (**Figure 4**)
  - The sibling group 13-17 years identified "*My parents being stressed or unhappy*" as the most difficult aspect to deal with (**Figure 4**)

Figure 3. Responses to questions designed to identify difficult aspects

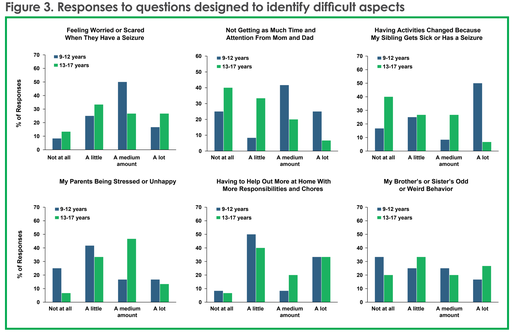
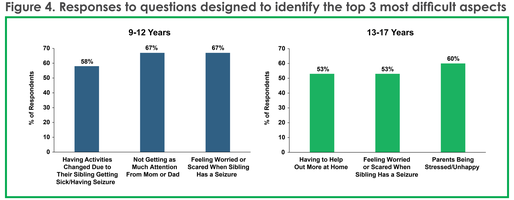


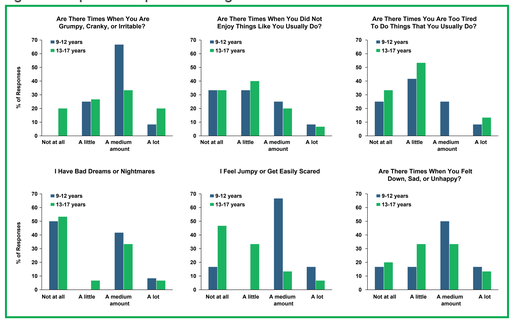
Figure 4. Responses to questions designed to identify the top 3 most difficult aspects



"% of Respondents" represents the percentage of combined totals for "*A medium amount*" and "*A lot*."

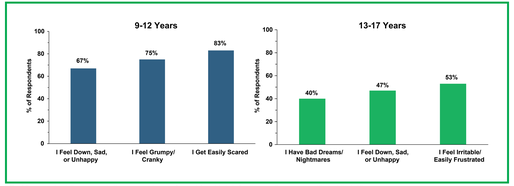
- The distribution of responses to questions designed to elicit responses about emotions associated with having a sibling with severe epilepsy are shown in **Figure 5**
  - The top emotion identified by siblings 9-12 years was being "*easily scared*" (**Figure 6**)
  - The top emotion identified by siblings 13-17 years was "*feeling irritable or frustrated*" (**Figure 6**)

Figure 5. Responses to questions designed to elicit emotions



Note: The questions were worded slightly differently on the age-specific questionnaires. For example, the sibling group 9-12 years was asked to respond to "*I get easily scared*," whereas the sibling group 13-17 years responded to "*I feel jumpy or get easily startled (easily scared)*."

Figure 6. The most common emotions described in siblings 9-12 years and 13-17 years



"% of Respondents" represents the percentage of combined totals for "*A medium amount*" and "*A lot*."

Other important interim findings from the Sibling Voices Survey in these 2 age groups included:

- In the sibling group 9-12 years
  - When asked "*How comfortable do you feel when talking to others about your brother's or sister's diagnosis?*" 45% responded "*A medium amount*" and no one responded "*I feel very comfortable*"
  - 75% indicated that they were most likely to talk to family members about their sibling's diagnosis
- In the sibling group 13-17 years
  - 94% reported that they were "*fairly comfortable*" or "*very comfortable*" speaking with others about their affected sibling's diagnosis
  - In addition to talking with parents (87%), 47% reported that they also talk to their friends about their sibling's diagnosis
- Both sibling groups expressed concern regarding their affected sibling's mortality
  - In the sibling group 9-12 years, 83% expressed fear that their sibling might die
  - In the sibling group 13-17 years, 73% reported that their parents or others have talked with them about the possibility of their sibling's mortality and 43% indicated that they were "*somewhat stressed/worried*" to "*extremely stressed/worried*" about this possibility

DISCUSSION AND CONCLUSIONS

- Although it is well established that caring for a child with Dravet syndrome, Lennox-Gastaut syndrome, or other severe epilepsy has considerable impact on caregivers, the impact on unaffected siblings has not been well studied
  - In a recent survey of caregivers of patients with Dravet syndrome, 74% reported that they had concerns about the emotional impact on unaffected siblings (1)
  - In that survey, when caregivers were asked to provide concerns in the open response section, the second most common concern was impact on the sibling, which was ranked well above the parent's own anxiety, depression, or feelings of isolation
- This interim analysis from the Sibling Voices Survey provides insight into the potentially significant impact that these diseases have on unaffected siblings and how they perceive the issues that involve themselves and their family
- Despite being an interim analysis, these findings suggest that caregiver concerns about the emotional impact on siblings are justified and warrant further investigation to identify true incidence rates of psychological issues among siblings in order to support the need for potential intervention and strategies for these family members
- The Sibling Voices Survey will remain active through the end of 2017, and further analyses will follow

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