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Assessing Quality of Life in Siblings of Children With Severe Epileptic Encephalopathies: A Comparative Analysis of Sibling Self-Reports and Parental Perception of Sibling Experiences

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INTRODUCTION

- Severe epileptic encephalopathies (EEs) of childhood (eg, Dravet syndrome [DS], Lennox-Gastaut syndrome [LGS]) often result in developmental delays, neurocognitive impairment, and motor deficiencies[1]
- EEs place substantial humanistic burdens on the entire family unit[2-4]
  - Impact on other siblings ranked second among the top caregiver concerns (74% of 154 caregivers)[5]
  - Parental surveys report concern for siblings being at heightened risk of psychosocial problems[6]
- Limited information is available from the sibling perspective on how growing up with a patient with an EE affects the mental/emotional well-being of siblings

OBJECTIVE

- Survey-based study that compares parental and sibling responses to questions designed to assess the psychosocial impact of growing up with a sibling with an EE

METHODS

Survey Development and Recruitment

- Survey questions were developed and beta-tested in consultation with communities of patients with DS and LGS and healthcare professionals
  - Study protocol approved by Western Institutional Review Board (IRB; Puyallup, WA)
  - Open for responses from July 19–December 31, 2017
- Participants were recruited through social media, patient advocacy websites, and patient community events
- Participants responded to 1 of 4 online, IRB-approved, cohort-specific surveys
  - Respondents or caregivers/parents gave consent to participate
  - Cohorts: parents, siblings 9-12 years old (y/o), siblings 13-17 y/o, and adult siblings ≥18 y/o
  - Adult siblings answered both currently and retrospectively

Survey Design and Analysis

- Quality-of-life (QoL) metrics used visual analog scale scores, with 0-10 or 0-3 response options
- Four questions assessed potential depressed mood
- Two questions assessed potential anxiety symptoms
- Additional questions assessed QoL parameters

RESULTS

Sibling Voices Survey: Respondents and Demographics

- N=248 submissions (**Table 1**)
- Parent and sibling respondents represented 115 and 107 patients with EEs, respectively (**Table 2**)

Table 1. Demographic Characteristics of Sibling Voices Survey Respondents

	Sibling Respondent Age Group			
Characteristic	9-12 y/o	13-17 y/o	Adult Siblings	All Siblings
n	24	17	79	120
Sex, n (%)				
Male	12 (50)	11 (65)	23 (29)	46 (38)
Female	12 (50)	6 (35)	56 (71)	74 (62)
Age, years Mean (range)	10 (9-12)	14 (13-15)	24 (18-63)	21 (9-63)
Family size Mean (range)	5 (4-8)	5 (4-10)	4 (1-30)	4 (1-30)
No. of children/family Mean (range)	4 (2-6)	4 (2-8)	3 (0-10)	3 (0-13)

NA, not applicable; y/o, years old.

Parent Respondents	
Characteristic	Parents
n	128
Sibling sex[a]	
Male	106 (54)
Female	92 (46)
Sibling age, years Median (range)[a]	12 (0-39)
Relationship, n (%)	
Mother	116 (91)
Father	10 (8)
Other	2 (<2)

[a]Some parents did not fill out surveys for all siblings in their family.

Table 2. Demographic Characteristics of Patients With an EE

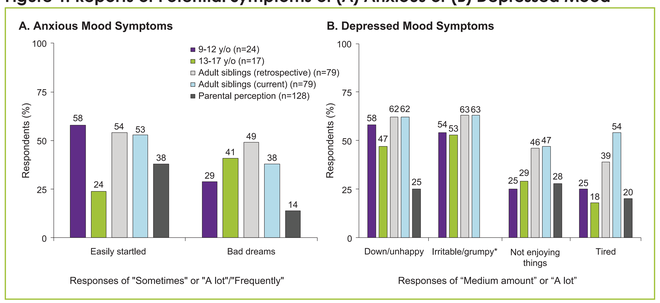
	Patients With an EE Corresponding to:	
Characteristic	Parent Respondents (n=128)	Sibling Respondents (n=120)
Patients, n	115	107
Sex, n (%)		
Male	53 (46)	44 (41)
Female	62 (54)	63 (59)
Age, years Mean (range)[a]	10 (1-50)	16 (1-61)
Patient diagnosis		
Dravet syndrome	47 (41)	41 (38)
Lennox-Gastaut syndrome	13 (11)	13 (12)
Other EE	55 (48)	53 (50)

[a]Excludes 2 deceased patients with an EE (both cohorts).
EE, epileptic encephalopathy; y/o, years old.

Impact on Emotional Well-Being

- Parents perceived less impact on having bad dreams (14%) and being unhappy (25%) than siblings reported (bad dreams: 29%, 41%, and 38%-49% for 9-12 y/o, 13-17 y/o, and adult siblings; being unhappy: 58%, 47%, and 62%, respectively) (**Figure 1**)

Figure 1. Reports of Potential Symptoms of (A) Anxious or (B) Depressed Mood

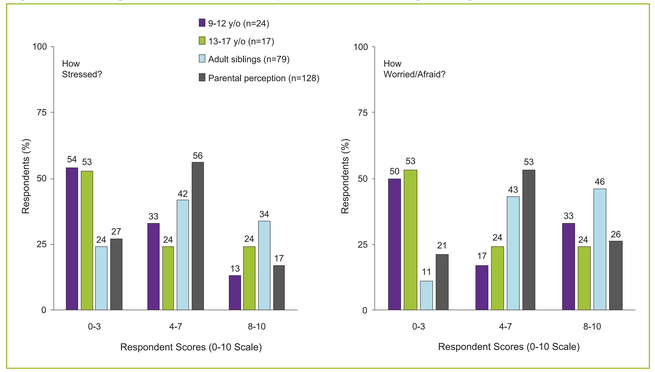


y/o, years old.

*Parents were not asked this question.

- Siblings 9-12 and 13-17 y/o reported lower ratings for stress and worry/fear (stress, scores 0-3: 54% and 53%, respectively; worry/fear, scores 0-3: 50% and 53%, respectively) than parents perceived (27% and 21%, respectively, in the 0-3 range) (**Figure 2**)

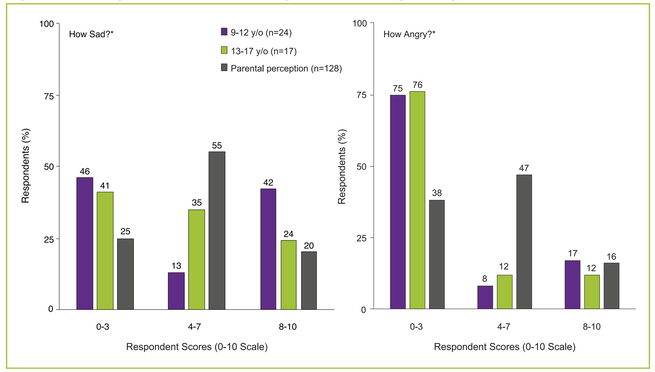
Figure 2. Ratings of Stress and Worry/Fear Over Sibling's Diagnosis



0-10 scale: 0, "not at all"; 5, "somewhat"; 10, "extremely."
y/o, years old.

- Substantial differences were observed in ratings for sadness and anger between younger siblings and parental perception (**Figure 3**)

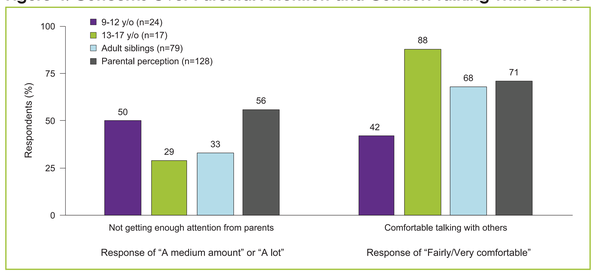
Figure 3. Ratings of Sadness and Anger Over Sibling's Diagnosis



*Adult siblings were not asked this question.
0-10 scale: 0, "not at all"; 5, "somewhat"; 10, "extremely."

- Nearly 2-fold difference seen between parental perceptions and at least 1 sibling cohort in the following metrics (**Figure 4**):
  - Not getting enough attention from parents
  - Comfort level in talking with others about sibling's diagnosis

Figure 4. Concerns Over Parental Attention and Comfort Talking With Others



CONCLUSIONS

- The Sibling Voices Survey data support recent survey studies that report parent/caregiver concerns regarding the emotional impact of EEs on siblings[4,6]
- Important differences exist between parental perception and sibling reports on:
  - Mood symptoms (eg, anxious mood, depressed mood)
  - Degree of sadness, anger, worry/fear, and stress
  - Comfort level talking with others about the sibling's diagnosis and concerns over amount of parental attention received
- Results highlight the need for increased awareness and ongoing assessment of the potential need to support siblings of patients with an EE
- Clinicians should discuss the potential for sibling depressed mood and anxiety with parents
- Further research is warranted to better understand how to identify those siblings who may be struggling with depression and/or anxiety and the best methods for intervention

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DISCLOSURES

LDB, ARG, BSG, CS: Employee, Zogenix; Stock ownership, Zogenix.
LS: Consultant, Zogenix.

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