

Impact of Severe Rare Childhood Epilepsy on Siblings Under 18 Years of Age

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INTRODUCTION

- Severe epileptic encephalopathies (EEs) of childhood are pharmacoresistant and result in developmental delays, neurocognitive impairment, and motor deficiencies¹
- Caregivers experience substantial humanistic and economic burdens due to the comprehensive care requirements of children with EEs²⁻⁴
- Concerns about the emotional impact on siblings of a brother/sister with an EE were reported by 74% (114/154) of caregivers responding to a recent survey²
- Quality of life of the entire family is affected, especially for siblings^{5,6}
- Impact on sibling mental health and social development may be under-recognized by the medical community

PURPOSE

- The Sibling Voices Survey evaluated the psychosocial impact of growing up with a sibling affected by Dravet syndrome (DS), Lennox-Gastaut syndrome (LGS), or other severe EEs

METHODS

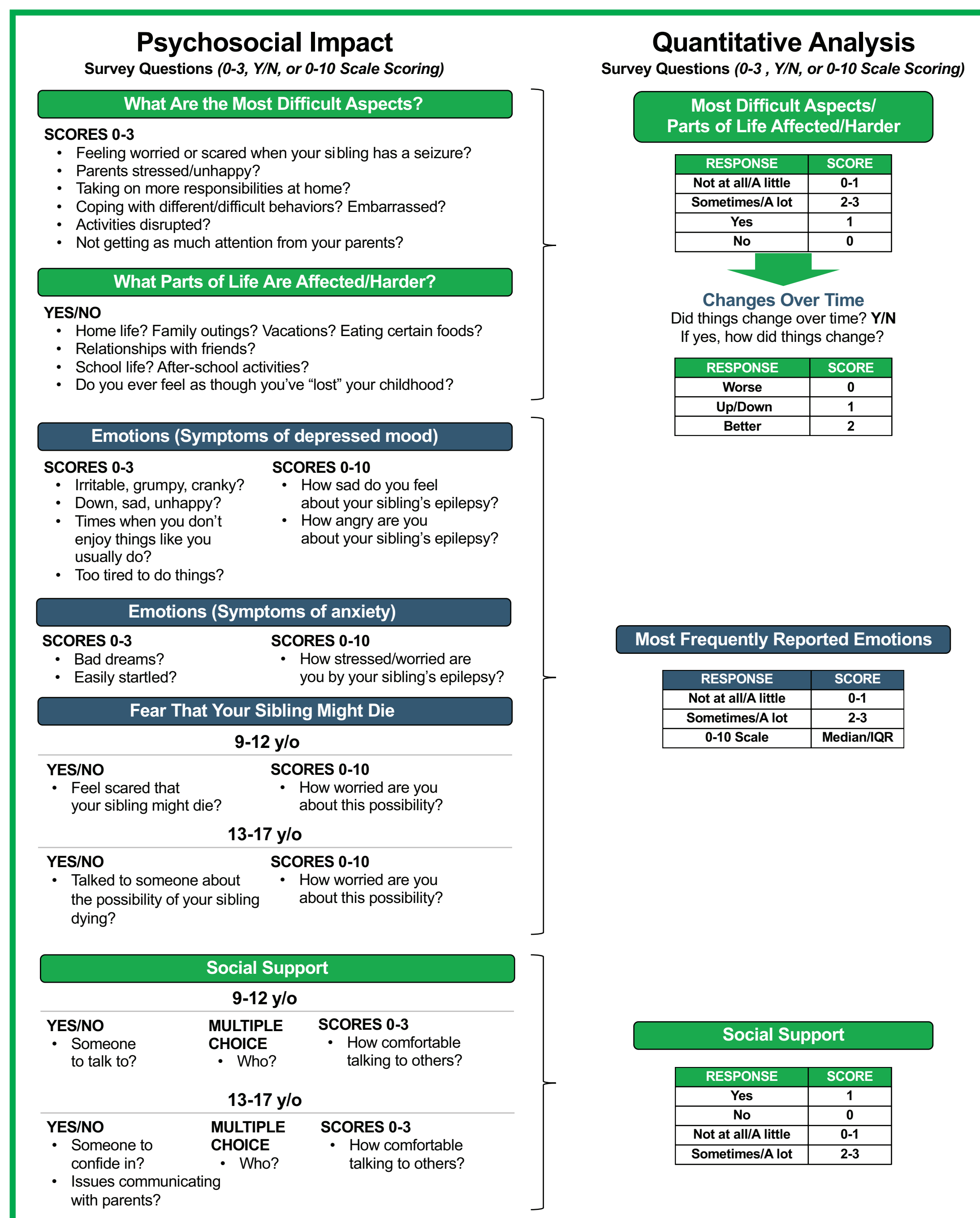
Survey Development and Recruitment

- Four online surveys were developed in consultation with DS and LGS patient communities and healthcare professionals (www.siblingsstudy.com)
 - Questions were refined based on beta testing in small groups of parents and siblings
 - Cohort-specific questions were designed for siblings 9-12 years old (y/o), 13-17 y/o, and ≥18 y/o and for parents (siblings ≥18 y/o and parents not reported here)
- Study protocol was approved by Western IRB (Puyallup, WA) on July 14, 2017
- Survey responses were solicited between July 19, 2017 and December 31, 2017 (5.5 months)

Survey Design and Analysis

- Survey questions were designed to evaluate psychosocial parameters (Figure 1)
- Quantitative analysis was by visual analog scores

Figure 1. Survey Questions and Scoring Criteria



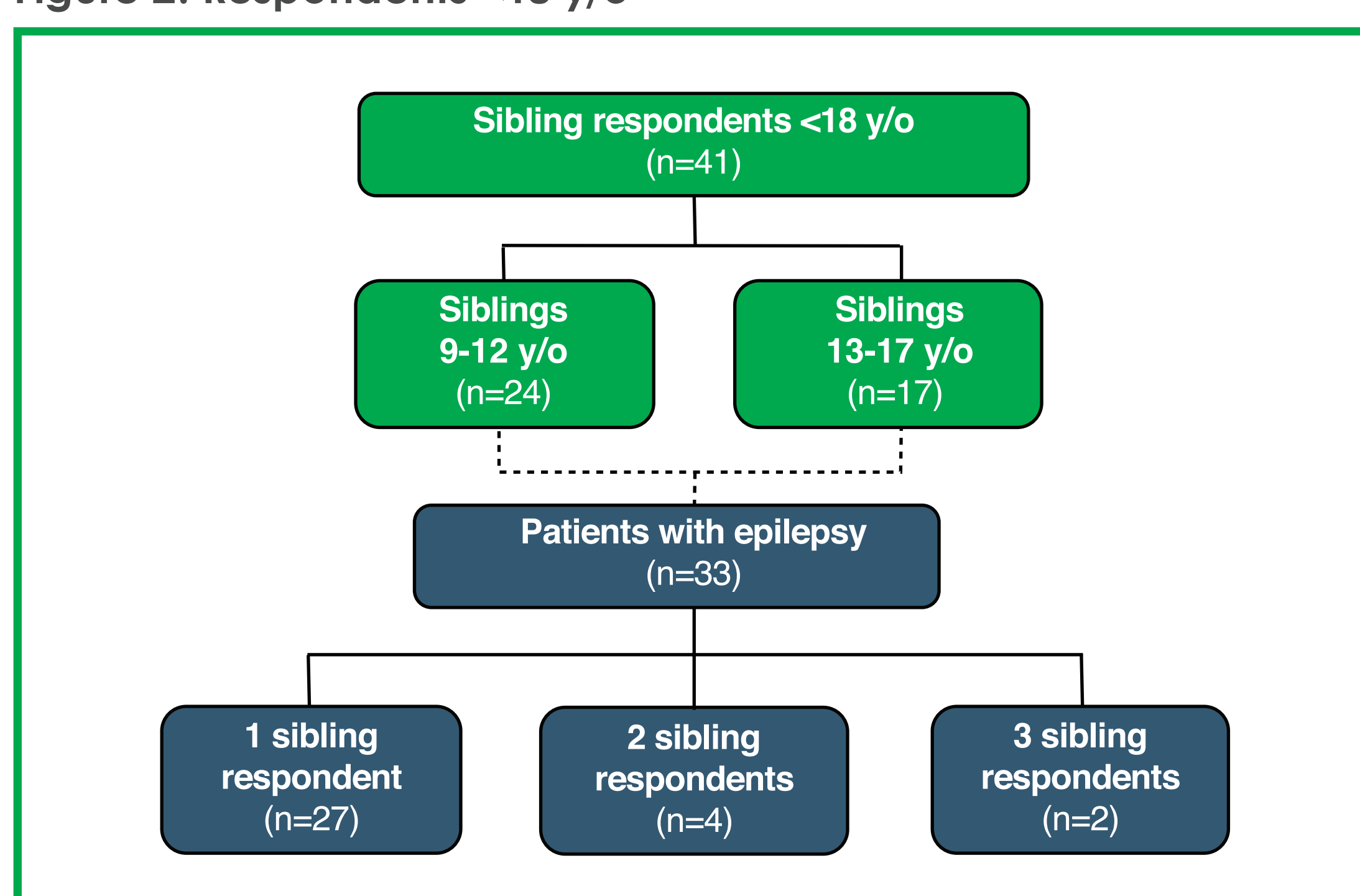
IQR, interquartile range; Y/N, yes/no; y/o, years old.

RESULTS

Sibling Voices Survey Respondents

- N=248 survey respondents; siblings <18 y/o (n=41) are shown in Figure 2

Figure 2. Respondents <18 y/o



Respondent Demographics

- Sibling <18 y/o respondents (Table 1)
 - 9-12 y/o (n=24), including 2 sets of twins and 1 set of triplets
 - 13-17 y/o (n=17)
 - Families (n=33), with 1-3 siblings per family
- Patients with epilepsy corresponding to sibling respondents (Table 2)
 - 75% (9-12 y/o cohort) and 46% (13-17 y/o cohort) of siblings with EE had Dravet syndrome

Table 1. Demographic Characteristics of Sibling Survey Respondents <18 y/o (n=41)

Characteristic	Age Group	
	9-12 y/o	13-17 y/o
n	24	17
Sex, n (%)		
Male	12 (50)	11 (65)
Female	12 (50)	6 (35)
Twin sibling, n (%)	2 (8)*	0 (0)
Age, years, mean (range)	10 (9-12)	14 (13-15)
School grade, mean (range)	5 (3-7)	9 (8-11)
Family size, mean (range)	5 (4-8)	5 (4-10)
No. of children/family, mean (range)	4 (2-6)	4 (2-8)

*Excludes a set of 3 triplet siblings who selected "no" for "twin sibling"; y/o, years old.

Table 2. Demographic Characteristics of EE Patients (n=33)

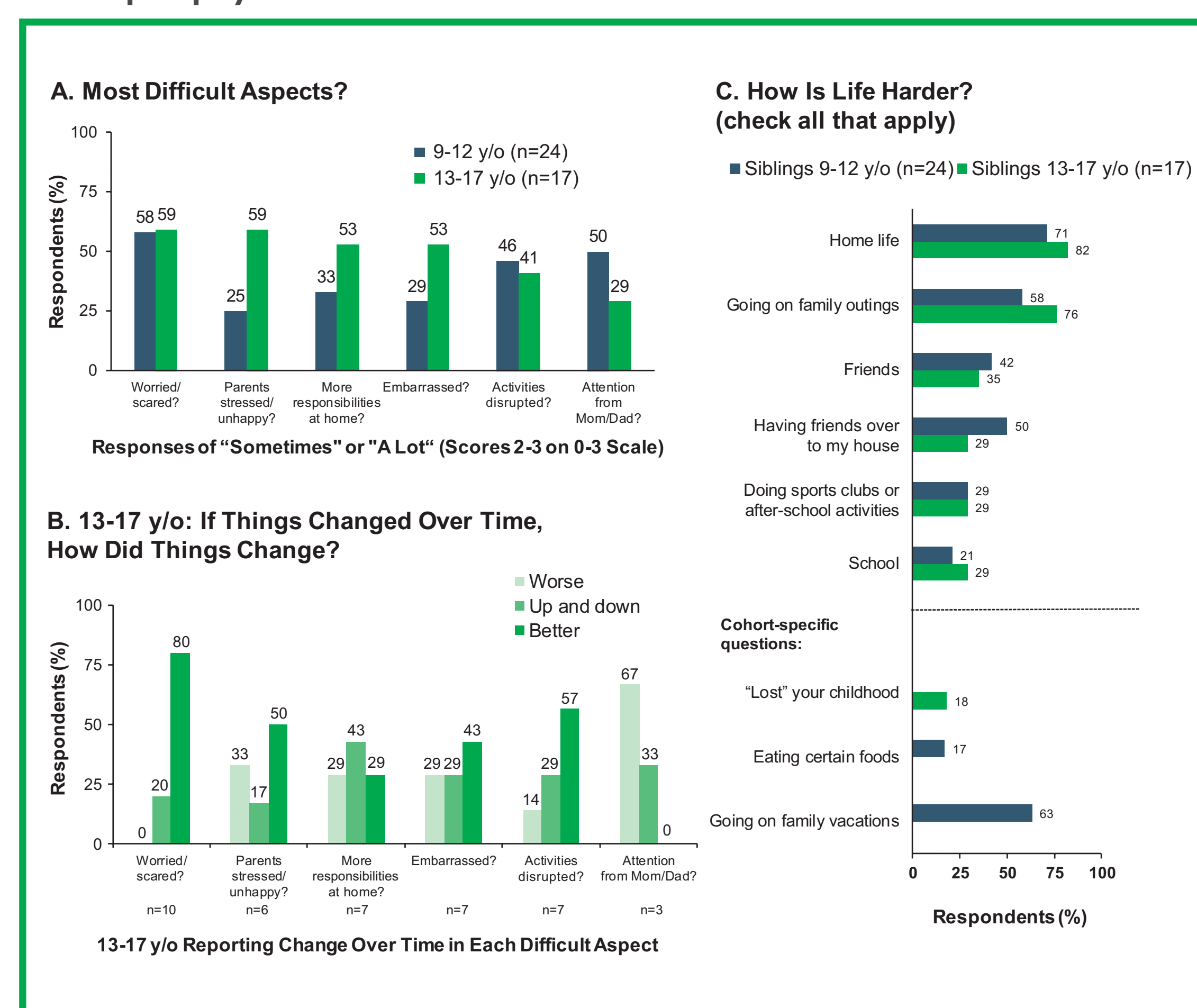
Characteristic	Sibling Age Group	
	9-12 y/o	13-17 y/o
Patients (n)	20	13
Sex, n (%)		
Male	7 (35)	5 (38)
Female	13 (65)	8 (62)
Age, years, mean (range)	7 (2-15)	8 (1-16)
Patient diagnosis, n (%)		
Dravet syndrome	15 (75)	6 (46)
Lennox-Gastaut syndrome	0 (0)	1 (8)
Other EE	5 (25)	6 (46)

EE, epileptic encephalopathy; y/o, years old.

Difficult Aspects

- The most difficult aspects of having a sibling with an EE differed among cohorts (Figure 3A)
 - Top concerns of 9-12 y/o: worried/scared; less parental attention; activities disrupted
 - Top concerns of 13-17 y/o: parental stress; worried/scared; more responsibilities at home; embarrassing behaviors
- 13-17 y/o siblings tended to perceive an improvement over time in all aspects except parental attention and responsibilities at home (Figure 3B)
- Difficulty with relationships with friends, home life, and school life were comparable among cohorts, with slightly more emphasis on friends in the younger cohort and slightly more emphasis on home/family and school in the older cohort (Figure 3C)

Figure 3. Most Difficult Aspects Experienced by Siblings of Patients With Epilepsy

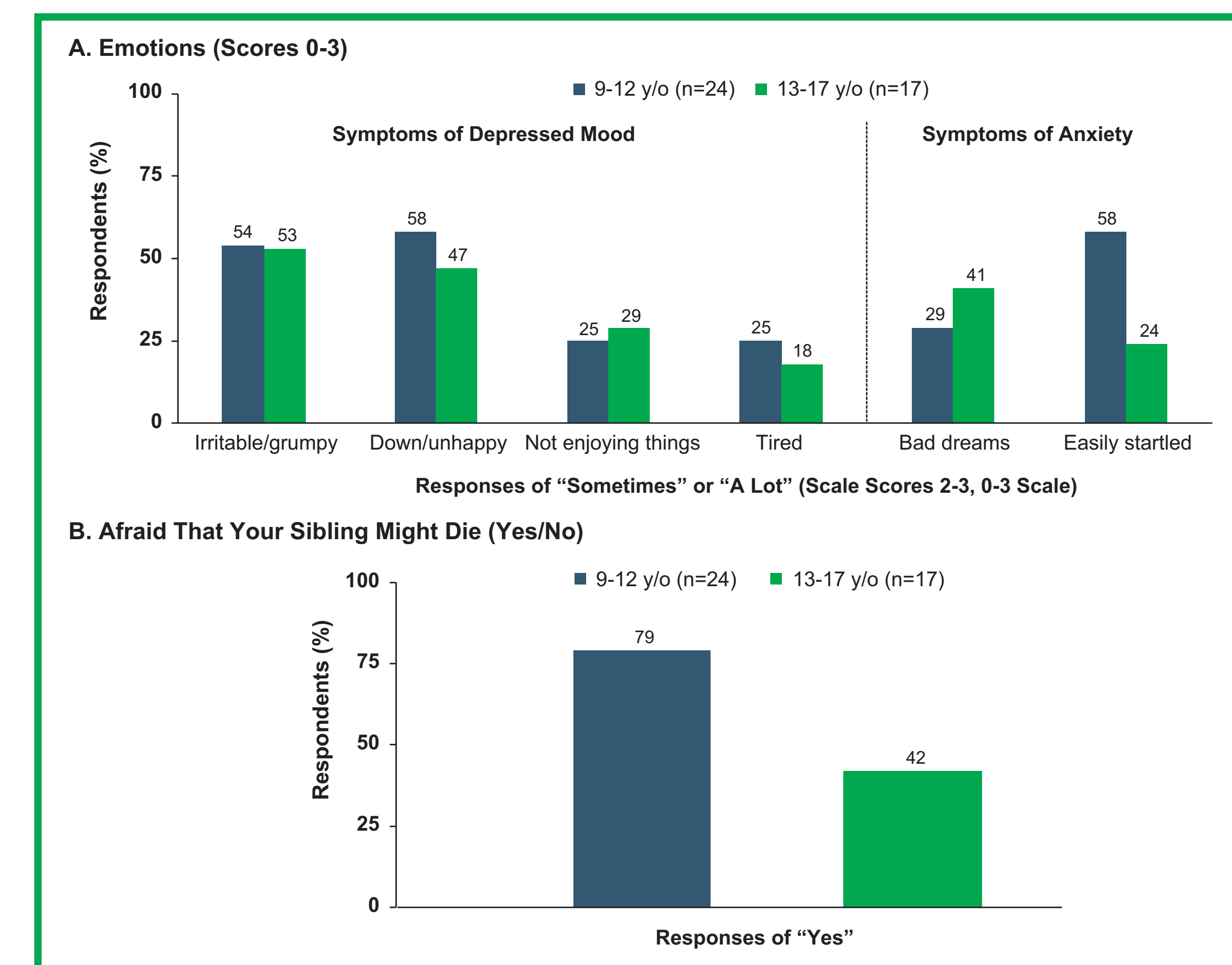


y/o, years old.

Emotions

- Emotions (symptoms of depressed mood or anxiety) varied among cohorts
 - Most frequent emotions reported by 9-12 y/o: easily startled, unhappiness, grumpiness (Figure 4A)
 - Most frequent emotions reported by 13-17 y/o: irritability, unhappiness, bad dreams (Figure 4A)
 - Greater degree of sadness and lower degree of anger than worry/fear or stress over sibling's diagnosis in both cohorts (median [IQR] on a 0-10 visual analog scale):
 - Sadness: 9-12 y/o, 6 [2-10]; 13-17 y/o, 5 [2-7]
 - Anger: 9-12 y/o, 1 [0-4]; 13-17 y/o, 2 [0-3]
 - Worry/Fear: 9-12 y/o, 4 [1-9]; 13-17 y/o, 3 [1-7]
 - Stress: 9-12 y/o, 3 [1-5]; 13-17 y/o, 3 [1-7]
- Cohort-specific questions addressing the fear that their sibling with epilepsy might die
 - 9-12 y/o: 79% (19/24) reported fearing their sibling might die (Figure 4B); of those responding "yes", 42% (8/19) reported thinking about this sometimes, often, or all the time
 - 13-17 y/o: 71% (12/17) reported that someone had discussed the possibility of their sibling dying; of those responding "yes," 42% (5/12) reported somewhat to extreme stress/worry (>5 on a 0-10 scale)

Figure 4. Emotional Self-Reports of Siblings With Epilepsy

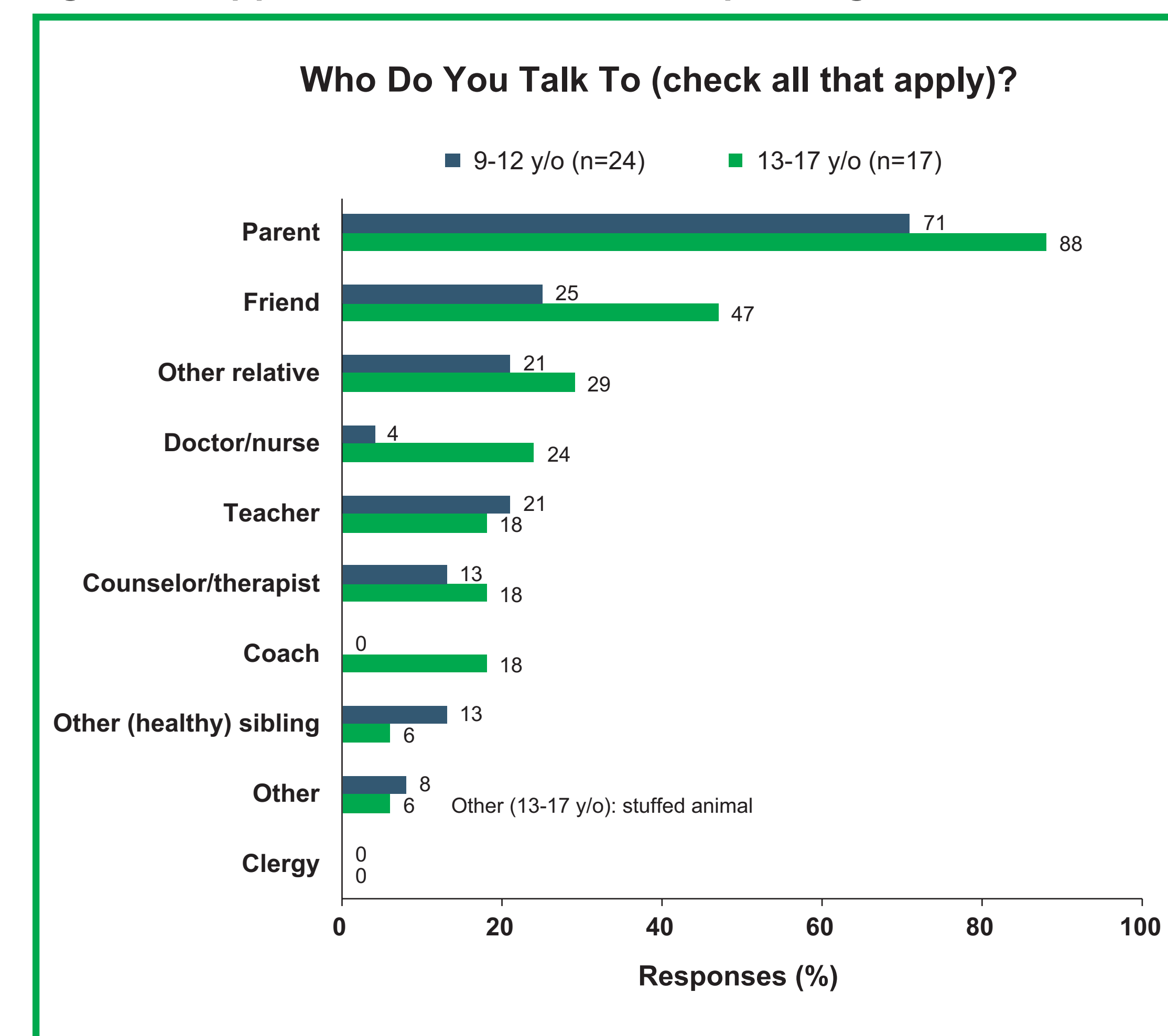


y/o, years old.

Social Support

- More 9-12 y/o (58%; 14/24) than 13-17 y/o (12%, 2/17) reported feeling uncomfortable talking about their sibling's diagnosis (scores 0-1 of 0-3)
- More 13-17 y/o (94%; 16/17) than 9-12 y/o (79%; 19/24) reported having someone to confide in or talk to about their sibling's diagnosis
- Siblings spoke with family members and friends most frequently (Figure 5)

Figure 5. Support Networks Described by Siblings



y/o, years old.

CONCLUSIONS

- The Sibling Voices Survey identified sibling-specific concerns about growing up with a brother/sister with epilepsy
- These results confirm and extend previously reported findings suggesting that caring for a child with DS, LGS, or other severe EE adversely affects the entire family unit²⁻⁴
- Supportive services for families with a child who has epilepsy could improve family psychosocial dynamics and functioning by addressing areas such as:
 - Stresses at home, school, and socially
 - Effects on emotional health such as insistent anxieties and fears or changes in mood
 - Social support networks most likely to be of help

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DISCLOSURE

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

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