

CDKL5 Caregiver Severity Scale (CCSA)

Participant Name: _____

Doctor Name: _____

For Study Personnel Use, Only:

Coordinator Code: _____

Date: _____

GENERAL QUESTIONS (2)

1. Full name of Parent/Guardian

Completing Form: _____

2. Date of Completion: _____ / _____ / _____
MM DD YYYY

TREATMENTS/THERAPIES QUESTIONS (4)

1. How many antiseizure medications is your child currently taking to control seizures? *(Please do not include any rescue medications, Vagal Nerve Stimulator, antiseizure medication used for sleep or ketogenic diet)*

- None
- One antiseizure medication
- Two antiseizure medications
- Three antiseizure medications
- Four antiseizure medications
- Five or more antiseizure medications

2. Is your child currently on the ketogenic, modified Atkins or low glycemic diet for seizure control?

- Yes
- No

Treatments/Therapies (cont.)

3. How often did your child receive the following therapies on average in the past 6 months?

	None	Less than 4 times per month	4 times per month (weekly)	5-8 times per month (up to twice weekly)	9 or more times per month (up to three or more times per week)
Physical Therapy					
Speech Therapy					
Augmentive Communication Therapy (Please report even if it was administered as a part of Speech Therapy) ACT is a therapy that uses other devices to help non-verbal children communicate. (ex. iPads, Tobii, switches, and picture boards)					
Vision Therapy					
Occupational Therapy					
Feeding Therapy					
Music Therapy					
Other Therapy 1 _____ _____					
Other Therapy 2 _____ _____					
Other Therapy 3 _____ _____					

Treatments/Therapies (cont.)

4. Thinking about the last 30 days, how helpful would you say have your child's therapies have been altogether? (if therapies have been utilized)

- Very helpful, my child is definitely gaining skills
- Quite helpful, my child is maintaining all skills and might be gaining slowly some skills
- Somewhat helpful, my child is maintaining all skills
- Minimally helpful, my child is mostly maintaining skills but might be losing some skills
- Not at all helpful, my child is definitely losing some skills

EPILEPSY QUESTIONS (4)

1. Has your child ever had a seizure in their life?

- Yes
- No

2. In the past 30 days, what has been your child's longest seizure-free period?

- My child has not had any seizure in the past 30 days
- Seizure free for greater than 2 weeks, but less than 30 days
- Seizure free for greater than 1 week, but less than 2 weeks
- Seizure free for greater than 1 day, but less than 1 week
- Has had seizures every day for the past 30 days

3. In the past 30 days, how often have your child's seizures become disruptive to his/her normal routine? This includes ALL seizure types.

- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half of the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

Epilepsy (cont.)

Convulsive Seizures: Tonic, Tonic-clonic, or Clusters of drops or jerks or spasms that are disruptive and bothersome to patient or family. Seizures with multiple phases should be counted as a single seizure. (A cluster of seizures would be seizures occurring one right after another without returning to the child's typical function) **Non-Convulsive Seizures:** Please provide your best estimate. Include ONLY for NON-CONVULSIVE: Absences (unresponsiveness not interrupted by touch) and Auras (pre-seizure activity) that do not lead to a convulsion, **Isolated Epileptic Spasm, Myoclonic Jerk or Single Drop** that DO NOT cluster: You would count single seizures, not part of clusters or seizures with multiple phases. Each seizure should last a few seconds or less. **Prolonged Seizures:** defined in this survey as continuous convulsive seizure lasting more than 5 minutes without return to their typical function between seizures.

4. In the past 30 days how many days has your child experienced one or more of the following seizures?

	N/A - Never had a seizure	No days in the last 30 days	Only a few days in the last 30 days	Fewer than half of the days in the last 30 days	More than half of the days in the last 30 days	Nearly every day in the last 30 days	Every day in the last 30 days
--	---------------------------------	-----------------------------------	--	---	--	---	-------------------------------------

Convulsive Seizures							
Non-Convulsive Seizures							
Isolated Epileptic Spasm or Myoclonic Jerk or Single Drop							
Prolonged Seizures							

4a. In the past 30 days, on the days your child had seizures, how many individual seizures did your child have on average per day?

Convulsive Seizures: Tonic, Tonic-clonic, or Clusters of drops or jerks or spasms that are disruptive and bothersome to patient or family. Seizures with multiple phases should be counted as a single seizure. (A cluster of seizures would be seizures one right after another without returning to the child's typical function)

- 1
- 2-5
- 6-10
- 11-15
- Too many to count

COMPLETE ONLY IF
CONVULSIVE
SEIZURES

ALERTNESS/INTERACTION QUESTIONS (6)

1. How alert has your child been over the last 30 days?

- My child has been very alert
- My child is alert most of the time
- My child is alert some of the time
- My child rarely alert

2. How interactive with family members or care providers has your child been over the last 30 days?

- My child has been very interactive
- My child is interactive most of the time
- My child is interactive some of the time
- My child is rarely interactive
- My child is never interactive

3. How well has your child made eye contact with you in the last 30 days?

- Direct eye contact, for total duration of an interaction
- He/she makes direct or indirect eye contact, holds it for at least 10 seconds, but not the total duration of an interaction
- He/she makes eye contact, but is usually less than 10 seconds
- He/she makes eye contact, but out the corner of his/her eye
- He/she doesn't really seem to look at me

4. How often was your child so sleepy during the daytime that it was disruptive to their normal routine in the last 30 days?

- N/A - my child is never so sleepy during the day that it is disruptive
- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

Alertness/Interaction (cont.)

5. How well do you think your child has been able to see over the last 30 days (Use your best judgement)?

- My child has been able to see everything in their environment
- My child has sometimes struggled to see objects that are small, far away, or when there is poor contrast
- My child can only see bright colors with contrasting background colors well
- My child sees lights well, but not other objects
- My child sees very little

6. On average over the last 30 days, how many good days did your child have? A “good” day may be defined as a day that is minimally disrupted by seizures or a day where your child is engaged, interactive, and able to fully participate in activities of daily living, such as finishing therapies throughout the day.

- Every day has been good in the last 30 days
- Nearly every day in the last 30 days has been good
- More than half of the days in the last 30 days have been good
- Less than half of the days in the last 30 days have been good
- Only a few days in the last 30 days have been good
- No days in the last 30 days have been good

BEHAVIOR QUESTIONS (3)

1. How often did your child have spells of irritability, regardless of cause in the last 30 days?

- N/A - My child has never been irritable
- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half of the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

Behavior (cont.)

COMPLETE ONLY IF
**ANY IRRITABILITY
OCCURRED**

1a. How easy or difficult was it, in general, to console your child during those spells of irritability?

- Very easy, my child calms down on his/her own
- Slightly easy, my child is pretty easy to calm down
- Moderately, sometimes things help, sometimes they don't
- Slightly difficult, it can be pretty hard to calm my child down
- Very difficult, inconsolable and nothing I do seems to help

2. How often has your child self-harmed him/herself, regardless of intent or purpose in the last 30 days?

- N/A - My child has never harmed him/herself
- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half of the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

3. How often has your child been aggressive, regardless of intent or purpose, in the last 30 days?

- N/A - My child has never been aggressive
- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half of the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

FEEDING QUESTIONS (2)

1. What texture of food was your child able to eat in the last 30 days?

- My child can eat and chew food of any texture
- Food have to be coarsely chopped
- Food have to be finely chopped
- Foods have to be pureed or mashed
- My child only gets mall tastes of food for pleasure
- My child does not take any food by mouth

2. Which answer most accurately describes your child's G-tube/GJ-tube status?

- * My child does not have a G-Tube/GJ-Tube
- * My child has a G-Tube/GJ-Tube but does not routinely use it for nutrition (used for meds or as backup)
- * My child gets their nutrition both through the G-Tube/GJ-Tube and eating by mouth
- All or most of my child's nutrition are provided through their G-Tube
- All of most of my child's nutrition are provided through their J-Tube
- My child receives nutrition through intravenous (IV) feeding (my child does not receive nutrition through their gut)

2a. How well was your child able to swallow in the last 30 days?

- Oral feeding with normal swallowing, no choking/gagging
- Oral feeding with occasional choking/gagging
- Oral feeding but it takes more than 30 minutes to eat meal or occasional choking/gagging possible
- Oral feeding with frequent choking/gagging
- Unable to safely swallow

**COMPLETE ONLY IF
YOU CHOSE A
STARRED RESPONSE
(*)**

OTHER CLINICAL FINDING QUESTIONS (12)

1. Did your child have signs of abnormal breathing while awake, such as breath-holding, deep breathing or rapid breathing that was not associated with seizures in the last 30 days?

- No
- Yes, occasionally
- Yes, daily
- Yes, daily and occasional cyanosis (blueness around mouth or face)
- Yes, daily and frequent cyanosis (blueness around mouth or face)

2. How often did your child grind their teeth during the day in the last 30 days?

- N/A - My child has never grinded their teeth or their teeth have not yet developed
- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half of the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

3. Which best describes your child's toileting habits in the last 30 days?

- My child is fully toilet trained
- My child is toilet trained during the day but still wears diapers during the night
- My child is toilet training and still wears diapers at all times but mostly uses the toilet
- My child is toilet training and still wears diapers at all times but rarely uses the toilet
- My child uses diapers only

4. Has your child had issues with reflux in the last 30 days?

- Unknown
- No
- Yes, but symptoms are controlled, no medications (just diet, etc)
- Yes, but symptoms are controlled on medications as needed
- Yes, but symptoms are controlled, on daily medications
- Yes, despite treatments this remains a frequent struggle

Other Clinical Findings (cont.)

5. In your opinion, how often has your child had a hard time falling asleep at night in the last 30 days?

- N/A - My child hasn't had a hard time falling asleep at night
- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half of the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

6. How often did your child wake up in the night after falling asleep in the last 30 days? (Please note: All children wake up during the night. (Please count only those times when your child cried or was disruptive enough to need your attention))

- N/A - My child has never woken up in the night after falling asleep
- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half of the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

7. When your child woke up during the night in the last 30 days, how long did it usually take to calm your child after they woke up and needed your attention?

- Less than 10 minutes, my child usually when right back to sleep
- 10-30 minutes
- 31-60 minutes
- About 1-2 hours
- More than 2 hours

Other Clinical Findings (cont.)

8. How often was your child awake during the night after falling asleep but did not need your attention in the last 30 days? (Please note: Please count only those times when your child was awake but not disruptive.)

- N/A - Child not monitored after I go to sleep
- No days in the last 30 days
- Only a few days in the last 30 days
- Fewer than half of the days in the last 30 days
- More than half of the days in the last 30 days
- Nearly every day in the last 30 days
- Every day in the last 30 days

9. Did your child have a reduced response to pain in the last 30 days?

- No, my child had a normal response to pain
- Yes, my child had a delayed response to minor pain
- Yes, my child had no response to minor pain or a delayed response to major pain
- Yes, my child had neither a response to minor pain or a delayed response to major pain

10. How often have you had to give a rescue medication for a single Prolonged Seizure or increased frequency of seizures in the last 30 days?

- N/A - No rescue medication given in the last 30 days
- Once in the last 30 days
- Twice in the last 30 days
- 3 times in the last 30 days
- 4 times in the last 30 days
- 5 or more times in the last 30 days

11. Did your child stay in the hospital overnight because of seizures in the last 30 days?

- Yes
- No

Other Clinical Findings (cont.)

12. Has your child had a serious bacterial infection requiring treatment or medicine, such as pneumonia or urinary tract infection in the last 30 days?

Yes

No

12a. What type of care did this infection require? If there was more than one infection in the last 30 days, please answer for the most infection. Please check all that apply.

Called my Primary Care Physician (PCP)

Visited my Primary Care Physician (PCP) or Urgent Care (UC)

Went to the Emergency Department (ED)

My child was admitted to the hospital

My child needed a stay in the Intensive Care Unit (ICU)

COMPLETE ONLY IF
YES TO #12