



**NEURONAL CEROID LIPOFUSCINOSIS (NCL) STUDY GROUP
UNIFIED BATTEN DISEASE RATING SCALE**

All items must be completed. Use U if information is Unavailable. Use N if Information is Not Applicable.

SUBJECT NO. INITIALS (First, Middle, Last) SITE NO.

DATE INFO OBTAINED (mm/dd/yyyy)

I. PHYSICAL ASSESSMENT

1. SPEECH CLARITY

1.

0 = normal

1 = unclear, no need to repeat

2 = must repeat to be understood

3 = mostly incomprehensible

4 = anarthria

**1A. ABNORMAL REPETITIVE
SPEECH SOUNDS**

1A.

0 = none

1 = sometimes

2 = most of the time

3 = constant

4 = anarthric

2. TONGUE PROTRUSION

2.

0 = maintains full protrusion for 10 seconds

1 = maintains full protrusion for more than 5 seconds

2 = maintains full protrusion for less than 5 seconds

3 = cannot fully protrude tongue

4 = cannot protrude tongue beyond lips

3. VISUAL ACUITY

3.

0 = normal

1 = mildly impaired

2 = finger counting only

3 = light/dark perception

4 = blind

4. PASSIVE MOTION-ARMS Right Left
4a. 4b.

0 = normal tone/full range

1 = mildly increased tone/full range

2 = moderately increased tone/full range

3 = markedly increased tone/incomplete range

4 = minimal or no passive range of motion

5. PASSIVE MOTION-LEGS Right Left
5a. 5b.

0 = normal tone/full range

1 = mildly increased tone/full range

2 = moderately increased tone/full range

3 = markedly increased tone/incomplete range

4 = minimal or no passive range of motion

6. PASSIVE MOTION-NECK 6.

0 = normal tone/full range

1 = mildly increased tone/full range

2 = moderately increased tone/full range

3 = markedly increased tone/incomplete range

4 = minimal or no passive range of motion

7. POWER-ARMS Right Left
7a. 7b.

0 = full power

1 = pronator drift/mild weakness

2 = moderate weakness/able to actively resist

3 = severe weakness/able to overcome gravity

4 = paralysis/unable to overcome gravity



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8. POWER-LEGS Right Left
 8a. 8b.

- 0 = full power
- 1 = mild weakness
- 2 = moderate weakness/able to actively resist
- 3 = severe weakness/able to overcome gravity
- 4 = paralysis/unable to overcome gravity

9. HAND TAPS (table or thigh) Right Left
 9a. 9b.

- 0 = normal for age
- 1 = mild slowing and or reduction in amplitude
- 2 = moderately impaired; definite and early fatiguing; may have occasional arrests in movement
- 3 = severely impaired; frequent hesitation in initiating movements or arrests in ongoing movements
- 4 = cannot perform the task

10. MAXIMAL DYSTONIA (UPPER EXTREMITIES) Right Left
 10a. 10b.

- 0 = absent
- 1 = slight/intermittent
- 2 = mild/persistent or moderate/intermittent
- 3 = moderate/persistent or marked/intermittent
- 4 = marked/prolonged

11. NORMAL SPONTANEOUS MOVEMENTS 11.

- 0 = normal
- 1 = minimally reduced (could be normal)
- 2 = mildly diminished
- 3 = moderately diminished
- 4 = markedly diminished, or absent

12. GAIT 12.

- 0 = normal gait
- 1 = small steps and/or slow
- 2 = walks with difficulty
- 3 = requires assistance

4 = cannot walk

13. RETROPULSION PULL TEST 13.

- 0 = normal
- 1 = recovers spontaneously, may take a step back
- 2 = would fall if not caught
- 3 = tends to fall spontaneously
- 4 = cannot stand

14. HEEL STOMPING Right Left
 14a. 14b.

- 0 = normal
- 1 = mild slowing or reduced amplitude
- 2 = definite and early fatiguing or occasional arrests in movement
- 3 = frequent hesitation in initiating movement or arrests in ongoing movement
- 4 = cannot perform task

15. MOTOR TICS OR STEREOTYPIES 15.

- 0 = absent
- 1 = rare
- 2 = mild/common or moderate/intermittent
- 3 = moderate/common
- 4 = marked/prolonged

16. MYOCLONUS 16.

- 0 = absent
- 1 = rare
- 2 = mild/common or moderate/intermittent
- 3 = moderate/common
- 4 = marked/prolonged

17. REST TREMOR 17.

- 0 = absent
- 1 = mild amplitude and infrequently present
- 2 = mild amplitude and usually present
- 3 = moderate amplitude and usually present
- 4 = marked amplitude and usually present



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18. TREMOR WITH MAINTAINED POSTURE OR ACTION 18.

- 0 = absent
- 1 = mild amplitude with action
- 2 = moderate amplitude with action
- 3 = moderate amplitude with action or sustention
- 4 = marked amplitude with action or sustention

19. DYSMETRIA (Finger-to-Nose) 19.

- 0 = normal
- 1 = mild irregularity
- 2 = moderate irregularity
- 3 = marked irregularity
- 4 = unable to hit target

20. APPENDICULAR CHOREA 20.

- 0 = absent
- 1 = slight/intermittent
- 2 = mild/common or moderate intermittent
- 3 = moderate/common
- 4 = marked prolonged

21. WEIGHT(lbs.) 21. .

22. MOTOR EXAMINER 22.

II. SEIZURE ASSESSMENT

23. GENERALIZED TONIC/CLONIC SEIZURES: AVERAGE FREQUENCY 23.

- 0 = none
- 1 = fewer than one per 6 months
- 2 = between one per 3 months and one per 6 months
- 3 = between one per month and one per 3 months
- 4 = between one per week and one per month
- 5 = between one per day and one per week
- 6 = more than one per day

24. GENERALIZED TONIC/CLONIC SEIZURES: POST-ICTAL PERIOD 24.

- 0 = none/not-applicable
- 1 = less than 1 minute
- 2 = between 1 and 10 minutes
- 3 = between 10 minutes and 1 hour
- 4 = between 1 hour and 3 hours
- 5 = more than 3 hours

25. ATONIC SEIZURES: AVERAGE FREQUENCY 25.

- 0 = none
- 1 = fewer than one per 6 months
- 2 = between one per 3 months and one per 6 months
- 3 = between one per month and one per 3 months
- 4 = between one per week and one per month
- 5 = between one per day and one per week
- 6 = more than one per day

26. MYOCLONIC SEIZURES: AVERAGE FREQUENCY 26.

- 0 = none
- 1 = fewer than one per 6 months
- 2 = between one per 3 months and one per 6 months
- 3 = between one per month and one per 3 months
- 4 = between one per week and one per month
- 5 = between one per day and one per week
- 6 = more than one per day

27. COMPLEX PARTIAL SEIZURES WITHOUT GENERALIZATION AND/OR ABSENCE: AVERAGE FREQUENCY 27.

- 0 = none
- 1 = fewer than one per 6 months
- 2 = between one per 3 months and one per 6 months
- 3 = between one per month and one per 3 months
- 4 = between one per week and one per month
- 5 = between one per day and one per week
- 6 = more than one per day



All items must be completed. Use U if information is Unavailable. Use N if Information is Not Applicable.

28. COMPLEX PARTIAL SEIZURES WITHOUT GENERALIZATION: POST-ICTAL PERIOD 28.

- 0 = none/not-applicable
- 1 = less than 1 minute
- 2 = between 1 and 10 minutes
- 3 = between 10 minutes and 1 hour
- 4 = between 1 hour and 3 hours
- 5 = more than 3 hours

29. SIMPLE PARTIAL SEIZURES: AVERAGE FREQUENCY 29.

- 0 = none
- 1 = fewer than one per 6 months
- 2 = between one per 3 months and one per 6 months
- 3 = between one per month and one per 3 months
- 4 = between one per week and one per month
- 5 = between one per day and one per week
- 6 = more than one per day

30. SIMPLE PARTIAL SEIZURES: AVERAGE DURATION OF EVENT 30.

- 0 = none/not-applicable
- 1 = less than 1 minute
- 2 = between 1 and 10 minutes
- 3 = between 10 minutes and 1 hour
- 4 = between 1 hour and 3 hours
- 5 = more than 3 hours

31. FREQUENCY OF INJURY RELATED TO SEIZURES 31.

- 0 = never
- 1 = sometimes
- 2 = usually
- 3 = always

32. MAXIMAL LEVEL of CARE FOR SEIZURE COMPLICATIONS (due to any seizure type/past 6 months) 32.

- 0 = none/no care required
- 1 = first aid at home
- 2 = paramedic called

3 = emergency department visit

33. HOSPITALIZATION REQUIRED FOR TREATMENT OF SEIZURES (due to any seizure type/past 6 months) 33.

- 0 = none/not applicable
- 1 = once
- 2 = more than once

34. ANTICONVULSANT ADJUSTMENT REQUIRED TO CONTROL SEIZURES IN PAST MONTH 34.
(1 = Yes, 2 = No)

35. SEIZURE ASSESSOR 35.

III. BEHAVIORAL ASSESSMENT (past month)

36. SAD MOOD
36a. Frequency 36b. Severity

- | | |
|-------------------|--------------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |

37. APATHY
37a. Frequency 37b. Severity

- | | |
|-------------------|--------------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |

38. ANXIETY
38a. Frequency 38b. Severity

- | | |
|-------------------|--------------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |



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39. AGGRESSION TOWARD OTHERS

39a. Frequency 39b. Severity

- | | |
|-------------------|--------------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |

40. AGGRESSION TOWARD SELF

40a. Frequency 40b. Severity

- | | |
|-------------------|--------------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |

41. STEREOTYPED/REPETITIVE BEHAVIOR

41a. Frequency 41b. Severity

- | | |
|-------------------|--------------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |

42. COMPULSIONS

42a. Frequency 42b. Severity

- | | |
|-------------------|--------------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |

43. AUDITORY HALLUCINATIONS

43a. Frequency 43b. Severity

- | | |
|-------------------|--------------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |

44. OBSESSIONS

44a. Frequency 44b. Severity

- | | |
|---------------|----------|
| 0 = never | 0 = none |
| 1 = sometimes | 1 = mild |

- | | |
|-------------------|--------------|
| 2 = frequent | 2 = moderate |
| 3 = almost always | 3 = severe |

45. MEDICATION REQUIRED FOR BEHAVIOR 45.
(1 = Yes, 2 = No)

46. BEHAVIORAL ASSESSOR 46.

CONTINUE ON NEXT PAGE



All items must be completed. Use U if information is Unavailable. Use N if Information is Not Applicable.

IV. CAPABILITY ASSESSMENT ASSUMING NORMAL VISION (answer as if child's vision were normal)

47. SCHOOL 47.

- 0 = unable to attend special needs classroom
- 1 = requires special needs classroom
- 2 = marginal ability in mainstream classroom
- 3 = normal ability in mainstream classroom

48. CHORES 48.

- 0 = unable to do even simple chores
- 1 = able to do simple chores with help
- 2 = able to do simple chores independently
- 3 = able to do all age appropriate chores independently

49. PLAY 49.

- 0 = unable to play even simple games
- 1 = able to play simple games with help
- 2 = able to play simple games independently
- 3 = able to play age appropriate games independently

50. ADL 50.

- 0 = total care
- 1 = gross tasks only
- 2 = minimal impairment
- 3 = normal

51. CARE LEVEL 51.

- 0 = full time skilled nursing
- 1 = chronic care at home
- 2 = home

V. CAPABILITY ASSESSMENT GIVEN ACTUAL VISION

52. SCHOOL 52.

- 0 = unable to attend special needs classroom
- 1 = requires special needs classroom
- 2 = marginal ability in mainstream classroom
- 3 = normal ability in mainstream classroom

53. CHORES 53.

- 0 = unable to do even simple chores
- 1 = able to do simple chores with help
- 2 = able to do simple chores independently
- 3 = able to do all age appropriate chores independently

54. PLAY 54.

- 0 = unable to play even simple games
- 1 = able to play simple games with help
- 2 = able to play simple games independently
- 3 = able to play age appropriate games independently

55. ADL 55.

- 0 = total care
- 1 = gross tasks only
- 2 = minimal impairment
- 3 = normal

56. CARE LEVEL 56.

- 0 = full time skilled nursing
- 1 = chronic care at home
- 2 = home

57. CAPABILITY ASSESSOR 57.



All items must be completed. Use U if information is Unavailable. Use N if Information is Not Applicable.

VI. NCL HISTORY (TO BE COMPLETED BY FIRST RATER)

Instructions: For each symptom, ask if child has experienced the symptom and if so, obtain approximate age of onset in years and months (e.g., age 4 1 / 2 should be coded as 04 for years and 06 for months). For each symptom reported as experienced, the rater should rank in order of onset beginning with 1. If child has not experienced symptom, code as N for not applicable.

Symptom	Experienced Symptom 0 = No, 1 = Yes	Onset Age		Rater Ranking Order of Onset (1 = first, 8 = last)
		Years	Months	
58a. Loss of vision	58a. <input type="checkbox"/>	58a1. <input type="text"/> <input type="text"/>	58a2. <input type="text"/> <input type="text"/>	58a3. <input type="checkbox"/>
58b. Motor difficulties	58b. <input type="checkbox"/>	58b1. <input type="text"/> <input type="text"/>	58b2. <input type="text"/> <input type="text"/>	58b3. <input type="checkbox"/>
58c. Cognitive difficulties	58c. <input type="checkbox"/>	58c1. <input type="text"/> <input type="text"/>	58c2. <input type="text"/> <input type="text"/>	58c3. <input type="checkbox"/>
58d. Behavioral difficulties	58d. <input type="checkbox"/>	58d1. <input type="text"/> <input type="text"/>	58d2. <input type="text"/> <input type="text"/>	58d3. <input type="checkbox"/>
58e. Seizures	58e. <input type="checkbox"/>	58e1. <input type="text"/> <input type="text"/>	58e2. <input type="text"/> <input type="text"/>	58e3. <input type="checkbox"/>
58f. Weight loss/feeding difficulties	58f. <input type="checkbox"/>	58f1. <input type="text"/> <input type="text"/>	58f2. <input type="text"/> <input type="text"/>	58f3. <input type="checkbox"/>
58g. Sleep disturbance	58g. <input type="checkbox"/>	58g1. <input type="text"/> <input type="text"/>	58g2. <input type="text"/> <input type="text"/>	58g3. <input type="checkbox"/>
58h. Other (Specify) _____ _____ _____	58h. <input type="checkbox"/>	58h1. <input type="text"/> <input type="text"/>	58h2. <input type="text"/> <input type="text"/>	58h3. <input type="checkbox"/>

59. COMMENTS _____

60. NCL HISTORY RATER 60.

VII. CLINICAL SUMMARY

61. ASSESSOR'S GLOBAL IMPRESSION OF CHANGE 61.

SINCE LAST ASSESSMENT

- 1 = much better
- 2 = somewhat better
- 3 = about the same
- 4 = somewhat worse
- 5 = much worse
- N = not applicable (never seen before)



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62. CLINICAL GLOBAL IMPRESSION – SEVERITY OF SEIZURES 62.

1 = none
 2 = minimal
 3 = mild
 4 = moderate
 5 = severe

63. CLINICAL GLOBAL IMPRESSION – COGNITIVE FUNCTION 63.

1 = no impairment
 2 = minimally impaired
 3 = mildly impaired
 4 = moderately impaired
 5 = severely impaired

64. CLINICAL GLOBAL IMPRESSION – BEHAVIOR 64.

1 = no impairment
 2 = minimally impaired
 3 = mildly impaired
 4 = moderately impaired
 5 = severely impaired

65. CLINICAL GLOBAL IMPRESSION – MOOD 65.

1 = no impairment
 2 = minimal distress
 3 = mild distress
 4 = moderate distress
 5 = severe distress

66. CLINICAL GLOBAL IMPRESSION – MOTOR 66.

1 = no impairment
 2 = minimally impaired
 3 = mildly impaired
 4 = moderately impaired
 5 = severely impaired

67. CLINICAL GLOBAL IMPRESSION – OVERALL 67.

1 = no impairment
 2 = minimally impaired
 3 = mildly impaired
 4 = moderately impaired
 5 = severely impaired

68. CLINICAL SUMMARY RATER 68.