# Unified Batten Disease Rating Scale

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

<table>
<thead>
<tr>
<th>SUBJECT NO.</th>
<th>INITIALS (First, Middle, Last)</th>
<th>SITE NO.</th>
</tr>
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<tbody>
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<thead>
<tr>
<th>DATE INFO OBTAINED (mm/dd/yyyy)</th>
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</table>

## 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

4. 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

5. 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

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

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

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

10. MAXIMAL DYSTONIA
    (UPPER EXTREMITIES)
    0 = absent
    1 = slight/intermittent
    2 = mild/persistent or moderate/intermittent
    3 = moderate/persistent or marked/intermittent
    4 = marked/prolonged
    Right 10a.  Left 10b.

11. NORMAL SPONTANEOUS MOVEMENTS
    0 = normal
    1 = minimally reduced (could be normal)
    2 = mildly diminished
    3 = moderately diminished
    4 = markedly diminished, or absent

12. GAIT
    0 = normal gait
    1 = small steps and/or slow
    2 = walks with difficulty
    3 = requires assistance
    4 = cannot walk

13. RETROPULSION PULL TEST
    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
    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
    Right 14a.  Left 14b.

15. MOTOR TICS OR STEREOTYPIES
    0 = absent
    1 = rare
    2 = mild/common or moderate/intermittent
    3 = moderate/common
    4 = marked/prolonged

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

17. REST TREMOR
    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
18. TREMOR WITH MAINTAINED POSTURE OR ACTION

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

19. DYSMETRIA (Finger-to-Nose)

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

20. APPENDICULAR CHOREA

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

21. WEIGHT (lbs.)

22. MOTOR EXAMINER

II. SEIZURE ASSESSMENT

23. GENERALIZED TONIC/CLONIC SEIZURES: AVERAGE FREQUENCY

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

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

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

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

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

   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

   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

   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

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

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

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

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

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

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

35. SEIZURE ASSESSOR
   III. BEHAVIORAL ASSESSMENT (past month)

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

   0 = never
   1 = sometimes
   2 = frequent
   3 = almost always

   0 = none
   1 = mild
   2 = moderate
   3 = severe

37. APATHY
   37a. Frequency 37b. Severity

   0 = never
   1 = sometimes
   2 = frequent
   3 = almost always

   0 = none
   1 = mild
   2 = moderate
   3 = severe

38. ANXIETY
   38a. Frequency 38b. Severity

   0 = never
   1 = sometimes
   2 = frequent
   3 = almost always

   0 = none
   1 = mild
   2 = moderate
   3 = severe
All items must be completed. Use U if information is Unavailable. Use N if Information is Not Applicable.

<table>
<thead>
<tr>
<th>39. AGGRESSION TOWARD OTHERS</th>
<th>40. AGGRESSION TOWARD SELF</th>
<th>41. STEREOTYPED/REPETITIVE BEHAVIOR</th>
<th>42. COMPULSIONS</th>
<th>43. AUDITORY HALLUCINATIONS</th>
<th>44. OBSESSIONS</th>
<th>45. MEDICATION REQUIRED FOR BEHAVIOR</th>
<th>46. BEHAVIORAL ASSESSOR</th>
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IV. CAPABILITY ASSESSMENT ASSUMING NORMAL VISION (answer as if child’s vision were normal)

47. SCHOOL
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
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
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
0 = total care
1 = gross tasks only
2 = minimal impairment
3 = normal

51. CARE LEVEL
0 = full time skilled nursing
1 = chronic care at home
2 = home

V. CAPABILITY ASSESSMENT GIVEN ACTUAL VISION

52. SCHOOL
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
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
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
0 = total care
1 = gross tasks only
2 = minimal impairment
3 = normal

56. CARE LEVEL
0 = full time skilled nursing
1 = chronic care at home
2 = home

57. CAPABILITY ASSESSOR
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.

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Experienced Symptom</th>
<th>Onset Age</th>
<th>Rater Ranking</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0 = No, 1 = Yes</td>
<td>Years</td>
<td>Months</td>
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<tr>
<td>58a. Loss of vision</td>
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<td>58b. Motor difficulties</td>
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<td>58c. Cognitive difficulties</td>
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<td>58d. Behavioral difficulties</td>
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<td>58e. Seizures</td>
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<td>58f. Weight loss/feeding difficulties</td>
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<td>58g. Sleep disturbance</td>
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<td>58h. Other</td>
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59. COMMENTS

60. NCL HISTORY RATER

VII. CLINICAL SUMMARY

61. ASSESSOR’S GLOBAL IMPRESSION OF CHANGE 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)
All items must be completed. Use U if information is Unavailable. Use N if Information is Not Applicable.

62. CLINICAL GLOBAL IMPRESSION – SEVERITY OF SEIZURES
   1 = none
   2 = minimal
   3 = mild
   4 = moderate
   5 = severe

63. CLINICAL GLOBAL IMPRESSION – COGNITIVE FUNCTION
   1 = no impairment
   2 = minimally impaired
   3 = mildly impaired
   4 = moderately impaired
   5 = severely impaired

64. CLINICAL GLOBAL IMPRESSION – BEHAVIOR
   1 = no impairment
   2 = minimally impaired
   3 = mildly impaired
   4 = moderately impaired
   5 = severely impaired

65. CLINICAL GLOBAL IMPRESSION – MOOD
   1 = no impairment
   2 = minimal distress
   3 = mild distress
   4 = moderate distress
   5 = severe distress

66. CLINICAL GLOBAL IMPRESSION – MOTOR
   1 = no impairment
   2 = minimally impaired
   3 = mildly impaired
   4 = moderately impaired
   5 = severely impaired

67. CLINICAL GLOBAL IMPRESSION – OVERALL
   1 = no impairment
   2 = minimally impaired
   3 = mildly impaired
   4 = moderately impaired
   5 = severely impaired

68. CLINICAL SUMMARY RATER