

INFANTILE PARKINSONISM - DYSTONIA RATING SCALE

for infants and young children

NON-MOTOR SYMPTOMS

Based on caregiver's report of the last 3 months

	Absent	Rarely (< once/month)	Sometimes (≤ once/week)	Often (>once/week)	Most days
<u>AUTONOMIC DYSFUNCTION</u>					
1. Excessive diaphoresis, temperature instability	0	1	2	3	4
2. Nasal congestion, stridor, audible breathing (nasal, laryngeal/pharyngeal)	0	1	2	3	4
3. Gastrointestinal dysmotility	0	1	2	3	4
4. Sleep disturbance	0	1	2	3	4
5. Other: myosis and ptosis, blood pressure abnormalities, heart rhythm abnormalities, etc	0	1	2	3	4

MOOD DYSFUNCTION

6. Emotional lability Disproportionate distress (e.g. crying, fear, fussing, agitation) with common activities	0	1	2	3	4
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MOTOR SYMPTOMS - SIGNS OF PARKINSONISM-DYSTONIA

OCULOGYRIC CRISES

Attacks of intermittent or sustained eye deviation, with or without associated abnormal posturing of the face, limbs, neck or trunk (not seizures). Based on caregiver's report of the last 3 months

7. **Severity of the crises** (if severity varies, rate the maximum severity)
- 0 None
 - 1 Eye deviation only
 - 2 Eye deviation plus dystonia of lower face and/or neck
 - 3 Eye deviation plus dystonia of trunk and/or limbs (with or without involvement of lower face and neck)
 - 4 Eye deviation plus body dystonia, associated with difficulty breathing, sweating, other systemic symptoms
8. **Duration of typical oculogyric episodes**
- 0 None
 - 1 Less than 1 hour
 - 2 1-2 hours
 - 3 2-4 hours
 - 4 > 4 hours
9. **Frequency of oculogyric episodes**
- 0 None
 - 1 Sporadic: less than once a month
 - 2 Mild: 1-3 times a month
 - 3 Moderate: at least once a week
 - 4 Severe: more than 3 times a week

BRADYKINESIA

10. GLOBAL SPONTANEITY OF MOVEMENT (BODY BRADYKINESIA)

Observation of spontaneous movements while lying down, shifting positions, sitting supported or unsupported, playing with toys, or rising from a chair (if applicable).

- 0 Absence of hypokinesia
- 1 Spontaneous movements mildly decreased in quantity
- 2 Spontaneous movements moderately decreased in quantity
- 3 Spontaneous movements markedly decreased. Patient may initiate a few purposeful movements
- 4 No movements. Patient may make occasional non purposeful stretches

11. FACIAL EXPRESSION (HYPOMIMIA)

Observation of facial appearance at rest and when crying or stimulated

- 0 Normal facial expression
- 1 Mild decreased facial expression: decreased blink rate, fewer movements around the mouth and eyes
- 2 Moderate decreased facial expression
- 3 Severe decreased facial expression: very reduced eye blinking, facial expression around the mouth and the eyes noted only during intense emotions. Lips may be parted most of the time
- 4 Expressionless: No eye-blink and no facial expression around the mouth and the eyes. Lips parted most of the time

12. UPPER LIMB MOVEMENTS: SPONTANEOUS

Observation of quantity of spontaneous movements of the upper limbs while lying down, sitting supported or unsupported, playing with toys. If there is asymmetry, score the more affected limb

- 0 Normal spontaneous upper limb movements
- 1 Spontaneous movements mildly decreased in quantity
- 2 Spontaneous movements moderately decreased in quantity
- 3 Spontaneous movements markedly decreased. Patient may initiate a few purposeful movements
- 4 No movements. Patient may make occasional non purposeful stretches

13. UPPER LIMB MOVEMENTS: VOLUNTARY

Evaluate latency of movement initiation and speed and amplitude of movement execution. In the case of young infants and/or uncooperative patients evaluate these characteristics during reaching, grasping and/or manipulating objects. If there is asymmetry, score the more affected limb.

- 0 Normal voluntary upper limb movements
- 1 Mild slowness of initiation and/or execution of voluntary movement
- 2 Moderate slowness of initiation and/or execution of voluntary movement
- 3 Severe slowness of initiation and/or execution of voluntary movement
- 4 No voluntary movements

14. LOWER LIMB MOVEMENTS: SPONTANEOUS

Observation of quantity of spontaneous movements of the lower limbs while lying down, shifting positions, sitting, playing with toys. If there is asymmetry, score the more affected limb

- 0 Normal spontaneous lower limb movements
- 1 Spontaneous movements mildly decreased in quantity
- 2 Spontaneous movements moderately decreased in quantity
- 3 Spontaneous movements markedly decreased. Patient may initiate a few purposeful movements
- 4 No movements. Patient may make occasional non purposeful stretches

TREMOR (rest, postural or action)

15. Distribution of tremor

Add 1 point for each body part where tremor is present

- 0 None
 - 1 Upper limb(s)
 - 1 Lower limb(s)
 - 1 Tongue
 - 1 Head
 - 1 Other
- Total: _____ /5

16. Severity of tremor

Score the most affected body part

- 0 No tremor
- 1 Minimal tremor
- 2 Mild: fine tremor visible only at close proximity
- 3 Moderate: tremor visible from several steps away
- 4 Severe: wide amplitude coarse tremor

RIGIDITY

17. Distribution of rigidity

Add 1 point for each body part where rigidity is present

- 0 None
 - 1 Upper limb(s)
 - 1 Lower limb(s)
 - 1 Neck
- Total: _____ /3

18. Severity of Rigidity (Score the most affected body part)

- 0 Normal: No rigidity
- 1 Slight: Full range of motion easily achieved OR in a cooperative patient rigidity detected only with activation maneuver
- 2 Mild: Full range of motion achieved with some effort OR in a cooperative patient mild rigidity detected without activation maneuver
- 3 Marked: Partial range of motion achieved with effort OR in a cooperative patient marked rigidity detected without activation maneuver
- 4 Severe: Little or no passive movement possible OR in a cooperative patient severe rigidity detected without activation maneuver and full range of motion is not achieved.

DYSTONIA

19. Facial, ocular and/or oromandibular dystonia (e.g. blepharospasm, upward eye deviation, grimacing, jaw opening or closing dystonia, tongue thrust)

- 0 None
- 1 Dystonia present when patient initiates a voluntary action
- 2 Dystonia present when sustaining a posture and/or triggered by manipulation during exam
- 3 Dystonia at rest (while patient is lying down or when sitting with support)
- 4 Dystonia at rest with fixed posture

20. Cervical, laryngeal and/or truncal dystonia (e.g. spasmodic dysphonic, torticollis, opisthotonus)

- 0 None
- 1 Dystonia present when patient initiates a voluntary action
- 2 Dystonia present when sustaining a posture and/or triggered by manipulation during exam
- 3 Dystonia at rest (while patient is lying down or when sitting with support)
- 4 Dystonia at rest with fixed posture

21. Dystonia of the upper limbs (if there is asymmetry, score the more affected limb)

- 0 None
- 1 Dystonia present when patient initiates a voluntary action
- 2 Dystonia present when sustaining a posture and/or triggered by manipulation during exam
- 3 Dystonia at rest (while patient is lying down or when sitting with support)
- 4 Dystonia at rest with fixed posture

22. Dystonia of the lower limbs (if there is asymmetry, score the more affected limb)

- 0 None
- 1 Dystonia present when patient initiates a voluntary action
- 2 Dystonia present when sustaining a posture and/or triggered by manipulation during exam
- 3 Dystonia at rest (while patient is lying down or when sitting with support)
- 4 Dystonia at rest with fixed posture











MOTOR SYMPTOMS - OTHER MOTOR SIGNS

AXIAL HYPOTONIA

23. Head lag

Pull infant to sit by the wrists or place the patient in the sitting position and observe head posture











- 0 Head in front of body when patients is pulled to sit OR straight in midline head while in sitting position
- 1 Patient lifts head in line with body when patient is pulled to sit OR while in sitting position patient has head tilted slightly to the side, backward or forward
- 2 Patient is able to lift head slightly when pulled to sit OR while in sitting position patient has head tilted moderately to the side, backward or forward
- 3 Patient tries to lift head but it drops back when pulled to sit OR while in sitting position patient has head tilted markedly to the side, backward or forward and is able to briefly move head towards midline
- 4 Head drops and stays back when patient is pulled to sit OR while in sitting position patient has head tilted markedly to the side, backward or forward and is unable to move head

	0	1	2	3	4
Pull to sit					
Sitting position					

24. Truncal hypotonia

Hold the child in ventral suspension or place the infant in a sitting position and observe the posture of the trunk

- 0 Back straight, head above the body on ventral suspension OR straight trunk while in sitting position
- 1 Back straight and head in line on ventral suspension OR trunk slightly curved or bent to side while patient in sitting position
- 2 Back slightly curved on ventral suspension OR trunk moderately curved or bent to side while patient in sitting position
- 3 Back curved more than the head on ventral suspension OR markedly curved or bent to side while patient in sitting position and briefly attempts to straighten back
- 4 Back and head curved on ventral suspension OR markedly curved or bent to side while patient in sitting position and is unable to straighten back

	0	1	2	3	4
Ventral suspension					
Sitting position					

25. MOTOR DELAY

- 0 Normal motor development
- 1 Mild motor delay and/or Gross Motor function classification system I or II
- 2 Moderate motor delay and/or Gross Motor function classification system III
- 3 Severe motor delay and/or Gross Motor function classification system IV
- 4 No or minimal motor acquisitions and/or Gross Motor function classification system V

DYSKINESIA

Is the patient currently taking L-dopa, dopaminergic agonists, MAO inhibitors, COMT inhibitors, anticholinergics? (circle one):

Yes

No

Dyskinesia refers to involuntary hyperkinetic jerky or writhing movements. The phenomenology may be consistent with chorea, athetosis, ballism, myoclonus, hyperekplexia. Stereotypies and tics are not included.

Specify the type of dyskinesia observed _____

26. Severity of dyskinesia

Consider amplitude of movements and their impact on function, including interference with purposeful movements, positioning, settling to sleep, or other activities of daily living.

- 0 None
- 1 Minimal and/or transient low amplitude hyperkinetic movements without clinical relevance
- 2 Mild low amplitude hyperkinetic movements that don't cause functional impairment or interference of routine care. Not associated with discomfort
- 3 Moderate low or high amplitude hyperkinetic movements causing some functional impairment or interference of routine care. Associated with some discomfort
- 4 Severe low or high amplitude hyperkinetic movements causing functional impairment or interference of routine care. Associated with prominent discomfort

27. Duration of dyskinesia during examination

- 0 None
- 1 Transient dyskinesia reported by parents, but absent at the time of the assessment
- 2 Mild dyskinesia: present less than 25% of the entire examination period
- 3 Moderate dyskinesia: present 26-75% of the entire examination period
- 4 Severe dyskinesia: present > 75% of the entire examination period

28. Body parts involved

Add score for every part of the body where dyskinesia is noted

- 0 None
- 1 Face
- 1 Arm(s)
- 1 Leg(s)
- 1 Trunk and/or neck
- 1 Other

Total: _____/5