INFANTILE PARKINSONISM-DYSTONIA RATING SCALE
INSTRUCTIONS FOR RATERS

The IPDRS has 3 parts (subscales):

1) Non-motor symptoms subscale,
2) Motor symptoms subscale
3) Dyskinesia subscale

1) NON-MOTOR SYMPTOMS SUBSCALE

This subscale evaluates the presence of manifestations suggestive of autonomic dysfunction and emotional lability. It integrates caregiver-reported information obtained through interviews, and may also include the rater's clinical observations and/or chart review.

A set of structured questions for the caregiver is provided below. Caregivers are asked to report whether any of the issues mentioned in the questions have occurred within the past 3 months If the answer is 'NO,' the item is scored as 0. If the answer is 'YES,' caregivers are then asked to indicate the frequency as indicated in the scoresheet.

Five groups of autonomic dysfunction manifestations are evaluated including: 1) sudomotor function and thermoregulation; 2) respiratory secretomotor function; 3) gastrointestinal motor function, 4) sleep and 5) other.

Questions to assess the presence of sudomotor function

- Does your child seem to sweat a lot?
- Has your child had unexplainable high fever?
- Has your child had a low temperature, below 96°F or 33.5°C?

Questions to assess respiratory secretomotor function

- Does your child experience nasal congestion or a stuffy nose even when he/she is not sick?"
- Does your child make noises when breathing?

Questions to assess gastrointestinal motor function:

- Does your child vomit after meals?
- Does your child have colic or abdominal pain?
- Does your child tend to be constipated? Does your child strain during bowel movement? Does your child use laxatives?
- Does your child have bouts of diarrhea?

Questions to assess Sleep

- Does your child have difficulty falling asleep?"

- Does your child wake up frequently during the night (more than once)?"
- Does your child feel sleepy during the day or need more than one nap?"
- Does your child hold his/her breath during sleep?

Other Symptoms:

- Does your child have episodes of low or high blood pressure?
- Does your child have abnormalities in cardiac rhythm, such as being too fast, too slow, or irregular?
- Has your child fainted after standing up from sitting or lying position? Has your child fainted while passing urine? Has your child fainted while coughing?

Emotional Lability:

- Does your child manifest disproportionate distress with common activities?
- Common activities may include: 1) anything out of the ordinary; 2) introduction to new stimuli (e.g., new food, new toy, new person); 3) motor activities such as therapy sessions, interacting with people outside the immediate family, or an outing."
- Disproportionate distress may be expressed through mood (e.g., crying, irritability, fussing, grimacing, verbalizations, agitation, fear) or motor activity (e.g., moving away, spitting out new food, pushing away a new toy)."

2) MOTOR SYMPTOMS SUBSCALE

This subscale evaluates the presence of the cardinal features of parkinsonism, as defined by the Movement Disorder Society, bradykinesia, tremor, and rigidity; as well as oculogyric crises, dystonia, axial hypotonia, and motor development. All items are scored by the clinician, with oculogyric crises being assessed based on the caregiver's report.

Bradykinesia

To score these features in infants and young children, observe spontaneous and voluntary movements. Spontaneous movements are scored for the total body, face, upper limbs, and lower limbs, while voluntary movements are scored for the upper limbs. If asymmetries are observed, the most affected body part is scored.

- <u>Rating of Spontaneous Movements of the Total Body, Upper Limbs, and Lower Limbs:</u>
  Observe spontaneous movements while the child is lying down, shifting positions, sitting (supported or unsupported), playing with toys, or rising from a chair (if applicable). Observe velocity, frequency, amplitude, smoothness and fluidity of movements taking into account the child’s developmental age
  Spontaneous movements are considered **mildly decreased** when there is a slight reduction in movement frequency, with movements being slower and/or smaller in amplitude. They are

considered **moderately decreased** when spontaneous movements are noticeably less frequent, slower, and/or reduced in amplitude. Spontaneous movements are considered **severely decreased** when they are markedly diminished in quantity, with the child producing only occasional purposeful actions. **No spontaneous movements** are observed when no movements are present, and only non-purposeful stretches may occur.

- Rating of facial expression: Observe facial expression at rest and when crying or stimulated. Observe eye-blink frequency, movements around the mouth and the eyes.
  Facial expression is considered **mildly decreased** when there is a mild reduction in facial expression, a slightly reduced blink rate, and fewer movements around the mouth and the eyes. Facial expression is considered **moderately decreased** when there is a more noticeable reduction in facial movements. Facial expression is **severely decreased** when there is markedly reduced eye blinking, with facial expressions around the mouth and eyes occurring only during intense emotions. In addition, the lips may be parted most of the time. Facial expression is considered **expressionless** when there is an absence of eye blinking and no facial expression around the mouth and eyes, with the lips parted most of the time.
- Rating of voluntary upper limb movements: Observe upper limb movements during reaching, grasping, and/or manipulating objects. In attentive and cooperative patients rapid alternating movements can be evaluated, including opening-closing hands, finger tapping, and pronation-supination of the forearm. Assess latency of movement initiation and the speed and amplitude of movement execution.
  Voluntary upper limb movements are considered **mildly decreased** when there is mild slowness in the initiation and/or execution of movements that does not interfere with motor tasks. They are considered **moderately decreased** when there is moderate slowness in the initiation and/or execution of movements that interferes with motor tasks but still allows task completion. They are considered **severely decreased** when there is severe slowness in the initiation and/or execution of movements, interfering with motor tasks to the extent that the task cannot be completed or requires repeated attempts. **No voluntary movements** are observed when no voluntary movements are present.

Tremor

Tremor is a rhythmic back-and-forth or oscillating involuntary movement about a joint axis. All types of tremor (rest, postural, or action) are included in this rating. To score tremor observe the patient while is quietly lying down, sitting, or being held in a parent's arms, as well as while playing and manipulating objects. In cooperative patients, postural and kinetic tremor can be assessed as part of the neurologic exam by having the patient hold a posture with the upper limbs and performing the finger-to-nose maneuver. The distribution of tremor is scored by assigning a score of 1 for each body part where tremor

is present. The severity of tremor is scored based on the maximum amplitude observed in the most affected body part at any time.

Rigidity

Rigidity is characterized by bidirectional resistance to passive movement, independent of velocity, and it is not specific to a particular task or posture. To score rigidity perform slow passive movement of major joints of the upper and lower limbs with the patient at rest, in a relaxed position.

Score rigidity when there is increased resistance independent of posture and speed of movement, and/or when there is maintenance of a limb position after passive movement. Instruct the patient to relax as much as possible as you test for rigidity. Score the most affected body part. In older and cooperative patients, first test without an activation maneuver. If no rigidity is detected, use an activation maneuver such as fist opening/closing, or foot tapping in the contralateral limb. The distribution of rigidity is scored by assigning a score of 1 for each body part where rigidity is present. The severity of rigidity is scored in the most affected body part.

Dystonia

Dystonia is characterized by involuntary sustained or intermittent muscle contractions causing twisting and repetitive movements or abnormal postures or both. The presence of dystonia and its severity is scored in 4 body areas including: 1) Facial, ocular and/or oromandibular, 2) Cervical, laryngeal and/or truncal, 3) Upper limbs 4) Lower limbs.

The severity of dystonia is rated based on the conditions under which it occurs: action-induced, postural/ triggered by manipulation, at rest, or at rest with a fixed posture. If limb dystonia is asymmetric, score the more affected side. Ideally, dystonia should be scored in the patient's usual state, not during an oculogyric crisis.

Oculogyric crises

Oculogyric crises are attacks of intermittent or sustained eye deviation, with or without associated abnormal posturing of the face, limbs, neck, or trunk. They are not epileptic phenomena. This item is based on the caregiver's report from the past three months and integrates caregiver-reported information on severity, duration, and frequency of the episodes. Caregivers are read the content of each score to ensure understanding of each response. Severity of oculogyric crises is based on the presence of dystonia in other body parts beyond the ocular region, and the presence of associated systemic symptoms. Duration is rated based on the length of typical episodes, ranging from less than 1 hour to more than 4 hours) and frequency is scored based on the number of occurrences per week, ranging from less than once a month to more than three times weekly.

Axial hypotonia

Axial hypotonia refers to decreased muscle tone affecting the axial musculature and/or decreased postural tone. Note that hypotonia must not be related to muscle weakness in the setting of neuromuscular disorder. Cervical and truncal tone are assessed separately.

- Cervical tone is assessed by pulling the infant towards sitting posture by traction on both wrist and observing the alignment of the head in relation to the body and the ability to lift the head. In case that the patient is able to sit, cervical tone is assessed by placing the patient in a sitting position, with or without support, and observing the alignment of the head in relation to the body and the ability to move the head.
- Truncal tone is assessed by holding the infant horizontally under the belly and observing the posture and alignment of the back and head. In case that the patient is able to sit, with or without support, place the patient in the sitting position and observe the posture of the trunk and head and the ability to straighten the back.

Motor development

Motor development is based on the patient's motor function relative to typical function expected for age. Consider normal age of acquisition of head control up to 4 months, sitting without support up to 9 months, standing alone up to 17 months and walking alone up to 18 months
https://www.who.int/childgrowth/standards/motor milestones/en/
Gross motor function can also be rated based on functional mobility level per The Gross Motor Function Classification System (GMFCS)
https://canchild.ca/system/tenon/assets/attachments/000/000/058/original/GMFCS-ER_English.pdf

3) DYSKINESIA SUBSCALE

The Dyskinesia subscale evaluates involuntary hyperkinetic movements, including phenomenology consistent with chorea, athetosis, ballism, myoclonus, and hyperekplexia. Dystonia is not included since this movement disorder is assessed under the Motor Manifestations of Parkinsonism-Dystonia. Stereotypies and tics are also not included.

Chorea is characterized by an ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments. Athetosis is a slow, continuous, involuntary writhing movement that prevents maintenance of a stable posture. Ballism is defined as chorea that affects proximal joints such as shoulder or hip. Myoclonus is a sequence of repeated, often nonrhythmic, brief shock-like jerks due to sudden involuntary contraction or relaxation of one or more muscles. Hyperekplexia is characterized by an exaggerated response to minor stimuli that are mostly acoustic and tactile.

Dyskinesias in infantile Parkinsonism-dystonia may either be part of the complex neurological presentation or a side effect of dopaminergic treatment, such as L-dopa, dopaminergic agonists, MAO

inhibitors, COMT inhibitors, and anticholinergics. A "Yes" or "No" response is required to confirm if the patient is taking these medications. The type of dyskinesia observed needs to be specified. The severity of dyskinesia is rated by considering the amplitude of movements, their impact on functionality, interference with daily activities, and any discomfort they cause. The duration of dyskinesia is assessed at the end of the examination, based on the percentage of time they are present. The distribution of dyskinesias is scored by assigning a score of 1 for each body part affected.

INSTRUCTIONS FOR SCORING

Each subscale (Non-Motor, Motor, and Dyskinesia) should be scored separately.

In each subscale, each item (except for the items scoring distribution of tremor, rigidity, and dyskinesias) employs a 5-level response format, where a score of 0 indicates the absence of a manifestation and a score of 4 represents its maximum severity.

The items scoring distribution of tremor and dyskinesias assign one point for each body part involved, with a score of 0 indicating absence of the manifestation and a score of 5 representing the maximum number of body parts affected.

The items scoring distribution of rigidity assign one point for each body part involved, with a score of 0 indicating absence of the manifestation and a score of 3 representing the maximum number of body parts affected.

The total score of the Dyskinesias subscale needs to be interpreted in the context of the patient's clinical situation. In patients with infantile parkinsonism-dystonia syndrome, dyskinesias can be intrinsic to the disease. In those on dopaminergic treatment, dyskinesias can be treatment-induced

The timing of assessments using the IPDRS scale is flexible and left to the clinician’s judgment. For infants with expected developmental progress, evaluations every 3–6 months are appropriate, while for older patients, shorter intervals may not provide additional value. Additionally, since some scale items assess frequency over the past 3 months, at least 3 months should pass between two IPDRS scoring sessions

Below is a list of items for each subscale, along with the minimum and maximum scores for each item:

SUBSCALE	Minimum score	Maximum score
1.NON-MOTOR SYMPTOMS		
Sudomotor and thermoregulation	0	4
Respiratory secretomotor function	0	4
Gastrointestinal motor function	0	4
Sleep	0	4

Other	0	4
Emotional lability	0	4
TOTAL	0	24
2.MOTOR SYMPTOMS		
BRADYKINESIA		
Global spontaneity of movement	0	4
Facial expression	0	4
Upper limb movements: spontaneous	0	4
Upper limb movements: voluntary	0	4
Lower limb movements: spontaneous	0	4
TREMOR		
Distribution of tremor	0	5
Severity of tremor	0	4
RIGIDITY		
Distribution of rigidity	0	5
Severity of Rigidity	0	4
DYSTONIA		
Facial, ocular and/or oromandibular dystonia	0	4
Cervical, laryngeal and/or truncal dystonia	0	4
Dystonia of the upper limbs	0	4
Dystonia of the lower limbs	0	4
OCULOGYRIC CRISES		
Severity of the crises	0	4
Duration of typical oculogyric episodes	0	4
Frequency of oculogyric episodes	0	4
AXIAL HYPOTONIA		
Head lag	0	4
Truncal hypotonia	0	4
MOTOR DEVELOPMENT		4
TOTAL	0	78
3.DYSKINESIAS		
DOPAMINERGIC TREATMENT	YES	NO
	L-dopa	COMT inhibitors
	Dopamine agonists	Anticholinergics
	MAO inhibitors	Others
TYPE DYSKINESIAS	Chorea	Athetosis
	Ballism	Myoclonus
	Hyperekplexia	
Severity of dyskinesia	0	4
Duration of dyskinesia	0	4
Body parts involved	0	5
TOTAL	0	13

References

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