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Expanded Disability Status Scale (EDSS) in Multiple Sclerosis

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ABSTRACT

Multiple sclerosis has an increasing prevalence and incidence. There are many articles showing that early treatment can prevent possible disability. Expanded disability status scale assessmenthas great importance both in pivotal studies and in clinical practice to evaluate treatment efficacy. For this reason, this review has been written to be well known and not to miss the details.

Keywords: Disability, EDSS, functional score, multiple sclerosis

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Introduction

Multiple sclerosis (MS) is the most common chronic inflammatory demyelinating disease of the central nervous system, affecting approximately 3 million people worldwide (1). It can be seen in almost any age range, although it is more common in the 20-40 age range and about 3 times more common in women. In our country, MS female/male patient ratio is 2.5/1 (2). The prevalence of MS is 41-61/100.000 (3).

The expanded disability status scale (EDSS) is the most commonly used scale in patients with MS. EDSS is a very effective method in reflecting disability (4). EDSS assessment is a non-linear assessment and is a scale in which MS is evaluated between 0 and 10, where normal neurological examination is 0 and MS-related death is 10 (5). Although EDSS is widely used in clinical studies and patient follow-up, it has some limitations. Increases of 1 point have different meanings in each point transition. The evaluation of functional systems (FS) is complex and subjective. It is insensitive and distant to the evaluation of cognitive functions and especially upper extremity functions between 4.0-6.5 EDSS. The contribution of cerebral functions to EDSS scores is very limited. In contrast, the contribution of pyramidal and cerebellar functions to the score is significant. EDSS includes an ambulation-based evaluation after 4.0 (6).

In an article published by Lublin (7) in 2014, the disease should be phenotyped according to active and progressive status. A numerical equivalent of disability has been adopted in terms of determining the degree of the disease, treatment change, or possible progressive process. For this reason, the EDSS is used in MS (8). In many pivotal studies, the primary endpoint is EDSS. Simultaneously, attack-related worsening and non-attack-related worsening are also determined by the increase in EDSS in patient follow-up (9).

Jean Martin Charcot described cognitive impairment in MS as markedly impaired memory, slowed conceptualization and impaired intellectual functioning (10). The EDSS was defined by

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Kurtzke (11). This article uses this article, which is still valid in clinical practice. In contrast to Charcot, they suggested that cognitive impairment in individuals with MS is seen in 3% of patients and that cognitive impairment occurs in patients with high rates of physical disability (11). This detail may perhaps explain the lack of emphasis on cognitive functions in the EDSS. Time is an important determinant of the nature of MS, so in a study investigating the predictive significance of time and cognitive status for EDSS, it was reported that although no cognitive test was predictive of EDSS in the early period, information processing speed was predictive of EDSS at 5-year follow-up, and both information processing speed and visuospatial ability were predictive of EDSS score at 6-8 years (12). Studies evaluating disease subtypes show that progressive MS is cognitively different from relapsing-remitting MS (13,14,15). In a study in which EDSS was categorized as <4 or ≥4, it was shown that the cognitive performance of the group with a low EDSS score was significantly better than the group with an EDSS ≥4 (16).

It is impossible to calculate the EDSS without knowing the functional scores. Although there are many digital calculation methods, these programs cannot perform some conversions. Functional scoring is explained in detail below. The tables were made by me and taken from the MS reference book (17).

Functional Systems

Pyramidal Functions

0. Normal

1. Abnormal findings without disability,

2. Minimal disability,

3. Mild or moderate paraparesis or hemiparesis; severe monoparesis,

4. Marked paraparesis or hemiparesis; moderate quadriparesis; or monoplegia,

5. Paraplegia, hemiplegia,

6. Quadriplegia.

The pyradimal function scoring table is given below (Table 1).

Table 1. Pyramidal functions scoring

One extremity	Score
Normal physical examination	0
Only if there are findings	1
If the motor force is 4/5	2
If the motor force is 2 or 3/5	3
If the motor force is 0 or 1/5	4
Two extremities	**Score**
If the motor force is 4/5	2
If the motor force is 3/5	3
If the motor force is 2/5	4
If the motor force is 0 or 1/5	5
Three extremities	**Score**
If the motor force is 4/5	3
If the motor force is 3/5	4
If the motor force is 2/5	5
Four extremities	**Score**
If the motor force is 4/5	3
If the motor force is 3/5	4
If the motor force is 2/5	5
If the motor force is 0 or 1/5	6

Cerebellar Functions

0. Normal,

1. Abnormal findings without disability,

2. Mild ataxia,

3. Middle truncal or limb ataxia,

4. Network ataxia, all extremities,

5. Inability to make coordinated movements due to ataxia.

The cerebellar function scoring table is given below (Table 2).

Table 2. Cerebellar function scoring

Body posture	Score
Normal physical examination	0
Only if there are findings	1
Romberg positivity	2
Moderate - hard	3
Hard	4
Walking	**Score**
Mild	2
Moderate - hard	3
Hard	4
Extremity	**Score**
Normal physical examination	0
Only if there are findings	1
Mild	2
Moderate	3
Hard (3-4 extremities)	4
Cannot act in coordination	5
Cannot be evaluated	X

Brain Stem Functions

0. Normal,

1. Findings only,

2. Moderate nystagmus or other mild disabilities,

3. Severe nystagmus, marked loss of extraocular power, or moderate disability of other cranial nerves,

4. Significant dysarthria or other significant disability,

5. Loss of the ability to swallow or speak.

The brain stem functions scoring table is given below (Table 3).

Table 3. Brain stem functions scoring

Extraocular movement score	Score
Only if there is a symptom [limitation in barely noticeable emergency obstetric hysterectomy (EOH) (no patient complaints)] 1	1
Mild (patient has complaints with limitation in EOH that is barely noticeable or incomplete paralysis in eye movements that the patient is not aware of)	2
Moderate (patient-aware incomplete paralysis of eye movements or complete loss of movements in one direction of gaze in both eyes)	3
Severe (complete loss of movements in both eyes when looking in more than one direction)	4
Nystagmus score	**Score**
Only signs or mild (gaze-evoked nystagmus)	1
Moderate (no nystagmus in primary position but persistent nystagmus at 30 degrees vertical/ horizontal gaze)	2
Severe persistent nystagmus in primary position or affecting vision, in all directions gaze	3
Complete internuclear ophthalmoplegia with very marked persistent nystagmus or persistent nystagmus in the abducting eye 3	4
Dysphagia score	**Score**
Only if there is a finding	1
Mild (difficulty drinking fluids)	2
Moderate (difficulty swallowing liquid solid foods)	3
Heavy (persistent difficulty swallowing, can eat puree)	4
Absent	5
Dysarthria score	**Score**
1 only if there is a finding	1
Mild (patient-aware dysarthria)	2
Moderate (dysarthria impairing intelligibility in everyday speech)	3
Heavy (unintelligible speech)	4
Complete loss (unable to speak)	5
Hearing Loss score	
7th cranial nerve	**Score**
Only if there are findings	1
Mild (facial weakness of which the patient is aware)	2
Moderate [incomplete facial paralysis (difficulty closing the eyes - must close them at night or difficulty closing the mouth - drooling)]	3
Heavy	4
5th cranial nerve	**Score**
Only if there are findings	1
Mild (patient-aware numbness)	2
Moderate [impaired sharp/crunch distinction in branches 1, 2, or 3 of the trigeminal nerve or trigeminal neuralgia (at least 1 pain attack in the last 24 h)]	3
Severe (inability to distinguish between sharp/ sharp or complete loss of sensation in all branches of the trigeminal nerve)	4

Sensory Functions (1982 Revision)

1. Decreased vibration or drawing only in one or both extremities,

2. Slightly reduced sensation of touch, pain, or position in one or both extremities, and/or moderately reduced, vibration in one or both extremities; or vibration deficit in 3-4 extremities alone (e.g. drawing shapes),

3. Moderate decreased sensation of touch, pain, or position in one or two extremities, and/or mainly loss of vibration; or mild touch, pain and/or moderate impairment of all proprioceptive tests in 3-4 extremities,

4. Markedly decreased sense of touch, pain, or loss of proprioception in one or two extremities, singly or in combination; or moderate loss of touch, pain and/or severe loss of proprioception in more than two extremities,

5. Loss of sensation (mainly) in one or both extremities; or moderate loss of sensation of touch, pain and/or proprioception in most of the parts of the body below the head,

6. Mainly loss of sensation in the sub-cranial parts.

The sensory function scoring table is given (Table given below 4, 5).

Table 4. Sensory functions

Position 1-2	Score
Mild	2
Moderate	3
Absent	5
Position 3-4	**Score**
Moderate	3
Hard	4
Absent	5
Figure drawings 1-2	**Score**
Mild	1
Absent	5
Figure drawings 3-4	**Score**
Mild	2
Moderate	3
Hard	4
Absent	5
Vibration 1-2	**Score**
Mild	1
Moderate	2
Absent	3
Vibration 3-4	**Score**
Light	2
Moderate	3
Hard	4
Absent	5
Pain 1-2	**Score**
Mild	2
Moderate	3
Hard	4
Absent	5
Pain 3-4	**Score**
Mild	3
Medium	4
Hard	5
Absent	6
Heat sensation 1-2	**Score**
Mild	1
Absent	5
Heat sensation 3-4	**Score**
Mild	2
Moderate	5
Absent	6

Table 5. Sensory functions converting scores

Vibration 3-4 moderate impairment + position 3-4 moderate impairment: 3 vibration 3-4 lost + position 3-4 lost: 4
Vibration 1-2 lost + pain-heat 1-2 lost + position 1-2 lost: 5 vibration 3-4 lost + pain-heat 3-4 lost + position 3-4 lost: 6

Bladder-bowel Functions

0. Normal,

1. Mild pause in urination (urgency), a feeling of urinary urgency or urinary retention,

2. Moderate urinary urgency, urinary urgency, urinary urgency, retention in the bowel or bladder, or rare incontinence,

3. Frequent urinary incontinence,

4. The need for almost continuous indwelling catheterization,

5. Loss of bladder function,

6. Loss of bladder and bowel function,

The bladder bowel function scoring table is given (Table 6 given below).

Conversion: Bladder-bowel FS grade 6-5

Table 6. Bladder bowel functions scoring

Catheter	Score	Converted
Intermittent	3	3
Always	5	4
Bowel	**Score**	**Converted**
Mild	1	1
Severe	2	2
Intervention	3	3
Missing	5	4
Bladder - incontinence	**Score**	**Converted**
Mild	1	1
Moderate (infrequent)	2	2
Moderate (frequent)	3	3
Missing	5	4

Visual (or Optical) Functions

0. Normal,

1. Scotoma with corrected visual acuity better than 20/30,

2. Maximum corrected visual acuity in the worse eye between 20/30-20/59,

3. Extensive scotoma in the worse eye, or a degree of visual field reduction but maximum-corrected visual acuity between 20/60 and 20/99,

4. Significant reduction in visual field and maximum-corrected visual acuity between 20/100 and 20/200 in the worse eye; maximum visual acuity 20/60 or less in grade 3 plus good eye,

5. Maximum corrected visual acuity less than 20/200 in the worse eye; maximum visual acuity 20/60 or less in grade 4 plus the better eye,

6. Fifth degree plus maximum visual acuity of 20/60 or less in the better eye.

The optical function scoring table is given (Table given below 7, 8).

Contribution of visual FS degrees to EDSS

6—4

5—3

4—3

3—2

2—2

1—1

Table 7. Optical functions scoring

Disk pallor	Score
0	0
1	1
Scotoma	**Score**
0	0
Small scotoma	1
Big scotoma	3

Table 8. Optical functions scoring 2 (visual acuity scoring)

Visual acuity		Score
Healthy eye	Damaged eye	
-	1.0	0
-	>0.67	1
-	0.67-0.34	2
-	0.33-0.21	3
-	0.2-0.1	4
<0.33	0.21-0.33	4
-	<0.1	5
<0.33	0.2-0.1	5
<0.33	<0.1	6

Cerebral (or Mental) Functions

0. Normal,

1. Mood disorder only (does not affect DSS score),

2. Slight decrease in mental function,3. Moderate impairment of mental function,4. Severe impairment of mental function (moderate chronic brain syndrome),

5. Dementia or chronic brain syndrome - severe or incompetent.

The mental function scoring table is given below (Table 9).

Table 9. Mental functions scoring

Mental	Score
Mood change	1
Mild	2
Moderate	3
Hard	4
Dementia	5
Fatigue	**Score**
Mild	1
Moderate - hard	2

Scoring

0.0: Normal neurological examination (grade 0 in all FS, including cerebral grade 1)

1.0: No disability, minimal findings (grade 1) in one FS (except cerebral grade 1)

1.5: No disability, minimal findings (grade 1) in more than one FS (except cerebral grade 1)

2.0: Minimal disability in one FS (one FS grade 2; others 0 or 1)

2.5: Minimal disability in two FS (two FS grade 2; others 0 or 1)

3.0: Moderate disability in one FS (fully ambulatory patient)

One FS grade 3, the others 0 or 1

Mild disability in 3 or 4 FS (3/4 FS grade 2, others 0 or 1)

3.5: Fully ambulatory patient, but moderate disability in one FS

One grade 3 + one or two FS grade 2

Five FS grade 2 (others 0 or 1)

4.0: Fully ambulatory patient (can walk around 500 metres unassisted and without rest) grade 4 severe disability in one FS (others 0 or 1)

Combination of lower grades, exceeding the limits of the previous steps

4.5: Can walk 300 meters without assistance or rest

The fully ambulatory patient unassisted for close to most of the day, able to work full time, grade 4 on one FS (others 0 or 1)

Combination of lower grades, exceeding the limits of the previous steps

5.0: Can walk approximately 200 meters without assistance or rest; the disability is severe enough to prevent him/her from fully conducting daily activities

Grade 5 in one FS (others 0 or 1)

Combinations exceeding low grades

5.5: Can walk approximately 100 meters without assistance or rest;

The disability was severe enough to prevent daily activities

Grade 5 alone in a FS (others 0 or 1)

Combinations exceeding low grades

6.0: Intermittent or unilateral fixed support required to walk approximately 100 meters with or without rest

Combinations of 3 or more degrees of impairment in more than two FS

6.5: Fixed bilateral support required to walk 20 meters without rest; combinations of 3 or more degrees of impairment in more than two FS

7.0: Cannot walk beyond 5 meters even with assistance;

Wheelchair-dependent

Turns the wheels by itself and can move into the wheelchair by itself

May spend approximately 12 h or more per day in a wheelchair

Grade 4 or more in one FS; rarely pyramidal grade 5

7.5: Cannot take more than a few steps;

Wheelchair-dependent

Assistance with the transition to a wheelchair may be required

Turns the wheelchair itself

Cannot spend the whole day in a standard wheelchair

Motorized wheelchair may be required

Grade 4 in more than one FS

8.0: Mainly bed/chair dependent, or can ambulate in a wheelchair

Can spend most of the day out of bed; can do most of his/her own work

Multiple grades 4 and above in FSs

8.5: Bedridden most of the day; can use arm(s) effectively to some extent

Multiple grades 4 and above in FSs

9.0: Hopelessly bedridden patient; can communicate and eat

Most of the FSs have a rating of 4 and above

9.5: Completely hopeless, bedridden patient; unable to communicate effectively or swallowing and eating impaired

10.0 Death

Practical Approaches to Ambulation

- Asymptomatic
- Can walk normally, but fatigue and exhaustion occur in situations requiring athletic performance
- Unassisted walks 300≤ >500 m (EDSS: 4.5-5)
- Can walk 200≤ >300 m without support (EDSS: 5)
- Can walk 100≤ >200 m without support (EDSS: 5.5)
- The unassisted walking distance was less than 100 m (EDSS: 6)
- Can walk more than 50 m with unilateral support (EDSS: 6)
- Can walk more than 120 m with bilateral support (EDSS: 6)
- Can walk up to 50 m with unilateral support (EDSS: 6.5)
- Can walk at least 5 and up to 120 m with bilateral support (EDSS: 6.5)
- Usually wheelchair-bound, cannot walk more than 5 m even with support, can switch to a wheelchair (EDSS: 7)
- Requires assistance for wheelchair use, cannot take more than a few steps even with support, requires assistance for transfer (EDSS: 7.5)
- Usually bed and chair bound, can spend most of the day out of bed, uses hands actively, needs help self-care (EDSS: 8)
- Spends most of the day in bed (EDSS: 8.5)
- Bedridden, able to communicate and feed (EDSS: 9.0)
- Bed-dependent, unable to communicate, feed and chew (EDSS: 9.5)

In Summary

- A FS 1 EDSS 1
- Multiple FS 1 (1+1+) EDSS 1.5
- One FS 2 EDSS 2
- Two FS 2 (2+2) EDSS 2.5
- A FS 3 or (2+2+2+2) or (2+2+2+2+2) EDSS 3
- A FS 3+2 or (3+2+2+2) or (2+2+2+2+2+2) EDSS 3.5
- An FS 4 or (EDSS; above 3.5) EDSS 4

• Unassisted 300-500 meters EDSS 4.5

• Unassisted 200-300 meters EDSS 5

• Unassisted 100-200 meters EDSS 5.5

From 6.0 points onward, the patient's need for support is recorded

• Unilateral support EDSS 6

• 2 sided support EDSS 6.5

From 7.0 onwards, wheelchair and gradual bed dependency

• Communicates bedridden after 8.5

• 9.5 EDSS cannot communicate

• 10.0 death

EDSS Calculation with Samples (18)

First, detailed patient examination of must be performed. Then the FS score is determined. Necessary changes are made in the systems that need conversion. The EDSS score is calculated based on a FS and ambulation.

Functional System Score Calculation

• Neurological examination: Visual acuity; left eye; 0.1 (20/200), right eye; 1.0 (20/20) visual FS score: 4 (after conversion: 3),

• Neurological examination: Visual acuity; left eye; 0.1 (20/200), right eye; 0.8 (20/25) left eye defects from childhood. Visual FS score: 1 (after conversion: 1),

• Neurological examination: Visual acuity; left eye; 0.05 (20/400), right eye; 0.8 (20/25) visual FS score: 5 (after conversion: 3),

• Neurological examination: Persistent nystagmus (primary) in the primary position, internuclear ophthalmoparasis (middle) in the left eye, clinically detectable dysarthria (mild) Brainstem FS score: 3,

• The patient has a clone in the right lower extremity, live deep tendon reflexes in the lower extremities, muscle strength is complete in all muscle groups. Pyramidal FS score: 1,

• The patient's right lower extremity 2/5 muscle strength, right upper extremity 3/5 muscle strength, live reflexes in lower extremities, plantar response extensor on the right. Pyramidal FS score: 4,

The patient cannot walk more than a few steps due to lower extremity ataxia. Have only trunkal ataxia when sitting. There is a mild tremor in the upper extremities cerebellar FS score: 4,

The patient had no complaints. Slightly reduced vibration sensation in the lower extremities. Other sensory examination findings were within normal limits Sensory FS score: 1,

• The patient had Lhermitte's complaint and mild depression sensory FS score: 0, cerebral system score: 1,

• Patient needs bladder catheterization several times a week, constipation problem is present, occasional manual intervention is required. Bowel and bladder FS score: 3 (after conversion: 3).

Conclusion

EDSS is a scoring system that is known by every neurologist but is not applied in practice. In this article, we want to address the EDSS approach in practice.

Ethics

Peer-review: Externally peer-reviewed.

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