

**Article abstract**—One method of evaluating the degree of neurologic impairment in MS has been the combination of grades (0 = normal to 5 or 6 = maximal impairment) within 8 Functional Systems (FS) and an overall Disability Status Scale (DSS) that had steps from 0 (normal) to 10 (death due to MS). A new Expanded Disability Status Scale (EDSS) is presented, with each of the former steps (1,2,3 . . . 9) now divided into two (1.0, 1.5, 2.0 . . . 9.5). The lower portion is obligatorily defined by Functional System grades. The FS are Pyramidal, Cerebellar, Brain Stem, Sensory, Bowel & Bladder, Visual, Cerebral, and Other; the Sensory and Bowel & Bladder Systems have been revised. Patterns of FS and relations of FS by type and grade to the DSS are demonstrated.

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# Rating neurologic impairment in multiple sclerosis: An expanded disability status scale (EDSS)

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In 1955 I described “a new scale for evaluating disability in multiple sclerosis,”<sup>1</sup> later known as the Disability Status Scale (DSS), devised to evaluate isoniazid as a possible treatment.<sup>2</sup> This scale was also used in the first multicentered, randomized, placebo-controlled, double-blind trial of MS therapy,<sup>3</sup> which refuted our original claim, a decision with which we had to concur from our later experience.<sup>4</sup> The DSS had 10 grades or steps beyond 0 (normal), extending to status 10 (death due to MS). The scale was “intended to measure the maximal function of each patient as limited by . . . neurologic deficits,”<sup>1</sup> and it was based on neurologic examination.

The DSS was later made half of a bifid rating system, the other part “being a series of grades in each of eight functional groupings. . . . In each portion, there is a numerical rating which is mutually exclusive in its category, and the higher the number, the greater is the dysfunction. Only objectively verifiable defects due to multiple sclerosis as elicited upon neurologic examination are included. Symptoms are discarded.”<sup>5</sup>

The functional groups, later called Functional Systems (FS), were Pyramidal (P), Cerebellar (Cl), Brain Stem (BS), Sensory (S), Bowel & Bladder (BB), Visual (V), Cerebral or Mental (Cb), and Other or Miscellaneous (O) Functions. All save the last

were graded from 0 (normal) to maximal impairment (grade 5 or 6); the “Other” FS was dichotomous, with 0 as none and 1 as any present. Approximate equivalents for the DSS steps were also provided. The Functional Systems were mutually exclusive in terms of neuroanatomy, but together comprised all neurologic abnormalities on examination that can be attributed to MS lesions. The FS were not additive; each FS could be compared over time only with itself, and for this reason it was necessary to retain the DSS for overall comparisons of the same patient at different examinations.

The FS were modified in 1965 by changing the Sensory scale from 0-5 to 0-6 and redefining the upper grades for Bowel & Bladder.<sup>6</sup> As will be seen below, the Sensory System is again being revised, and Bowel & Bladder has a new step.

This two-part system of assessing neurologic impairment in MS has been used in several studies, and it has been proposed for adoption as one part of a tridimensional scheme for a “minimal data set” in MS, which will be discussed below. However, some investigators believe the DSS is too insensitive to change in the middle ranges, and have urged division of step 7 into two parts. Further, while the DSS was considered satisfactory in several treatment trials in acute bouts, it was thought that there should be more

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room for change in studies of chronic MS.

For these reasons, an Expanded DSS (EDSS) is now presented. It provides, for each step from 1 through 9, two steps that together add up to the same step of the original DSS. This division relies even more heavily on the standard neurologic examination as encoded in the Functional Systems. In fact, it is fully defined in the lower ranges by the FS grades. For this reason, before presenting the Expanded DSS, we need to consider the Functional Systems.

**Functional Systems.** The grades for each of the Functional Systems are defined in appendix A. They are identical with those provided in 1965<sup>6</sup> except for the new Sensory and Bowel & Bladder Systems. The frequency of involvement in each system at admission to the hospital for an early bout of MS in one series is described in table 1.<sup>7</sup>

Recall that each FS is independent of the others, yet together they reflect all neurologic impairment in MS. There are over 1.3 million possible patterns of involvement by FS type and grade. However, if we consider each System as just involved (1) or not involved (0), then neurologic impairment can be defined by an eight-digit binary number. For example, a patient with Pyramidal, Cerebellar, and Sensory signs, the other Systems normal, would be described as 1101 0000. There are then only 256 possible patterns (2<sup>8</sup>) into which a patient can fall. From the same series as in table 1, there are described the most common patterns to be expected if lesions in one system were independent of lesions in the others (table 2). These expected frequencies compare well with those actually observed for the same specific patterns.<sup>8</sup> One-half of the patients fell into one of only 14 patterns, and 1/4 into one of only 4 patterns.

Several points of clarification may be in order for the Functional Systems. Pyramidal, Cerebellar, Sensory, and Bowel & Bladder functions all refer to impairment of body parts below the head *only* (regardless of the site of the lesions), and Brain Stem functions have always referred to impairment "attributable to lesions of supra- and intersegmental tracts subserving cranial nerves 3 through 12, together with involvement of these nuclei or their intramedullary fibers. These, therefore . . . encompass pseudobulbar palsies and scanning speech . . . in addition to the so-called cranial nerve functions."<sup>5</sup>

For each FS and the DSS, the rule remains: "Where criteria for the precise grade are not met, the nearest appropriate category is utilized."<sup>5</sup> Thus Pyramidal grade 5 would be used rather than 4 for one who is almost paraplegic. Whatever the specific grade definition, then, "almost" or "practically" can be prefixed. One method for difficult decisions is to "bracket" the likely grade and then cone down on the most applicable.

**The Expanded Disability Status Scale.** The EDSS (appendix B) will be discussed under con-

**Table 1. Percentage frequency of involvement according to Functional Systems (FS) from neurologic examinations at admission to hospital for an early bout of MS; Army WW II series\***

Functional Systems (FS)	% involved	Total N known
Pyramidal (P)	84.9	511
Cerebellar (CII)	76.9	481
Brain Stem (BS)	73.0	514
Sensory (S)	55.2	478
Bowel & Bladder (BB)	22.6	517
Visual* (V)	33.9	425
Cerebral-total <sup>‡</sup> (Cb)	20.7	487
Cerebral-mentation <sup>§</sup>	2.9	487
Other (O)	14.9	523

\* From Kurtzke et al, Acta Neurol Scand 1972;48:19-46.  
 \* Neuropathic signs either/both eyes; see \*.  
 ‡ Includes mood changes only (step 1).  
 § Steps 2+ on the scale.

**Table 2. Patterns of involvement by Functional System (FS) from neurologic examinations at admission to hospital for an early bout of MS; Army WWII series\***

Rank <sup>†</sup>	Pattern <sup>‡</sup>	No. of cases		Cumulative p <sup>§</sup>	
		O	E	O	E
1	1111 0000	31	28.92	0.093	0.086
2	1110 0000	29	28.74	0.179	0.172
3	1111 0100	12	15.32	0.215	0.218
4	1110 0100	16	15.23	0.263	0.263
5	1101 0000	14	9.37	0.304	0.291
6	1100 0000	8	9.32	0.328	0.319
7	1011 0000	6	8.78	0.346	0.345
8	1010 0000	7	8.72	0.367	0.371
9	1111 0010	15	7.78	0.412	0.395
10	1110 0010	8	7.73	0.436	0.418
11	1111 1000	11	7.09	0.469	0.439
12	1110 1000	4	7.05	0.481	0.460
13	0111 0000	1	6.18	0.484	0.478
14	0110 0000	9	6.15	0.510	0.497
15-256	all other	164	168.63	1.000	1.000
256	Total	335	335.01	1.000	1.000

\* Adapted from Kurtzke, Acta Neurol Scand 1970;46:493-512.  
 † Rank order of expected frequency of specific pattern, based upon product of individual observed frequencies with hypothesis of independence for all patterns where E ≥ 5.0; O = observed and E = expected frequency.  $\chi^2_{14} = 20.58, p > 0.10$  for O versus E.  
 ‡ Involved (1) or not involved (0) for P, CII, BS, S, BB, V, Cb, O in cited order; cases with complete information on all 8 FS.  
 § Cumulative proportion (p) of total, observed (O), and expected (E) patterns.

**Table 3. Percentage frequency distribution of Functional System (FS) grades according to DSS steps. I: DSS 1-2\***

DSS 1-2 FS	FS grades							100% = (N)
	0	1	2	3 (percentages)	4	5	6	
P	51.7	35.3	13.0	—	—	—	—	(207)
CI†	65.8	14.1	20.1	—	—	—	NA	(199)
BS	48.5	29.9	14.7	6.9	—	—	NA	(204)
S‡	79.6	8.9	11.5	—	—	—	NA	(191)
BB‡	90.7	6.4	2.9	—	—	—	NA	(204)
V§	68.2	18.2	13.6	—	—	—	—	(22)
Cb	91.9	7.6	-----0.5-----	—	—	—	NA	(198)
O	86.6	13.4	NA	NA	NA	NA	NA	(307)

\* Data from some 2,000 exams in 20 years among 527 males, Army WWII series.  
† Excludes those with Pyramidal grade 3+.  
‡ 1961 scales.  
§ VA Hospital series (N = 392).  
— No cases.  
NA Not applicable; step(s) not in scale.

secutive groupings of the original DSS. For this expansion, we have had to make more finite and arbitrary distinctions than in the original scale.

**DSS Step 0.** As before, this defines the normal neurologic examination—regardless of symptoms. Therefore, all FS are grade 0, except for Cerebral System grade 1. Cerebral “grade 1 refers to mood aberrations such as euphoria or depression, which may not be a primary effect of the disease process, but this is hoped to represent that stage of brain damage when alterations of personality or emotional control are the sole features.”<sup>5</sup> For DSS step 0 and step 1, Cerebral grade 1 is treated as a 0.

**DSS Steps 1-2.** These steps refer to minimal objective abnormality, with step 1 as signs without impaired function. Table 3 shows the distribution of FS grades for DSS 1-2 from an overview of some 20 years’ follow-up examinations in 527 men with MS, our Army WW II series.<sup>8</sup> The ratio of step 2 to 1 was about 2:1. The DSS scores in this series were not strictly delimited by the FS equivalents described here. Nevertheless, the low frequency of involvement is evident; this was essentially limited to FS grades 1 and 2 except for the 7% in Brain Stem grade 3. The FS scales used here and below are the 1961 variants for Sensory and Bowel & Bladder.

**EDSS Step 1.0** is limited to one FS grade 1, excluding Cerebral grade 1, with all others grade 0.

**EDSS Step 1.5** is defined as two or more FS grade 1, again excluding Cerebral grade 1, but no grade above 1 in any FS.

**EDSS Step 2.0** is limited to one FS grade 2, others grade 0 or 1.

**EDSS Step 2.5** is limited to two FS grade 2, others grade 0 or 1.

Note that it is irrelevant which FS are involved, and from table 3, it is likely to be any of them except Bowel & Bladder or Cerebral.

**DSS Steps 3-4.** These steps still refer to mild disorder, not sufficient to impede normal activities of

daily living or work in most situations. However, a concert pianist, a pilot, or a steeplejack would doubtless not be able to function as usual and still be ascribable to these steps. Full ambulation—meaning ability to be up and about all day and to walk usual distances without resting—characterize these steps. Impaired ambulation of any degree should not occur with FS grades defining DSS step 3. There is some overlap of FS in steps 4 and 5. Table 4 delineates the distribution of FS grades for DSS 3-4. The ratio of step 3 to 4 was about unity. Only rarely was grade 4 attained. We begin to see the predominance of Pyramidal involvement, closely followed by Cerebellar and Brain Stem.

**EDSS Step 3.0** is limited to one FS grade 3, or three or four FS 2, others being 0 or 1.

**EDSS Step 3.5** is limited to one FS grade 3 plus one or two grade 2, or two FS grade 3, or five FS grade 2, others being grade 0 or 1.

**EDSS Step 4.0** consists of combinations just exceeding two grade 3, or one grade 3 plus two grade 2, or five grade 2; or one FS grade 4 alone, all others being grade 0 or 1. At this point, the ambulation/work/daily activity abilities start to take precedence over the precise FS grades. With FS that exceed the criteria for EDSS step 3.5, there must be, for step 4.0, full ambulation (including ability to walk without aid or rest for some 500 meters), and ability to carry out full daily activities to include work of average physical difficulty.

**EDSS Step 4.5** has the same minimal FS grade requirements as step 4.0. The patient must be able to walk without aid or rest for some 300 meters and to work a full day in a position of average difficulty. The patient is up and about most of the day, but some limitation of full activity separates this from step 4.0.

**DSS Steps 5-6.** The patient is not ordinarily housebound and can walk. Seldom is a full work day possible without special provisions. The original DSS 5 was defined as “maximal motor function

**Table 4. Percentage frequency distribution of Functional System (FS) grades according to DSS steps. II: DSS 3-4\***

DSS 3-4 FS	FS grades							100% = (N)
	0	1	2	3 (percentages)	4	5	6	
P	18.5	19.9	25.4	35.1	1.1	—	—	(664)
CII <sup>†</sup>	26.5	11.6	45.1	16.9	—	—	NA	(623)
BS	29.8	27.1	23.9	19.0	0.2	—	NA	(652)
S <sup>‡</sup>	49.4	6.5	31.7	12.2	-----0.2-----	—	NA	(596)
BB <sup>‡</sup>	77.4	7.1	11.8	3.7	—	—	NA	(650)
V <sup>§</sup>	60.6	2.8	12.8	9.2	-----4.6-----	—	—	(109)
Cb	80.3	16.8	-----2.9-----	—	—	—	NA	(615)
O	84.8	15.2	NA	NA	NA	NA	NA	(784)

\* Data from some 2,000 exams in 20 years among 527 MS males, Army WWII series.  
<sup>†</sup> Excludes those with Pyramidal grade 3+.  
<sup>‡</sup> 1961 scales.  
<sup>§</sup> VA Hospital series (N = 392).  
 — No cases.  
 NA Not applicable; step(s) not on the scale.

**Table 5. Percentage frequency distribution of Functional System (FS) grades according to DSS steps. III: DSS 5-6\***

DSS 5-6 FS	FS grades							100% = (N)
	0	1	2	3 (percentages)	4	5	6	
P	2.1	6.6	8.5	49.5	32.8	0.5	—	(424)
CII <sup>†</sup>	5.6	2.5	24.9	56.7	10.3	—	NA	(358)
BS	19.2	23.1	30.2	26.0	1.5	—	NA	(407)
S <sup>‡</sup>	29.8	7.2	40.3	22.4	-----0.3-----	—	NA	(362)
BB <sup>‡</sup>	59.3	10.5	17.1	10.1	1.0	2.0	NA	(398)
V <sup>§</sup>	60.8	6.4	15.2	11.2	-----6.4-----	—	—	(125)
Cb	72.6	20.7	-----5.0-----	—	0.8	0.8	NA	(362)
O	72.1	27.9	NA	NA	NA	NA	NA	(459)

\* Data from some 2,000 exams in 20 years among 527 MS males, Army WWII series.  
<sup>†</sup> Excludes those with Pyramidal grade 3+.  
<sup>‡</sup> 1961 scales.  
<sup>§</sup> VA Hospital series.  
 — No cases.  
 NA Not applicable; step(s) not in scale.

walking unaided up to several blocks,” and for 6 it was “assistance required for walking.”<sup>1</sup> There is generally some impairment in usual daily activities. Table 5 indicates for these steps the increasing frequency and severity of FS involvement, particularly Pyramidal and Cerebellar systems, with Brain Stem and Sensory not far behind. The ratio of step 5 to 6 was about 1.7:1. The principal discrimination among these four new EDSS steps rests with walking; the patient’s statements about walking are ordinarily acceptable, but direct observation—and on more than one occasion—may be required. We are after “usual best function” here, and neither supramaximal nor insufficient efforts at performance. The FS equivalents are advisory and not prescriptive for these and higher steps.

EDSS Step 5.0 requires ambulation for about 200

meters without aid or rest. Disability is severe enough to impair full daily activities, eg, to work a full day without special provisions. Usual FS equivalents are one grade 5 alone, others 0 or 1, or combinations of lesser grades that will usually exceed those specified for EDSS step 4.0.

EDSS Step 5.5 requires ambulation for some 100 meters without aid or rest. Other criteria are inability to work part-time (about ½ day) without special provisions. Usual FS equivalents are as in step 5.0. Note the arbitrary distances for walking ability.

EDSS Step 6.0 requires assistance to walk about 100 meters. This may mean resting, the use of unilateral aids (cane, crutch, or brace) at most times, or the intermittent use of bilateral aids. The assistance of another person also counts as “with aid.” The

**Table 6. Percentage frequency distribution of Functional System (FS) grades according to DSS steps. IV: DSS 7-9\***

DSS 7-9 FS	FS grades							100% = (N)
	0	1	2	3 (percentages)	4	5	6	
P	0.7	1.3	1.0	3.0	40.6	45.9	7.5	(305)
Cl†	1.0	—	4.0	23.2	49.5	22.2	NA	(99)
BS	17.9	12.1	16.8	32.8	15.7	4.6	NA	(280)
S‡	28.1	4.6	33.6	29.9	-----3.7-----	-----	NA	(217)
BB‡	20.3	3.6	12.7	27.9	7.6	27.9	NA	(276)
V‡	54.1	4.4	8.1	17.0	-----	16.3-----	-----	(135)
Cb	67.9	17.4	-----5.8-----	-----	4.7	4.7	NA	(172)
O	57.9	42.1	NA	NA	NA	NA	NA	(328)

\* Data from some 2,000 exams in 20 years among 527 MS males, Army WWII series.  
† Excludes those with Pyramidal grade 3+.  
‡ 1961 scales.  
§ VA Hospital series (N = 392).  
— No cases.  
NA Not applicable; step(s) not on the scale.

primary measure for this step is the ability to walk with help for about 100 meters. Usual FS equivalents are combinations with more than two FS grade 3+.

**EDSS Step 6.5** requires assistance to walk about 20 meters without resting by means of aids (canes, crutches, braces, or people), which are generally bilateral and generally constantly necessary. Usual FS equivalents are as in 6.0—combinations with more than two FS grade 3+. A person who cannot walk 20 meters is functionally almost nonambulatory and should be considered close to DSS 7.

**DSS Steps 7-9.** These are the severely involved patients who are almost invariably limited to wheelchair or bed. Table 6 demonstrates the marked shift to the right for FS grade involvement, particularly those functions having to do with ambulation. This behavior of groups of MS patients lends validity to a scoring system that stresses ambulation in the higher ranges; only in the most severe will the loss of upper limb and head functions be added. The ratio of the steps here was about 1.4:1:1.

The original definition of DSS step 7 was “restricted to wheelchair (able to wheel self and enter and leave chair alone). . . . It does not include the patient who is tied in the chair and perambulated.”<sup>1</sup> Conversely, ability to walk short distances is not sufficient to qualify for step 6. The arbitrary limit for “short distances” is taken here as about 5 meters. This provides some leeway between EDSS step 6.5 (20 meters) and 7.0 (5 meters). As with the other grades, assignment is to that closest to his performance.

**EDSS Step 7.0** defines essential restriction to wheelchair with inability to walk beyond about 5 meters even with aid. Patients can transfer alone (with mechanical aids if needed) and wheel the standard wheelchair; are able to be up and about in the

chair some 12 hours a day; with the chair, are not housebound and may even be employed. Usual FS equivalents are combinations with more than one FS grade 4+; rarely, Pyramidal grade 5 alone.

**EDSS Step 7.5** describes inability to take more than a few steps and, essentially, restriction to wheelchair. With or without aid, these patients can transfer. They can wheel themselves, but cannot carry on in standard wheelchair a full day. They may require motorized wheelchair for ability to be up and about in the chair. Usual equivalents are combinations with more than one grade 4+.

**EDSS Step 8.0.** The original DSS 8 definition was “restricted to bed but with effective use of the arms . . . ; he can usually feed himself and perform part of his toilet.”<sup>1</sup> In our setting, it has been standard procedure to get bed patients into chairs as much as possible, so that the horizontal posture was not a requirement for “bed patient.” This (to me) obvious point has led to some confusion as to requirements for DSS 8.

**EDSS Step 8.0** is defined as bed patients who may be in chair or (passively) in wheelchair for much of the day, and it is so specified in appendix B. Primarily, though, they retain many self-care functions and generally have effective use of the arms. Usual FS equivalents are combinations, generally grade 4+ in several systems.

**EDSS Step 8.5** are the bed patients who in daytime generally cannot tolerate prolonged periods in chair and are more often in bed, unless tied in the chair. Primarily, they still have some effective use of one or both arms and can perform some self-care functions, but less than for step 8.0. Usual FS equivalents are as in step 8.0.

**EDSS Step 9.0** are the “helpless bed patients” who, however, can communicate and eat. They cannot perform self-care functions (such as feeding). Usual FS

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