including a large area of abnormal uptake at the base of the skull, consistent with metastatic disease. Plain radiographs demonstrated extensive erosive lesions at the base of the skull. Needle biopsy of the hepatic lesion demonstrated large-cell undifferentiated carcinoma with "squamoid" features. After tracheostomy and gastrostomy, palliative chemotherapy and radiation therapy were given.

Discussion. The genioglossus, an extrinsic tongue muscle, is stronger than the intrinsic tongue muscles.[3] When the tongue is forcibly protruded, action of the weaker intrinsic tongue muscles may be masked. The intrinsic muscles that turn the tip of the tongue are the superior and inferior longitudinal muscles.[3,4] Unilateral contraction of these muscles shortens the tongue ipsilaterally, turning the tip to that side.

Afer unilateral denervation, the protruded tongue deviates to the weak side. However, the tip of the nonprotruded, unilaterally weak tongue can be turned to the normal side, but not to the weak side. This pattern on protrusion, a sign of unilateral extrinsic muscle weakness, has been well described and illustrated. The sign of intrinsic muscle weakness in the unilaterally weak, nonprotruded tongue may be known to some experienced clinicians, but has not been illustrated in the neurologic literature.

Finally, a unilaterally weak tongue protrudes the cheek on the weak side by using the contralateral genioglossus muscle, not by using ipsilateral intrinsic tongue muscles to turn the tongue into the cheek.

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A neurologic rating scale (NRS) for use in multiple sclerosis

Article abstract—A neurologic rating scale (NRS) has been developed for clinical assessment of MS patients. The scale has been tested on 250 MS patients. Assignment of the NRS score is based on assessment of each component of the neurologic examination and accurately reflects overall neurologic function. Clinical exacerbations are evident as significant deviations from baseline scores. There was close interexaminer correlation, with the range of variability no greater than 2.6%. The NRS is a simple, reliable, and sensitive scale that can be used with other objective measurements of neurologic function, such as neurophysiologic studies, in the clinical assessment of MS patients.

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Jack C. Sipe, MD; Robert L. Knobler, MD, PhD; Sherry L. Braheny, MD; George P.A. Rice, MD; Hillel S. Panitch, MD; and Michael B.A. Oldstone, MD

Multiple sclerosis presents unique and difficult problems in the long-term assessment of patients because there are neither diagnostic tests nor reliable laboratory indicators of disease activity. Changes in clinical status have been the principal means for evaluating improvement and assessing new forms of therapy. Several rating scales have been developed to assess neurologic disability and function in attempts to quantify changes in clinical status.[1-5] Refinements of these rating scales have also been proposed.[6-8] The scales are based primarily on activities of daily living rather than on the standard neurologic examination, and they are insensitive to many changes in neurologic function; some are too elaborate for efficient use.

Increased interest in clinical trials[10-13] demonstrates the need for a simple, reliable, sensitive, and clinically reproducible scale. We describe a neurologic rating scale (NRS) that was developed in conjunction with a standard neurologic examination protocol.

Materials and methods. *Patients.* We studied 250 MS patients who fulfilled the conventional diag-

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nostic criteria.[9,14,15] Twenty-four patients with the exacerbating-remitting form of MS were enrolled in a clinical treatment protocol and were examined at least 15 times each year for 3 years. Other patients were examined at 3-month intervals or more frequently during periods of clinical fluctuation.

Neurologic examination protocol. A neurologic history and examination form (figure 1) was completed by a neurologist experienced in the evaluation of MS patients. Normal neurologic function was graded as zero, with 1+, 2+, 3+, and 4+ indicating increments of activity (mild, moderate, severe, or maximally increased), and grades of −1, −2, −3, and −4 indicating decrements of functional activity (mild, moderate, severely reduced, or absent). The grading system was applied to mental status, cranial nerves, motor system, sensation, and tendon reflexes.

Neurologic rating scale (NRS). The assignment of points in the NRS directly reflects the examiner's clinical assessment of each component in the neurologic examination (table 1). An intact system receives the full "normal" point value, with a progressive loss in points for mild (−1; 1+), moderate (−2; 2+), or severe (−3, −4; 3+, 4+) involvement. Severe (−3; 3+) and maximal (−4; 4+) deficits are scored as severe on the NRS. A category for important subjective symptoms such as bladder, bowel, or sexual dysfunction was incorporated, because there is no simple way to measure central autonomic function. The total point distributions for the several systems are specifically weighted for common fluctuating neurologic abnormalities of MS, such as visual, motor, sensory, and cerebellar signs. Tendon reflexes and Babinski responses, more often present than other signs, are less emphasized. Disorders of cognition, affect, and mood are also included. The final score is obtained by noting the assigned points in each of the columns and adding the subtotals. A neurologically normal individual would have a score of 100 points.

Results. The NRS has been applied to 250 MS patients. Serial NRS scores for six MS patients are provided (figure 2). The numerical value for each patient encounter measures residual neurologic function. Clinically significant exacerbations were manifested as negative deviations from baseline values, and clinical improvement led to an increase on the NRS scale (eg, figure 2, patient A). Kurtzke Disability Status Scores (DSS)[4] provide a correlation with the status of the patient but were less sensitive to clinical changes than were NRS scores (figure 2, patients A-F).

Four neurologists independently scored the NRS

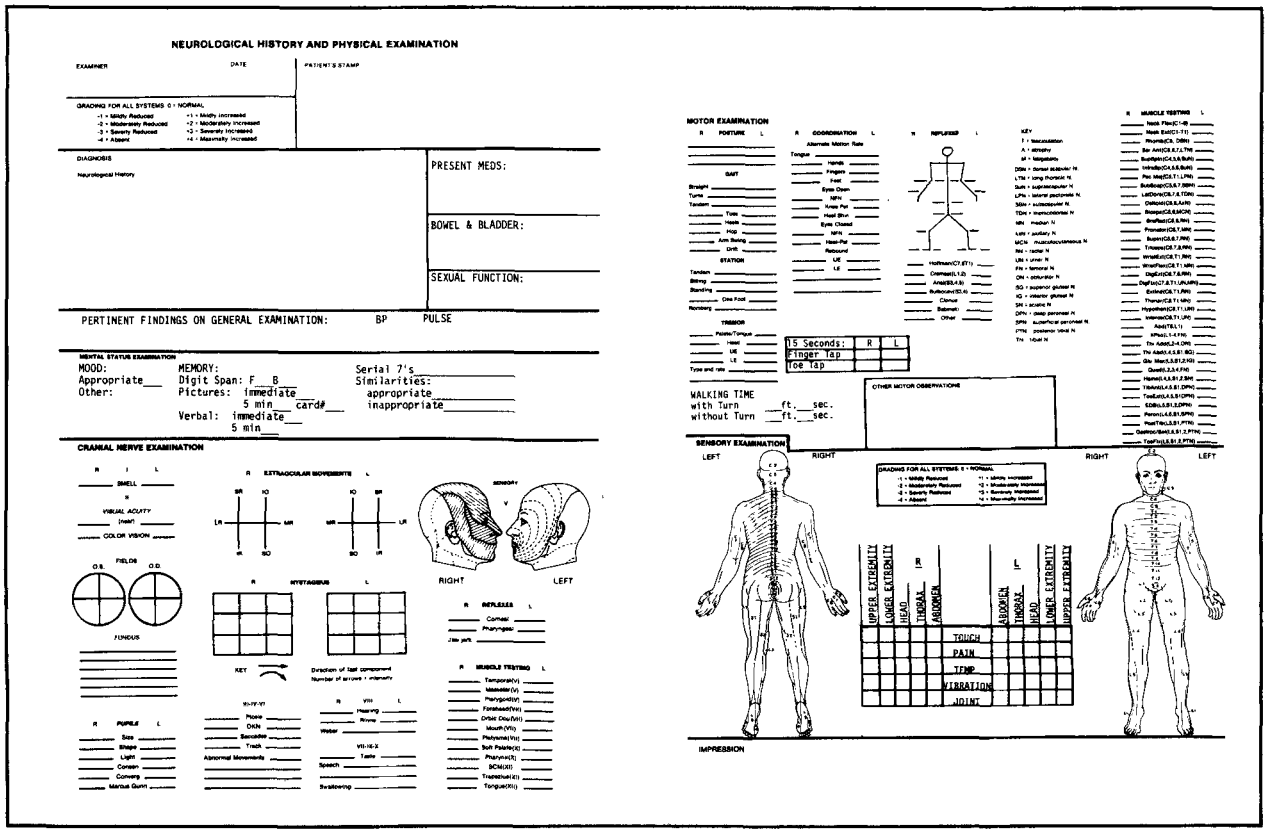

Figure 1. Neurologic history and examination form completed by the examining neurologist at each patient encounter. The grading convention for all systems is noted at the upper left corner of the form.

Table 1. Scripps Neurological Rating Scale (NRS) worksheet*

System Examined		Maximum Points	Normal	Degree of Impairment Mild	Mod.	Severe
Mentation and Mood		10	10	7	4	0
Cranial Nerves:	Visual Acuity	21	5	3	1	0
	Fields, Discs, Pupils		6	4	2	0
	Eye Movements		5	3	1	0
	Nystagmus		5	3	1	0
Lower Cranial Nerves		5	5	3	1	0
Motor:	RU	20	5	3	1	0
	LU		5	3	1	0
	RL		5	3	1	0
	LL		5	3	1	0
DTRS:	UE	8	4	3	1	0
	LE		4	3	1	0
Babinski:	R; L (2 ea)	4	4	—	—	0
Sensory:	RU	12	3	2	1	0
	LU		3	2	1	0
	RL		3	2	1	0
	LL		3	2	1	0
Cerebellar:	UE	10	5	3	1	0
	LE		5	3	1	0
Gait; Trunk and Balance		10	10	7	4	0
Special Category: Bladder/Bowel/Sexual Dysfunction		0	0	−3	−7	−10
Totals		100				
Neurological Rating Scale Score						

* Points assigned for each component of the neurologic examination are subtotaled, and points for autonomic dysfunction are subtracted, leaving the final (NRS) score.

for five individual patients, using the neurologic history and examination form that had been completed by an examining physician; the resulting NRS scores were in close agreement (table 2), with a range of variability less than 2.6%.

Discussion. The MS NRS has been introduced and tested as a clinical indicator in the evaluation of patients with MS. It provides a rapid summation of neurologic function as objectively measured by the neurologic examination (figure 1) and, in practical terms, provides a convenient quantitative base of information (table 1 and figure 2) for neurologic functions of MS patients who are followed serially.

NRS scores are more sensitive indicators of clinical change than the Kurtzke DSS, allowing for rapid recording of clinical changes that may not be identified in the DSS (figure 2). Isolated new clinical findings during an exacerbation, such as internuclear ophthalmoplegia, can make up to a 10-point change in the NRS score without altering the DSS. For example, patients D and E (figure 2) had 17-point and 20-point declines in NRS scores, while DSS scores did not change. Similarly, the NRS score of patient A (figure 2) revealed a 17-point improvement, while the DSS was unchanged. Analyses based on the DSS alone may mask important changes in neurologic function.

The NRS is not intended to replace the DSS, but is a more sensitive, complementary clinical method. It is suited for clinical studies with serial observation and may be used with other objective measurements such as neurophysiologic studies, spinal fluid examinations, or other laboratory data. The NRS can be used in either a prospective or retrospective manner, depending on the study design. Because it is simple,

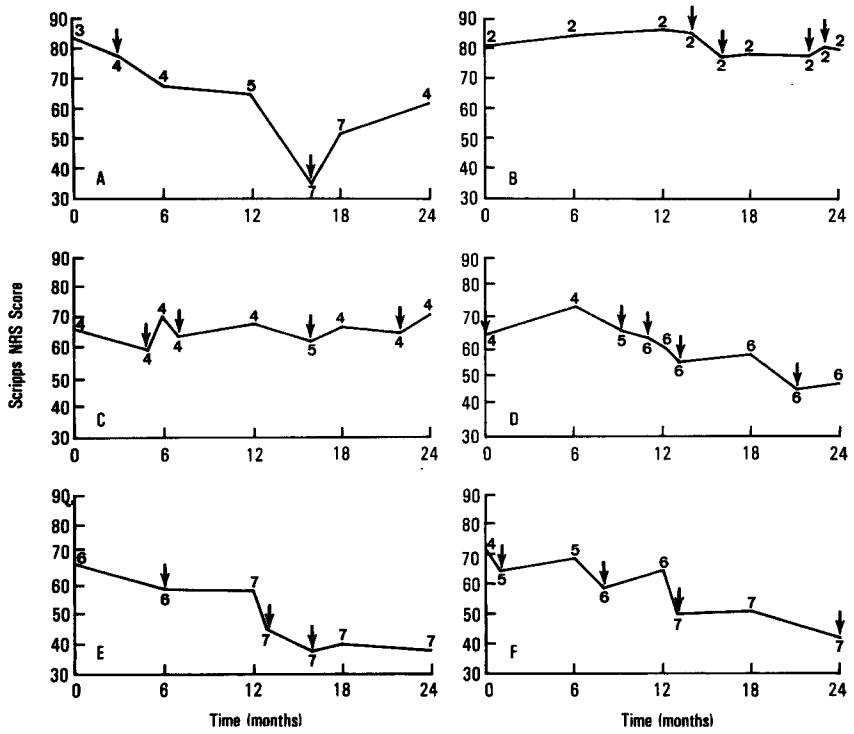

Figure 2. Composite illustration of the clinical course for six patients (A-F). The solid line represents the NRS score, and arrows indicate clinical exacerbations of MS. Kurtzke DSS scores are indicated numerically below *the line during exacerbations and* above *the NRS line during periods of clinical stability or improvement.*

Table 2. Comparison of Scripps NRS scores by four neurologists*

Pt	Dr. 1	Dr. 2	Dr. 3	Dr. 4	Mean ± SD
1	98	96	98	94	96.5 ± 1.9
2	86	81	81	84	83.0 ± 2.5
3	65	67	65	64	65.3 ± 1.3
4	74	70	72	68	71.0 ± 2.6
5	53	56	54	52	53.8 ± 1.7

* Each physician independently scored the same neurologic examination (previously recorded by another physician) for each of five individual patients.

the NRS may also be used to follow the course of disease or the effectiveness of treatment in non-research patients.

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Transient cataplexy after removal of a craniopharyngioma

Article abstract—We studied a patient with cataplexy secondary to a surgical lesion that involved the perichiasmal hypothalamus. We believe that this lesion interfered with the hypothalamic mechanism for *timing* sleep and wakefulness, whereas the pontine mechanism for *generating* sleep cycles remained relatively intact.

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Narcoleptic patients (whether falling asleep at night or inappropriately during the day) frequently begin sleep with a rapid eye movement (REM) period. Cataplectic attacks are polygraphically indistinguishable from REM sleep and often develop into full-blown REM sleep episodes.[1,2] The etiology is unknown but may become better understood by study of symptomatic cases that follow anatomically discrete structural lesions of the brain, as in the following case.

Case report. A 14-year-old girl was admitted in March 1966 for evaluation of metabolic bone disease; she had slipped femoral capital epiphyses at age 12 and an atraumatic left tibial fracture at age 14. She had developed normally, except that she was always below average in height. Occasional frontal headaches had appeared at age 13, and she required glasses for reading. Menstruation had not occurred. She weighed 49.8 kg (75th percentile) and was 140 cm in height (5th percentile). Pubic hair was present, and there was early breast development without areolar pigmentation. Examination was normal except for diminished visual acuity in the right eye (20/80) and bitemporal visual field defects. Skull films revealed an enlarged sella turcica (3 cm in maximal dimension), demineralized posterior clinoid processes, and extensive suprasellar calcification. EEG showed paroxysmal bursts of bilaterally synchronous high-voltage theta activity without lateralizing or focal features.

At surgery, a huge tumor was found to occupy the suprasellar region. The entire optic chiasm was forced upward to the left, the right optic tract appeared destroyed, and the mass extended upward in the midline behind the chiasm, pushing the third ventricle upward and backward. The tumor was removed completely; pathologic examination revealed craniopharyngioma and hypothalamic neurons. Postoperatively, her visual acuity was 20/500 in the right eye and 20/200 in the left eye, and she had a left homonymous hemianopia.

She was discharged taking phenytoin. In the next year, she required thyroxine, Pitressin Tannate, cyclic estrogens, and hydrocortisone during periods of stress or infection. In 1967, she returned to school. Her full-scale IQ was 108.

Description of sleep disturbance. In October 1968, she began to fall asleep in class, sometimes suddenly and without warning. In December 1970, she had "fainting" spells that lasted 60 to 90 seconds with apparent unconsciousness and loss of postural tone. These increased in frequency from once weekly to several daily by early 1971. In some attacks, she had hallucinoid dreams. There was no family history of sleep disturbance. Examination was unchanged, except for appearance of bilateral optic atrophy; she weighed between 55 and 65 kg and was 148 cm in height. EEG revealed frequent paroxysmal bursts of diffuse bilaterally synchronous high-voltage theta activity; spike discharges were occasionally seen not confined to the operative area. Her symptoms were unchanged after trials of phenytoin, phenobarbital, and ethosuximide in varying dosages and combinations.

She was readmitted in February and April of 1971. In the attacks, she lost postural tone without warning. If she were standing, she would gradually fall to the floor; if she were in bed, her head would fall back onto the pillow. Eyes were closed, but rapid movements of the globes could be seen beneath the lids. Tendon reflexes were not obtained. No movements, incontinence, injury, cyanosis, diaphoresis, or altered pulse or respiration were seen. She would awaken immediately if spoken to or pinched, apparently aware that she had had a spell. She often reported "bad dreams," incorporating the awakening

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