The Clinical Dementia Rating (CDR) : Current version and scoring rules

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Stiff-man syndrome treated with intrathecal baclofen

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Stiff-man syndrome is a rare, progressive motor disorder causing disabling muscle rigidity and spasms, which is often associated with insulin-dependent diabetes mellitus. In patients with this syndrome, Solimena et al[1,2] documented the presence of autoantibodies against glutamic acid decarboxylase (GAD), an enzyme selectively concentrated in pancreatic beta cells and CNS neurons secreting the major inhibitory neurotransmitter γ-aminobutyric acid (GABA). Although plasmapheresis has been tried,[3] diazepam remains the most effective treatment; it acts on the GABA receptors, making them more sensitive to GABA.[4] Unfortunately, as the disease progresses, diazepam becomes less effective and severe spasms occur. Baclofen is a direct GABA-B agonist that does not require GABA secretion to activate its receptor. Thus, baclofen would logically be a useful medication for treating stiff-man syndrome if it could be provided in sufficiently high concentration to the spinal cord receptors. We have treated severe spasticity of spinal origin with intrathecal baclofen and found that it significantly reduces spasms and rigidity when oral medications failed.[5] We now report similar success with two patients with stiff-man syndrome.

Patient 1. A 42-year old man reported 7 years of progressive rigidity and spasms that began in his trunk musculature and gradually involved both legs and made walking difficult. Diazepam had afforded initial relief, but as the disease progressed, he became disabled and unable to work at his job as a machinist. Pain became worse as the intensity and frequency of spasms increased. He gradually elevated his diazepam dose to 140 mg/d, without significant improvement. At age 34, he developed diabetes mellitus and was placed on insulin. His only neurologic abnormalities were demonstrated on motor examination. He had increased muscle tone in his axial muscles, with a pronounced lumbar lordosis. Mild sensory stimulation or volitional movements would set off severe spasms in his back and legs. His reflexes were normal and a Babinski sign was not present.

A lumbar puncture was performed and 75 μg baclofen (Ciba-Geigy, Basel, Switzerland) was injected. One hour later, his rigidity and spasms completely disappeared and he was able to walk normally. By 8 hours, the effect was decreasing, and by 24 hours he returned to baseline. A programmable drug pump and catheter (SynchroMed, Medtronic, Inc., Minneapolis, MN) going to the lumbar subarachnoid space was implanted, and his spasms were completely controlled with baclofen 120 μg/d. Over the next 2½ years, the dose of intrathecal baclofen was gradually increased to 1,200 μg/d to suppress his spasms; diazepam has been reduced to 20 mg/d, but is still necessary in low dose. He has had an episode of meningitis requiring antibiotics and replacement of the pump and catheter system, and twice the catheter has dislocated from the subarachnoid space and needed replacement. Each time he stopped receiving intrathecal baclofen his spasms recurred.

Patient 2. A 63-year-old woman was diagnosed with stiff-man syndrome in 1988. At that time, she had a one-year history of progressive spasms in her back and legs and difficulty with gait, and she required a walker. Pain occurred during attacks of spasms. On motor examination, she had hyperextension of the lumbar spine and severe spasms at the hips and knees, without weakness, atrophy, or fasciculations. The reflexes were normal and a Babinski sign was not present. An extensive work-up was normal except for antibodies to GAD in her CSF (by Dr. DeCaricelli at Yale University). Diazepam was given and helped initially, but a year later she was bedridden and having markedly increased pain. Intrathecal preservative-free methylprednisolone and systemic immunotherapy with azathioprine and plasmapheresis were tried without significant effect. By 1992, she had developed ankylosis at her hip, knees, and feet, secondary atrophy of her leg muscles, and severe pain due to her spasms. Oral narcotics only provided partial relief, and on numerous occasions she was admitted for intravenous narcotics.

Intrathecal baclofen was tried, and 75 μg produced excellent pain relief, stopping the spasms for 4 hours. A drug pump was then implanted, with a constant infusion of 120 μg/d over the last 9 months. She has good control of her spasms and pain. She remains bedridden because of lower-extremity muscle atrophy and ankylosis.

Discussion. Both patients meet the criterion for stiff-man syndrome outlined by Lorish et al.[6] Diazepam was initially effective, but as the disease progressed even high doses (>90 and >140 mg/d) were inadequate. Since diazepam works by potentiation of the effect of endogenously released GABA on the GABA receptor, disease progression may reflect lower GABA availability as GABAergic neurons are lost. Baclofen is a direct agonist of GABA-B receptors and does not require endogenous GABA. This may explain its usefulness in our two patients. Unfortunately, the second patient developed severe muscle and skeletal changes, and these changes have not reversed. Her major problem of painful spasms has been controlled. The first patient was treated earlier in his course and fared better; he continues to do well after 2½ years of intrathecal baclofen treatment.

Intrathecal baclofen is now approved for the treatment of spasticity of spinal origin, and is available at many centers. On the basis of our experience with these two patients, we recommend testing patients with stiff-man's syndrome with a lumbar subarachnoid 75-mcg baclofen bolus injection when oral diazepam is no longer providing adequate symptomatic relief. If successful, a drug pump can be implanted for chronic administration. If our observations on two patients are confirmed in other patients with stiff-man syndrome, it will be the first example of direct replacement of a neurotransmitter by intrathecal infusion.

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The Clinical Dementia Rating (CDR): Current version and scoring rules

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The Washington University Clinical Dementia Rating[1] (CDR) is increasingly used in longitudinal studies and clinical trials for staging the severity of Alzheimer's disease (AD). The CDR is derived from a semistructured interview with the patient and an appropriate informant and rates impairment in each of six cognitive categories (Memory, Orientation, Judgment and Problem Solving, Community Affairs, Home and Hobbies, and Personal Care) on a five-point scale in which none = 0, question-

Table. Clinical Dementia Rating (CDR)

	Impairment				
	None 0	Questionable 0.5	Mild 1	Moderate 2	Severe 3
Memory	No memory loss or slight inconstant forgetfulness	Consistent slight forgetfulness; partial recollection of events; "benign" forgetfulness	Moderate memory loss; more marked for recent events; defect interferes with everyday activities	Severe memory loss; only highly learned material retained; new material rapidly lost	Severe memory loss; only fragments remain
Orientation	Fully oriented	Fully oriented except for slight difficulty with time relationships	Moderate difficulty with time relationships; oriented for place at examination; may have geographic disorientation elsewhere	Severe difficulty with time relationships; usually disoriented to time, often to place	Oriented to person only
Judgment and Problem Solving	Solves everyday problems and handles business and financial affairs well; judgment good in relation to past performance	Slight impairment in solving problems, similarities, and differences	Moderate difficulty in handling problems, similarities, and differences; social judgment usually maintained	Severely impaired in handling problems, similarities, and differences; social judgment usually impaired	Unable to make judgments or solve problems
Community Affairs	Independent function at usual level in job, shopping, and volunteer and social groups	Slight impairment in these activities	Unable to function independently at these activities although may still be engaged in some; appears normal to casual inspection	No pretense of independent function outside home Appears well enough to be taken to functions outside a family home	Appears too ill to be taken to functions outside a family home
Home and Hobbies	Life at home, hobbies, and intellectual interests well maintained	Life at home, hobbies, and intellectual interests slightly impaired	Mild but definite impairment of function at home; more difficult chores abandoned; more complicated hobbies and interests abandoned	Only simple chores preserved; very restricted interests, poorly maintained	No significant function in home
Personal Care	Fully capable of self-care		Needs prompting	Requires assistance in dressing, hygiene, keeping of personal effects	Requires much help with personal care; frequent incontinence

Score only as decline from previous usual level due to cognitive loss, not impairment due to other factors.

Assignment of CDR

(material added to the original rules shown in italic type)

Use all information and make the best judgment. Score each category as independently as possible. Mark in only one box for each category, rating impairment as decline from the person's usual level due to cognitive loss alone, not impairment due to other factors such as physical handicap or depression. *Occasionally, the evidence is ambiguous and the clinician's best judgment is that a category could be rated in either one of two adjacent boxes, such as mild (1) or moderate (2) impairment. In that situation, the standard procedure is to check the box of greater impairment.*

Aphasia is taken into account by assessing both language and nonlanguage function in each cognitive category. If aphasia is present to a greater degree than the general dementia, the subject is rated according to the general dementia. Supply evidence of nonlanguage cognitive function.

The global CDR is derived from the scores in each of the six categories ("box scores") as follows: Memory (M) is considered the primary category and all others are secondary. CDR = M if at least three secondary categories are given the same score as memory. Whenever three or more secondary categories are given a score greater or less than the memory score, CDR = score of majority of secondary categories on whichever side of M has the greater number of secondary categories. However, when three secondary categories are scored on one side of M and two secondary categories are scored on the other side of M, CDR = M.

When M = 0.5, CDR = 1 if at least three of the other categories are scored 1 or greater. If M = 0.5, CDR cannot be 0; it can only be 0.5 or 1. If M = 0, CDR = 0, unless there is impairment (0.5 or greater) in two or more secondary categories, in which case CDR = 0.5.

Although applicable to most AD situations, these rules do not cover all possible scoring combinations. Unusual circumstances that occasionally occur in AD and may be expected in non-Alzheimer's dementia as well are scored as follows:

(1) With ties in the secondary categories on one side of M, choose the tied scores closest to M for CDR (eg, M and another secondary category = 3, two secondary categories = 2, and two secondary categories = 1; CDR = 2).

(2) When only one or two secondary categories are given the same score as M, CDR = M as long as no more than two secondary categories are on either side of M.

(3) When M = 1 or greater, CDR cannot be 0; in this circumstance, CDR = 0.5 when the majority of secondary categories are 0.

able = 0.5, mild = 1, moderate = 2, and severe = 3. (Note: Personal Care has no questionable impairment level.) From the six individual category ratings, or "box scores," the global CDR is established by clinical scoring rules where CDR 0 = no dementia and CDR 0.5, 1, 2, or 3 indicates questionable, mild, moderate, or severe dementia. (Although alternative scoring algorithms exist, use of the clinical rules permits comparability with previously determined CDR scores.) The usefulness of the

CDR may result from several factors: (1) it is clinically based (ie, independent of psychometric test scores); (2) the six categories used for rating dementia severity are directly linked to validated clinical diagnostic criteria[2] for AD; (3) it has high inter-rater reliability for physicians[3] and nonphysicians[4]; and (4) an expanded and more quantitative version of the scale can be achieved by summing the ratings in each of the six categories to provide the Sum of Boxes.[5]

The CDR has been modified slightly over the years, as experience permitted resolution of ambiguities. These refinements primarily have been to sharpen the distinction between specific severity levels within a category. The first revision[6] separated Community Affairs box scores 2 and 3, eliminated the possibility that Home and Hobbies could be "well maintained" for box score 0.5, and removed vague modifiers ("if any" from box score 0.5 in Community Affairs and "occasional" from box score 1 in Personal Care). The next revision[7] distinguished Orientation box scores 0 and 0.5; other changes were slightly reworded descriptions for Orientation box scores 1 and 2 and for Judgment and Problem Solving box score 1, deletion of the modifier "may still" from the box score 1 description of normal appearance in Community Affairs, and substitution of "slight" for "mild" or "only doubtful" to describe 0.5 impairment for Memory, Judgment and Problem Solving, and Community Affairs.

A new version of the CDR more appropriately uses information regarding performance of financial transactions for rating Judgment and Problem Solving rather than Community Affairs. The new version is presented here (table) for interested readers, along with improved clinical scoring rules for the global CDR (material added to the original rules[1] is shown in italic).

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'Reappearance' of congenital nystagmus after minor head trauma

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Congenital nystagmus (CN) is typically horizontal, conjugate, and is noticed at birth or in early infancy. Although the appearance of CN does not usually change much throughout life, fluctuations in the amplitude of the nystagmus are common. We report a patient with congenital nystagmus that was first noticed at age 6, "resolved" at age 7, and became clinically apparent again after a minor closed-head injury at age twelve.

Case report. A 12-year-old boy was evaluated for reappearance of nystagmus after a minor head injury. His parents first noticed a head-tilt when he was 2 to 3 years old. Ophthalmic evaluation at age 5 showed no clinical evidence of nystagmus. Pendular nystagmus with excellent visual acuity (20/20 OU) and normal fusional amplitudes were found at age six. CT of the brain was reportedly normal except for sinus disease. The nystagmus was no longer visible at age seven. In the interim, he was an honor student and was active in sports. Two months before his examination, he was struck in the back of the head with a baseball. There was no loss of consciousness. He was brought to the hospital for evaluation. Nystagmus was noted by the ambulance driver, and it subsequently persisted. There were no other neurologic sequelae from the head injury.

Since the injury, the patient needed to position his head to see clearly, and had difficulty catching a baseball. His parents noticed an intermittent head-turn while he was doing close work.

The patient had asthma, but no other medical, ophthalmologic, or neurologic conditions. Several family members had strabismus; there was no known nystagmus in the family.

Neuro-ophthalmic examination revealed normal visual acuity, color vision (Hardy-Rand-Ritter plates), pupils, visual fields, fundi, and intraocular pressures. The extraocular movements were full. The eye movements were videotaped and reviewed. Horizontal pendular-appearing nystagmus was present in primary gaze. The amplitude was variable and, at times, no nystagmus was visible. In lateral horizontal gaze, the nystagmus had a jerk component, beating in the direction of gaze. No definite null position was seen, even with head-tilt. The amplitude of the nystagmus seemed greater at distance than at near, and there was poor generation of horizontal optokinetic nystagmus (OKN). "Inverse" OKN was not observed. There was no observable change in the nystagmus with monocular occlusion, and no head-tilt, head-nodding, or head oscillation was seen. The neurologic examination was normal.

Eye movement recordings. Eye movements were recorded using the infrared reflection technique. Full-system bandwidth was direct current to 100 Hz. The predominant waveform, jerk nystagmus with extended foveation, allowed foveation periods from 150 to 400 msec. In the figure, A and B show the position and velocity records during 4 seconds of fixation on a 0.2° diameter light-emitting diode. Note the periods of extended foveation despite the frequent bias reversals. In the figure, C shows the phase planes of both eyes during this 4-second interval, also indicating well-developed foveation within the foveation window.[1] The ±0.5° window has been extended ±0.1° to allow for the 0.1° radius of the target. The foveating saccades from both directions bring the target within this effective foveation window of ±0.6° and ±4°/sec. Pendular waveforms were not observed during the recording. There was a broad neutral zone with frequent bias reversals within ±20° of primary gaze. Convergence had a minimal damping effect on the nystagmus. Smooth pursuit was normal at low velocities, but of low gain at velocities of 20°/sec and above. The amplitude of the nystagmus was variable throughout the record, and ceased completely with inattention. There was no evidence of a superimposed acquired nystagmus.

Discussion. As is typical of persons with CN, our patient did not experience oscillopsia. His well-developed foveation periods allowed him to enjoy excellent visual acuity most of the time. High-acuity foveation periods require that the eye position and target position coincide with minimal retinal slip. During smooth pursuit, the ability to match eye position to target position during foveation periods declines with increasing target velocity.[2] In our patient, low pursuit gain with pursuit movements of 20°/sec or more accounted for his intermittent blurred vision, especially when trying to fixate on targets moving rapidly across his visual field. Although his nystagmus was not apparent in infancy, the presence of excellent vision and lack of oscillopsia suggested that the nystagmus was not an acquired type; the recorded CN waveforms confirmed this.

Our patient also demonstrated several atypical clinical features of CN: his nystagmus was not noticed until age 6, his waxing and waning course was unusual, and his nystagmus reemerged after minor head trauma. The amplitude of CN can vary with many factors, including head position, eye position, febrile illness, and anxiety. Nystagmus with typical CN waveforms has developed in adolescence or adulthood without any