The “Spray Can” Sign

Validation of a Clinical Observation in Chronic Inflammatory Demyelinating Polyneuropathy

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Background: The presentation of chronic inflammatory neuropathies is variable. The decision regarding when to intervene with treatment is ideally determined by identifying early markers of loss of function.

Objective: To test the hypothesis that an observation of functional impairment, defined by a patient with demyelinating neuropathy, can be used as a reproducible and reliable measure of improvement with intravenous immune globulin.

Design: A 28-year-old woman presented with a chronic inflammatory demyelinating polyneuropathy. Her first complaint was the inability to use her deodorant spray because of hand weakness. A calibrated pincer gauge fixed on top of her usual spray can was used to objectively test finger flexion. Tip grip and lateral pinch were also measured. A calibrated dynamometer was used to measure grip strength.

Results: Power and precision grip force were reproducible in normal control subjects by means of the spray can test. This test proved to be a reliable indicator of reduced muscle strength in the patient and improved after treatment with intravenous immune globulin.

Conclusions: The spray can test objectively quantified the daily function, nominated by the patient, of operating an aerosol can. This measurement, drawn from a functional loss observed by the patient, proved to be a portable and reliable indicator of decline and recovery in chronic inflammatory demyelinating polyneuropathy.

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INTRAVENTOUS (IV) immune globulin has been previously shown to be an effective and safe treatment for selected patients with acquired demyelinating polyneuropathy.1,2 There are few scales specifically designed for assessment of function in patients with chronic inflammatory demyelinating polyneuropathy (CIDP). In the setting of clinical practice, the frequency of treatments is usually determined by the patient's subjective analysis of decline and subsequent improvement after treatment. This is often based on the loss of a specific function, such as walking a certain distance, that can be quantified by means of standardized measurements. Few clinical studies have correlated specific subjective observations of weakness, as defined by individual patients, with an objective reproducible measurement.

Comparative studies between IV immune globulin treatment and other therapeutic modalities are accordingly limited by the variable degree and extent of weakness among individual patients. The challenge is therefore to maximize the sensitivity of the test measurement, while maintaining uniformity for statistical purposes within the study population and limiting the time and personnel required to perform the measurements.

REPORT OF A CASE

A 28-year-old woman presented in May 1995 with a 4-month history of progressive weakness of her arms and legs. Her first symptom was an inability to hold and spray her deodorant and hairspray because of weakness of the flexor digitorum profundus and superficialis muscles of her index finger and the intrinsic muscles of her hand. Her medical history was otherwise unremarkable.

Neurologic examination at first presentation showed marked weakness of upper and lower limb muscles. The weakness was most marked in the distal musculature. Manual muscle testing
showed grade 4/5 weakness in infraspinatus and biceps and 3/5 in wrist flexors and extensors and in the small muscles of her hands. Hip flexors were grade 4/5 and ankle dorsiflexors were 4-/5 on the left and 4/5 on the right. Plantar flexors were 4/5 on the left and 5−/5 on the right. She was areflexic throughout. She had reduced sensation to light touch and pinprick below the knees bilaterally and mild reduction of vibration sensation to the ankles bilaterally.

The cerebrospinal fluid showed an elevated protein level (145 mg/dL) and was otherwise normal. Electrophysiologic studies confirmed the presence of a demyelinating sensory and motor polyneuropathy. There was abnormal temporal dispersion in the median and ulnar nerves at the forearm bilaterally, and in the tibial and peroneal nerves in the lower extremities. Conduction velocity was significantly reduced throughout (eg, 25 m/s in the right median forearm, 37 m/s in the left peroneal, and 26 m/s in the right tibial), and distal motor latencies were markedly prolonged. F responses were absent in the peroneal and tibial nerves bilaterally. Sensory nerve conduction velocities were similarly slowed. Other possible causes of neuropathy were excluded. A diagnosis of chronic inflammatory demyelinating neuropathy was made.3

The patient began a 5-day course of IV immune globulin (0.4 g/kg per day), and her strength gradually improved during the following 2 to 3 weeks to a point where she could walk normally and could reengage in all activities of daily living. She reported that the first indicator of improvement was a return of ability to spray her deodorant can.

At most recent follow-up, 7 years after diagnosis, the patient was in full-time employment as an accountant and was undergoing a 3-day infusion of IV immune globulin (0.4 g/kg per day) at 6- to 8-week intervals. Each relapse was characterized by a decline in upper and lower extremity function, with difficulty walking and poor fine movements in the upper extremities. She used the loss of ability to spray her deodorant as an indicator of her need for treatment and noted that recovery on each occasion was heralded by an improvement in her ability to perform this task.

We tested the patient’s observation that her reduced ability to operate her aerosol can heralded a decline in her clinical function, and whether this loss of function could be used as an indicator of IV immune globulin response.

We developed a method to measure the strength required to operate her aerosol can and constructed a system whereby hand function and strength were assessed before, during, and 3 weeks after IV immune globulin treatment. A calibrated pincer gauge (B&L Engineering, Tustin, Calif)4,5 was fixed on top of the spray can and the patient was asked to simulate spraying the can and press as hard as she could with her index finger (Figure 1). The same spray can was used throughout testing. Maximal power of hand grip was also measured with a calibrated dynamometer,4 and tip grip and lateral pinch grip strength were measured with the calibrated pinch gauge (Table).

The reliability of our measure was assessed by means of an age-matched group of 9 healthy females.4,5 The mean of 3 measurements was taken on 2 occasions (with a 1-hour interval between the occasions). All statistical analysis was performed with SPSS (SPSS Inc, Chicago, Ill).

Before commencement of IV immune globulin treatment, power and precision grip forces were reduced compared with normative data for age-matched female controls.4,5 Improvement in hand strength occurred after IV immune globulin therapy. Improvements in “spray can strength” were consistently concordant with improvements in hand, tip, and lateral pinch grip strength with successive cycles of IV immune globulin therapy. Figure 2 depicts muscle strength during 3 cycles of treatment. Peak recovery approximated 50% to 75% of normal hand strength (Table).

The spray can test gave consistent and repeatable results when performed on the same arm in the same healthy subject on different occasions. There was a significant intraclass correlation coefficient for the mean of the 3 measurements on 2 occasions (P<.001; r = 0.94 for the right arm and r = 0.88 for the left arm).7

![Figure 1. The spray can apparatus. A calibrated pincer gauge (B&L Engineering, Tustin, Calif) was fixed on top of the spray can.](http://archneur.jamanetwork.com/pdfaccess.ashx?url=/data/journals/neur/6988/)

### RESULTS

<table>
<thead>
<tr>
<th>Muscle Strength, lb</th>
<th>Patient</th>
<th>Control Subjects*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grip</td>
<td>47.3</td>
<td>74.5 ± 13.9</td>
</tr>
<tr>
<td>Spray can</td>
<td>10.2</td>
<td>13.6 ± 1.9</td>
</tr>
<tr>
<td>Tip pinch grip</td>
<td>8.8</td>
<td>10.3 ± 2.3</td>
</tr>
<tr>
<td>Lateral pinch grip</td>
<td>11.6</td>
<td>18.0 ± 2.6</td>
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</tbody>
</table>

*Mean ± SD.
Measurement scales are important tools to determine effective outcomes after treatment. Although rating scales are used most extensively in clinical trials, validated objective measurements are also valuable in a clinical setting where the efficacy of a therapeutic intervention may differ between patients, or where the efficacy of the treatment in individual patients is doubtful. Not all patients with CIDP respond to IV immune globulin, and in some cases, the treatment becomes less effective over time. Furthermore, the required frequency of treatments varies from patient to patient. In the absence of a specific scale that accounts for these variables, the determination of the frequency of treatments and/or the decision to seek alternate treatments can be difficult.

An ideal scale should be valid, reproducible, efficient, sensitive to change in the underlying condition, and specific to the pathophysiology of the condition. Few validated scales have been designed specifically for CIDP. Those that have been used most frequently in clinical trials include generic disability scores and a combination of objective functional assessments. These scales are limited to an extent by their failure to account for the asymmetry of CIDP in the generation of composite disability scores, by the potential for interobserver and intraobserver bias and impaired reliability on testing-retesting, and by the nonquantitative nature of describing functional impairment.

The Neuropathy Impairment Scale (previously called the Neurologic or Neuropathy Disability Score, or NDS) or variations of this have been widely used to measure neurologic deficit in CIDP as well as many other neuropathies. It has been validated and has a high degree of reproducibility. It is, however, a global score of muscle weakness and reflex and sensory abnormality; although it is a good measure of global neuropathic abnormality, it is time consuming and may lack sensitivity for detecting early asymmetric focal weakness indicative of impending relapse and need for treatment as in our case.

More recently, the Inflammatory Neuropathy Cause and Treatment (INCAT) Disability Scale was designed specifically for neuropathy. The scores of this scale range from 0 to 5 each for upper and lower limbs and are based on a combination of symptoms and functional impairment. The overall score is based on the sum of disability in arm and leg. Although specific for the pathophysiology, this scale has not been fully validated, and the disability measurements may be subject to different interpretations by different observers. Furthermore, the scale partially depends on defined functional impairment, eg, symptoms of impairment in zipping and buttoning, washing or brushing hair, using a knife and fork together, and handling small coins, rather than on functions that may be of particular importance to the individual patient. Moreover, the determination of whether an action is impaired is to an extent subjective, which may limit the sensitivity of the test. Other scales have attempted to address the symptomatic component of disability by use of a rating system in which patients are examined with respect to a series of defined tasks (eg, 9-hole peg test, maximal grip strength, etc). However, such rating scales have not always been adequately validated or objectively verified. For example, there is no correlation between grip strength and performance in the 9-hole peg test in stroke patients, nor is there a correlation between muscle grip strength and the ability to open a jar in a normal population. Furthermore, scales that are scored as “mild,” “moderate,” or “severe” are limited by the risks of observer bias and should not be used as primary end points, as different observers will define these measures differently.

Given the limitations of existing clinical rating scales for CIDP, we sought to determine whether the observation of functional decline made by a patient whose condition had been stabilized with IV immune globulin for more than 5 years could be validated and used as an objective measure of early decline and recovery of function. Our patient based her decision to seek repeat infusions of IV immune globulin on the loss of her ability to operate her aerosol deodorant. During the early years of her treatment, she had noted that the decline in her ability to operate her deodorant was a reliable early indicator of impending relapse, and that recovery of this function signaled a general improvement in muscle strength. This specific functional impairment was therefore used by the patient as a surrogate marker of her disease activity. We sought to evaluate the reliability and validity of this observation, with a view
to developing an individually tailored scale that could be used in the future to predict the efficacy of her treatment. We demonstrated that objective measurement of strength required to operate the aerosol correlated well with other accepted quantitative tests.

We have shown by this series of observations that it is possible to objectively quantify a daily function nominated by a patient, such as operating an aerosol can, and to subsequently use this function as a reliable indicator of both decline and recovery in IV immune globulin–responsive demyelinating neuropathy. That a subjective functional improvement can be directly translated into an objective and reproducible test is important in the development of clinical trials for patients with demyelinating neuropathy who are stable with IV immune globulin therapy, and for whom it is desirable to explore the efficacy of other non–blood-based therapeutic modalities. We submit that such an approach, which is focused on functional loss as defined by the patient and validated by test-retest and across control subjects, can be used effectively to evaluate the efficacy of new treatments against the efficacy of repeated infusions with IV immune globulin. We further submit that this approach could be developed for more general use as a sensitive, valid, and reliable scale for measurement of outcome in CIDP or to complement other scales.

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REFERENCES


