Dysfunction scores

# Polyneuropathy dysfunction scores

**P J Dyck, P C O'Brien**

Three polyneuropathy scores are described, which seem to be valid and sensible measures to score dysfunction and disability in patients with generalised motor neuropathies

In this issue of the *Journal of Neurology, Neurosurgery and Psychiatry* are three closely related reports on scores for assessing the functional disability of patients with polyneuropathies. A further purpose may be to assess the severity of the polyneuropathy itself. Most patients studied had immune polyneuropathies and generalised weakness. In the first one, Graham and Hughes[1] describe ''a new peripheral neuropathy activities measure, the Overall Neuropathy Limitations Scale (ONLS)'', a slight modification of the Overall Disability Sum Score (ODSS) ***(see p 973)***. From responses of patients to questions about limitations of acts of daily living and by observation of these activities, a sum score of limitations is derived. Activities such as ''wash and brush hair, turn a key in the lock, difficulty in walking'' and other motor activities are included. The patient and observer (when the activities were witnessed) judged each activity: not affected, affected but not prevented or prevented. The investigators report good inter-rater reliability, good correlation with results of the ODSS and 36-item Short Form (SF-36) Physical Component Summary scores. The performance of this limitations score was good, whether the activity was observed or unobserved. They list simplicity as its favourable feature.

In a second study by the same two investigators,[2] the limitations of walking and running were assessed (Walk-12—a previously published multiple sclerosis scale). The patient completes a pencil and paper questionnaire on the limitations of ability in walking, running, climbing stairs and balancing. The choices for the responses are given as degrees of limitations: not at all, a little, moderately, quite a bit and extremely. The items and severities are combined into an overall score. On the basis of this study, also mainly on patients with immune polyneuropathies and generalised weakness, the scale had strong performance characteristics ***(see p 977)***.

In the third study, Merkies and Schmitz[3] compare the ODSS to other disability scores—that is, Guillain–Barré syndrome, Rankin, 10-m walking test and 9-hole peg test. They report that the performance of ODSS was superior to that of the other disability scores ***(see p 970)***.

To put the three studies into a larger context, we ask how well the three scores reflect overall severity of a patient's functional disability related to polyneuropathy or to severity of the peripheral neuropathy itself at a point in time. Are they suitable for all varieties of neuropathy? Should they be used as primary or secondary outcome measures in controlled clinical trials? In epidemiological surveys? In medical practice? Previous reviews have discussed some of these issues.[4–8]

How should the severity of a patient's polyneuropathy be expressed? The severity of a patient's polyneuropathy is obviously the sum of a patient's symptoms, neurological signs, test abnormalities, dysfunctions and other adverse outcomes. The scales outlined here seem to provide a comprehensive and sensible evaluation of the acts of daily living, walking, running and climbing stairs, which seem to be good measures of disability in patients with polyneuropathies with generalised weakness. They are probably not suitable measures for sensory, autonomic or sensorimotor polyneuropathies, which are a large group of infectious, inflammatory, metabolic, deficiency, toxic and immune disorders.

Are these scales good primary outcome measures for use in prospective controlled clinical trials? The investigators of these three reports may wish to use these scales for this purpose, but we think there may be better primary outcome measures. For primary outcome measures, we prefer direct measures of symptoms, weakness, sensory loss, autonomic dysfunction and nerve tests directly attributable to disease of the nerves. To illustrate, attributes of nerve conduction, especially the use of composite scores of nerve conduction (eg, Σ5NC nds), are especially useful as primary measures, because they are objective (results cannot be willed by the patient), are attributable to disease of the nerves and nothing else, are sensitive and specific, correlate with neurological deficit and provide continuous measures over a broad range of severities.[9 10] We have also found that composite scores of nerve conduction (and pulse variation with deep breathing or the Valsalva manoeuvre) have the characteristic of being a monotone measure (they show a noticeable trend of worsening over time, given that such worsening occurs).[10] Another characteristic that is needed, at least for a multicentre trial, is generalisability to multiple centres. This characteristic is the degree to which a clinical instrument gives the same test value at different participating centres. The low inter-rater variability of some of the described tests is promising in this regard, but it needs to be tested in more and disparate medical centres.

By contrast, disability scales are not direct measures of nerve dysfunction or impairment. Other diseases (eg, connective tissue or joint disease), poor volition, medicolegal gain and psychological changes may affect the performing of acts of daily living. It is for this and other reasons that the US Social Security Administration suggests that disability should be based on a doctor's judgement based on examination of impairment (changed function or structure).[11]

What is the special role and value of disability scales? They provide additional characterising information on the benefits (or lack) of the intervention in patients' lives. They indicate whether the treatment makes a real difference in the lives of the patients and from their perspective. It is, therefore, an important independent measure of the meaningfulness of the intervention.

Are these scales suitable for office medical practice (eg, to monitor severity of neuropathy) so that immune-modulating treatment can be increased or decreased? We think they may serve this purpose. But here also we prefer to use summed scores of neurological signs (eg, Neuropathy Impairment Score) or summed attributes of nerve conduction expressed as normal deviates (eg, Σ5NC nds), because they are more direct measures of nerve dysfunction and are more objective, sensitive and specific.

In conclusion, the three polyneuropathy scores described here seem to be valid and sensible measures to score dysfunction and disability in patients with generalised motor neuropathies. They may not be ideal for sensitively recognising or tracking severity of sensorimotor, focal or multifocal neuropathies. We think there are other more sensitive and objective primary outcome measures for use in controlled trials, but that they

may be excellent secondary outcome measures, as they provide a meaningful measure of motor dysfunction for acts of daily living.

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Subarachnoid haemorrhage

# Subarachnoid haemorrhage in patients ⩾75 years: clinical course, treatment and outcome

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In most Western societies, the percentage of patients ⩾60 years has been increasing over the past century. This has become a challenge both economically and socially. Medical science, which initially increased life expectancy, now faces the task of predicting outcome early in an unexpected disease such as subarachnoid haemorrhage (SAH). This is difficult, particularly in the geriatric population ⩾75 years. The study by Nieuwkamp *et al* ***(see p 933)***[1] deals with this important issue in a cohort of 170 patients from two different institutions, who were studied retrospectively. Overall favourable outcome was seen in 15% of patients in the age group. Predictors of unfavourable outcome were poor clinical condition on admission, with recurrent haemorrhage and subdural extension of the haematoma.

In the past, it has been hypothesised that SAH should not be treated after the age of 60 years.[2] Although later studies have shown that patients may benefit from being treated in the seventh and eighth decades of life,[3] it has remained controversial at what age treatment cannot improve the outcome anymore. The role of concomitant medical disease may also be important, but it is yet to be discussed in the literature adequately. In addition, predictors of favourable outcome have not been defined prospectively. Most neurosurgeons are hesitant to treat patients ⩾75 years as intensely as younger patients, because unfavourable outcomes are more likely. This may have produced a bias in the retrospective analysis and the results may reflect what has been expected from the beginning. For future investigations, some of the following considerations may be helpful.

The geriatric population is different from the younger adult population. As shown by other investigators, the rupture rate of aneurysms is higher and the outcome is more likely to be unfavourable.[3 4] Important studies like the International Study of Unruptured Intracranial Aneurysms that investigated the natural history of unruptured aneurysms, however, did not stratify their analysis into different age groups.[5]

No prospective randomised controlled trials (RCTs) have looked into the subset of the geriatric population. In general, elderly patients have been treated in the same manner as other adults. Early treatment of aneurysms has been shown to be beneficial for adult patients after SAH. So long as geriatric patients are not accepted as representing a special subset similar to paediatric patients, there probably never will be an RCT. At this point, it is hard to justify that elderly patients should be randomised in trials stratified into treating aneurysms either by clipping or coiling, or by non-interventional or non-operative conservative treatment. In