Quality-of-life scales for neurologic diseases

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Originally released May 5, 2009; last updated April 4, 2014; expires April 4, 2017

Introduction

Overview

Neurologic diseases can have profound impact on all areas of a patient’s life, including cognitive, physical, sensory, emotional, and social functioning. Because few of these diseases are curable, treatment tends to focus on limiting disease progression and relieving symptoms. Therefore, health-related quality-of-life (HRQL) outcomes that assess functional ability and well-being are important for evaluating research and clinical practices. HRQL information is best obtained by asking patients directly, using validated questions with standardized response options. HRQL measurement tools are readily available and can easily be incorporated into practice without adding burden to the clinician. In this article, the authors review the value of collecting HRQL information, the types of HRQL measures that can be used for neurologic diseases, considerations to use when selecting a measure, and recent initiatives that can make HRQL assessment in neurology practical and clinically useful. New HRQL measurement tools, guidance documents, and potential uses of HRQL measures are presented.

Key points

• HRQL assessment provides patient-centered information that can be useful for research, clinical care, and performance monitoring.
• Selection of HRQL measures should be guided by the purpose of the assessment, the setting in which it will be used, and the characteristics of the measure.
• NIH-funded measurement systems, such as Neuro-QOL, provide standard assessment tools that can enable cross-disease comparisons along with disease-specific results.

Rationale for measuring HRQL

Neurologic diseases may affect any aspect of cognitive, physical, sensory, emotional, and social functioning. Whether acute or insidious, stable or progressive, such disorders tend to have chronic and long-lasting impact. Because few neurologic diseases are curable, medical care tends to focus on limiting disease progression, symptom management, and maximizing quality of life. These objectives are sometimes not well-reflected in traditional clinical measures such as lab and radiological results. In fact, although these traditional tests remain essential for disease and treatment monitoring, they fail to capture many of the outcomes that are important to patients. They may miss patients’ direct experience of how the disease or its treatment has affected them. Patients frequently care most about HRQL outcomes because those are the ones that reflect their symptom experience and their ability to function meaningfully in their environment. After dealing with the distress associated with diagnosis and treatment demands, patients must appraise their situation and adapt to changes imposed by their condition. Symptoms and functional abilities drive the patient’s perception of the burden of that disease on themselves and their families. It is, therefore, important to assess HRQL to obtain that perspective, which can be surprisingly different from standard clinical severity measures. Some potential advantages of conducting formal HRQL evaluation include:

• Identifying symptoms or concerns that might not be spontaneously reported.
• Improving patient-provider communication.
• Increasing patient participation in the clinical decision-making process.
• Engaging patients in selecting from clinically equivalent treatment options.
• Monitoring response to treatment or change over time.
• Monitoring performance/quality of care.

Routine collection of HRQL information from patients is still relatively uncommon, though the recent publication of guidance documents (International Society for Quality of Life Research 2011; Snyder et al 2012) may facilitate increased collection efforts. Barriers to implementation include logistical difficulties, lack of infrastructure, and provider uncertainty about the clinical value of collecting HRQL information (Rose and Bezjak 2009). However, studies in general medical practice and specialty areas such as oncology show it can be feasible, efficient and acceptable to both
patients and providers and improves communication. A standard, computerized, HRQL questionnaire before a clinic visit with a summary of the results for the patient and/or provider immediately prior to the clinical encounter significantly increases discussion of HRQL issues, particularly those that are less observable (eg, social functioning) or of a diffuse quality (eg, fatigue) and heightens physician awareness of patients' HRQL (Detmar et al 2002; Velikova et al 2002).

Systematic evaluation of the impact of HRQL assessment on treatment and on patient involvement in decision making is in its early stages, with results being inconsistent (Valderas et al 2008). Nevertheless, HRQL information can potentially help move the scope of interventions beyond clinical impairment and closer to patient-centered care. For instance, attempting to establish good seizure control with minimal adverse medication effects is essential to treating epilepsy. Epilepsy also impacts psychological, social, and school work functioning. It is associated with academic underachievement in children, un- or under-employment in adults, restricted social activity, stigma, and psychological distress for children and adults (Baker 2002; Sabaz et al 2003). Knowledge of these other effects provides clinicians with additional targets for intervention as well as other potential indicators of treatment success (Ronen et al 2003). Although direct questioning about each of these areas during the actual clinical encounter is possible, collecting this information via HRQL measures that are completed and scored prior to the encounter makes it more feasible.

In the research setting, HRQL outcomes are generally accepted as valid indicators of how well a treatment or intervention works. HRQL outcomes can complement and enrich findings based on more traditional measures. For example, in a randomized, double-blind comparison of lamotrigine and valproate for the treatment of seizures, patients taking lamotrigine were significantly more likely to report improvements in their health, levels of energy or fatigue, mood, and social functioning (Sackellares et al 2002). This occurred even though both groups showed equivalent levels of seizure control. Additional controlled studies have found minor HRQL benefit of lamotrigine over other antiepileptic drugs when used for partial seizures (Marson et al 2007a) but not general seizures (Marson et al 2007b). There are insufficient good-quality data on HRQL effects of antiepileptic drugs to draw any firm conclusions (Wilby et al 2005), but this example does illustrate how such information can be useful during medical decision making. It is also important during the drug and treatment approval process and when making decisions about allocation of health care resources.

**What is HRQL?**

HRQL, a narrower concept than general quality of life (QOL), has been defined as "the extent to which one's usual or expected physical, mental, and social well-being are affected by a medical condition or its treatment" (Cella 1992). Although multiple definitions of HRQL exist, almost all share the concepts of subjectivity and multidimensionality. Subjectivity acknowledges that only patients, or valid proxies, can provide a complete understanding of the effects of a disease or treatment on their lives. Multidimensionality is a reminder that any or all of life's domains can be affected by health or its absence. Full appreciation of illness and treatment impact may require assessment of several important life domains, even those not directly affected by the specific condition or treatment. These domains can be broad (eg, physical well-being) or further subdivided into increasingly specific constructs (eg, mobility, fine motor function).

**Special considerations when assessing HRQL in neurologic illness**

Neurologic illness can cause mental and physical limitations that make it challenging to obtain patients' subjective report of their experience. Cognitive and communication difficulties, as well as burden of disease, can restrict a patient's ability to complete HRQL questionnaires. Therefore, measure characteristics such as length, complexity, and response format can be especially important to consider when choosing an HRQL tool to use with these populations. In addition, some measures may not discriminate well among individuals with poor function, thus, making them of limited use for people with severe neurologic disease. Children's participation may be limited by these same factors and also by their age or developmental level. Rather than excluding these individuals, the use of proxy respondents should be considered, provided that potential biases and limitations of proxies are taken into account. Both the adult and pediatric literature suggests that there is greater agreement between proxy and patient ratings when rating observable functioning (eg, physical HRQL) and less for more subjective dimensions such as social functioning, pain, and emotional well-being (Eiser and Morse 2001; Sneeuw et al 2002; Oczkowski and O'Donnell 2010). However, a recent systematic review suggests that rating dimensions that are clinically relevant (eg, physical function for children with muscular dystrophy) may result in greater agreement, even for subjective areas (Upton et al 2008).
Types of HRQL measurement approaches

Once the decision is made to assess HRQL, the next step is to decide which instrument to use. This can be a challenging task. Although many measures of HRQL are available, no gold standard exists. The choice of a measure depends on the setting, the purpose for which it will be used, and the characteristics of the instrument itself. Suggested criteria for instrument review include conceptual model, reliability, validity, responsiveness, interpretability, burden, availability of alternate forms (eg, child, adolescent, proxy), and cultural and language adaptations (Anonymous 2002). Content areas covered by the instrument, and how those areas were derived, may be important considerations (Waters et al 2009). Additional criteria, such as age-appropriateness, are applicable to selection of pediatric instruments (Eiser and Morse 2001; Ravens-Sieberer et al 2006).

Most measures of HRQL used to assess neurologic disease are one of two types: generic or targeted. Generic measures include questions that are general enough to be applicable to healthy and clinical populations. Targeted measures may be targeted towards specific diseases (eg, epilepsy), domains (eg, cognition, fatigue), or interventions (eg, radiation therapy). Each type has particular strengths, which are listed below.

<table>
<thead>
<tr>
<th>Table 1. Characteristics of Generic and Targeted HRQL</th>
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<tbody>
<tr>
<td>Provide general/healthy population norms</td>
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<tr>
<td>Allow cross-disease comparisons</td>
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<tr>
<td>Useful in resource allocation and cost-effectiveness analyses</td>
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<tr>
<td>Questions highly relevant (acceptable) to patients</td>
</tr>
<tr>
<td>Comprehensive coverage of important domains or areas</td>
</tr>
<tr>
<td>Sensitive to changes in health status or function</td>
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</table>

A frequently advocated third “modular” approach to HRQL measurement combines a generic “core” measure with a targeted instrument or instruments. A modular instrument can be specifically constructed for this purpose, or can select among existing separate generic and targeted measures. This approach has the advantages of both while minimizing limitations of each. A potential downside is the increased time and burden associated with a longer, combined instrument.

**Generic HRQL measures.** Generic measures have been used to describe the health status or HRQL of people with many common neurologic disorders. For adults, the most commonly used measure is the Medical Outcomes Study Short Form-36 (Ware and Sherbourne 1992). The SF-36 evaluates 8 domains: Physical functioning, role limitations due to physical effects of illness, social functioning, pain, energy and vitality, general health perceptions, role limitations due to emotional effects of illness, and mental health. A single additional item assesses change in health status during the past 12 months. A new generic measurement system available since 2006 is the Patient Reported Outcomes Measurement Information System (PROMIS), which offers a flexible range of assessment options for common symptoms and functional problems such as pain, fatigue, depression, anxiety, sleep, physical function, and social function. PROMIS is an NIH Roadmap initiative to create a publicly available and sustainable system for measuring patient reported outcomes across diseases and conditions (Cella et al 2007).

After an extensive review of HRQL measures for children, Eiser and Morse (Eiser and Morse 2001) recommended 3 generic tools for use in chronic illness: the Child Health Questionnaire (Landgraf et al 1996), the Pediatric Quality of Life Questionnaire (Varni et al 1999), and the Health Utilities Index (Feeny et al 1998). The Child Health Questionnaire consists of 14 scales with both self-report and parent versions available. However, the length of the self-report version (87 items) limits its practicality. The Pediatric Quality of Life Questionnaire (www.pedsqol.org) is a modular instrument available in child, adolescent, and parent forms. The generic core scales consist of 23 items measuring physical, emotional, school, and social functioning. Additional disease- and condition-specific modules are available, with the pediatric brain tumor, cerebral palsy, and neuromuscular disease modules of particular relevance to neurology. The Health Utilities Index (including multiple versions: HUI, HUI1, HUI2, HUI3) provides measures of health status and a preference-based health-related quality-of-life utility score. A 2006 review recommended consideration of newer generic instruments that have been developed according to established methodology (Ravens-Sieberer et al 2006), such as the KIDSCREEN (Ravens-Sieberer et al 2008) and DISABKIDS (Baars et al 2005), both of which are modular in approach and have been used internationally. Solans and colleagues provide a useful review of the characteristics and
psychometric properties of generic and disease-specific pediatric HRQL measures (Solans et al 2008).

**Targeted measures.** Targeted measures may focus on a disease, one or more HRQL domains, or specific interventions. These categories are not mutually exclusive and a disease-specific instrument may also be domain-specific. Disease-specific measures evaluate aspects of HRQL that are particularly applicable to that condition. For example, targeted instruments to evaluate HRQL in stroke include questions about language and communication problems; memory, thinking and cognitive function; vision; physical functioning (upper extremity function, mobility); and work productivity that generic instruments frequently omit (Buck et al 2000; Golomb et al 2001). Domain- or symptom-specific measures evaluate one area of HRQL and can range from the broad (eg, social well-being) to the more specific (eg, fatigue).

The last decade has seen tremendous growth in the development and application of disease-specific measures for particular neurologic conditions including, for adults, Alzheimer disease, amyotrophic lateral sclerosis, brain tumors, epilepsy, multiple sclerosis, Parkinson disease, spinal cord injury, and stroke. Instrument development has been slower for children and adolescents, likely because of the many additional theoretical and methodological issues involved. Most pediatric measure development activity has occurred in epilepsy (Eiser and Morse 2001a; Eiser and Morse 2001b; De Civita et al 2005; Ravens-Sieberer et al 2006).

Space does not permit a review or listing of all available HRQL measures. Online databases of patient-reported outcome measures such as PROQOLID and test compilations and reviews such as the Handbook of Neurologic Rating Scales (Herndon 2006) can be additional sources of HRQL measures. An NINDS initiative, the Common Data Elements (CDE) project, can also be helpful in measure selection. The overarching goal of the CDE project is to standardize data collection in neurology clinical research. After reviewing available instruments, CDE working groups made HRQL measurement recommendations for the following diseases: Amyotrophic lateral sclerosis, pediatric and adult epilepsy, headache, Huntington disease, multiple sclerosis, Parkinson disease, stroke, spinal cord injury, and traumatic brain injury. Recommendations for other diseases are likely to follow.

The tables below list common (in some cases only) targeted measures for specific neurologic diseases and for symptoms associated with more than one disease. When found, sources for critical reviews of measures (usually both disease-specific and generic) used with those disease populations have also been included.

### Table 2. Pediatric Disease-Specific HRQL Measures for Major Neurologic Diseases

<table>
<thead>
<tr>
<th>Disease-specific measure</th>
<th>Review articles</th>
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<tbody>
<tr>
<td><strong>Attention deficit hyperactivity disorder</strong></td>
<td>None identified</td>
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<tr>
<td>None identified</td>
<td>None identified</td>
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<tr>
<td><strong>Autism spectrum disorder</strong></td>
<td>None identified</td>
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<tr>
<td>None identified</td>
<td>None identified</td>
</tr>
<tr>
<td><strong>Brain tumors</strong></td>
<td>(Klassen et al 2010)</td>
</tr>
<tr>
<td>• Pediatric Functional Assessment of Cancer Therapy–Childhood Brain Tumor Survivor (PedsFACT-BrS) (Lai et al 2007)</td>
<td></td>
</tr>
<tr>
<td>• Pediatric Quality of Life Inventory Brain Tumor Module (PedsQL™) (Palmer et al 2007)</td>
<td></td>
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<tr>
<td><strong>Duchenne muscular dystrophy</strong></td>
<td>None identified</td>
</tr>
<tr>
<td>• Life Satisfaction Index for Adolescents (LSIA) (Reid and Renwick 1994)</td>
<td></td>
</tr>
<tr>
<td>• Pediatric Quality of Life Inventory Neuromuscular Module (PedsQL™) (Davis et al 2010)</td>
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<tr>
<td><strong>Cerebral palsy</strong></td>
<td>(Carlon et al 2010)</td>
</tr>
<tr>
<td>• Caregiver Priorities and Child Health Index of Life with Disabilities (CPCCHILD; proxy) (Narayanan et al 2006)</td>
<td></td>
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<tr>
<td>• Cerebral Palsy QOL-Child (CP QOL Child; proxy and child) (Waters et al 2007)</td>
<td></td>
</tr>
<tr>
<td>• Pediatric Quality of Life Inventory Cerebral Palsy Module (PedsQL™) (Varni et al 2002)</td>
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Epilepsy

- Quality of Life in Pediatric Epilepsy (child and parent/proxy) (Arunkumar et al 2000)
- Quality of Life in Childhood Epilepsy Questionnaire (QOLCE, proxy) (Sabaz et al 2000)
- Quality of Life in Epilepsy Inventory - Adolescents (QOLIE-A-48) (Cramer et al 1999)

Table 3. Adult Disease-Specific HRQL Measures for Major Neurologic Diseases

<table>
<thead>
<tr>
<th>Disease-specific measure</th>
<th>Review articles</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Attention deficit disorder</strong></td>
<td>None</td>
</tr>
<tr>
<td>ADHD Quality-of-Life Scale (AAQoL) (Brod et al 2006)</td>
<td></td>
</tr>
<tr>
<td><strong>Amyotrophic lateral sclerosis</strong></td>
<td>(Heffernan and Jenkinson 2005; Epton et al 2009)</td>
</tr>
<tr>
<td>Amyotrophic Lateral Sclerosis Assessment Scales (ALSAQ-40 and 5) (Jenkinson et al 1999a; Jenkinson and Fitzpatrick 2001)</td>
<td></td>
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<tr>
<td><strong>Alzheimer disease and dementia</strong></td>
<td>(Ettema et al 2005; Scholzel-Dorenbos et al 2007)</td>
</tr>
<tr>
<td>Cornell Brown Quality of Life in Dementia (Ready et al 2002)</td>
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<tr>
<td>Quality of Life in Alzheimer Disease (QOL-AD) (Logsdon et al 2002)</td>
<td></td>
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<tr>
<td>Dementia Quality of Life Instrument (D-QOL) (Brod et al 1999)</td>
<td></td>
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<tr>
<td>DEMQOL and DEMQOL-Proxy (Smith et al 2007)</td>
<td></td>
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<tr>
<td><strong>Brain tumors</strong></td>
<td>(Gilbert et al 2000)</td>
</tr>
<tr>
<td>EORTC-30 - Brain Module</td>
<td></td>
</tr>
<tr>
<td>Functional Assessment of Cancer Therapy-Brain (FACT- BR) (Weitzner et al 1995)</td>
<td></td>
</tr>
<tr>
<td><strong>Epilepsy</strong></td>
<td>(Leone et al 2005)</td>
</tr>
<tr>
<td>Well-Being Scale (Collings 1990)</td>
<td></td>
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<tr>
<td>Liverpool Quality of Life Battery (Baker et al 1991)</td>
<td></td>
</tr>
<tr>
<td>Quality of Life Assessment Schedule (Kendrick and Trimble 1994)</td>
<td></td>
</tr>
<tr>
<td>Epilepsy Surgery Inventory (Vickrey et al 1992)</td>
<td></td>
</tr>
<tr>
<td>Quality of Life in Epilepsy Instruments (QOLIE; 86-, 31- and 10-items) (Devinsky et al 1995; Cramer et al 1998).</td>
<td></td>
</tr>
<tr>
<td><strong>Huntington disease</strong></td>
<td>(Ho et al 2004)</td>
</tr>
<tr>
<td>Huntington Quality of Life Instrument (H-QoL-I) (Clay et al 2012)</td>
<td></td>
</tr>
<tr>
<td><strong>Migraine</strong></td>
<td>(Becker 2002)</td>
</tr>
<tr>
<td>Migraine Disability Assessment Scale (MIDAS) (Lipton et al 2001)</td>
<td></td>
</tr>
<tr>
<td>Headache Impact Test-6 (Bayliss et al 2003)</td>
<td></td>
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<tr>
<td>Migraine-Specific Quality of Life Questionnaire-Revised (MSQ)(Version 2.1) (Martin et al 2000)</td>
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</tr>
<tr>
<td>Migraine Specific Quality-of-Life (MSQOL) instrument (McKenna et al 1998)</td>
<td></td>
</tr>
<tr>
<td><strong>Multiple sclerosis</strong></td>
<td>(Gruenewald et al 2004; Mitchell et al 2005; Riazi 2006)</td>
</tr>
<tr>
<td>MS Quality of Life Inventory (MSQLI) (Ritvo et al 1997)</td>
<td></td>
</tr>
<tr>
<td>MS Quality of Life (MSQOL-54) (Vickrey et al 1995)</td>
<td></td>
</tr>
<tr>
<td>Functional Assessment of Multiple Sclerosis (FAMS) (Cella et al 1996)</td>
<td></td>
</tr>
</tbody>
</table>
Neuromuscular disease
   Individualized Neuromuscular Quality of Life questionnaire (INQoL) (Vincent et al 2007)

Parkinson disease
   • Parkinson's Disease Questionnaire-39 item version (PDQ-39) (Peto et al 1995)
   • Parkinson's Disease Quality of Life Questionnaire (PDQL) (De Boer et al 1996)
   • Parkinson's Impact Scale (PIMS) (Calne et al 1996)
   • Parkinson's Disease Quality of Life Instrument (PDQUALIF) (Welsh et al 2003)

Spinal cord injury
   • Life Situation Questionnaire-Revised (LSQ-R) (Krause and Anson 1997)
   • Spinal Cord Injuries Quality of Life Questionnaire (SCIQL-23) (Lundqvist et al 1997)

Stroke
   • Stroke Impact Scale (SIS) (Duncan et al 1999)
   • Stroke Specific Quality of Life Scale (SS-QOL) (Williams et al 1999); SS-QOL-12 (Post et al 2011)
   • Newcastle Stroke-specific Quality of Life measure (NEWSQOL) (Buck et al 2000)

Traumatic brain injury
   • Quality of Life after Brain Injury (QOLiBRI) (von Steinbuechel et al 2005)

Table 4. Symptom-Specific HRQL Measures

Symptom-specific measure                                      | Review articles
---                                                             | ---
**Dizziness and vertigo**                                      | ---
   • Dizziness Handicap Inventory (DHI) (Jacobson and Newman 1990) | (Duracinsky et al 2007)
   • UCLA Dizziness Questionnaire (UCLA-DQ) (Honrubia et al 1996) | ---
   • Vertigo Handicap Questionnaire (Yardley and Putman 1992)    | ---

**Fatigue**                                                    | (Dittner et al 2004)
   • Fatigue Severity Scale (FSS) (Krupp et al 1989)            | ---
   • Fatigue Assessment Instrument (FAI) (Schwartz et al 1993)   | ---
   • Fatigue Impact Scale (FIS) (Fisk et al 1994)               | ---
   • Functional Assessment of Chronic Illness Therapy-Fatigue (FACIT-F) (Cella et al 2002) | ---

**Pain**                                                       | (McDowell 2006)
   • Brief Pain Inventory (BPI) (Cleeland 1989)                 | ---
   • McGill Pain Questionnaire (Melzack 1975)                   | ---

**Sexual function**                                            | (Corona et al 2006)
   Derogatis Interview for Sexual Functioning-self-report (DISF-SR) (Derogatis 1997) | ---

**Sleepiness**                                                 | (Bailes et al 2006)
   • Stanford Sleepiness Scale (SSS) (Hoddes et al 1973)       | ---
   • Epworth Sleepiness Scale (ESS) (Johns 1991)                | ---
   • Functional Outcomes of Sleep Questionnaire (FOSQ) (Weaver et al 1997) | ---
The Neuro-QOL measurement system

Despite the existence of many HRQL measures for neurologic disorders, HRQL evaluation in neurology remains challenging. Lack of agreement on the best measure to use and the uncertain psychometric properties of some existing HRQL measures can make the task of measure selection overwhelming and can lead to confusion and limited understanding of disease impact in any broader context because results of most studies aren't comparable.

Recognizing this challenge, the National Institute of Neurological Disorders and Stroke (NINDS) initiated a multiyear, multisite project to develop a bilingual (English and Spanish), clinically relevant, and psychometrically robust HRQL tool responsive to the needs of the neurology community that would ultimately also be appropriate to use in clinical settings. The instruments that were developed through this effort, the Neuro-QOL measurement system, can be found at www.neuroqol.org.

Conceptually and practically linked to the more generic PROMIS effort, Neuro-QOL is comprised of 13 adult and 8 pediatric item banks (sets of questions comprehensively covering a specific symptom or functional area) and 4 scales to assess HRQL in people with neurologic disorders. These instruments assess the areas listed in Table 5. Initial validation studies have been conducted in stroke, multiple sclerosis, amyotrophic lateral sclerosis, Parkinson disease, adult and pediatric epilepsy, and muscular dystrophy. Available in English and Spanish, they are suitable for use with people 8 years of age and older. Like PROMIS, Neuro-QOL includes questions about issues that are common across selected neurologic diseases (generic banks) along with targeted banks and scales that evaluate symptoms, concerns, or issues only relevant to persons within specific disease groups. This approach enables cross-disease comparisons (by comparing generic items that are given to individuals across conditions) and evaluation of condition-specific HRQL issues (by reviewing targeted items administered only to individuals with the condition in question).

<table>
<thead>
<tr>
<th>Table 5. HRQL Areas Assessed By Neuro-QOL</th>
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</thead>
<tbody>
<tr>
<td><strong>Physical function</strong></td>
</tr>
<tr>
<td>Adult</td>
</tr>
<tr>
<td>• Lower extremity (mobility)</td>
</tr>
<tr>
<td>• Upper extremity (fine motor, ADLs)</td>
</tr>
<tr>
<td>• Sleep disturbance</td>
</tr>
<tr>
<td>• Fatigue</td>
</tr>
<tr>
<td>Pediatric</td>
</tr>
<tr>
<td>• Lower extremity (mobility)</td>
</tr>
<tr>
<td>• Upper extremity (fine motor, ADLs)</td>
</tr>
<tr>
<td>• Pain</td>
</tr>
<tr>
<td>• Fatigue</td>
</tr>
<tr>
<td><strong>Mental function</strong></td>
</tr>
<tr>
<td>Adult</td>
</tr>
<tr>
<td>• Depression</td>
</tr>
<tr>
<td>• Anxiety</td>
</tr>
<tr>
<td>• Stigma</td>
</tr>
<tr>
<td>• Positive affect and well-being</td>
</tr>
<tr>
<td>• Applied cognition (general concerns)</td>
</tr>
<tr>
<td>• Applied cognition (executive function)</td>
</tr>
<tr>
<td>• Emotional and behavioral dyscontrol</td>
</tr>
<tr>
<td>• Communication difficulty</td>
</tr>
<tr>
<td>Pediatric</td>
</tr>
<tr>
<td>• Depression</td>
</tr>
<tr>
<td>• Anxiety</td>
</tr>
<tr>
<td>• Stigma</td>
</tr>
<tr>
<td>• Anger</td>
</tr>
<tr>
<td>• Applied cognition (general concerns)</td>
</tr>
<tr>
<td><strong>Social function</strong></td>
</tr>
<tr>
<td>Adult</td>
</tr>
<tr>
<td>• Ability to participate in social roles and activities</td>
</tr>
<tr>
<td>• Satisfaction with social roles and activities</td>
</tr>
<tr>
<td>Pediatric</td>
</tr>
<tr>
<td>• Social relations (interactions with peers)</td>
</tr>
<tr>
<td>• Social relations (interactions with adults)</td>
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</tbody>
</table>

Details of Neuro-QOL development can be found on the Neuro-QOL website and in several articles (Cella et al 2011; Cella et al 2012; Gershon et al 2012; Lai et al 2012). In brief, Neuro-QOL followed established procedures for self-report measure development. These included gathering extensive patient, caregiver, and expert input via focus groups, interviews, and surveys; an iterative item generation, review, and selection process; and several phases of field testing. The first waves of field testing (conducted online with general population and clinical panels) were for the purpose of collecting the data needed for item response theory (IRT) analyses. Additional field testing with 694 patients and 197 proxies was conducted at multiple clinical sites across the United States for the purpose of validating brief versions of the instruments for use in the clinical groups initially targeted by the Neuro-QOL effort.
Neuro-QOL’s utilization of modern test development models, such as IRT, provides the instruments with certain advantages; these include the ability to be brief yet still precise and valid (Chang and Reeve 2005). Using IRT methodology, sets of items can be calibrated along a continuum that covers the full range of the construct to be measured. From this calibrated set, or “item bank,” items can be selected to make up “short forms” (typically consisting of 6 to 8 questions) that fit the goals of the user. For example, a user wishing to assess locomotion in a group of patients who tended to have poor or very poor mobility might select items that cluster near the lower end of the motor bank. Item banks are also the basis for Computerized Adaptive Testing (CAT). This is a specialized type of computer-based testing that allows for frequent assessments, immediate feedback, and precise evaluation of patients at the individual level while placing minimal burden on patients (Chang and Reeve 2005; Hahn et al 2006). Users can administer short, unique tests to every individual, with reliability and scores equivalent to longer, fixed-length assessments. Item response theory also allows comparisons of patients and questionnaire items across multiple instruments through a mechanism of equating the instruments along a common measurement continuum (McHorney and Cohen 2000). Thus, a given score on a Neuro-QOL measure can be directly linked to what that individual would score on any existing HRQL measure that has been equated to Neuro-QOL.

**Relationship of Neuro-QOL to other HRQL measures.** Affiliated projects are currently underway to extend the Neuro-QOL system to spinal cord injury, traumatic brain injury, and Huntington disease populations by adding disease specific items or domains as appropriate. Neuro-QOL is not a static system, and it is anticipated that it will be extended to additional diseases in the future. The generic Neuro-QOL measures can be linked to the parallel PROMIS measures. Neuro-QOL has included items from most of these banks into its system. These common items will allow direct comparisons between Neuro-QOL and PROMIS scores and results in both clinical and research settings.

**Conclusion**

There are many available questionnaires to measure quality of life in neurologic disease. Some are generic and offer an ability to compare across conditions and even to nonclinical populations; others are more specifically tailored to address specific symptoms and treatment effects of targeted neurologic diseases. Both types can add important information to the clinical encounter and to clinical research while being minimally burdensome to patients, clinicians, and researchers. Selection of a measure should be guided by the goals of the assessment and its psychometric and other properties. Guidance documents for the use of HRQL measures in clinical (International Society for Quality of Life Research 2011; Snyder et al 2012) and research settings (Butt and Reeve 2012) can help in selection. Although the existence of so many measures suggests that clinicians and researchers are likely to find some measure to fit their needs, the lack of a uniformly accepted instrument limits comparability and communication about scores in the research and clinical communities. Neuro-QOL, a NINDS initiative, combines generic and targeted quality-of-life assessment in ways that enable comparability across diseases and disease specificity within the same system. Future adoption and improvements on options such as Neuro-QOL should help improve the way neurologists communicate about outcomes that are best measured by asking patients.

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**References especially recommended by the author or editor for general reading.**

**Profile**

**Age range of presentation**

0-01 month  
01-23 months  
02-05 years  
06-12 years  
13-18 years  
19-44 years  
45-64 years  
65+ years  

**Sex preponderance**

male=female  

**Family history**

none  

**Heredity**

none  

**Population groups selectively affected**

none selectively affected  

**Occupation groups selectively affected**

none selectively affected  

**Associated disorders**

Alzheimer disease  
Amyotrophic lateral sclerosis  
Attention deficit disorder  
Brain tumors  
Cerebral palsy  
Duchenne muscular dystrophy  
Epilepsy  
Huntington disease  
Migraine  
Multiple sclerosis  
Neuromuscular disease  
Parkinson disease  
Spinal cord injury  
Stroke  
Traumatic brain injury  

**Other topics to consider**

Alzheimer disease  
Amyotrophic lateral sclerosis
Attention deficit hyperactivity disorder
Cerebral palsy
Duchenne muscular dystrophy
Epilepsy
Huntington disease
Migraine
Multiple sclerosis
Parkinson disease
Rating scales of movement disorders

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