INTRODUCTION

Assessing the health-related quality of life (Hr-QoL) of patients with atypical parkinsonism disorders is important yet no validated measures are currently available. The atypical parkinsonian disorders are chronic progressive conditions, which not only shorten life expectancy but affect many aspects of patients’ and their carers’ lives. No curative treatment for these disorders is available, and management of these patients largely has to concentrate on amelioration of symptoms, such as falls, immobility, autonomic features or dysphagia, activities of daily living and (in)dependence, and patients’ social and emotional well-being; in short, the improvement of patients’ quality of life. Assessment of patients with atypical parkinsonism has concentrated on objective measures such as mortality and clinical evaluation of impairment and physical functioning, supplemented by laboratory test results. However, a large literature shows that patients’ own assessments of their health, their preferences, and their views regarding health often differ significantly from physicians’ objective assessments (1). Where possible, treatment decisions should focus on health outcomes of value to the individual patient.

Scales to measure Hr-QoL, fully psychometrically tested and validated, are now used in a number of clinical and research contexts. Some types of Hr-QoL scale yield information that, combined with economic data, can be used to assess the cost benefit of health interventions and to inform decisions about the allocation of scarce health care resources. There are currently no validated measures to assess Hr-QoL in patients with atypical Parkinsonian disorders.

This chapter starts with a general overview of some of the conceptual and methodological issues relating to the measurement of subjective health assessment and Hr-QoL. Section two gives an overview of Hr-QoL instruments that have been used in Parkinson’s disease (PD) and the impact of PD on Hr-QoL. The third section addresses what is known about the Hr-QoL of patients with atypical parkinsonism, in particular multiple system atrophy (MSA) and progressive supranuclear palsy (PSP). The chapter concludes with some comments about future research in this area.

HEALTH-RELATED QUALITY OF LIFE: CONCEPTUAL AND METHODOLOGICAL ISSUES

What Is Quality of Life?

Although the definition of this somewhat elusive term is still occasionally discussed in the literature, there is general consensus on some fundamental points. First, although the phrases “quality of
life,” “health-related quality of life,” and health status are used somewhat interchangeably, there is broad agreement that, in the medical context, Hr-QoL should be regarded as a multidimensional construct (2), comprising physical, psychological, and social well-being. Within these three broad dimensions, most Hr-QoL scales have items on physical health and functioning, activities of daily living, mental health (e.g., perceived stigma, anxiety, depression), social activities, family relationships, and cognitive functioning. Because of these multiple factors, it has been argued that although it may be helpful to derive a summary index of Hr-QoL, the different aspects of Hr-QoL as measured by the scale domains should also be presented separately in order to better understand the precise impact of interventions (3).

Second, since quality of life is highly subjective, any appraisal of Hr-QoL should rely, where possible, on the perception of the individual patient. Many groups of patients cannot, however, assess their own Hr-QoL, e.g., those with severe dementia, and there is a growing literature on the use of proxy ratings.

Third, no quality of life instrument can comprehensively cover all aspects of Hr-QoL. Although some scales attempt to comprehensively assess all aspects of Hr-QoL, such instruments are often lengthy and burdensome and so are not feasible in clinical practice, particularly where patients have disabling conditions. Therefore, most measures focus on a limited number of specific aspects of Hr-QoL. The choice of instrument will be determined by the precise aim of the study.

Finally, in order to provide meaningful data for research and clinical practice, Hr-QoL measures need to be carefully developed and validated and there is now a large literature on the validation and psychometric properties that need to be demonstrated before the scientific community will accept that an instrument has been shown to be appropriately validated. Before considering psychometric testing in more detail, it is useful to consider next why Hr-QoL might be measured.

Why Assess Health-Related Quality of Life?

Though Hr-QoL measures have been developed for a number of reasons, two basic aspects of health care underlie most of the questions that Hr-QoL appraisals set out to answer: outcome of treatment and cost. As discussed above, Hr-QoL has emerged as an important outcome that incorporates patients’ views of their health. Also, since no country in the world can afford to do all that it is technically possible to do to improve the health of its citizens, the need has arisen for some system of setting priorities. The assessment of the Hr-QoL of patients with atypical parkinsonian disorders will become increasingly important if and when new drug treatments and other therapies for these disorders are developed. Trials will need to address the benefit of therapeutic interventions and measure change of symptoms in relation to Hr-QoL.

HEALTH-RELATED QUALITY OF LIFE MEASURES: DEVELOPMENT AND VALIDATION

All clinical assessment measures need to be shown to be valid and reliable. In addition, self-completed measures for patients with a disabling disease need to be short and feasible. Instrument developers must test the psychometric properties of a new instrument, which is a labor-intensive exercise, involving a series of studies to obtain data on the performance of the measure in different situations. For a comprehensive review of the statistical procedures, see Streiner and Norman (4). In brief, validity is how well the instrument measures what it purports to measure. There are various statistical procedures for testing different aspects of an instrument’s validity. The terminology is somewhat confusing but Streiner and Norman provide a useful guide to the various types (e.g., face validity, construct validity, criterion validity, concurrent validity, and predictive validity). Reliability assesses whether the same measurement can be obtained on other occasions and concerns the amount of error inherent in any measurement. Two basic tests are the internal consistency of a test, measured by coefficient alpha, and test–retest reliability where scores taken on two occasions are compared. Sensi-