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Health related quality of life in people with hereditary neuromuscular diseases: An investigation of test–retest agreement with comparison between two generic questionnaires, the Nottingham health profile and the short form-36 items

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Abstract

The present work attempts to define reproducibility, test–retest and internal consistencies of two standardised tools that measure health related quality of life (HRQoL), specifically as they apply to hereditary neuromuscular disease (HNMD): the Nottingham health profile (NHP) and the medical outcome study 36-item short-form questionnaire (MOS SF-36). A cross sectional survey of 108 hereditary neuromuscular disease patients completed the questionnaires consecutively in the course of multidisciplinary consultations in Reims between April 2002 and February 2005. The results of the study confirm the acceptability of using generic questionnaires such as the Nottingham health profile and the SF-36, and show good reliability for these instruments. For both instruments, reproducibility (test–retest) appears excellent for the physical dimensions explored, and satisfactory for the mental dimensions. There is nonetheless a need for health related quality of life measures validated for neuromuscular disease patients. Health related quality-of-life (HRQoL) measures provide information on how patients assess their health and the care provision they are offered.

Keywords: Neuromuscular diseases; Questionnaires; Quality of life; Muscular dystrophies

1. Introduction

On the basis of epidemiological studies, it can be seen that around 30,000 people in France are affected by hereditary neuromuscular disease (HMND) [1–3]. Caring for this patient population involves finding ways to ensure that their perceived health related quality of life is fully taken into account. Life expectation among these patients has increased considerably following medical progress and improvements in the way they are followed for via specialised multidisciplinary centers. Measuring health related quality-of-life (HRQoL) is fast becoming a major issue in medicine, in particular for chronic pathologies involving disability. Despite this, very few publications have studied the HRQoL of patients suffering from neuromuscular disease [4–12].

In contrast to the 'objective' criteria that are the basis for most medical decisions, HRQoL is a concept which integrates 'subjective' aspects, as perceived by the patient him/herself. The aim is to take the patient's point of view on his or her state and satisfaction of health as the Refs. [13–19].

World health organisation (WHO) specified that 'health is a state of physical, mental and social well being, and not merely the absence of disease and infirmity' [20]. This definition incorporates some aspects of the concept of HRQoL [21]. HRQoL measure assess the extent to which

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health impact an individual's ability to function and his/her state psychological well-being and general satisfaction with physical, psychological and social domains of life. The major dimensions of most traditional generic HRQoL instruments (including NHP and SF-36) take account of impairments (symptoms, subjective complaints, sensations, etc.), functional states (physical status, emotional and intellectual functioning, social and role participation), and general health perceptions (satisfaction with health, perceived well-being). Researchers have developed a range of HRQoL instruments designed to measure this concept. Such instruments are frequently self-administered questionnaires. Their scientific value depends on their psychometric properties (reliability, various forms of validity, sensitivity etc.…) [22].

Two categories of questionnaire can be distinguished: HRQoL questionnaires said to be 'generic' (not specific to a particular pathology) and those said to be 'disease specific' or 'targeted'.

Since there are no instruments that are specific to HNMD, it appeared worthwhile exploring some psychometric properties of two self-administered generic measures of HRQoL among patients examined in the multidisciplinary consultation unit for neuromuscular disease in Reims University Hospital Center. The two instruments used are internationally recognised and have been validated in the general population: the NHP [23] and the MOS SF-36 [24].

The work had several objectives: (i) to determine the acceptability of using these two generic HRQoL measures in a sample of neuromuscular disease patients; (ii) to compare reproducibility (test–retest) and concept domains of the two instruments.

2. Materials and methods

The work involved data collection over the period from April 2002 to February 2005. In the Champagne–Ardenne and Sud Picardie regions in France, the pathologies encountered among HNMD patients are: Duchenne and Becker muscular dystrophy, myotonic muscular dystrophy type 1 (Steinert), facio-scapular-humeral dystrophy (FSHD), limb girdle muscular dystrophy (LGMD), congenital muscular dystrophy (CMD) and spinal muscular atrophy (SMA). The patients suffering from HNMD included in the study were all registered in the neuromuscular disease consultation unit in Reims University Center Hospital. They agreed to take part in the study via the approved informed consent procedure. The study complies with the Helsinki recommendations. Illiterate patients or those with reading difficulties were not included in the study. Following informed consent, the questionnaires were completed by the patients at the time of the multidisciplinary consultation. All patients underwent a complete clinical examination in the course of the consultation and all were over 15 years of age. The minimum age for completion of the SF-36 is 14 [24]. We made the same empirical hypothesis that the age over 15 is valid for the NHP.

Table 1
Common domains to the both generic questionnaires of health related quality of life NHP and MOS SF-36

SF-36	NHP
8+1 Domains 36 items	6 Domains 38 items
– ***Physical Functioning*** (=**PF**) 10 items	– ***Physical mobility*** 8 items
– Role physical (=**RP**) 4 items	Xxxxxxxxxx
– ***Bodily pain*** (=**BP**) 2 items	– ***Pain*** 8 items
– General health (=**GH**) 5 items	Xxxxxxxxxx
– ***Vitality*** (=**VT**) 4 items	– ***Energy*** 3 items
– ***Social functioning*** (=**SF**) 2 items	– ***Social isolation*** 5 items
– ***Role emotional*** (=**RE**) 3 items	– ***Emotional reaction*** 9 items
– Mental health (=**MH**) 5 items	Xxxxxxxxxx
– Change health (=**CH**) 1 items	Xxxxxxxxxx
xxxxxxxxxx	Sleep 5 items

Two generic HRQoL scales were used: the French language version of MOS SF-36 [24] and the NHP in its French version [23], the ISPN (Indice de Santé Perceptuelle de Nottingham) (Table 1).

The SF-36 provides scores dimension by dimension. There is no global score, but rather a multi-composite score profile. For some items, the response choices are dichotomous and for others the choices are spread over a Likert scale comprising between three and six points. The score 0 corresponds to the 'worst possible state of health' and 100 to the 'best possible state of health'. The response choices for the items in the NHP are dichotomous (yes/no), and the instrument also yields a multi-composite score. In the present study NHP responses (for items belonging to the physical mobility dimension) were weighted and recoded according to recommendations made by post [25,26] and validated by the present team [27], so as to minimise missing data from this population of subjects with restriction of mobility. To enable the comparison of mean scores per dimension between the two questionnaires, the NHP scores were subtracted from 100, since the scoring pattern in this instrument works in reverse fashion to that of the SF-36. Hence the scores presented in this study range from 0 to 100, 0 corresponding to the worst perceived health status and 100 to the best.

The questionnaires were first completed by the subjects recruited at the time of their multidisciplinary consultations. To calculate the test–retest reliability of the instruments,