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Review article

Health-related quality of life in chronic inflammatory neuropathies: A systematic review

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ABSTRACT

Chronic inflammatory neuropathies represent a heterogeneous group of disorders which affect patients' functional status and quality of life. We conducted a systematic review of the scientific literature on the effects of both disease and treatment interventions on health-related quality of life (HRQoL) in this patient population. The available data are limited, as few studies have systematically considered HRQoL in patients with inflammatory neuropathies. Moreover, in treatment trials, HRQoL measures have exclusively been used as secondary outcome measures. There is some evidence suggesting that baseline pre-treatment HRQoL reports are lower in patients with chronic inflammatory neuropathy than in age and gender-matched controls. Following treatment interventions, improvements in self-reported measures were consistently documented in the physical domain of HRQoL, which in turn correlated with improvements in traditional strength and functional scales. The impact of available treatments on the quality of life of patients with inflammatory neuropathies remains largely under-investigated. Interestingly, recent, although limited evidence from generic HRQoL measures may partly or completely contradict the results found with the primary, traditional outcome measures used (rituximab for anti-MAG neuropathy; immunoglobulins versus corticosteroids for chronic inflammatory demyelinating polyneuropathy). Similarly, HRQoL measures may suggest superiority, rather than equivalence, of certain drug administration methods (subcutaneous over intravenous immunoglobulins). Further research is needed to assess HRQOL in patients with untreated chronic inflammatory neuropathies in comparison to normative values, as well as precisely quantify treatment benefit. The role of both generic and disease-specific HRQoL measures in the evaluation of patients with chronic inflammatory neuropathies is also worthy of further consideration.

Contents

1. Introduction

Health-related quality of life (HRQoL) is a concept that reflects subjective individual perceptions of the effects of an illness and its treatment on physical, mental and social aspects of life [1]. As a result, HRQoL appears to be directly indicative of reduced level of patients' well-being from their disease. Changes may otherwise indicate genuine

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and meaningful effects of treatments administered. Therefore, use of HRQoL scales has been proposed as an appropriate method to ascertain actual effects on patients' lives, resulting from disease as well as from therapeutic interventions, and should at least complement other health status measurements in neuromuscular conditions. Although an earlier study, which will be detailed as part of the current review, had demonstrated the Medical Outcome Study 36-item short form health status scale (SF-36) Questionnaire as a potentially valuable instrument in inflammatory neuropathies [2], the extent to which HRQoL evaluations have been used subsequently in the research setting has appeared variable.

Chronic inflammatory neuropathies are a broad and heterogeneous group of conditions manifesting in focal, multifocal or generalized sensory and/or motor deficits, evolving in a progressive or relapsing and remitting manner. In chronic inflammatory neuropathies, different therapies have demonstrated favourable effects on muscle strength and/or neurological function. How relevant these can be in actually improving the day-to-day living of treated patients may however be difficult to ascertain. Furthermore, baseline pre-treatment quality of life characteristics are uncertain in this group of disorders and how that may compare with normative values is unknown. We conducted a systematic review of the literature relating to use of instruments measuring HRQoL in chronic inflammatory neuropathy, focusing in particular on chronic inflammatory demyelinating polyneuropathy (CIDP) and multifocal motor neuropathy (MMN). We aimed to specifically analyse the use of HRQoL measures in the studied populations, in relation to baseline function as well as therapeutic benefit and its clinical correlates.

2. Materials and methods

Our search methodology followed the standard guidelines for systematic literature reviews outlined in the PRISMA statement [3]. We conducted a Medline search of all English language articles published between 1966–October 2014 on HRQoL in all forms of chronic inflammatory neuropathies. We used Medline with the search MeSH terms “inflammatory neuropathy”, as well as “chronic neuropathy”, “paraproteinaemic demyelinating neuropathy” (“PDN”), “POEMS” (“Polyneuropathy, Organomegaly, Endocrinopathy, M-Protein, Skin”) syndrome, “CANOMAD” (“Chronic Ataxic Neuropathy with Ophthalmoplegia, M-protein and Disialosyl antibodies”) syndrome, each combined with “quality of life” and “health status”. Reference lists of each retrieved paper were screened for additional relevant publications. All papers discussing specifically ascertainment of HRQoL in any aspect of baseline clinical manifestations, treatment effects or disease monitoring, were analysed. Articles were included irrespective of disease subtype or course, number of patients studied, main purpose of the analysis considered, type of monitoring otherwise performed/described in addition to HRQoL, or type of therapeutic intervention utilized. Papers considered were read in full-text version and analysed in detail in relation to HRQoL evaluations, results and conclusions. Reference lists of retrieved articles were searched for any additional relevant publications in the field. The findings are presented here in a descriptive manner following the chronological development of this rapidly expanding field.

3. Results

We retrieved a total of 23 articles which had descriptions of use of instruments measuring HRQoL in patients diagnosed with chronic inflammatory neuropathy. The reviewed articles, summarized in Table 1, showed heterogeneity in terms of study focus. Some studies described the results of a cross-sectional analysis of HRQoL in patients with CIDP. In other studies, changes in HRQoL were described as part of a therapeutic intervention for CIDP or MMN, with intravenous immunoglobulin (IVIg) treatment versus corticosteroids/placebo, or subcutaneous immunoglobulin (SCIg) versus IVIg/placebo, as a secondary outcome measure. In few studies, HRQoL was evaluated as part of the IVIg-related benefit in small treated cohorts, outside the setting of a drug trial. Finally in a single recent study, HRQoL was used as secondary outcome measure in a trial setting of rituximab versus placebo for anti-myelin-associated glycoprotein (MAG) antibody neuropathy.

The first relevant study was published in 2001 and presented the results of a multicentre double-blind cross-over randomized controlled trial (RCT) of IVIg versus oral corticosteroids, in 25 patients with chronic inflammatory neuropathy [4]. Both treatments provided significant, but not significantly different, improvements at 2 weeks in the primary outcome measure, the 11-point Inflammatory Neuropathy Cause and

Table 1
Studies on health-related quality of life in chronic inflammatory neuropathies (Medline search of English language articles 1966–2014).

Authors	Publication year	Chronic inflammatory neuropathy subtype(s)	Study type	Number of participants	Main outcome/finding regarding HRQoL
Hughes et al. [4]	2001	CIDP	Therapeutic	25	No improvement on IVIg or steroids
McCrone et al. [5]	2003	CIDP	Therapeutic	25	More improvement on IVIg than on steroids
Merkies et al. [2]	2002	CIDP, PDN	Cross-sectional	31	Lower than in controls; Improvement with treatment
Padua et al. [6]	2004	CIDP, MMN	Prospective cohort	11	Improvement with IVIg
Padua et al. [7]	2005	CIDP, MMN	Prospective cohort	11	Improvement with IVIg
Garssen et al. [8]	2004	CIDP	Interventional: Physical Training	4	Improvement with training
Hughes et al. [9]	2008	CIDP	Therapeutic	117	Improvement with IVIg
Merkies et al. [10]	2009	CIDP	Therapeutic	117	Improvement with IVIg
Merkies et al. [11]	2010	CIDP	Therapeutic	117	Correlation with strength, grip, disability
Harbo et al. [12]	2008	CIDP	Cross-sectional	14	Impairment; Correlation with isokinetic strength
Harbo et al. [13]	2009	CIDP	Therapeutic	11	Improvement with IVIg
Harbo et al. [14]	2009	MMN	Therapeutic	9	SCIg equivalent to IVIg
Eftimov et al. [15]	2009	MMN	Therapeutic	10	SCIg equivalent to IVIg
Misbah et al. [16]	2011	MMN	Therapeutic	8	Improvement on SCIg
Cocito et al. [17]	2012	CIDP	Therapeutic	5	SCIg equivalent to IVIg
Cocito et al. [18]	2013	CIDP	Therapeutic	10	Improved with 20% SCIg compared to 16% SCIg
Braine et al. [19]	2012	MMN	Therapeutic	16	SCIg equivalent to IVIg
Nobile-Orazio et al. [20]	2012	CIDP	Therapeutic	45	IV Steroids equivalent to IVIg
Mahdi-Rogers et al. [21]	2014	CIDP, MMN, PDN	Economic, HRQoL	106	Impaired compared to average Unrelated to treatment received
Maxwell et al. [22]	2013	CIDP, PDN	HRQoL, Social	39	Impaired compared to normal
dos Santos et al. [23]	2014	CIDP	HRQoL, cognitive	41	Impaired compared to non-matched normal subjects
Niermeijer et al. [24]	2007	PDN	Therapeutic	35	Improved with cyclophosphamide + prednisone
Léger et al. [25]	2013	PDN (anti-MAG neuropathy only)	Therapeutic	54	Significantly improved physical domains with Rituximab