Living with ALS
Perspectives of patients and next of kin

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To my family

ABSTRACT

ALS is a neurodegenerative disease without curative treatment. The knowledge of the relationship between patients and their next of kin with respect to quality of life (QoL) is deficient. The overall aim of this thesis is to describe different perspectives of QoL of patients with ALS and their next of kin, and to describe strengths and hindrances in the manageability of their daily lives.

The participants were recruited from Sahlgrenska University Hospital in Gothenburg, Sweden. In the quantitative studies I–III, 35 couples participated. Fourteen patients and thirteen next of kin participated in the qualitative study (IV).

Few changes were found over time in studies I and III, but in patients, there was a decreased rating in some of the physical subscales and in general health in the health-related QoL (HRQoL). The ratings in those subscales were worse in patients than in next of kin, even though next of kin also gave a decreased rating in some of the physical and mental subscales. Next of kin estimated individual QoL to be worse than patients did. No changes were found over time in anxiety, depression, or individual QoL. The ratings in discrete pairs were often similar, indicating that if one person felt bad, the other one did also. Even though the pairs gave relatively good ratings of QoL, study II showed that QoL was worse than in a subset of the general population. Study IV found a constant fluctuation between factors that facilitated and hindered the manageability for each individual person, as well as similarities and differences between patients and their next of kin.

QoL was worse in our participants compared with the general population and did not change much over time. The similarities and differences between the patients and next of kin show the need to offer them physical, psychosocial, and existential support, both together and individually, to ensure the best possible QoL. The knowledge that the manageability can change from one moment to another makes it necessary to meet the individuals with a wide perspective and to support them in the situation in which they are currently living.

Keywords: ALS, anxiety, coping, depression, manageability, next of kin, patient, sense of coherence, QoL, well-being

ORIGINAL ARTICLES

This thesis is based on the following papers, which will be referred to in the text by their roman numerals.

I Olsson AG, Markhede I, Strang S, Persson LI. **Well-being in patients with amyotrophic lateral sclerosis and their next of kin over time.** *Acta Neurol Scand. 2010;121:244–250. DOI. 10.1111/j.1600–0404.2009.01191.x (2009).*

II Olsson AG, Strang S, Persson LI. **Quality of life, anxiety and depression in ALS patients and their next of kin.** *Manuscript – Submitted.*

III Olsson AG, Markhede I, Strang S, Persson LI. **Differences in quality of life modalities give rise to needs of individual support in patients with ALS and their next of kin.** *Palliat Support Care. 2010;8:75–86.*

IV Olsson AG, Graneheim Hällgren U, Persson LI, Strang S. **Fluctuation in the everyday life of living with ALS in both patients and next of kin.** *Manuscript – Submitted.*

ABBREVIATIONS

ALS	Amyotrophic Lateral Sclerosis
ALSFRS	Amyotrophic Lateral Sclerosis Functional Rating Scale
ALSFRS-R	Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised
FALS	Familial amyotrophic lateral sclerosis
FTLD	Frontotemporal lobar dementia
HADS	Hospital Anxiety and Depression Scale
HADa	Hospital Anxiety and Depression Scale – Anxiety
HADd	Hospital Anxiety and Depression Scale – Depression
HRQoL	Health-related quality of life
LMN	Lower motor neuron
MMSE	Mini-Mental State Examination
MND	Motor neuron disease
SALS	Sporadic amyotrophic lateral sclerosis
SEIQoL-DW	Schedule for the Evaluation of Individual Quality of Life – Direct Weighting
SF-36	Short-Form Health Survey
BP	Bodily pain
GH	General health
MCS	Mental component summary
MH	Mental health
PCS	Physical component summary
PF	Physical functioning
RE	Role emotional
RP	Role physical
SF	Social functioning
VT	Vitality
SOC	Sense of coherence
SWB	Subjective well-being
QoL	Quality of life
UMN	Upper motor neuron
VAS	Visual Analogue Scale
WHO	World Health Organization

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INTRODUCTION

This thesis focuses on changes in different perspectives of quality of life (QoL), such as health-related QoL (HRQoL), individual QoL, well-being, anxiety, and depression, among patients with amyotrophic lateral sclerosis (ALS) and their next of kin in relation to the physical deterioration due to the disease, and how those participants manage the situation of living with the disease.

The incidence in ALS is around 1.5-2.7/100 000, but since the disease does not have any cure and since it is fatal, the survival time is limited, with a prevalence of around 2.7-7.4/100 000 (1, 2). The lifetime risk of getting the disease by the age of 70 years has traditionally been estimated at 1/1000, but nowadays it seems to be around 1/400 (1).

Multidisciplinary and palliative teams have been developed to maintain the QoL and improve the care of patients with ALS and their families (1). Palliative care should be applied as early as possible in the course of any chronic, ultimately fatal, illness, and is an approach that improves the QoL in patients and their families (3).

Different results have been shown in studies of patients with ALS and their caregivers, but many studies show that the psychological and individual QoL, anxiety, and depression due to the disease and the physical disability do not change considerably (4-12). However, the complexities of everyday life seem to affect the individuals and their ways of managing the situation (13-21).

In this thesis, patients with ALS and their next of kin, treated at Sahlgrenska University Hospital, Gothenburg, Sweden, participated in two longitudinal and two cross-sectional studies. Self-estimates of health-related QoL (HRQoL), individual QoL, well-being, anxiety, and depression in patients and their next of kin were compared with data of the decreased physical function in the patients. Different instruments were used to study multiple parameters, to illustrate several perspectives of their QoL. Patients and their next of kin were also interviewed about their ability to manage the living situation.

Quality of life is a broad concept that includes physical, psychological, social, and spiritual dimensions (22, 23). Different perspectives were, therefore, examined using several instruments, as well as interviews, in this thesis. The results increase the understanding of what it is like to live with ALS as a patient and as a next of kin. It may hopefully lead to better treatment and care of the whole families.

BACKGROUND

ALS

Motor neuron diseases (MND) include different disease entities, such as progressive muscle atrophy, primary lateral sclerosis, bulbar motor neuron disease, and amyotrophic lateral sclerosis (ALS) (24). Symptoms in ALS have been described by Bell (1824), Aran (1850), Duchenne (1851), and Cruveilhier (1853), but it was in 1869 that Jean Martin Charcot first described it as the disease entity ALS (1).

ALS is a progressive, lethal neurodegenerative disease. Different symptoms arise, depending on the localization of the damage in the nervous system. Damage of upper motor neurons (UMN) gives rise to, for example, progressive symptoms and signs such as weakness, spasticity, and hyperreflexia while damage to lower motor neurons (LMN) gives rise to symptoms and signs such as muscular atrophy, weakness, fasciculation, and hyporeflexia (24). Speech and swallowing problems can be traced to lesions of both upper and lower motor neurons (25).

The World Federation of Neurology has developed criteria for the diagnosis of amyotrophic lateral sclerosis. To be diagnosed with ALS, the patient needs to have signs of LMN degeneration as revealed by clinical, electrophysiological, or neuropathological examination, signs of UMN degeneration at clinical examination, and a progressive spread of signs in one or more regions. Other diseases that can account for the signs of motor neuron degeneration must be excluded. ALS or suspicion of ALS is classified as definite, probable, possible, or suspected ALS (26). El Escorial criteria of definite, probable, and possible ALS have been found to be closely associated with neuropathological confirmation of the disease and are reliable for the clinical diagnosis (27).

The disease is sporadic and of unknown cause in 90-95% of cases (sporadic ALS, SALS), while 5-10% is of genetic origin, so-called familial ALS (FALS) (1, 25, 28). The incidence is around 1.5-2.7/100 000, and the prevalence is around 2.7-7.4/100 000 (1, 2, 25, 29). In Sweden, there is no central register of patients with ALS, but corresponding data from most of the neurological units in Sweden showed a prevalence of 5.4/100 000 in 2003 (30).

Mean age of onset is described as 55-70 years, but the disease can start at any age in adult life (1, 31). The incidence is slightly higher in males than in females, M:F ratio 1.3:1-1.6:1 (1, 25, 29). Median survival from onset to death is two to three years for bulbar onset and three to five years for limb onset (1), but another study found that median survival from first symptom of bulbar onset

was just under two years, whereas, it was a couple of months over two years in spinal onset (32).

Around 5-10% of people with ALS survive more than 10 years (32, 33). Respiratory insufficiency is the most common cause of death (24, 34), and in contrast to preconceptions of it leading to a painful choking death, death is often composed and still (34). Since many patients worry that they will choke to death, it is important to inform them that most patients lapse into a terminal hypercapnic coma with a peaceful death during sleep (25).

Although ALS is an incurable disease (1, 33), many of the symptoms are treatable. Multidisciplinary and palliative care can do much to help the families from onset until the patient passes away. The goal of the efforts should be to improve the QoL, help the families with their needs, and help them to maintain their autonomy as long as possible (1, 25).

QUALITY OF LIFE (QoL)

In diseases with no cure, the care needs to be focused on the maintenance and improvement of QoL, with good palliative care (7). Measures of the QoL generate valuable information about the person, symptoms and consistencies of the disease. Such knowledge makes it easier to give priority to adequate forms of support and improve the care (35).

There are many different definitions and descriptions of QoL. The World Health Organization (WHO) has defined QoL as 'individuals' perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns' (p.1405, ref.36). The individual's physical health, psychological state, level of independence, social relationships, personal beliefs, and relationships to salient features of the environment are included in the concept (36).

Another definition of QoL is that it is 'a multidimensional evaluation of an individual's current life circumstances in the context of the culture and value systems in which they live and the values they hold. QoL is primarily a subjective sense of well-being encompassing physical, psychological, social, and spiritual dimensions. In some circumstances, objective indicators may supplement or, in the case of (people) unable to subjectively perceive, serve as a proxy assessment of QoL' (p.219, ref.23).

The four dimensions – physical, psychological, social, and spiritual – are together with the two indicators well-being and functional status included in the concept of QoL (23). Another description of four broad dimensions in QoL are physical, functional, emotional, and social well-being (37).

A good QoL is often described as having a positive psychological outlook and emotional well-being, physical and mental health, and a capacity to be physically activity with ability to do what one wants to do; having good relations with family and friends; taking part in social activities and recreation; living in a safe neighbourhood with good facilities and services; having good economic security; and being independent (38).

In this thesis, the concept of QoL (23) has shaped the structure of the chosen instruments in the studies. The HRQoL instrument SF-36, the Hospital Anxiety and Depression Scale (HADS), and the modified Visual Analogue Scale (VAS) can in different ways be used to evaluate the physical, psychological, and social dimensions. The individual QoL instrument SEIQoL-DW makes it possible for the participants to choose at each study visit the areas in life most important to them. By this method, all four dimensions, including spiritual, can be examined. The indicators well-being and functional status (23) are also included in these scales, and the functional status is measured in the disease-specific physical functional scales (ALSFRS-R and Norris scale).

Health-related quality of life (HRQoL)

The concept of QoL has sometimes been criticized as too general to be useful in health care, and it has been suggested that global QoL be differentiated from HRQoL (22). According to WHO, health is a state of physical, mental, and social well-being (39), and it is included in the concept of HRQoL, since it is not just an absence of disease, but also points to persisting positive factors in life (40).

HRQoL is described as evaluating health status, attitudes, values, and perceived levels of satisfaction and general well-being in relation to specific health status or life from the individual's perspective. Measures in HRQoL often include some domains of physical, social, and role functioning, as well as mental and general health (41), and thus, HRQoL is a dimension of wider QoL (38). HRQoL is modified by social opportunities, perceptions, functional states, and impairments due to disease, injuries, treatments, or policy (40).

Individual quality of life

Individual QoL examines the individual person's perspective of his or her QoL. Instead of using standardized questionnaires, the person presents what he or she thinks is important in life and how well it works (42). Since QoL is individual in nature, and only the person himself can judge his or her QoL, it is important to examine the individual QoL among the participants in a study (40).

Well-being

QoL is a broad concept with both subjective and objective components. Often well-being refers to the subjective components of QoL, whereas, functional capacity often refers to the objective components of QoL (23). Satisfaction with life is also included in the concept of QoL, as a subjective assessment and a hallmark of well-being (23, 43). Although well-being often is included in definitions of QoL, it cannot be considered synonymous with QoL, since it is subjective; whereas, QoL can be both subjective and objective (23).

Subjective well-being (SWB) includes various evaluations with regard to people's lives, events happening to them, their bodies and minds, and the circumstances in which they live (43). SWB has been described as a positive concept containing dimensions of happiness, satisfaction in life, morale, self-esteem, and sense of coherence. It has been described as comprising a balance between positive and negative impacts. Psychological well-being has been described as emphasizing the existential challenges in life, such as personal development, self-actualization, coping strategies, growth, and mastery (38).

Quality of life in ALS

Studies examining QoL and mood changes such as depression and anxiety in patients with ALS are often studied at one single time in life. Choice of instruments and methods are partly responsible for different results (44-48). QoL in patients often relates to the psychosocial and existential aspects, suffering, social support, sense of burden, and hopelessness, rather than to physical functioning (7), and in spite of physical limitations, the QoL often is estimated as good (5, 12, 49-51). The physical functional scales usually only correlate with physical components in QoL instruments and not with the mental components (4, 9). However, some ALS-specific HRQoL measures decreases in parallel with the increasing physical deterioration due to the disease (52). The QoL in caregivers often relates to social problems, relationship satisfaction, spirituality, and religiousness (53).

It has been shown that both patients and their caregivers estimate worse mental QoL than the general population does (54), but the opposite also occurs, that is, results indicating that the mental QoL can be moderately better in patients than in the general population over time (4). Further, QoL and depression have been found to be fairly good in both patients and their caregivers (51). It has been shown that patients have better individual QoL than their caregivers (48), but on the other hand, another study showed that caregivers have better QoL than patients (55).

Depressive symptoms affect QoL (56) but neither depression nor anxiety are as prevalent or severe as might be expected, and if they do arise, the symptoms are

often mild in patients with ALS (5, 6, 45, 47, 57, 58). Another study shows the opposite, that severely and mildly depressive symptoms are relatively common (59). Few differences have been found from the perspective of gender, although one study shows that men have higher levels of depressive symptoms than women (47).

The burden of caregivers increases with the patients' worsening disability. Increased burden does also correlate with caregivers' estimates of QoL and prevalence of depression. Patients' and their caregivers' estimates of depression have been found to relate to each other (55). However, depressive symptoms are few or non-existent in caregivers, but nearly half of the caregivers have anxiety, and that is also related to patients' anxiety (19).

Different results have been found in the individual QoL instrument SEIQoL-DW in patients with ALS and their caregivers. The instrument has levels between 0 and 100; higher estimates denote better QoL. Some studies have found that the mean value for the QoL of patients is around 73-76 (46, 48), with better estimates than their caregivers (mean 65), and a mean value of 70 in healthy couples (48). However, the median values have also been found to be as bad as 46 in patients and 56 in caregivers, while the median is 84 in healthy spouses. The physical dysfunction correlates to the individual QoL in patients, but does not affect the individual QoL in caregivers (60). In contrast to the last-cited study, it also has been shown that patients' individual QoL does not correlate with their physical and functional status (50). Many areas of importance in the SEIQoL-DW are selected equally by patients and their caregivers, such as family, social activities, finances, spirituality, and health, as well as accommodation, leisure activities, mobility, relationships, and profession (8, 50, 60).

Over time, the HRQoL decreases, while the individual QoL is stable (8), and the physical components in QoL instruments and in physical functional scales decrease, while the mental components often are constant during the course of the disease (4, 5, 9-11). Neither QoL nor symptoms of depression have been shown to change in patients over time, while caregivers had worse estimates in ratings of depression over time, even if it was still not classified as depression. The QoL was unchanged also in caregivers over time (5). Few studies examine the perspective of gender, however one study found that there were no differences by gender in patients or caregivers (60).

SENSE OF COHERENCE (SOC)

A salutogenic viewpoint insists that a person shall not be seen as just healthy or sick, but rather as being in a multidimensional continuum between health and ill-health. Sense of coherence (SOC) is the core to the answer of the salutogenic

question. SOC is an important factor in maintaining health, and it is defined as 'a global orientation that expresses the extent to which one has a pervasive, enduring though dynamic feeling of confidence that (1) the stimuli deriving from one's internal and external environments in the course of living are structured, predictable, and explicable; (2) the resources are available to one to meet the demands posed by these stimuli; and (3) these demands are challenges, worthy of investment and engagement' (p.19, ref.61). The three components comprehensibility, manageability, and meaningfulness are included in the concept of the SOC. Comprehensibility refers to the extent to which one can experience internal and external stimuli as understandable, that is, as information that is ordered, sustained, structured, and obvious, rather than as noise – chaotic, disturbed, random, unexpected, and mysterious. Manageability focuses on the resources that are available to meet the demands of the individual. A high manageability allows the person not to feel like a victim of the circumstances. Meaningfulness implies that the person has something in life that engages him or her and is important to both an emotional and cognitive sense (61), which means that it is possible to find meaning in a situation, in spite of stressful conditions (62).

When a person is afflicted with a serious disease it is important to find the ability to manage the situation. Antonovsky's theory of SOC (61) was applied in Paper IV, because it reveals important aspects of health and because all three components of comprehensibility, meaningfulness, and manageability were considered to be of interest, even though only manageability was studied in this thesis. The reasons that only manageability and not comprehensibility and meaningfulness were analysed in the present study are that the earlier studies in the thesis focused on QoL, and in the last study, it was desirable to study how patients and next of kin managed their life situation. If all three components of SOC had been included, it had also made the analysis diluted and superficial. Manageability might also contribute more than the other components could to developing concrete strategies for health care professionals to help patients and next of kin to manage their situation. The other two components will be treated in future studies.

The term coping is also a usual concept that could have been used, since it implies the capacity to use thoughts and behaviours to handle both internally and externally taxing demands to manage the situation (63, 64). However, we wanted to illuminate all three components of the SOC, even though just one is examined in this thesis.

Nursing theory

A theory related to coping and manageability is the theory of care of Patricia Benner and Judith Wrubel, which concerns and supports this thesis. They stress

that a disease may lead to a disorder in the individual's accustomed functioning, and areas that earlier have been taken for granted stop working. Stress leads to feelings of threat, loss, or challenge, and it needs to be processed through sorrow, constructions or new proficiencies. To manage the disorder, the individual needs to master it. One's own opinion of special interests and an understanding of the background, proficiencies, and practical functioning determine what a person experiences as generating stress and what possibilities he or she has available to overcome it. Health promotion needs to be based on the individual's experiences of the situation, what that person finds important, and what coping mechanisms he or she can bring to bear. It should be based on the individual's own resources, but since the individual also is dependent on the support of family, friends and others, health professionals need to take into account both the social circumstances and the culture (63, 65).

Manageability and coping in ALS

Maintaining of personal integrity in spite of the ongoing changes and adaptation to the consequences of the disease is important in the daily lives of patients with ALS (18). The changes in their lives, with loss of independence and control, may cause self-esteem and self-confidence to be undermined (16, 18), but self-esteem can also be sustained, if new goals in life are set up that convey an optimistic perspective on life. Passive and active strategies, like denial or change of common support structures (18), engaging in social activities (17), and focusing on what can be achieved rather than what is no longer possible, help in the managing (15). Even though there is uncertainty about the future, patients try to find coping strategies to manage the situation (17). They cycle between great optimism and despair, and they sometimes feel loss or breakdown of self, fear of the future, denial, and a feeling of living with a surreal notion of time (15). The unknown future frightens and it is not meaningful to plan for the future. The fear of being dependent is ever-present. Loss of control is perceived as huge; it involves control of the situation, the environment, and the future (16). However, patients often find coping strategies that give them meaning in life (7).

Patients with ALS often find that children and grandchildren contribute to the meaning of life and give them strong motives for not giving up, but such relationships also cause feelings of guilt, because their life is passing away (13). The need to confide in someone is small, and patients and their caregivers find the value in doing so in different ways (14). Faith can help patients to cope and accept the illness and loss. They can feel that they need to be in control, but also that they feel less control. It is important to feel dignity, to be treated with respect as a person and not just as a *sick* person. Support from families and social and health care services are very important in coping (21).

Caregivers to patients with advanced progressive illness can feel threats, such as uncertainty about the future, their own health problems, changes in immobility in their lives because of the disease, and the worry of the patient about how they are judged when they participate in their social life. Resources for care develop when they can accept the situation, when they are taking care of themselves, by, for instance, doing something of their own, if they feel supported, if they feel shared responsibility, and if the patients are able to show that they value what the caregivers do for them, or just have feelings of getting on with it (66).

The needs of the family members encompass psychological, spiritual, and physical support, intimacy with the patient, knowledge of the prognosis and proximity to death, and recognition for having helped the patient (67). The risk of decreased health increases with increased burden. The greater the number of hours the caregiver spends on care, the higher the risk of decreased health of their own (19, 68). Friends and family are important factors in the ability to cope with the future. There are feelings of lack of knowledge, lack of empathy, and inflexibility in the provision of services by health care and home care professionals (19). Family caregivers need to cope and adjust to the situation, so the family can live as normally as possible. They need to establish as much control over life as possible, and they need a transition to the terminal stage of the disease process and to their role as caregiver to a dying loved one (20).

RATIONALE FOR THE STUDY

This thesis examines different perspectives of QoL and manageability in both patients with ALS and their next of kin. Most other studies on the QoL in ALS have not studied pairs of patients and next of kin together, and especially, not in longitudinal studies during part of the course of the disease. The pairs in Papers I and III were followed up regularly at intervals of 4–6 months. Since more will have happened with the disease at longer intervals, we preferred to do so, instead of examining them at shorter intervals.

When patients and their next of kin are studied together, it is possible to gain a wider perspective of their QoL and their ability to manage the disease and the situation in life. Since the whole family is affected by the changes in life situation during the course of the disease, studying the parts of the pair together makes it possible to find similarities and differences between them, which also makes it easier to find strategies to help them, both as individuals and as a pair.

If the reactions and the strain weighing upon the patient and next of kin are revealed, it hopefully will be easier for the pair to meet each other in the situation and see the possible different perspectives they hold with respect to the disease and life. Findings in this thesis might improve the understanding and strengthen the awareness of the need for support and of how resources can be applied to maximize the effect of the care for both the patient and the next of kin from the time of diagnosis until the end of life.

AIMS

The overall aim of this thesis is to describe different perspectives of QoL of patients with ALS and their next of kin, and to describe strengths and hindrances in the manageability of their daily lives.

SPECIFIC AIMS

The specific aims of the papers were:

Paper I: To elucidate the well-being of both patients and their next of kin, how they estimate each other throughout the part of the course of the disease, as well as to find out whether well-being is related to the declining physical function of the patient.

Paper II: To examine self-estimates of HRQoL, anxiety and depression in patients with ALS and their next of kin, and to compare these results with a subset of the Swedish population.

Paper III: To examine HRQoL, individual QoL and self-estimates of anxiety and depression in patients with ALS and their next of kin in relation to patients' physical function over time.

Paper IV: To illuminate aspects that facilitates and hinders the manageability of living with ALS in both patients and next of kin.

MATERIAL AND METHODS

STUDY DESIGN

To understand the situation of living with ALS from the perspective of both the ill person and the next of kin, multi-method designs with both quantitative and qualitative methods were used. There are limitations to every method, but by using different procedures with both quantitative and qualitative methods, a phenomenon can be studied from different aspects, which can increase the understanding and strengthen the research (69).

The studies conducted with questionnaires aim to show quantitative, comparable data, whereas, the qualitative study illuminates the components of the life situation, which makes the understanding deeper, since the interviewed person uses his or her own thoughts and words to describe the situation.

The studies in this thesis were descriptive, paired, cross-sectional, and longitudinal. In Paper I, well-being was examined over time, and the results were compared between patients and their next of kin and with the decline of the physical function in the patients. In Paper II, the HRQoL in patients and their next of kin was compared to a representative subset of the general population at one single study time. In Paper III, the HRQoL with questionnaires and the individual QoL with semi-structured interviews were studied over time in patients and their next of kin. The results were compared within the pairs and related to the physical function of the patients. In Paper IV, the manageability of the situation of living with ALS in both patients and their next of kin was studied with a qualitative, descriptive study, using content analysis. Studies included in the thesis are shown in Table 1.

Table 1. Studies included in the thesis.

	Study design	Subjects	Instruments	Data analysis
Paper I	Descriptive, longitudinal	35 patients, 35 next of kin	VAS, ALSFRS-R, Norris scale, MMSE	Fisher's test for paired comparisons, Mann-Whitney test, Wilcoxon signed rank test, Spearman rho correlation
Paper II	Descriptive, cross-sectional	35 patients, 35 next of kin	SF-36, HADS, ALSFRS-R, Norris scale, MMSE	Mann-Whitney test, Wilcoxon signed rank test, Spearman rho correlation, Pitman permutation test, t-test
Paper III	Descriptive, longitudinal	35 patients, 35 next of kin	SF-36, HADS, SEIQoL-DW, ALSFRS-R, Norris scale, MMSE	Fisher's test for paired comparison, Mann-Whitney test, Wilcoxon signed rank test, Spearman rho correlation
Paper IV	Descriptive, cross-sectional	14 patients, 13 next of kin	Interviews in depth	Qualitative content analysis

SETTING

The cohort of patients in all studies included patients diagnosed with definite or probable ALS, treated by the ALS/MND team at Sahlgrenska University Hospital, Gothenburg, Sweden. Next of kin were closely related to the patients.

PARTICIPANTS

Papers I–III

The participants were recruited by the author at the Department of Neurology, Sahlgrenska University Hospital in Gothenburg, Sweden. Included patients had been diagnosed with probable or definite ALS according to El Escorial criteria (26). The inclusion and exclusion criteria were the same in studies I-III, and the participants were included in all three studies at the same time.

Included patients were being treated by the ALS/MND team at Sahlgrenska University Hospital and were recruited from January 2006 to March 2007. Since studies I and III were longitudinal, the collection of data stopped at December 2007.

The patients were asked to choose the next of kin closest to them, and those people were then asked to participate in the studies. Both patients and their next of kin had to be physically and psychologically able to consent to participate.

Patients in the terminal phase of the disease were not included because of difficulties due to the severity of the illness and because it would be difficult to do follow-up in the longitudinal studies.

Forty-seven consecutive patients were considered possible participants according to the inclusion and exclusion criteria. However, six patients were excluded; they did not wish their next of kin to participate, as the studies were designed to compare estimates of patients and their next of kin. One of these patients also had signs of cognitive impairment, according to the MMSE test. No other patient had signs of cognitive impairment. One man was excluded because he could not understand and speak the Swedish language adequately. Four men and one woman did not want to participate. In total, thirty-five patients and thirty-five next of kin were included in the first three studies (Table 2.)

Table 2. Number of participants at each study visit and exit from study (f. = female, m. = male).

Participants	**First visit**	**Second visit**	**Third visit**	**Fourth visit**
Patients	35 (f.18, m.17)	32 (f.15, m.17)	26 (f.13, m.13)	9 (f.5, m.4)
Next of kin	35 (f.19, m.16)	31 (f.17, m.14)	26 (f.14, m.12)	9 (f.5, m.4)
Cause of dropouts				
Revoked consent		1 patient		
Too ill to participate		2 patients, 1 next of kin	2 patient	1 patient
Deceased			4 patients	2 patients
Not eligible for follow up within projected time				14 patients

Thirty of the next of kin were married or cohabited with the ALS patient, three were children, one was a sister, and one had previously been married to the patient. The latter five next of kin all had close contact and a close relationship with the respective patient. The social system in Sweden provides services to patients with decreased ability to perform activities of daily living (ADL) with personal assistance, home care, or housing in a nursing home. All patients in these studies lived at home during the study collection. The extent of assistance performed by the next of kin and by the social system varied from no help at all to assistance 24 hours a day. It was appropriate to use the term next of kin, rather than caregiver, because not all of them actually had a major role in assisting the patient in their daily life.

The mean age of patients was 63.4 years (median 64 years, range 28-84 years at entry). The mean age of their next of kin was 61.3 years (median 64.5 years, range 27-86 years).

In study II, data among the general Swedish population in SF-36 and HADS was obtained from two Swedish studies, so comparisons could be made (35, 70).

Paper IV

In Paper IV, patients with probable or definite ALS according to the El Escorial criteria (26) were recruited at the Department of Neurology, Sahlgrenska University Hospital in Gothenburg, Sweden. Included patients were being treated by the ALS/MND team at Sahlgrenska University Hospital, Gothenburg, Sweden. Next of kin were married to or cohabited with the patients.

All pairs were recruited among the participants in the previous studies in this thesis. All patients must have been diagnosed at least 6 months before entry to the study, as we considered it important that they realized the gravity of the disease. They should not have had any other mortal diseases. Their speech needed to be good enough to participate in interviews. Patients in the terminal stage of the disease were excluded. Of the 35 pairs who participated in the previous studies in the thesis, 19 pairs were eligible for the present study.

Purposeful sampling with 'maximum variation sampling' was used (69) with respect to participants' gender, age, psychosocial background, and physical function. The selection started by recruiting as many different variations as possible among the participants. After ten interviews with the patients and ten interviews with their next of kin, we began to find repetition in the responses. However, to avoid loss of possible additional information, the number of interviews was extended to fourteen patients (seven men and seven women) and thirteen next of kin (eight men and five women). The 14th next of kin declined to participate in the interview. The patients were between 42 and 80 years of age (median 67.5 years) and the next of kin were between 38 and 87 years (median 68 years). All patients lived at home and their needs for help in daily living varied from little help to help 24 hours a day.

PROCEDURE

Papers I–III

Data in Papers I–III were collected at the same study visits with the ALS/MND team at the Department of Neurology, Sahlgrenska University Hospital. The participants in all studies were asked to participate in the studies by verbal and written information provided at a regular visit to the physician. After around two weeks' consideration they were asked by the author to indicate whether they wanted to participate or not. At the start of the studies, they also gave verbal and written consent to participate.

The collection of data in studies I–III occurred between January 2006 and December 2007, but new recruitment stopped at March 2007, which also led to a collection of data for study II occurring between January 2006 and March 2007. The recruitment stopped at March 2007, because there would have been too few

follow-up visits with the newly recruited participants if we had had a longer recruitment. The follow-up visits in studies I and III were made every 4 to 6 months, with a goal of every 6 months, up to four times. Study II had just one study visit. Due to the progression of the disease, the number of participants for the follow-ups in studies I and III was reduced (Table 2).

Before the visits, the HRQoL instrument SF-36 and the Hospital Anxiety and Depression Scale (HADS) were sent home to the participants, who were requested to complete the questionnaires on their own before the following visit to the hospital. In studies I–III, at every visit, the physical function in patients was measured by a neurologist, using the physical functional scales ALSFRS-R and Norris scale. Patients and their next of kin were then interviewed by researchers from the ALS/MND team in separate rooms and asked to rate their well-being by the Visual Analogue Scale (VAS) and their individual QoL by the SEIQoL-DW. The participants delivered the questionnaires at the interview occasion.

At entry to the studies, all patients were tested with the Mini-Mental State Examination (MMSE), since some patients can be afflicted with dementia and cognitive failure. Due to functional disability caused by the disease, speech problem or loss of function in a hand or arm affected some patients. Patients with low numbers in their performance in the MMSE were discussed in the research group; as the low numbers were related to the physical handicap and not on cognitive dysfunction, they continued in the studies. Only one patient had real signs of cognitive dysfunction at the time of testing before inclusion, and she did not allow the next of kin to participate, and was thus excluded.

In study I, the patients and their next of kin estimated their own and the other person's well-being by the VAS over a period of time. First they estimated their own general, physical, and psychological well-being, and then they estimated what they thought the well-being of their next of kin was. Comparisons with the physical functional scales ALSFRS-R and Norris scale were made.

In study II, the patients and their next of kin estimated their own HRQoL by SF-36 physical component score (PCS) and mental component score (MCS) and by the Hospital Anxiety and Depression Scale (HADS) at one study visit. Comparisons were then made between each patient and next of kin and with a subset of the general Swedish population. Data from the general Swedish population were obtained from two other studies (35, 70). The total number of participants in the general Swedish population in both SF-36 and HADS does not agree with the numbers of male and female participants in the same population, which can be seen in the paper. There were probably participants who had not registered their gender in the study of the general population.

In study III the patients and their next of kin were studied over time. They estimated their own HRQoL using the SF-36 and the HADS. They also estimated their individual QoL using the SEIQoL-DW. Comparisons were also made with the physical functional scales ALSFRS-R and Norris scale.

Paper IV

Data were collected in spring 2007 by in-depth interviews, performed face to face by the author. The interviews lasted between 20 to 83 minutes (median 48). Interviews were performed, according to participants' preference, in an undisturbed room at their homes or at the hospital. The interviews were tape-recorded and notes were taken to support the verbal information.

METHODS OF DATA COLLECTION

Questionnaire

The health-related quality of life instrument (SF-36)

SF-36 is a generic health status questionnaire and one of the most widely used patient-based measures of HRQoL (41). SF-36 has been translated and standardized after Swedish standards. The instrument includes 36 items. Thirty-five of those are divided into 8 multi-item scales: physical functioning (PF), role physical (RP), bodily pain (BP), general health (GH), vitality (VT), social functioning (SF), role emotional (RE), and mental health (MH). The 36th item measures the health transition over the past year. The instrument has been validated and its reliability has been tested (71-73). The construction of the 8 scales in SF-36 was designed to achieve representation of health concepts from a multi-dimensional perspective and to measure the full range of health stages, including levels of well-being and personal evaluations of health. Each item has a score from 0 (worst possible health state) to 100 (best possible health state) (73). The SF-36 can be subdivided into two major dimensions of health: physical component summary (PCS) and mental component summary (MCS) (71).

The SF-36 was used in study II to compare the estimates between patients and their next of kin to a subset of the general Swedish population and in study III to compare the estimates of the pair over time.

The Hospital Anxiety and Depression Scale (HADS)

The HADS is a self-assessment scale with 14 questions in which the person estimates their own experience on two subscales containing questions about anxiety and depression. The rating used consists of a 4-level scale with values between 0 and 3 points per question. After summarizing all questions, 7 points or lower was considered to indicate absence of significant levels of anxiety or depression, scores between 8 to 10 indicated doubtful cases of anxiety or

depression, and scores between 11 and 21 indicated definite cases of anxiety or depression (74). The validity was tested well in a Swedish study (75).

The scale was used to examine patients' and their next of kin's estimates of questions about anxiety and depression. It was used in study II, where the estimates from the pairs were compared with a general Swedish population, and in study III, where the patients' and next of kin's estimates were followed and compared over time.

The Schedule for the Evaluation of Individual Quality of Life – Direct Weighting (SEIQoL-DW)

Individual QoL examines the individual person's perspective of his or her QoL. Instead of using standardized questionnaires, the person self-presents what he or she thinks is important in life and how well it works. The SEIQoL-DW is a semi-structured quality of life instrument that assesses the QoL from the individual's perspective (42).

First, the interviewee chooses the five most important areas in his or her life at the moment. Second, the person estimates how well these areas work, or how satisfied the person is with the areas, through a vertical visual analogue scale (VAS) between 0 and 100 for each area, from the worst possible to the best possible level. Finally, the person weights the five areas against each other with a circular disc. The disc has five colour tags, whereby each colour represents one chosen area. The disc has a larger backing disc that displays a scale from 0 to 100, which then converts the ratings to a total of 1, since the pieces are measured in percent. The interviewee regulates these pieces until the weighting feels right. By this procedure, a SEIQoL index is calculated by multiplying each area level by the corresponding area weight. The products are then added up across the five areas and the SEIQoL index is computed. The instrument is validated and reliability tested (42). Also in the group of patients with ALS, the instrument had a high validity and reliability (76).

The instrument has in study III been translated by two separate translators from the original English version to Swedish. Then those translated versions were compared, and one uniform Swedish version was retranslated into English. Independent professional translators compared that version with the original version. The retranslated English version was also sent for assessment to the original makers of the instrument, but no response was received. A final retranslated version was then translated to Swedish again. The instrument was tested and evaluated in a few patients before the start of the study.

The Visual Analogue Scale (VAS)

The VAS is a common instrument that is used in many clinical and research settings for measuring subjective phenomena. The scale contains a line, often 100 mm, measuring the intensity of a phenomenon. The endpoints of the scale are extreme absence of the phenomenon and a maximum intensity of the phenomenon. The normal mood often is located close to the midpoint in VAS (77). Although the scale has been criticized (78, 79), the reliability and validity have been shown to be good, and the scale is a practical, easily administered scale. With good instructions and repeated use of the scale, as in longitudinal studies, the method can be reasonable and practical. Data from VAS have been analysed by both parametric and nonparametric statistical tests, and it has been concluded that the choice of analysis generally makes little difference (77).

In Paper I the patients and their next of kin estimated their own general, physical, and psychological well-being on a 100 mm horizontal scale, where zero implies very bad well-being and 100 implies very good well-being. After those estimates, the patients and their next of kin estimated the general, physical, and psychological well-being of the other person in the pair.

The Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised (ALSFRS-R)

The ALSFRS is an instrument that measures the physical functional status. It was used to assess changes in the functional states in patients with ALS. The scale has an internal consistency and a test-retest reliability, and has been shown to be in good agreement between the visits (80). However, the ALSFRS has a weakness, as it gives a disproportionate weighting to limb and bulbar, as compared to respiratory, dysfunction. Therefore, the ALSFRS has been revised to ALSFRS-R, which also assesses dyspnoea, orthopnoea, and the need for ventilatory support. The internal consistency and construct validity is shown to be strong, and it is a good predictor of survival time. The scale measures fine and gross motor, bulbar, and respiratory function (81).

The ALSFRS had earlier been translated to Swedish to be used in another study (10). In this thesis the ALSFRS-R was translated from English to Swedish and compared with the earlier translation of ALSFRS, and then the ALSFRS-R was retranslated to English and back to Swedish again by a translator.

The ALSFRS-R was used and evaluated by a physician in the three first papers at every study visit.

The Norris scale

The Norris scale is a clinical measure scale that measures the physical functional status and can be used for monitoring changes in the course of ALS. It has 22 items examining bulbar, respiratory, trunk, arm, leg, and general domains involving reflexes, fasciculation, and muscle atrophy (24, 82). The scale also measures emotional lability, fatigability and leg rigidity (82). The Norris scale assigns equal importance to changes in tendon reflexes and muscle strength as it does to bowel and bladder function, which seldom are disturbed in ALS (80). The Norris scale is a reliable measure with a linear decline during the course of ALS (83).

The Norris scale was used and estimated in the three first papers at every study visit by the physician.

The Mini-Mental State Examination (MMSE)

Since 20-40% of patients with ALS have slight cognitive dysfunctions from impaired frontal executive function (1), and since 5–10% develop a frontotemporal lobar dementia (FTLD) (1, 33), it was of substantial interest to make a rough test on cognitive dysfunctions of the patients to exclude those with significant cognitive disturbances at the beginning of the studies in Papers I–III, as the patients might be expected to fall out of the studies at later times, due to inability to give adequate reports. The MMSE was used to exclude major cognitive difficulties in patients. The scale includes 30 questions with 30 points; a level under 24 points is judged as abnormal (84).

Qualitative interviews

Interviews lead to understanding of another person's world view (85).
In Paper IV, the interviews were semi-structured, with questions inspired by Antonovsky's components in SOC, that is, comprehensibility, manageability, and meaningfulness (61). The participants were asked to talk about their experiences and life situation. Main issues were 'How has the disease affected your life?' 'How do you manage the situation?' 'How do you experience your life situation now?' and 'What gives you meaning?' To get a wider understanding, the interviewer asked questions such as 'Can you tell us more about that?' 'What do you mean by that?' To reduce the risk of misunderstanding, the interviewer also repeated what the interviewee had said, and asked if it was correctly perceived.

DATA ANALYSIS

Statistical analysis (Papers I and III)

Papers I and III were longitudinal studies intended to interpret the results obtained by multiple testing of changes over time in the physical functional

scales (ALSFRS-R and Norris scale), in well-being (VAS), in HRQoL (SF-36), and in anxiety and depression (HADS). A regression coefficient was calculated for each patient and next of kin in each variant of multiple analyses. The coefficient describes the slope of the graph where x-values are the time of the successive visits (at 0, 6, 12, and 18 months) and y-values are the variables investigated. Every regression coefficient was given an equal weight, regardless of whether the participant attended the study sessions two, three, or four times. The participants who were used to calculate the regression coefficients were drawn from those who still remained in the study at visit two and onwards. Fisher's test for paired comparisons was used to test whether the coefficient was different from zero, that is, whether there was a change over time. The same kind of analysis was performed when patients and their next of kin estimated each other's well-being by VAS over time.

Paired comparisons between patients and their next of kin were analysed with the Wilcoxon signed rank test. The Mann-Whitney test was used to analyse differences between gender at entry to the studies and in analysis by regression coefficients of changes over time. Spearman's rho correlation coefficient was used to examine whether there were correlations between scales. Data were paired and two-tailed tested, and presented by mean and standard deviation. In paper I, the Bonferroni test was used to reduce the risk of mass significance problems. Depending on the data studied, different p-values were estimated to be significant. Since it also is a risk to use the Bonferroni test, as it can give falsely negative values, a level of $p < 0.05$ was considered significant in Paper III.

Statistical analysis (Paper II)

In Paper II, Wilcoxon's signed rank test was used to analyse paired comparisons between the patients and their next of kin. Correlations were analysed by Spearman's rho test, while the Mann-Whitney test was used to compare differences in relation to gender. Pitman's permutation test was used to analyse age dependence in SF-36 and HADS. Since data were not paired with the general Swedish population, the t-test was used as a non-paired test to compare results of the general Swedish population versus patients and their next of kin in SF-36 and HADS. The tests were two-tailed, and mean and standard deviation were calculated. A value of $p < 0.05$ was considered significant.

Qualitative content analysis (Paper IV)

Content analysis can be dated to the churches of the 17th century, but it was during the Second World War that it became better known for its use in both propaganda and newspapers (85).

Depending on the purpose of the study, content analysis can be used in both quantitative and qualitative methods. It is a method that systematically analyses written or verbal communication (85), focusing on subject and context, and dealing with similarities and differences between and within parts of the text (86). Qualitative content analysis is used for subjective interpretation of a text's content through a systematic classification process of coding and identifying themes (87).

Text can be analysed with respect to both manifest and latent content. Manifest content of a text is analysed through little or no interpretation, so that only the obvious is analysed. Latent content makes an interpretation of underlying meanings in a text (86, 88). A text involves multiple meanings, which can always be interpreted in different ways. It is important to be alert to that consideration when trustworthiness is discussed (85, 86).

In Paper IV, the text was subjected to qualitative content analysis, inspired by Krippendorff and Graneheim and Lundman (85, 86). The interviews were tape-recorded and transcribed verbatim by the first author (A.O.). The transcribed text was randomly double-checked against the tape recordings by the last author (S.S.).

The analysis was performed in several steps. First, the text was divided into meaning units, and every meaning unit related to same central content and context. The meaning units were condensed and labelled with a code. Due to the extensive material, the codes were sorted into three content areas, comprehensibility, manageability, and meaningfulness. The continued analysis focused on the content area of manageability, since manageability was the content area that could best provide answers to how the participants managed the situation, and since the text material as a whole was too large to be analysed together. The codes were sorted and abstracted to 19 subthemes, illuminating threads of meaning running through the codes. Through a process of reflection and discussion, the authors agreed on the subthemes. The subthemes seemed to represent manageability in relation to various actors and their perspectives: oneself, the family, others, and the authorities. Finally, arising from these perspectives, the 19 subthemes were abstracted into four themes. To ensure that the formulation of themes was trustworthy, the analytic process involved a back and forth movement between the whole and the parts of the text. All authors were involved in that process and agreement on themes was reached. This trustworthiness can be compared with instructs from other researchers (85, 86, 88). Examples of the analysis are shown in Table 3.

Table 3. Examples of the analysis in Paper IV.

Theme	**Fighting for support versus not accepting support**		
Subtheme	**Support creates feeling of safety, while pride and shame decrease acceptance of help**	**Support creates feeling of safety, while pride and shame decrease acceptance of help**	**Support creates feeling of safety, while pride and shame decrease acceptance of help**
Codes	Dual feelings of wanting and not wanting help	Dual feelings of wanting and not wanting help	Personal assistants give calm and safety
Condensation	Wants to have freedom, but doesn't want people running around at the home	Restraints if we need help from the outside	Calm and safety when the cohabitant has personal assistants
Meaningunit	… I don't want to have people here all the time // I don't want to pay the accounts when someone is beside me. // but at the same time, one maybe, I think it is, should help me a little.	... but then, one never knows how it will be. And then, of course, there might be restraints. Then we shall have help from the outside and it feels a little special too; unknown people will come here. How will it feel? I don't know. I can't… that side, but one has to accept it to get a life, some personal life, one must have some personal life, one wants to have that.	Then my cohabitant has personal assistants and then one can feel both calm and safety to do what I want to do.

ETHICAL CONSIDERATIONS

Patients and next of kin whose lives are affected by ALS can be affected physically, psychologically, emotionally, and socially. Therefore, it is important to respect the individual in the situation he or she lives in and proceed from that point to analyse the person's life situation. In this thesis, the investigator asked the participants at each single study visit if they wanted to continue to take part in the study. Since the investigator was connected to the participating clinic, it was very important that the participants felt they could leave study without fear of any changes in their future treatment and care.

Patients and next of kin participated voluntarily. They received verbal and written information, and gave their approval to participate. Participants who were in such a physical or psychological condition that they were not able to give informed consent were excluded. Also excluded were patients who were in a terminal stage of the disease, because it could be too taxing for them to participate. In the fourth study, participants had to have had the diagnosis for at least six months before entry into the study, since they needed to understand the disease and the situation they were in.

A person not involved in the care of the participants was available to talk with the participants, if any of them needed that support in connection with a study visit. No participant used that opportunity.

The four studies in this thesis received approval from The Regional Ethical Review Board in Gothenburg, Sweden (No 297-05).

RESULTS

SAMPLE CHARACTERISTICS

At entry to studies I–III, the mean age of the patients was 63.4 years (median 64 years, range 28-84 years) and the mean age of their next of kin was 61.3 years (median 64.5 years, range 27-86 years). There were 17 men and 18 women among the patients and 16 men and 19 women among their next of kin.

In study IV, 14 patients (7 men and 7 women) and 13 next of kin (8 men and 5 women) participated. The patients were between 42 and 80 years of age (median 67.5 years) and next of kin were between 38 and 87 years old (median 68 years).

PHYSICAL FUNCTIONAL SCALES

The physical functional scales ALSFRS-R and Norris scale showed a steady decline in the patients during the study time. Since patients with lower ratings in the physical functional scales fell off from the study, the mean values in the fourth visit were higher than in the third visit. However, the statistical analysis used was related to each patient, so the changes over time were specific for each person studied (Table 4).

Table 4. The functional rating scales. Columns 2 to 5 show mean value (+/- standard deviation) and number of participants. Column 6 shows mean value (+/- standard deviation) and number of participants for regression coefficients.

	Baseline	Second visit	Third visit	Fourth visit	Mean, SD, n for regr. coefficients	Changes over time
Functional rating scales						
ALSFRS-R (0-48) total	36.6 (7.6)/35	33.4 (8.5)/32	31.7 (9.4)/26	35.4 (9.1)/9	-0.59(0.63)/32	p<0.001*
Female	37.2 (8.2)/18	34.9 (7.8)/15	34.1 (7.9)/13	39.4 (8.5)/5	-0.44(0.54)/15	p=0.007
Male	35.9 (7)/17	32.2 (9.2)/17	29.1 (10.4)/13	30.5 (8.1)/4	-0.73(0.68)/17	p<0.001*
Norris (0-100) total	72.7 (17.8)/35	67.5 (16.7)/32	63.4 (18.9)/26	71.6 (18.4)/9	-1.07(1.37)/32	p<0.001*
Female	74.8 (18.7)/18	71.8 (15.5)/15	69.2 (14.8)/13	80.8 (13.2)/5	-0.77(1.16)/15	p=0.02
Male	70.5 (17)/17	63.8 (17.3)/17	57.6 (21.2)/13	60 (18.8)/4	-1.33(1.51)/17	p=0.002*

* Fisher's test for paired comparisons was used to analyse changes over time. P-values in the table indicate deterioration over time from baseline to the last visit for each patient/next of kin. With the Bonferroni test, P-values <0.002 indicate significant differences.

SUMMARY OF PAPER I

No changes were found in well-being over time within the whole group of patients or their next of kin. However, when the groups were separated by gender, there was a decrease in physical well-being ($p < 0.001$) and a tendency to reduction in the general well-being ($p < 0.05$) over time in male patients. No significant differences were found between genders in either patients or their

next of kin over time, even though it was a tendency that male patients estimated poorer physical well-being than female patients did.

At the first study visit, there was a tendency of poorer physical and psychological well-being ($p < 0.05$) in female next of kin than in male next of kin, but neither here were there any significant differences in gender.

Patients estimated physical well-being worse than next of kin at the first study visit. The ratings of patients were lower than those of next of kin at each time of the visits, even though no differences were found in estimates over time between them (Paper I, Table 2). The relationship between the estimates of each patient and next of kin in general and psychological well-being was fairly constant in each pair throughout the part of the course of the disease. Thus, if the rating was low in the patient, it was also low in the next of kin; if it was high in one of them, it usually was so in both parties. However, there were some pairs with a larger difference, indicating that one of them had a lower rating of well-being than the other.

When the pairs estimated each other's well-being, next of kin usually estimated the patient's general and psychological well-being worse than the actual self-estimates of the patient. There was a trend that next of kin also estimated the physical well-being worse than the actual self-estimate of the patient. In contrast, the patients estimated the well-being of their next of kin much the same as their next of kin did for themselves.

SUMMARY OF PAPER II

Not surprisingly, the patients had poorer ratings than their next of kin in the physical component summary (PCS) in the SF-36 ($p < 0.001$). However, no differences were found between them in the mental component summary (MCS) in the SF-36. In most pairs the estimates of the SF-36 MCS followed each other, so that if one of them estimated low ratings, the other one also did, and vice versa, as is also shown in the rating of well-being in Paper I.

Patients' ratings were lower than those of the subset of the general Swedish population in both SF-36 PCS and MCS. The next of kin had poorer ratings than the general Swedish population in SF-36 MCS. No differences in gender or age were found in either patients or the next of kin.

There were no differences between patients and their next of kin in HADa or HADd. A correlation was found between them in HADa ($p < 0.001$, $r = 0.566$**) but not in HADd. Most of the pairs of patients and next of kin gave equal estimates in the HADa; thus, if one of them gave low ratings, the other one also did, and vice versa.

Both the patients and their next of kin estimated lower ratings in HADa and HADd than did the general Swedish population. No differences were found in relation to gender or age, either in the group of patients or the group of next of kin.

The physical functional scales did, as expected, correlate with the SF-36 PCS. They also correlated with the patients' HADd. The physical functional scales did not correlate with any of the next of kin's scales.

SUMMARY OF PAPER III

Changes were found over time in the patients' subscales of PCS, PF, and GH in SF-36. Next of kin had changes over time in the subscales MCS, PF, BP, and RE. No changes were found over time in the global QoL score in SEIQoL-DW or in HADS, in either the patients or their next of kin, and there were no differences with respect to gender.

Patients estimated poorer PCS, PF, RP, and GH in SF-36 than did the next of kin at the first study visit. However, the next of kin had a poorer global QoL score in SEIQoL-DW than the patients. There were no differences in the changes over time in paired comparisons between patients and their next of kin in the SF-36, SEIQoL-DW, or HADS, except in SF-36 PF, where patients' physical functioning decreased. The differences found between the pairs at visit one were similar at later visits in the study. The patients still had poorer estimates in the SF-36 PCS and in the subscales of PF, RP, and GH, and their next of kin still had poorer estimates in the global score in SEIQoL-DW, which can be seen in the mean values (Paper III, Table 1).

The areas chosen in SEIQoL-DW did not change a great deal over time in either the patients or their next of kin. Both patients and their next of kin estimated relationships such as those with children/grandchildren, family, marriage and friends as important areas of life. However, the functioning/satisfaction were often better among patients than among their next of kin. There were also some differences between the pairs in the chosen areas. Patients estimated, for example, hobbies as being more important, whereas, next of kin estimated employment/education and travelling as more important.

Among the patients, ALSFRS-R correlated with PF and MH in SF-36 over time. The Norris scale correlated with patients PCS, PF, BP, and MH in SF-36 over time. No other correlations were found between the physical functional scales and SF-36, SEIQoL-DW, or HADS over time in patients or their next of kin, except between patients' ALSFRS-R and next of kin's PF.

SUMMARY OF PAPER IV

The themes in Paper IV reveal a constant fluctuation between factors that facilitated and factors that hindered manageability for the individual person, with similarities and differences between the groups of patients and their next of kin. When data were analysed, four perspectives arose, those of oneself, of family, of others, and of authorities. The latter included health care personnel and local authorities, as well as other authorities. Each perspective was accorded one theme of its own, which included both patients and their next of kin.

In the perspective of oneself, the theme *fluctuations in acceptance and burden* illustrated that both patients and their next of kin tried to accept the situation and live in the present, because it reduced the pain of thinking about the disease and the future. Patients focused on their health and on leading an active life; they felt that both their self-esteem and their activities were affected by the changes in integrity and autonomy. Forced passivity led to frustration, negative thoughts, isolation, and control issues. However, next of kin were affected by the burden, and that affected the fluctuation between opportunity, and lack, of time for themselves. They felt that their own ill health and their fear and hate of the disease decreased their ability to manage.

In the perspective of the family, the theme *fluctuations in support and disparate needs* illustrated that both patients and their next of kin felt that understanding and support within the family strengthened their ability to manage, while different communication needs decreased it. Patients also felt that feelings of guilt weakened it, whereas, the next of kin felt that feelings of being controlled by the patient and practical liabilities weakened it. Further, next of kin felt that their own creativity facilitated their common everyday life. Both patients and their next of kin were affected when under-aged children were involved. Patients derived strength from the children, but also worried about the children's vulnerability. Next of kin had qualms of conscience due to the children's vulnerability, and they worried about not providing enough support.

In the perspective of others, the theme *fluctuations in real presence and fear* illustrated that patients' own attitudes and speech problems controlled their communication with others. Both members of the pairs felt that real presence from others gave them strength. However, the manageability was decreased if patients felt that other people were afraid of the situation, and next of kin experienced decreased manageability if they felt fear of preconceptions and absence of support.

In the perspective of authorities, the theme *fighting for support versus not accepting support* made it evident that both patients and their next of kin felt that support created feelings of safety, while pride and shame decreased their

willingness to accept help. Patients felt that lack of insight among authorities made the manageability more difficult. Next of kin felt that help from the health and social services, outside and support for communication facilitated everyday living.

DISCUSSION

FINDINGS

In this thesis, the HRQoL with SF-36 and HADS in patients with ALS and their next of kin were compared with a subset of the general Swedish population. It was obvious that patients had worse estimates in both SF-36 and in HADS, while next of kin had worse estimates in the HADS and in the mental component score in SF-36. It has been shown that physical subscales in QoL are worse in patients compared with the general population, while the mental subscales were slightly better in patients (4). In contrast, it also has been found that mental subscales in QoL are lower in both patients and caregivers compared with the general population (54). Regardless of the physical function of the patients, the lower ratings in both patients and next of kin indicate that the consequences of the disease affect them, even though the QoL does not decrease much over time, as shown in Papers I and III. If there already are lower levels, it might be expected that such findings do not change much more over time.

Comparisons between Papers I and III showed that few changes were found in each patient or next of kin over time by the different instruments used in the studies. In well-being, anxiety, depression, and individual QoL, no changes were found in the patients and their next of kin. In the HRQoL instrument SF-36, some changes appeared in both patients and their next of kin, even though most estimates were constant over time. Since the disease has a continuous progression with a decreased physical functioning over time, it was not surprising that the estimates of the patients in PCS, PF, and GH in SF-36 also showed decreasing levels. Other studies also have found that physical subscales in QoL decrease over time, while mental subscales do not (4, 9, 11). The similarities between our findings and others' also strengthen the adequacy of the choice of instruments used in our studies.

It could have been expected that the physical well-being in the VAS also should have decreased in the whole group of patients. We found a decrease only in male patients, but comparisons between genders did not show any significant differences between them. The absence of deterioration in the whole group of patients may be attributed to patients' having already estimated lower levels in their physical well-being from the beginning and to their response to the situation having shifted, which can be compared with another study (89). As expected, and in agreement with other studies, in our study the physical functional scales ALSFRS-R and Norris scale showed decreased functions in the patients over time (4, 5, 58, 81).

The decreased estimates in SF-36 in Paper III among next of kin might depend on the physical and mental burden they take on, which also was found in Paper

IV. Other studies also have found that the caregivers' burden increases with the worsening of patients' disability (5, 55), which might lead to an increased risk of worsening of their health, and particularly, of their mental health (19). Both in Paper IV and in another study, it was obvious that next of kin needed time to themselves, as well as feelings of support from others, to be able to manage well. Health care professionals should promote and help them to take time for themselves. In comparison with the theory of care by Benner and Weber (63, 65), it is important to be aware of the family's situation and how they master the situation, since their ability to cope also affects the patient. These points emphasize the need to take care of the whole family, because the patient is less likely to manage successfully if those around the patient are not also managing well.

It might have been expected that the anxiety in patients and next of kin would be high, due to the incurable disease. However, in agreement with other studies, both anxiety and depression were mild and not very prevalent in most patients (5, 6, 45, 47, 57, 58). Also, next of kin, had in our studies relatively low estimates of depression, which also is confirmed by other studies (5, 19). One possible explanation of the low ratings in patients and their next of kin might be that they find ability to manage the situation, as is shown in Paper IV. There were both facilities and hindrances in the manageability. Perhaps they find a balance, even though there occasionally are feelings of anxiety and impediments to the manageability. The possibility of developing cognitive dysfunction and frontotemporal lobar dementia (FTLD) (1, 33) may also result in a lack of initiative and decreased emotional reaction to the disease and its consequences. Dementia is not common, but it needs to be observed. However, in our work, the MMSE excluded possible patients with broad cognitive defects.

With respect to gender, no differences between males and females were found over time in SF-36, HADS, or SEIQoL-DW. In well-being as measured by the VAS, male patients showed deterioration in physical well-being. However, when compared to female patients, no significant differences were found. More interestingly, there was a trend of lower estimates in female next of kin in both physical and psychological well-being compared to male next of kin. After Bonferroni test correction, there were no significant differences, but when studying the mean values, differences can be seen between male and female next of kin in all component scales in the VAS, where the women had worse estimates than the men. Possible explanations can be that male patients had a trend of worse physical function and that female next of kin probably are physically weaker than male next of kin, which could make it more difficult to take care of the ill one. The conclusion is, however, that even though there were some trends of differences with respect to gender in both patients and their next

of kin, more research about the perspective of gender is needed to substantiate or reject possible differences.

Between the groups of patients and next of kin, no differences were found in the SF-36, HADS, or VAS, except in physical well-being in the VAS and in the physical subscales PCS, PF, and RP, and in GH in SF-36 at visit 1 and thereafter. Apparently due to the disease, patients' estimates were worse than next of kin's in the physical parameters. Surprisingly, next of kin had worse estimates than patients in the individual QoL measured by the SEIQoL-DW. Possible explanations may depend on limitations in their own lives due to the burden, which was illustrated in Paper IV, as well as in other studies (5, 55). It has been found both that there are no differences in the QoL or depression between patients and their caregivers (51), and also that just caregivers had increased levels of depressive ratings, even though it was too low to be classified as a depression (5). In agreement with our study, it has been found that patients had better individual QoL than their caregivers (48). However, in our study the pairs were matched with each other, which they were not in the other studies (48, 51). Few longitudinal studies have examined the QoL in pairs, especially with equal numbers of patients and caregivers (5, 90).

It appears that the individual pairs' estimates were closely aligned in VAS, in HADS, and in some subscales in SF-36, while there were some differences between the pairs in other subscales in SF-36 and in the SEIQoL-DW. The results suggest that if either the patient or next of kin feels bad, the probability is high that the other person does also. This inference must be regarded with caution, as there were also differences between them expressed in studies I–III. Also in Paper IV, differences were found in how they managed their life situations. It is necessary to give the patients and their next of kin information about possible similarities and differences between them, so they can meet each other more openly. The results strengthen the notion that care needs to be directed to both the individual person and to the whole family together from the beginning of the disease to the end.

The SEIQoL-DW reveals what the individual person regards as important in life. In Paper III, both patients and next of kin chose relationships, especially with children/grandchildren, family, and friends, as very important areas in the individual QoL instrument SEIQoL-DW. They also valued health as an important area in life. Differences showed that patients rated hobbies and marriage higher, and next of kin appreciated employment/education more. Common areas of great importance for individual QoL have in other studies been shown to relate to health, family, social activities, finances, spirituality/religion, and physical and psychological well-being for both patients and their caregivers, and accommodations, leisure activities, mobility, and

relationships for patients (8, 50, 60). At the last visit in Paper III, patients rated mealtime, getting out, and independence as important. It is possible that the low functioning/satisfaction, especially in getting out and in independence, increased the importance of these areas. Mealtime seemed to be associated with relatively good functioning/satisfaction, but it is possible that the importance of the area depended on a possible feeling of insecurity about what would happen with swallowing and the physical function of being able to eat, independent of others. However, it can not be confirmed in this thesis, since it was not studied.

It is important to study if and how the individual QoL changes over time, so health care professionals can focus on helping the person where he or she is at the moment. Nowadays, most studies examining SEIQoL-DW are cross-sectional (46, 48, 50, 91, 92), and there is insufficient research over time among patients and their next of kin together. In Paper III, there were few changes over time in the chosen areas in SEIQoL-DW. Health care professionals can take note of what they should focus on when helping the individual and the family. To make the QoL as good as possible for the individual, it is important to find out what is important to him or her. Next of kin more often chose areas such as travelling and having personal time/freedom than patients did. These results can be compared with the burden and lack of personal time that was found in Paper IV. It seems as if next of kin have a large burden that also affects their individual QoL in the SEIQoL-DW. The importance of relationships reported in Paper III is also shown in Paper IV, where the themes were derived from, inter alia, the perspective of family, others, and authorities. If important areas in life have poor satisfaction or functioning, it probably will be more difficult to manage the situation; if important areas in life are working satisfactorily, it probably makes it easier to manage the situation. In Paper III, mean values in the SEIQoL-DW were 79–66 in patients and 64–69 in next of kin, which are in agreement with other studies (46, 48, 50, 91, 92). However, the median in patients' QoL has also been shown to be as low as 46, and in caregivers, 56 (60). In agreement with a longitudinal study with two-month intervals, the mean value does not decrease over time in patients (8).

In Paper I, patients and next of kin estimated each other's well-being in the VAS. Next of kin underestimated the patients' well-being, while patients estimated the well-being of their next of kin as the next of kin themselves did. Other studies have found tendencies for caregivers to under-estimate patients' QoL and tendencies that they estimated that patients were more affected of the disease than they actually were (51, 90), while it was found both that patients tended to over-estimate caregivers' QoL (51) and that they over-estimated caregivers' burden (90). Since it would be too much effort for the participants to estimate each other's measurements in the other scales, we did not expose them to that task.

In study IV, it was obvious that there were constant fluctuations between factors of life that facilitated and factors that hindered manageability for the individual patient or next of kin. There were also similarities and differences between the patients and next of kin. The affected self-esteem in patients who lost control and their use of passive and active strategies to try to manage the situation is confirmed by other studies (16-18). In comparison with other studies caregivers are affected by the burden of care giving (5, 55, 68). In our study, next of kin lost their sense of control because of the burden of always being available, of not getting any personal time, problems with setting limits, guilty feelings, and feelings of being controlled. Opposite to other studies (21, 67), the importance of using faith and spiritual support to improve manageability in patients and next of kin did not appear in either Paper III or IV in this thesis. Possible explanations can be that Sweden is a secularized country, with either a large proportion of non-believers or those who regard their beliefs as private. It is also possible that faith will be found to be important when meaningfulness is considered in a further study.

In the third paper, it was obvious that children and grandchildren were very important to both the patient and the next of kin. The fourth paper confirmed that fact, but it was also obvious that under-aged children caused feelings of worry, qualms of conscience, and concerns about not being able to support them. Few studies have examined the parents' perspective of having under-aged children (13), and further studies are necessary.

Both in Paper IV and in other studies (19, 21), it was found that care and support from the family and from health and social services are very important for managing the life situation in both patients and next of kin. Patients can both trust the professionals but also be insecure about their service entitlements, and caregivers can experience lack of knowledge and understanding by professionals (14, 17, 19). The situation is complex, since it appeared that patients and next of kin both felt that they needed to fight for support, at the same time as they were reluctant to accept support. There were both strengths and hindrances in the strategies within the same themes. It is important to be aware of the phenomenon that people change in their ability to manage from one moment to another; the individual person has to be supported where he or she is at the moment, but it is also important to help them find strategies that can assist them to move on.

METHODOLOGICAL CONSIDERATIONS

The overall aim of the thesis was to describe different perspectives of QoL in patients with ALS and their next of kin, and to describe strengths and hindrances in the manageability of their daily lives. The concept of QoL is broad, and by studying HRQoL, individual QoL, well-being, psychological aspects such as anxiety and depression in relation to the patient's decreased physical function over time, through comparison with the general population, and through interviews about the manageability of the life situation, deeper understanding could been obtained. The use of different instruments and methods of both quantitative and qualitative analysis in the same groups of participants made it possible to get a wider perspective and see differences and similarities between the patients and their next of kin. The possibility to enhance the care and support to both the individual and to the family were increased.

The focus of having paired data and studying both patients and next of kin throughout the studies also increases the knowledge about the family situation, since there are few studies that examine the pairs together from the different aspects and methods as were used.

Quality of life is a wide concept encompassing physical, psychological, social, and spiritual dimensions, as well as indicators of well-being and functional status (23). The aim of using the instruments that were selected for this thesis was to get wide perspectives of the QoL. SF-36 is a generic HRQoL instrument; HADS is often included in studies using HRQoL instruments, and it exposes signs of anxiety and depression among the participants; SEIQoL-DW is an individual QoL instrument that reveals signs of what is considered important to the person himself; and VAS showed well-being from different perspectives. These instruments, together with the physical functional scales ALSFRS-R and Norris scale, could give multidimensional evaluation of the QoL dimensions of physical, psychological, social, spiritual well-being, and functional status, depending on what the participant estimated.

It was an advantage to use these instruments, rather than more physically oriented QoL instruments or illness-related instruments, such as SIP or SIP/ALS-19. We excluded those types of instruments, partly because the physical functional scales in this thesis are related to and validated to ALS, and partly because many patients experience high emotional distress when using physically oriented instruments or illness-related instruments, since they already know that they have a serious, progressive disease (93, 94). The McGill Quality of Life Questionnaire (MQOL) could have been used (93, 95), but we preferred more commonly used instruments in ALS.

The instruments used have been tested for validity and reliability in earlier studies (93, 94, 96). However, our versions of the VAS and the SEIQoL-DW have not been validated and tested for reliability. The results would have been strengthened if these scales had been compared with a Swedish general population. On the other hand, it would have been difficult to find a representative population, and since SEIQoL-DW is an individual QoL instrument, the findings are needed and interesting, just because they are individual. It would probably not contribute much to study the general population. To reduce the risk of incorrect grouping with respect to age, the Pitman's permutation test was used in study II. Comparisons with the general population were not matched with respect to age and gender. Since the factors important to match in the population are unknown, a proper meaningful and reliable matching would not be possible to accomplish.

Further, in Paper II, we only compared the PCS and MCS in SF-36. It might have been of interest if we had used all the subscales, but on the other hand, the PCS and MCS cover important domains of physical and psychological QoL.

The numbers of investigated people in Papers I–III were deemed to be a number sufficient to be able to detect major differences and similarities of clinical importance. It is possible that a larger sample might have given other results, but the results in our studies, for the most part, are supported by the results of others.

The longitudinal studies were planned to be finished in December 2007, and therefore, 14 pairs were not seen for a fourth visit, as they were not eligible for the follow-up. To analyse the possible consequences of follow-up of a smaller cohort due to the loss of patients, we analysed and compared the 26 pairs who completed all the first three study visits with the whole group of 35 pairs. The results from the two groups were equivalent. Loss of participants in studies with severely debilitating and lethal diseases such as ALS is unavoidable, but in this thesis the chosen statistical methods compensated for the loss. However, prolonging the study would have increased the strength of the research, as it would have allowed some of the last 14 pairs to also participate in the fourth visit.

A follow-up every 4–6 months in the longitudinal studies was chosen, because we wanted to investigate the pairs at relatively long intervals. The longer time frame strengthens the possibility to find possible changes due to the course of the disease or due to effects of the burden of disease. Some other longitudinal studies had shorter intervals and paid much less attention to seeing both patients and next of kin/caregivers.

The fourth study estimated the manageability, which is included in the sense of coherence (SOC) (61). Due to the large amount of data, it was not possible to study all three components of comprehensibility, manageability, and meaningfulness, which are included in the SOC, since the analysis would have been diluted and superficial. All three components are important and it has seemed that meaningfulness is required for good manageability, and comprehensibility is needed to facilitate manageability (61). However, the focus on manageability in study IV had purpose to derive resolute guidelines for how health care professionals can help patients and their next of kin. Comprehensibility and meaningfulness will be examined in further studies that will complement the fourth study with important information.

Before the studies started, senior researchers working with QoL instruments were consulted about the methodology, strategy, and choice of instruments. A statistician was consulted in the first three studies, before the studies started and in connection with the analysis. All authors participated in the preparation of the studies and an external researcher participated in the fourth paper, to increase the competence and trustworthiness of the whole process. All papers have been reviewed in the research group.

Since some of the authors were connected to the participating clinic, it was very important for the participants to feel that their participation or lack of participation would not affect their treatment, the relationship with the ALS/MND team and the future care. It is possible that answers were affected by this circumstance, but since participants gave exhaustive descriptions of their life situations, and after the investigator discussed the circumstances with the participants, it is hoped that the risk was reduced.

CONCLUSIONS

This thesis was conducted to increase the understanding and knowledge about different perspectives of QoL, such as health-related QoL, individual QoL, anxiety, depression, and well-being in both patients with ALS and their next of kin. It was also done to increase the understanding of the complexity of finding strategies to manage the living situation, from the perspective of both a patient and a next of kin.

The major findings of the thesis were:

- Few changes were found in the QoL over time in patients and their next of kin, even though the physical function in patients decreased. Patients estimated decreased ratings in the physical subscales in HRQoL and in physical well-being, while next of kin estimated decreased ratings in some of the physical and some of the mental subscales in HRQoL.
- Even though the QoL was relatively good, both patients and their next of kin estimated lower ratings than the general population.
- Few differences were found in the QoL between patients and next of kin, but patients estimated lower ratings in the physical subscales in HRQoL and in physical well-being, while next of kin estimated lower individual QoL in SEIQoL-DW. The results indicate the importance of giving support and care to both the individual and the whole family.
- Estimates made by members of the individual pairs were often parallel, which suggests that if one of them feels bad, the other one does also, and vice versa.
- Patients' estimates of the well-being of their next of kin corresponded to those of the next of kin themselves, while next of kin estimated patients' well-being worse than patients themselves did.
- Relationships were the most important areas in life in the individual QoL of both patients and their next of kin.
- There were both facilitating factors and hindrances in the complex situation of managing the life situation for patients and their next of kin.
- There were both obvious similarities and differences in the facilities and hinders of manageability between patients and their next of kin.
- The four themes in the qualitative paper were derived from oneself, from the family, from others, and from the authorities.
- Large burden and decreased time to oneself diminished the individual QoL and manageability for the next of kin.

CLINICAL IMPLICATIONS

The findings in this thesis manifest a deeper understanding and knowledge about the complex situation of living with ALS as both a patient and as a next of kin. They will hopefully give support and guidance for how health care professionals can help the families to meet their needs.

Often patients and their next of kin estimate relatively equal QoL, which supports the view that health care professionals need to be extra observant of the whole family and their situation, if it appears that one member of the family feels bad. Since the patients' ability to master the situation also affects how well other family members can master the situation (63, 65), focus need to be concentrated on both the patients and their next of kin. There is a potential risk that if the situation in the family does not work, it will cause that the patients' situation deteriorates, with a possible increased burden on the health care system, since the self-care does not work. The possibility for the whole family to live in a situation they can manage and master benefits the patients, their next of kin, the health care system, and the community.

However, there were also some differences in the estimates of QoL and in the manageability between patients and their next of kin. These differences confirm the importance of giving individual support as well as support to the family members together. Individual support is important, since the person needs to be met, confirmed, and supported in the situation he or she is in at the moment. To increase the possibility to get a higher understanding in the pairs, it also is necessary to support them together. They need to get information, support, and supervision about what a situation can be like; patients and next of kin often have equal QoL, but there also can be some differences, especially in how they manage the complex situation in which they live. It hopefully can help them to understand the other person's situation and needs. Patients also need to be aware that next of kin often are very burdened in the situation. It is important to create opportunities to give them time on their own for recreation, so they can manage the situation over a long time.

The individual QoL instrument SEIQoL-DW can, in part, be used in clinical care to help the person himself, the next of kin, and the health care professionals to be aware of what is important in life and how it works. It may help all of them to understand and focus on the things that are important for the individual person. Clarification of the important areas in life may help to increase the ability to manage the situation.

Further, the family and the health care professionals benefit from knowing that the ability to manage the situation often can change from one moment to the next, and that experience of strengths and hindrances affect the manageability of

daily living. The person needs to get support, confirmation, and supervision in the situation he or she is in, but it also is necessary to help the person to see and find their own strategies to manage the situation and to move on.

Medical and nursing care can hopefully improve through the increased knowledge and understanding of the situation of living with ALS as a patient and as a next of kin, from the diagnosis to the end of life.

SVENSK SAMMANFATTNING
Att leva med ALS
Perspektiv från patienter och närstående

BAKGRUND
Amyotrofisk lateralskleros (ALS) är en neurodegenerativ sjukdom som kan ge kliniska symtom och undersökningsfynd som svaghet, spasticitet, hyper- eller hyporeflexi, muskelatrofi, fascikulationer, tal- och sväljningsproblem. I 90-95% av alla fall är orsaken till sjukdomen okänd medan 5-10% drabbas av ärftlig ALS. Idag finns det inget bot mot sjukdomen och oftast avlider patienten i en koldioxidretention inom 2-5 år beroende på symtomutveckling.

Livskvalitet och hanterbarhet vid ALS har undersökts i tidigare studier men det är få som har undersökt patienter och närstående i par och över tid med längre tidsintervall vilket är genomfört i denna avhandling.

SYFTE
Det övergripande syftet med avhandlingen var att beskriva livskvalitet utifrån hälsorelaterad livskvalitet, individuell livskvalitet, ångest, depression och välbefinnande hos patienter med ALS och deras närstående i relation till patienternas fysiska funktionsnedsättning samt att belysa styrkor och hinder i deras hanterbarhet i det dagliga livet.

METOD
Deltagarna i avhandlingen rekryterades från Sahlgrenska Universitetssjukhuset i Göteborg, Sverige. För deltagande i studierna skulle patienterna ha säker eller sannolik ALS. Trettiofem par av patienter och närstående deltog i de tre första kvantitativa delstudierna och 14 patienter och 13 närstående deltog i den fjärde kvalitativa studien.

Studie I och III var longitudinella med ett undersökningsintervall mellan 4 – 6 månader. Studie II var en tvärsnittsstudie. Både patienter och närstående skattade sin livskvalitet utifrån olika instrument för att en så bred bild som möjligt skulle framkomma. Delstudie IV var kvalitativ och deltagarna intervjuades utifrån frågor inspirerade av Antonovsky's teori om känsla av sammanhang (KASAM); det vill säga begriplighet, hanterbarhet och meningsfullhet (61). Intervjuerna analyserades med innehållsanalys.

RESULTAT
Det framkom få förändringar över tid i livskvaliteten hos patienter och närstående. Den fysiska funktionen, det fysiska välbefinnandet i VAS och en del

av de fysiska subskalorna i SF-36 försämrades över tid hos patienterna. De skattade även dessa subskalor sämre än närstående skattade deras fysiska hälsorelaterade livskvalitet. Närstående skattade försämring i deras hälsorelaterade livskvalitet över tid i både en del av de fysiska och de mentala subskalorna i SF-36. De skattade sämre individuell livskvalitet i SEIQoL-DW än vad patienterna gjorde. I övrigt fanns det inga försämringar över tid i grupperna av patienter och närstående över tid i ångest, depression, individuell livskvalitet eller välbefinnande.

Skattningen av livskvaliteten var ofta densamma hos paren av patienter och närstående vilket tyder på att om den ena mår dåligt så är risken stor att även den andra personen gör det. Det förekom dock även viktiga skillnader mellan dem. Deltagarna angav relativt god livskvalitet men studie II fann ändå att både patienter och närstående hade en sämre livskvalitet än en jämförd del av befolkningen.

Alla deltagare skattade relationer av olika slag som mycket viktiga områden i livet i SEIQoL-DW men det förekom även en del skillnader mellan vad patienter och närstående skattade som viktigt.

I den fjärde delstudien framkom en konstant fluktuation mellan aspekter av möjligheter och hinder i hanterbarheten hos den enskilde individen samt likheter och skillnader i hur patienter och närstående hanterade deras livssituation. Båda grupperna försökte leva i nuet och finna acceptans, patienterna försökte fokusera på deras hälsa och att leva ett aktivt liv. På grund av förändrad integritet och autonomi försvårades dock hanterbarheten med en försämrad självkänsla och inaktivitet. Den påtvingade passiviteten ledde till frustration, negativa tankar, isolering och kontrollbehov. Närståendes hanterbarhet försämrades av den stora bördan de fick bära och bördan försvårade till stor del möjligheten till egen tid. Känslor som rädsla, hat och egen ohälsa försvårade också hanterbarheten. Vidare upplevde både patienter och närstående att familjesituationen och kommunikationen dem emellan påverkade hanterbarheten mycket. Relationer med vänner och andra utomstående var också faktorer som påverkade dem. Upplevelsen av äkta närvaro från andra stärkte deras hanterbarhet men patienterna upplevde att detta försämrades om de kände att andra var rädda för situationen och närstående var själva rädda för förutfattade meningar från andra. De upplevde att frånvaro av andras stöd försämrade deras möjlighet till att hantera situationen. Både patienter och närstående upplevde att de kämpade för att få rätt stöd från sjukvård och samhälle samtidigt som de hade svårt för att acceptera och ta emot hjälp utifrån.

DISKUSSION

Livskvaliteten var relativt god hos patienter och närstående men den var ändå lägre än hos den jämförda andelen av befolkningen. Detta resultat tillsammans med fynd av att mycket av livskvaliteten skattades lika mellan patienter och närstående och att det fanns en del skillnader i både livskvalitet och i hanterbarhet styrker att patienter och närstående behöver både individuellt stöd och stöd tillsammans i familjen. Det individuella stödet är värdefullt för att individen själv ska få möjlighet att bli mött, bekräftad och stödd i den situation han eller hon befinner sig i för tillfället. Gemensamt stöd kan öka möjligheten till förståelse mellan parets olika sätt att se på livssituationen, vad som är viktigt och hur de kan hjälpas åt att hantera situationen. Eftersom de närståendes börda är så pass påtaglig är det även av stor betydelse att patienterna får vetskap om detta och att man tillsammans kan utforma möjligheter för egen tid och rekreation. De ofta likartade skattningarna i livskvaliteten visar även att sjukvårdspersonal behöver vara extra observant på hur hela familjen mår eftersom det tyder på att om en person mår dåligt ökar risken att en annan person i familjen också gör det.

Den djupare förståelsen av hanterbarheten och vetskapen att den kan förändras från en stund till en annan gör det möjligt att möta familjen och den enskilde ur ett vidare perspektiv och ge stöd, bekräftelse och handledning utifrån den punkt personen befinner sig i för tillfället. Det är samtidigt viktigt att även försöka hjälpa personen att finna strategier över hur han eller hon kan finna nya vägar till hanterbarhet.

Hälso- och sjukvården och andra delar av samhället kan förhoppningsvis utifrån den kunskap och förståelse som framkommit i denna avhandling förbättra omhändertagandet av både patienter med ALS och deras närstående från diagnos till livets slut.

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