Optimizing quality of care for patients with ALS and their family caregivers
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General introduction
1.1 Amyotrophic Lateral Sclerosis (ALS)

Amyotrophic lateral sclerosis (ALS) is a lethal, progressive motor neuron disease (MND) that mainly causes degeneration of the upper and lower motor neurons ultimately leading to paralysis and early death. Clinically ALS presents with considerable variability in onset, presentation of symptoms and disease progression, resulting in a complex and diverse disability profile. Respiratory failure is the most common cause of death in ALS. The median survival time after symptom onset is two-three years.\(^1,2\)

Due to progressive muscle weakness and other symptoms, patients with ALS experience increasing difficulties in the performance of activities of daily living and this may lead to restrictions in participation, impending loss of autonomy and quality of life.\(^3,5\) Dysfunctional motor neurons affect voluntary muscle activity, for example causing difficulties with sitting, walking, manipulating objects, chewing, swallowing, speaking and breathing. Besides isolated motor neuron involvement, patients with ALS and their families may be faced with cognitive deterioration and/or behavioural changes, which indicates that ALS is a multisystem neurological disease.\(^6\) About 5-10% of ALS is familial, the remaining 90% of patients is classified as having sporadic disease. It is estimated that the Dutch ALS population consists of about 1400 patients at any time. In the Netherlands, the average annual incidence rate is 2.8 per 100.000 person-years, and a prevalence rate of 10.3 per 100.000 individuals, which is similar to the most recently published prospective population-based registers in other countries.\(^7,8\)

1.2 Multidisciplinary care for patients with ALS and their informal caregivers

In the absence of a cure or medical interventions to stop the progression of muscle weakness, the focus of support for patients with ALS is on symptomatic, neuro-palliative rehabilitation care.\(^9,10\) The management of ALS varies considerably around the world despite the current recommended ‘gold standard’ for ALS care; expert multidisciplinary management.\(^11,12\) As recommended within the ALS guidelines from the American Academy of Neurology (AAN) and the European Federation of Neurological Societies (EFNS) multidisciplinary ALS care should be available for patients with ALS.\(^13,14\) In the Netherlands, multidisciplinary ALS care is available nationwide, and it is estimated that at least 90-95 percent of the Dutch patients with ALS receive care in conformity with these international guidelines, the Dutch consensus protocol for rehabilitative management in ALS and the multidisciplinary allied health practice guidelines for physical, speech and occupational therapy in ALS (http://www.als-centrum.nl/kennisbank/multidisciplinaire-als-richtlijn/).\(^7,13-15\)

The diversity in rate and degree of loss of muscle strength in different body regions, and the associated diversity and amount of disease symptoms, emphasize the urgency for evidence based multidisciplinary care for patients and their informal caregivers. The related range of impairments and limitations (e.g., related to nutrition, respiration, ambulation, cognition, well-being), that can present during the disease course, and related treatment choices further contribute to the complexity of ALS care. It is inherent that many different
health care professionals (Figure – The multidisciplinary ALS team), social services, agencies, authorities, companies, organizations and volunteers are involved.

In ALS, optimal timing of appropriate care interventions is of great importance for the patient and a serious challenge for their health care professionals. However, the progression of the disease and the functional limitations in time are currently difficult to predict, which hampers informing the patient about the expected disease course and the proactive provision of care interventions. Earlier studies attempted to construct prognostic algorithms for survival. However, prognostic tools for the functional course in ALS are lacking.

Figure The multidisciplinary ALS team.

1.3 The quality of ALS care
The current evidence base for the management of ALS indicates that care in a multidisciplinary setting is most effective. There is some evidence for extended survival, decreased medical complications by symptom treatment, increased use of assistive devices and improved QOL, related to attendance at specialized multidisciplinary clinics. Good clinical practice recommendations based on the consensus of experts are formulated with respect to the composition of the multidisciplinary team, the frequency of contact with the patient and caregiver and visits of the patient, and the communication and coordination between all service providers and care specialists involved.
From previous national and international research projects and daily clinical practice we know that patients with ALS and their informal caregivers report gaps in complex multidisciplinary and regular service provision. On the basis of data from patients, caregivers and health care professionals it is suggested that there is room for improvement in services with regard to coordination of care, symptom treatment, availability of aids and adaptations and caregiver support. Accordingly, to improve ALS care related to the provision of assistive devices and home adaptations (ADHA), it is necessary to investigate the patients’ experiences during the procurement process to gain insight in the bottlenecks of this care issue.

Complex ALS care, that fails to meet the care needs of patients and caregivers with respect to symptom burden, care coordination, mutual communication between patients, caregivers and their care professionals and caregiver strain, might be improved through intensive case management. As far as we could ascertain, there was no evidence for case management in patients with ALS. However, earlier non-ALS studies suggested case management as an innovative strategy to optimize complex care. Case management supporting tasks (e.g., family support, education, advocacy, psychosocial support and care coordination) in various complex long term conditions appeared to be beneficial on patient and caregiver outcomes like quality of life, quality of care and caregiver burden. Moreover, Mitsumoto et al. (2005) and Andersen et al. (2012) previously recommended additional research with respect to the increase of empirical evidence and improvement of the quality of multidisciplinary ALS care on symptom treatment, QOL and caregiver strain.

Informal caregivers usually hold a central position during the disease course of patients with ALS. After the diagnosis, most of the patients prefer to stay in their homes until the end of their life and caregivers make a valuable contribution to achieve this goal. However, informal caregivers of patients with ALS are vulnerable as caregiving exerts substantial strain that increases during disease progression. To optimize caregiver support, insight is needed into potentially modifiable factors associated with caregiver strain.

1.4 Aims of this thesis
The overall aim of this thesis is to optimize multidisciplinary ALS care through expanding our knowledge related to previously identified bottlenecks in complex ALS care – aids and adaptations, prognostication, quality of care, caregiver strain – and examining the care intervention case management in a mixed methods research project. Our mixed methods research project included a multicenter cluster randomized controlled trial (RCT) and a qualitative study with semi-structured interviews.
Chapter 1

The specific aims of this thesis

- To summarize the available evidence based on the scientific literature about prognostic factors for the course of the functional status of patients with ALS. (Chapter 2)

- To explore the experiences of patients with ALS during the application and provision process of assistive devices and home adaptations, and particularly to determine the problems they perceived during this procurement process. (Chapter 3)

- To study the effect of case management on patients’ quality of life (QOL), caregivers’ strain, and perceived quality of care (QOC) in patients with ALS and their caregivers. (Chapter 4)

- To explore the experiences of patients, their caregivers and health care professionals with case management additional to multidisciplinary ALS care. (Chapter 5)

- To identify factors for receptiveness to case management and what aspects of case management have additional value to the usual multidisciplinary ALS care. (Chapter 5)

- To identify patient and caregiver disease- and psychosocial-related factors associated with caregiver strain over time in ALS. (Chapter 6)

1.5 Outline of this thesis

Chapter 2 describes the results of a systematic review of evidence regarding prognostic factors for the rate of functional decline of patients with ALS, assessed with the ALS Functional Rating Scale. In chapter 3 we explored through a nationwide survey the experiences of patients with ALS during the application and provision process of assistive devices and home adaptations, and particularly determined the problems they perceived during this procurement process.

Chapter 4, 5 and 6 are based on results from the multicenter, cluster randomized controlled trial on the effectiveness of case management additional to usual multidisciplinary ALS care. During 12 months, patients with ALS and their most important informal caregiver received case management in addition to usual care or usual care alone. In chapter 4 we present the results of the care intervention case management on outcome measures patients’ quality of life, caregiver strain and patients’ and caregivers’ quality of care. The experiences of patients, their caregivers and health care professionals with case management during the RCT are explored through semi-structured interviews and described in chapter 5. Furthermore, aspects of case management that may have additional value to usual multidisciplinary ALS care and factors for receptiveness to case management are presented.
Data, collected during the RCT case management, are applied to analyse which (modifiable) patient and caregiver factors are associated with caregiver strain. We present the results of the (longitudinal) analyses in chapter 6.

In chapter 7 the main findings, strengths and limitations of this thesis are described. Finally, the implications of the study findings for the care in ALS are discussed and future perspectives are considered.
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