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**What are the palliative care needs
of people severely affected by
neurodegenerative conditions,
and how can a Specialist
Palliative Care Service best meet
these needs**

**A thesis submitted to the University of Kent
for the Degree of Doctor of Philosophy**

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Abstract

Background: There is increasing recognition of the need for service development for patients in the advanced stages of neurological conditions. This study explores the palliative care needs of people with advanced amyotrophic lateral sclerosis, multiple sclerosis, Parkinson's disease and related disorders, and assesses the impact of a new specialist palliative care service (SPCS)

Methods:. A mixed methods approach was adopted:

- a qualitative needs assessment using in-depth interviews with patients and their family carers and focus groups of professionals involved in the care of this population.
- a quantitative pilot-exploratory randomized and controlled trial (RCT) to assess the impact of a new SPCS designed to meet the palliative care needs of the study population – using a waiting list methodology comparing the immediate provision of SPCS with standard care .

Results:

- Qualitative study: 22 patients, 21 family carers and 11 professionals participated to the needs assessment. The content analysis showed a high prevalence of problems. Professionals confirmed the high burden of problems, were positive about the creation of a new SPCS and their knowledge of specific palliative care topics seemed lacking.
- Quantitative study: 50 patients, with 45 carers, participated in the exploratory RCT. The comparison between the groups (FT-ST) after 16 weeks revealed significant improvement for the SPCS group for quality of life and in four physical symptoms - pain, breathlessness, sleep disturbance and intestinal symptoms

Conclusions: This research confirms the high prevalence of physical symptoms, psychosocial issues and spiritual themes for people severely affected by advanced neurodegenerative disorders and that these can be helped by specialist palliative care. The input of a SPCS caused a significant improvement of the individual quality of life of the patients and improved symptom control for pain, breathlessness, quality of sleep and intestinal symptoms compared to standard best care alone.

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General summary

Background: Patients severely affected by progressive neurodegenerative conditions experience a high burden of physical symptoms as well as psychosocial and spiritual unmet needs. The family carers are exposed to an increasing burden of care as the patients' disability progresses. There is increasing recognition of the need for service development for patients in the advanced stages of neurological conditions, but little evidence of the efficacy of Specialist Palliative Care Services (SPCS) to meet the palliative care outcomes (PCO) in this population and how this can best be provided to address them.

This study explores the palliative care needs of people in advanced stages of amyotrophic lateral sclerosis, multiple sclerosis, Parkinson's disease and related disorders, assesses the impact of a new specialist palliative care service on the care of patients and, on the basis of findings, recommends options for a more tailored palliative care service provision for this population.

Methods: The overall methodology adopted followed a validated framework for the design and the evaluation of complex services to improve health. A mixed methods approach was adopted to address the research question:

1) a qualitative needs assessment using (*) in-depth interviews with patients and their family carers to explore the unmet needs and to capture the strategies adopted to overcome the problems and (*) focus groups to collect the experience of the professionals involved in the care of this population.

2) a quantitative pilot-exploratory randomized and controlled trial (RCT) to assess the impact of a new SPCS designed to meet the palliative care needs of the study population. The RCT adopted the waiting list methodology that compared the effects of the SPCS on group of patients immediately receiving the service (FT) with a control group receiving standard care (ST) that received the care after a period of 16 weeks.

Analysis: for the qualitative study a thematic content analysis aimed at finding the main groups of unaddressed needs was performed and the results were used to identify some PCO to be assessed in the following phase. For the quantitative study descriptive statistics were used to analyze the sample and groups comparison statistics plus a clinical judgement of significant impact were adopted to find differences between the groups for the trial outcomes.

Results: 1) Qualitative study: 22 patients, 21 family carers and 11 professionals participated to the needs assessment. There was a high prevalence of physical problems -movement impairment and oral-swallowing problems affected 100% of the interviewed patients, painful conditions and respiratory troubles 81%, intestinal symptoms 70%, urinary and sleep disorders 60%; the presence of psychological issues - anxiety, depression, abandonment and coping issues were reported by 70-100% of the participants; social isolation (100%); spiritual themes like difficulties to find a meaning in the experience of the disease and ambivalence related to faith, religiousness and spiritual support (50-88%). The existing services did not provide adequate home care support, were not equally distributed among the different diagnostic groups and did not help in end of life decisions. Professionals confirmed the high burden of problems faced by the neurological patients, were positive about the creation of a new SPCS and reported their difficulties to provide care outside the hospital when patients are too ill to be transported. Knowledge of specific palliative care topics, like symptomatic therapies for the advanced stages, seemed lacking.

2) Quantitative study: 50 patients, with 45 carers, participated to the explorative RCT. They were randomized 25 in the FT and 25 in the ST groups. At baseline no differences on demographics, diagnostic groups and trial outcomes were found between the groups. The comparison between the groups (FT-ST) after 16 weeks revealed that the FT patients had a statistical ($p < 0.01$) and clinical ($> 20\%$) significant improvement in individual Quality of Life and in four physical symptoms - pain, breathlessness, sleep disturbance and intestinal symptoms; clinically relevant advantage ($> 20\%$) for the social isolation of the patients; moderate clinical advantage (11-20%) for the urinary and the oral symptoms; one spiritual theme “finding a meaning in the experience of the disease” and the satisfaction about the received services for both patients and carers were found in the FT versus the ST. For all the other analyzed variables no differences appeared between the groups, though for some psychological issues the trend seemed favouring the patients who did not receive the SPCS. The caregivers’ burden of care was reduced very little in the FT versus the ST, but the variation is too small to be clinically or statistically significant (5%).

Conclusions: this original research provides an in-depth perception of the palliative care unmet needs of people severely affected by advanced neurodegenerative disorders. It confirms the high prevalence of physical symptoms, psychosocial issues and spiritual themes that can be helped by specialist palliative care. There has been a lack of adequate services available for this population in Turin city and its province. This is the first study to assess the impact of a SPCS on the palliative outcomes of patients severely affected by different neurodegenerative conditions and in their family carers. These are novel data from a population for which data exploring the impact of SPCS are still lacking and they show that the input of a SPCS caused a significant improvement of the individual QoL of the patients and improved symptom control for pain, breathlessness, quality of sleep and intestinal symptoms compared to standard best care alone. The findings of this research can be relevant for the developing clinical services for these patients. The adopted methodology was a feasible and reliable method to be followed for the design, development evaluation of new SPCS for patients with disease other than cancer.

1. Introduction

1.1 Presentation

The research was stimulated by the increased awareness of the need to look at the provision of palliative care for people with diseases other than cancer (NCHSPCS 2003, Addington-Hall, J. M. and Higginson 2001). FARO (Foundation for Assistance and Research in Oncology) is a charity of Turin, Italy, providing a multidisciplinary home palliative care service since 1989. In 2001 a 14 beds hospice opened, and both the home care service and the hospice facility were dedicated exclusively to people severely affected by cancer. FARO celebrated in 2009 its first 20 years of home care assistance during which the service took care for of more than 11.000 patients and their families. The multidisciplinary service consists of physicians expert in palliative medicine, trained nurses, psychologists, physiotherapists, a spiritual assistant and a group of volunteers. In 2006 the FARO administration board felt that other services, specifically dedicated to non cancer patients, should be developed. Following attendance at international seminar the role of palliative care in neurological disease was raised. This led to consideration of a new service for this patient group.

However there was little known of the needs and problems of this patient group in Italy and before a service was developed a fuller assessment was needed. Moreover it was felt that any service development should be fully evaluated before wider dissemination, to ensure it was acceptable to patients, families and staff, feasible, effective and cost effective.

These reasons led to a contact with my actual supervisor, Dr David Oliver, a world known expert in palliative care and neurological diseases, chair of the Task Force for Neurology of the European Association for Palliative Care (EAPC) and editor of the firsts published books on these subjects (Oliver, Borasio and Walsh 2006, Voltz et al. 2004).

A research project was set up with the double purpose of the development, assessment and evaluation of a new SPCS provided for FARO in Turin and a PhD research at the University of Kent. The present thesis collects both the service development process matched to the research framework and its methods and results.

1.2 The research question

The research question of this thesis is composed by two parts:

- What are the palliative care needs of people severely affected by neurodegenerative conditions?

and

- How can a Specialist Palliative Care Service (SPCS) best meet their unmet needs?

1.3 The research outcomes

1. answer to the research question
2. alongside the development of a new SPCS for patients severely affected by ALS/MND, MS, PD and related disorders in Turin, assess and evaluate the impact of this service on some important Palliative Care Outcomes.

1.4 General methodology

The general scheme adopted for the development and evaluation of the new SPCS for patients severely affected by ALS/MND, MS, PD and related disorders was the MRC framework for design and evaluation of complex interventions to improve health.

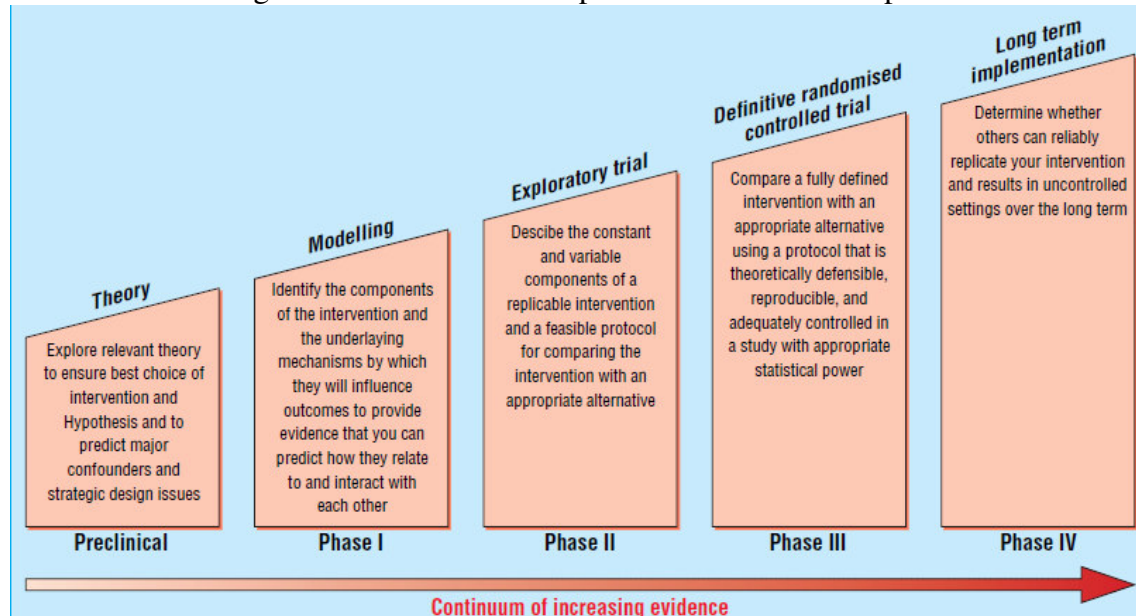


Figure 1: the MRC framework phases (Campbell *et al.* 2000)

A mixed methods methodology was chosen to answer the research question matching the studies with the different phases of the framework:

A literature review in the **preclinical** (fig.1) phase produced the evidence of the need for this research project by providing existing data on:

- prevalence and description of symptoms and psychosocial and spiritual issues for the different diagnostic groups
- how different health and social support systems impact on patients and their families quality of life.

The results of the literature review allowed to generate the choice of the methods of the research project (**theory**, fig.1) through:

1. The demonstration of how much the subjective reports from the patients and carers stories (qualitative data) can educate a forthcoming new care team about the fields in which their professional skills are to be strengthened. This induced the choice of a qualitative approach to explore the needs of patients and families with advanced ALS/MND, MS, PD and related disorders in order to better organize the following intervention on the real told experience of the potential participants (**modelling – phase 1**, fig.1). In this qualitative study, named Neu-Needs, in-depth interviews with patients and their family carers and focus groups with professionals involved in the participants' care, were used. The study aim was to:
 - explore the lived experience of the diseases,
 - the problems faced by patients and their families,
 - the coping strategies adopted,
 - the level of satisfaction about the various available services
 - and the congruence of the problems aroused in the interviews and focus groups with the previous published experiences.

2. A quantitative study (**explorative trial- phase 2**, fig.1) was chosen to evaluate the impact of a SPCS on some important Palliative Care Outcomes (PCO), most of which came out from the content analysis of the previous qualitative study.

This choice was induced by the clear feeling of the potential benefits that the skills of a SPCS could provide to this population, but, on the other hand, the absence of strong evidence of efficacy due to the lack of randomized controlled trials in this fields particularly in such an heterogeneous group of neurological patients.

The quantitative study, named Ne-Pal from the initials of neurology and palliative care, was an explorative randomized controlled study adopting the immediate referral to the

service (fast-track_FT) versus a 16-week wait (standard best practice_ST) methodology (Higginson, I. J. *et al.* 2008). Group comparison between FT and ST at baseline (T0) and after 16 weeks (T1) allowed to highlight which changes in the measured outcomes could presumably be due to the SPCS intervention, in other words to see if and where a SPCS made the difference on PCO versus a control group.

Main outcome measures were:

- Quality of life of patients
- Burden of care on family carers

Secondary outcomes were:

- Symptom load
- Psychosocial and spiritual issues
- Service satisfaction
- Feasibility of RCT in this population (Compliance, recruitment, completeness of follow-up, impact on survival)

2. Background: Literature Review

Palliative care services have been involved in the care of people with neurological disease since the early days of St Christopher's Hospice, which opened in 1967. In a letter in 1990 Dame Cicely Saunders, the founder of modern palliative care, wrote to Dr Mary Eleanor Toms that they had been caring for patients in terminal stages of Motor Neurone Disease since the opening of the hospice, and that their first patient was admitted in 1967 (Clark 2002, p. 322). Recently the interface between neurology, rehabilitation and palliative care has been explored and a framework indicating each specialty peculiar tasks and their overlapping fields of intervention has been published (Turner-Stokes, Sykes and Silber 2007, Turner-Stokes *et al.* 2007).

The present thesis explore the relation existing between palliative care and neurology by means of:

- a literature review
- a qualitative needs assessment
- a quantitative evaluation of a new SPCS for neurological patients

The first part of the thesis presents the results of the literature review aimed at exploring the existing evidences of specific themes of interest for this research project.

The literature review findings will be displayed in this order:

- Literature search methodology
- Definitions of palliative care and of specific concepts related to this topic
- Introduction to the involvement of palliative care for non cancer patients
- Palliative care in neurology.
- Introduction to the neurodegenerative disorders considered in this thesis
- The human needs, the palliative care needs and the palliative care unmet needs in neurology

2.1 Literature search methodology

The literature review was performed by searching the following electronic databases: Medline (Ovid, PubMed), Embase (Ovid), Cinhals, Psychinfo (Ovid), the Cochrane library.

The terms that were searched were:

1. General palliative care terms: Palliative care, Specialist Palliative Care, Terminal care, Terminally ill, Hospice, End of life, Death, Dying.
2. Disease related terms: Neurolog\$, Amyotrophic Lateral Sclerosis, Motor Neurone Disease, Parkinson\$, Multiple Sclerosis, Multiple System Atrophy, Progressive Supranuclear Palsy.
3. Outcome measures: Outcomes, Symptoms, Place of Care, Place of Death, Quality of Life, Caregivers, Carers, Needs Assessment, Service Evaluation.

Terms listed in the point 1 were combined using the Boolean term "OR", the same for the ones at point 2 and 3. The results of these 3 combination were then combined among each others using the term "AND".

The resulted papers where then scrutinized by reading the titles and then the abstracts. This procedure was performed in June 2006 and repeated in September 2007, October 2009 and January 2010.

Hand search of the reference list of the collected papers and of specific books was then implemented during the whole period of the research. Electronic sources added extra

hits as well as posters and other abstracts collected during participations to conferences on palliative care and neurology.

Overall 837 references were retrieved and catalogued into an electronic reference manager software (Endnote 9). Of these papers about 200 were then printed in full text, while for the remaining only the abstracts were analyzed.

2.2 Definitions of Palliative Care

The general definition of palliative care usually adopted is the following, from the World Health Organization:

WHO Definition of Palliative Care (W.H.O. 2002)

Palliative care is an approach that improves the quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual.

Palliative care:

- provides relief from pain and other distressing symptoms;
- affirms life and regards dying as a normal process;
- intends neither to hasten or postpone death;
- integrates the psychological and spiritual aspects of patient care;
- offers a support system to help patients live as actively as possible until death;
- offers a support system to help the family cope during the patients illness and in their own bereavement;
- uses a team approach to address the needs of patients and their families, including bereavement counselling, if indicated;
- will enhance quality of life, and may also positively influence the course of illness;
- is applicable early in the course of illness, in conjunction with other therapies that are intended to prolong life, such as chemotherapy or radiation therapy, and includes those investigations needed to better understand and manage distressing clinical complications.

Palliative care therefore is a complex concept with the aim of promoting both physical and psychosocial well-being. It is vital and integral part of all clinical practise, whatever the illness or the stage of the illness progression. The key principles underpinning palliative care which should be practised by all health professionals in primary care, hospital and other settings comprise:

- Focus on Quality of Life
- Whole person approach taking into account the person's past life experience and current situation
- Care which encompasses both the person with life-threatening disease and those individuals who matter to that person
- Respect for patient autonomy and choice (preferred place of care, access to specialist palliative care)
- Emphasis on open and sensitive communication, extending this to patients, informal carers and professional colleagues

Other definitions that can help to practically explore the palliative care field are the following:

1. *Palliative care is (NICE 2004)*

The active holistic care of patients with advanced progressive illness. Management of pain and other symptoms and provision of psychological, social and spiritual support is paramount. The goal of palliative care is achievement of the best quality of life for patients and their families. Many aspects of palliative care are also applicable earlier in the course of the illness in conjunction with other treatments.

2. *End of life care (DH-UK 2008)*

End of life (EoL) care has been defined as the care that:

Helps all those with advanced, progressive, incurable illness to live as well as possible until they die. It enables the supportive and palliative care needs of both patient and family to be identified and met throughout the last phase of life and into bereavement. It includes management of pain and other symptoms and provision of psychological, social, spiritual and practical support.

3. *Supportive care (NCPC 2006)*

Supportive care helps the patient and their family to cope with their condition and treatment of it – from pre-diagnosis, through the process of diagnosis and treatment, to cure, continuing illness or death and into bereavement. It helps the patient to maximise the benefits of treatment and to live as well as possible with the effects of the disease. It is given equal priority alongside diagnosis and treatment.

Supportive care should be fully integrated with diagnosis and treatment. It encompasses:

- Self help and support
- User involvement
- Information giving
- Psychological support
- Symptom control
- Social support
- Rehabilitation
- Complementary therapies
- Spiritual support
- End of life and bereavement care

The practice of palliative care will vary from area to area and there are profound differences across both Europe and the world in the way these principles have developed. Overall the developments have been:

Palliative medicine

Palliative Medicine is the medical speciality which concerns itself with the appropriate

medical care of patients with progressive disease (Mathew *et al.* 2003).

Palliative medicine has specialty status in just two European countries: Ireland and the UK. In five countries it is considered as a sub-specialty, for which a second certification is required: Poland, Romania, Slovakia and Germany and, recently, France. Some 10 other countries have started the process of certification for palliative medicine (Centeno *et al.* 2007) .

When palliative medicine became a specialty in the UK the Association of Palliative Medicine provided the definition (NCHSPCS 1995):

Palliative medicine is the appropriate medical care of patients with advanced and progressive disease for whom the focus of care is the quality of life and in whom the prognosis is limited (though sometimes may be several years). Palliative medicine includes consideration of the family's needs before and after the patient's death.

Palliative interventions are non-curative treatments given by specialists in disciplines other than specialist palliative care aimed at controlling symptoms and improving patient's QoL (e.g. palliative radiotherapy, chemotherapy, surgical procedures and anaesthetic techniques for pain relief).

Specialist Palliative Care Services: are those services with palliative care as their core speciality. These services are needed by a significant minority of people whose deaths are anticipated, and may be provided directly through specialist services, or indirectly through advice to a patient's present professional advisers or carers. These services provide physical, psychological, social and spiritual support and involve professionals with a broad range of skills including medical and nursing, social work, pastoral/spiritual care physiotherapy, occupational therapy, pharmacy and related specialties (Addington-Hall, J. M. and Higginson 2001 p.7, Mathew *et al.* 2003).

2.3 Specialist Palliative Care Services

The evidence for benefit from SPCS is sparse and limited by methodological shortcomings. In a recent review Zimmerman *et al.* found a small number (22) of randomized controlled studies on the SPCS impact on quality of life, satisfaction with care or cost effectiveness. Significant benefit from intervention was found in 4 of 13 studies assessing QoL and 1 of 14 assessing symptoms. A higher impact was shown in those studies whose primary outcome was family satisfaction with care. Also cost effectiveness is shown in 1 study out of 7. Authors found out limitations in randomization techniques, problems with recruitment, attrition and adherence (Zimmermann *et al.* 2008)

This is in contrast with Higginson's and Foley's opinion that conversely states that palliative care is a necessity, no longer a luxury, there's evidence of benefits and cost effectiveness, even though the argument that palliative care should be given because it

is cheaper is highly dangerous. Palliative care has to be provided in the most effective and efficient way (Higginson, I. and Foley 2009)

New evidence of palliative care efficacy in cancer patients have recently been published by Bakitas et al. Result of their RCT on SPCS versus standard care, measuring QoL, symptoms, mood and service utilization, show significant improvement from SPCS on QoL and depression, while symptoms and service utilization were not different in the 2 groups (Bakitas *et al.* 2009).

Improving awareness of the needs of patients dying from disorders other than cancer should lead to an increase in referrals to specialist palliative care services (Edmonds 2004)

Palliative care is no longer a luxury, but can now be defined as a necessity. Probably it is not as cheap as it was believed in the past, but certainly it has to be provided in the most efficient and effective ways (Higginson, I. and Foley 2009).

2.4 Palliative care for non cancer patients

2.4.1 Introduction

Every person with advanced, progressive and incurable illness should receive palliative care appropriate to their assessed needs (NCHSPCS 2003).

Meeting the palliative care needs of non-cancer patients presents a number of difficult and largely unresolved challenges including prognostic uncertainties, funding issues, boundary disputes between professionals and between services and a lack of relevant expertise.

In an era of increasing emphasis on evidence based health care, the lack of evidence in this area is striking (Addington-Hall, J. M. and Higginson 2001 p.8).

We know, from a study in 1963, that dying patients affected by non malignant conditions are less likely to have physical symptoms controlled than the ones with a diagnosis of cancer (Hinton 1963).

Cicely Saunders, founder of St. Christopher's Hospice and pioneer of modern palliative care, wrote that "terminal care should not only be part of oncology but geriatric medicine, neurology, general practice and throughout medicine" (Saunders and Baines 1983, in Addington-Hall, J. M. and Higginson 2001).

The rapid growth of specialist palliative care has led to improved care for the minority in direct receipt of these services and, as palliative care principles have permeated into the mainstream of cancer care, for the majority who do not receive these services. However, most people do not die from cancer. People who die from other diseases may also need improved symptom control, better nursing care, and more open communication about death and dying. Little is known about the needs of people who die from non-malignant diseases, the adequacy of existing services, or, indeed, the effectiveness of specialist palliative care for these patients. Better information is needed if palliative care services for non cancer patients are to develop in an appropriate and cost-effective way. In particular, the question of which non cancer patients might benefit from specialist palliative care needs to be addressed in order to target funding and services (Addington-Hall, J., Fakhoury and McCarthy 1998)

Patients who have palliative care needs can be grouped in several ways, by diagnosis, by symptoms or problems experienced or by type of service received. Each year on a

population of 1.000.000 in western countries, there are about 11.000 deaths 25% are caused by cancer and of these we know that around 65% will require specialist palliative care. A higher proportion (63%) die from non malignant conditions. Only some people with these conditions will require specialist palliative care, but it is still unknown how many (Higginson, I. 1997a). An Australian group studied needs of palliative care by estimating the size of potential palliative care population. They made three potential estimates to describe the characteristics of people eligible to receive palliative care in Western Australia. The Minimal Estimate included ten conditions that are clear indicators that palliative care is likely to be of benefit to patients. Deaths from these ten conditions constituted exactly 50% of all deaths. Slightly more than half of these were due to malignancies. Five other conditions (heart failure, renal failure, COPD, Alzheimer's disease and liver failure) represented sizeable proportions of this estimate. Much smaller proportions (less than 2%) of the deaths were due to the remaining four conditions (motor neurone disease, Parkinson's disease, HIV/AIDS and Huntington's disease). The authors concluded that palliative care should be available to patients dying of these conditions and these findings are supported both by the literature and among workers in the field (Rosenwax *et al.* 2005). An estimate based on symptom load of people who died of non malignant diseases suggested an increase of 79% in the expected caseload if a SPCS was fully available for non cancer patients. The authors stated that, based on these criteria, 16.8% of people dying for progressive non malignant conditions would require specialist palliative care (Addington-Hall, J., Fakhoury and McCarthy 1998).

A strong body of opinion argues that the skills and philosophies of palliative care should be extended to all care settings. The role of specialist in palliative medicine is to offer what has been learnt about palliation of malignant disease to those caring for patients with progressive, incurable, non-malignant conditions and to share and exchange best practise (O'Brien, T., Welsh and Dunn 1998).

Specialist Palliative Care Services (SPCS) are involved in the care of terminally ill cancer patients.

Strong evidence suggests that primary conventional care alone is inadequate for patients with advanced cancer. In a review is indicated that in hospital, hospice and community settings a multi-professional approach with specialist input is beneficial. SPCS improve satisfaction of both the patient with advanced cancer and their family or carers. Such teams are more able to identify and deal with patient and family needs, and to provide access to other services. There's also evidence of improved pain control and symptom management as result of the specialist approach. SPCS can effect cost by reducing the number of hospital inpatient days and time spent in out-of-home services (Hearn and Higginson 1998).

Non cancer patients seem to have lower chances to choose to die at home and to get access to home care, if it is true that cancer patients in higher socioeconomic groups were both more likely to die at home and to access home care (Grande, Addington-Hall and Todd 1998).

2.4.2 Palliative Care Needs

Prognosis is the medical answer to one of the most important need of people affected by life threatening conditions. Answer questions like "How long have I got?" or "What will it happen?" are among the most difficult tasks faced by health care professionals. Illness trajectories may be of help (Lunney *et al.* 2003, Lynn *et al.* 1996), but since diseases affect individuals in different ways, prognosis is often difficult to estimate. Because it seems that patients with specific diseases and their carers often have

common pattern of experiences, symptoms and needs as the illness progresses, the notion of diseases trajectories can help decision making and care planning that can be translated into improvements in outcomes for patients and their families (Murray *et al.* 2005). Even though prognostic models that attempt to estimate survival of less than 6 months have generally poor discrimination, reflecting the unpredictable nature of most non-malignant diseases, a number of generic and disease specific predictors are available for clinicians to identify older, non-cancer patients with palliative care needs. Simple, well validated prognostic models that provide clinicians with objective measures of palliative status are needed (Coventry *et al.* 2005).

Prognostic indicators for many progressive conditions have been published and are used in different countries to identify those patients requiring hospice care (The National Hospice 1996, Stuart 1999, Gold Standards Framework 2006, Grbich *et al.* 2005). The surprising question “Would I be surprised if this patient died within the next year?” can be a useful prompt for discussion and care planning with the patient. Collaboration between specialties to evolve effective symptom management and models of care for the increasing number of people living and dying with advanced progressive diseases and well-structured, coordinated and adequately resourced delivery of health and social care can be an effective response to most of this population needs (Kite, Suzanne and Hicks 2005).

Many symptoms experienced by cancer and non-cancer patients are similar: cancer patients’ symptoms may be more severe, but those of non-cancer patients tend to be more prolonged (O’Brien, T., Welsh and Dunn 1998, Addington-Hall, J., Altmann and McCarthy 1998). A review of the literature looking at the palliative care unmet needs of non-cancer patients (Luddington *et al.* 2001) found that symptom control was unsatisfactory in patients affected by chronic obstructive pulmonary disease (COPD) where it was defined as comparable if not more distressing than what reported by in lung cancer patients (Skilbeck, J. *et al.* 1998) and were likely to endure a long symptomatic course with two third of patients becoming severely disabled in the last year of life. Pain was as prevalent as in cancer in last year of life, but more frequent in last week of life and breathlessness was more frequent in non cancer than in cancer both in last year and last week of life (Addington-Hall, J., Altmann and McCarthy 1998). Breathlessness was defined as the most distressing symptoms at the end of life by both patients and their carers (Hockley, Dunlop and Davies 1988, Law 1997). Some professionals tend to see these patients’ need as less urgent due to the lengthy course of the diseases and nurses assume that patients with non cancer diagnosis are less likely to suffer from pain (Kurti and O’Dowd 1995).

Difficulties in coping with the deterioration of health status, decrease of independence, social isolation and family burden, limited resources available and difficulties in the access to community services, depression, acceptance of poor prognosis and concerns about the future are main findings in a research aimed at better understanding the unmet needs of people severely affected by chronic conditions (Fitzsimons *et al.* 2007). The authors put in evidence also the difficulties of talking openly with patients and families about the palliative nature of the treatments.

Psychosocial and emotional distress are described in several studies (Luddington *et al.* 2001, Hinton 1963, Hockley, Dunlop and Davies 1988). Striving for independence is a common need faced by patients affected by life limiting conditions (Cotterell 2008).

2.4.3 Specialist Palliative Care

The increased attention of the palliative care movement towards the care of non cancer patients is introducing the conviction that palliative care should be available to all on

the basis of need, not diagnosis (Addington-Hall, J. 1998, Addington-Hall, J. M. and Higginson 2001). Criteria for referral to specialist palliative care are usually advanced progressive disease, focus of care changed from curative to palliative, and limited prognosis. Appropriate reasons for referrals include symptom management, psychosocial issues of the patient or families and terminal care (Edmonds 2004).

Now the difficult is to determine which kind of service provision can better fit the needs of people severely affected by diagnosis other than cancer. Some authors suggest that specialist palliative care skills are perceived to be transferable to patients with non cancer diagnoses (Kite, S., Jones and Tookman 1999), and that clinicians and patient groups caring for patients with advanced non malignant disease have to work together with specialist palliative care services and with health commissioners to develop, fund and evaluate appropriate, cost-effective services which meet patient and family needs for symptom control and psychosocial support (Addington-Hall, J., Fakhoury and McCarthy 1998), but there is debate regarding how patient care is best delivered, because some authors fear that current models of specialist palliative care may not be the most appropriate for addressing the complex problems experienced by the many patients with a non-cancer diagnosis. They suggest that care should be structured around patient problems, viewing specialist palliative care as a service for those with complex end of life symptoms or problems (Field and Addington-Hall 1999, Skilbeck, J. K. and Payne 2005). If comprehensive palliative care is to be extended to non malignant diseases, existing models of care provision will need modification (Skilbeck, J. K. and Payne 2005). Models of care are to be adapted to the fluctuating nature of symptoms and a combined approach, with sharing of skills between disciplines, is required. There is also a concern of not eroding service provision for cancer patients (Traue and Ross 2005).

There is strong evidence that adding a multi-professional support team can provide a higher quality of care than conventional care alone. Specialist palliative home care services demonstrated their ability to keep patients at home for longer than when such services did not exist. They also resulted in lower costs for the health system and better symptom control for the patients. These studies included cancer and non-cancer patients. However there is little evaluation of new services for patients with progressive non-malignant conditions (Higginson, I. 1997a, p214-215, Higginson, I 1993). Further criticism has been raised when the impact of different models of specialist palliative care on patients quality of life was evaluated. Main concerns were about the lack of high scientific evidence of impact due to a lack of prospective RCT's, use of inappropriate outcome measurement tools and effectiveness of measures of quality of life to reflect the values of palliative care (Salisbury *et al.* 1999, Zimmermann *et al.* 2008). Research that evaluates the effect of general and specialist palliative care on psychosocial outcomes in non-cancer patients and their carers is needed (Coventry *et al.* 2005), as there is a need for randomized studies to evaluate different types of service configuration and delivery related to specific outcomes (Higginson, I. J. *et al.* 2003). It is imperative to listen to the experiences of patients and carers as a basis for developing interventions and guidelines for services (Seymour *et al.* 2003).

Professional specialists working in disciplines other than palliative care tend to be satisfied about the involvement of specialist palliative care in non cancer: an example is represented by the consultant physicians in south-east Wales that expressed a general enthusiasm for SPCS to be made available to patients other than cancer. The most appropriate form of service according to their point of view, is a system shared care and responsibility, and this was seen as a means of addressing concerns regarding the lack of disease specific expertise within the palliative care team (Dharmasena and Forbes 2001). However there is a need, for those palliative care professionals traditionally

evolved from oncology, to receive training in order to respond to the challenges of the different needs expressed by these patients and some concern about the limited understanding of non-malignant disease process, prognostic factors, appropriate medications (Fisher 2006, Wallwork 2000).

2.4.4 Facts and figures

The Gold Standards Framework agency, in its prognostic indicators paper (Gold Standards Framework 2006), indicates how just 25% of deaths in a developed country are due to cancer, a higher proportion is caused by organ failure (33%) and frailty or dementia (33%), and sudden death (9%). It has been suggested that there are different illness trajectories experienced by people at the end of life depending on their diagnosis, and these may be shown diagrammatically:

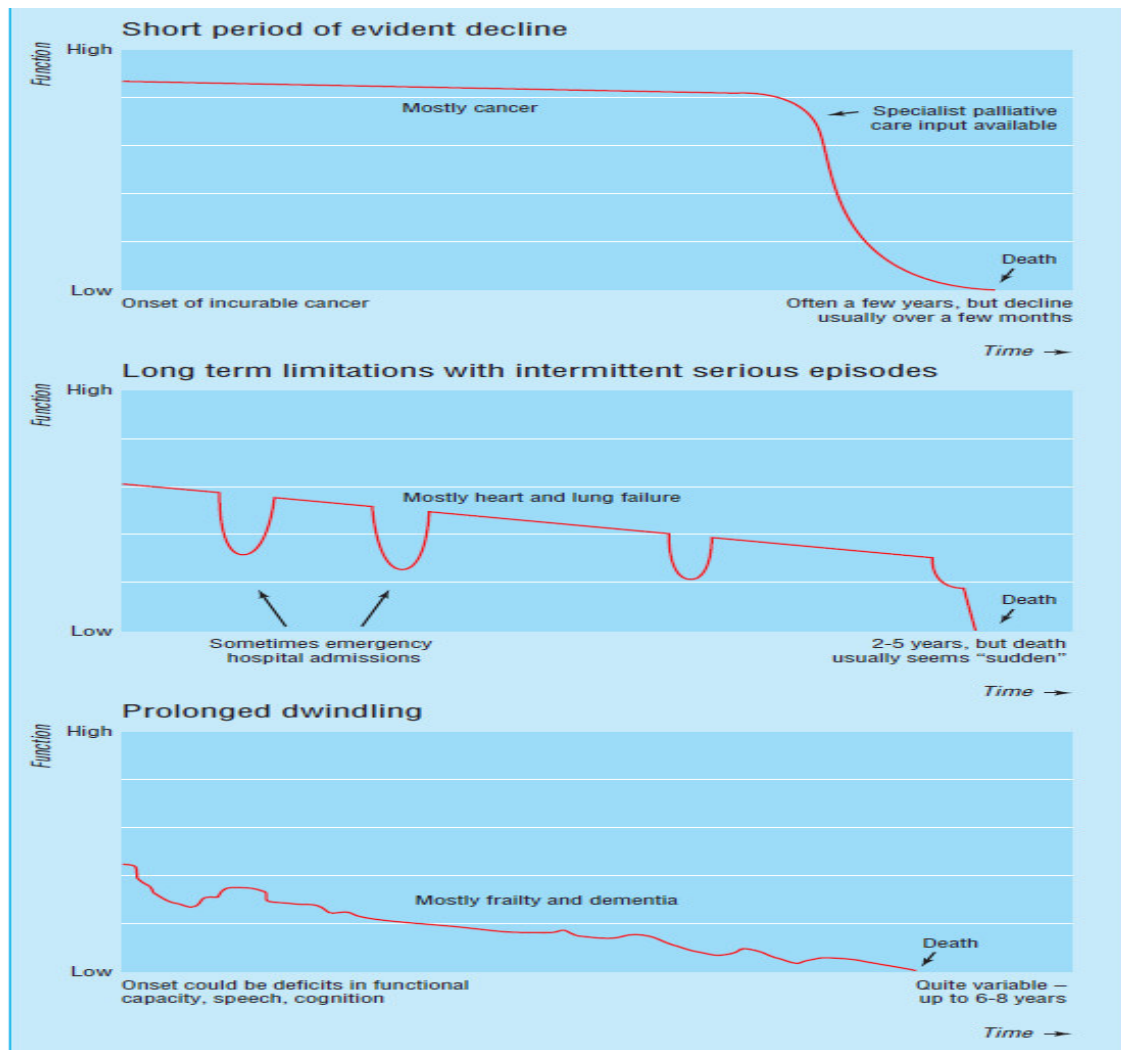


Figure 2: Illnesses trajectories at the end of life (Lunney *et al.* 2003)

From the analysis of the number of potential non cancer patients requiring palliative care and of their end of life trajectories it should be possible to estimate the increase work load of SPCS. It had previously been estimated that an increase of at least 79% in caseload would be expected if specialist palliative care services were made fully available to noncancer patients (Addington-Hall, J., Fakhoury and McCarthy 1998).

It is now a key principle in guidance on the commissioning of palliative care services for adults in United Kingdom that "It is the right of every person with a life threatening illness to receive appropriate palliative care wherever they are". (Addington-Hall, J. M. and Higginson 2001, p.7). In the same chapter the authors stated that in UK 96,7% of in-patient hospice and 96,3% of community palliative care services were used by cancer patients, despite the fact that 75% of death are caused by non-malignant conditions. Provision of palliative care services for patients with diagnosis other than cancer is both inequitable and inadequate and there is ample evidence that terminally ill patients with non-malignant diseases require palliative care (Addington-Hall, J., Fakhoury and McCarthy 1998, Wallwork 2000). Uptake of SPCS is significantly lower in people with non cancer diagnosis and in families with lower incomes. This is seen as an unmet need by their caregivers (Currow, David C. *et al.* 2008, Currow, D. C., Abernethy and Fazekas 2004). In a retrospective review of a service appears how in spite of 29% of referrals to the hospital palliative care team were for non-cancer patients, only 9% of the home care and 4% of hospice inpatient admissions were for patients with diagnosis other than cancer (Kite, S., Jones and Tookman 1999). A structured documentary review of the Health Improvement Plans 1999-2003 (HImPs) reveals that among the health authorities that have made strategic plans for palliative care, the vast majority are for people affected by cancer. Especially for non-cancer patients specialist palliative care is perceived as an "optional extra" rather than an integral and essential part of the overall supportive care strategy (Seymour, Clark and Marples 2002). More recently, in the report for the years 2006-2007 the National Council for Palliative Care published the National Survey of Patient Activity Data for Specialist Palliative Care Services. Overall 7.2% patients cared for a SPCS in the UK had a diagnosis other than cancer. The range of percentages of non-cancer patients in the different units ranged from 0% to 38%. A total of 28 units (20% of responding units) had over 10% non-cancer patients while 9 units only saw cancer patients (National Council for Palliative Care 2007).

In U.S.A. the situation is different: the National Hospice Organisation fact sheet for spring 1999 states that in 1995, 60% of hospice patients had cancer and 40% other diagnosis (Addington-Hall, J. M. and Higginson 2001, p. 8). Later figures reveal further change in this proportion as the National Hospice and Palliative Care Organization published in 2008 the "Hospice Care in America" which reported figures of palliative care assistance about the year 2007: of 2,400,000 deaths in USA in that year, 930,000 (38.8%) were under the care of a hospice programme. The median length of stay in a hospice was 20 days and the average length of service was 67.4 days. The primary diagnosis of these patients was cancer for 41.3%, and 58.7% had a non cancer diagnosis: heart diseases (11.8%), unspecified debility (11.2%), dementia (10.1%), lung disease (7.8%) and ALS\MND (2.3%) were the most frequent (NHPCO 2008).

2.4.5 The Italian situation

Palliative care is now well integrated in the health and social systems in most developed countries. In Italy a rapid growth of new hospices has happened in last ten years, but still most of them provide care for cancer patients only. Palliative care for non cancer patients has been advocated by many official statements and it is now part of the new Italian law on palliative care released by the Italian parliament in March 2010. Regional health systems already incorporated these principles for a long time. The Piedmont

Chapter 2. Background: Literature Review

Regional Health Commission produced guidelines on palliative care provision in 2002 stating that hospice care is indicated when patients are affected by progressive disease, in advanced stages, with a poor prognosis, for which no therapies aimed at the cure or stabilization are possible or appropriate. Neurodegenerative conditions are clearly included within these criteria, but often patients severely affected by these illness still do not have access to specialist palliative care in Italy.

Patients severely affected by these disorders are usually cared for by the neurologists who diagnosed their diseases, by rehabilitation specialist teams that help with disability problems and by primary care, involving general practitioners and district nurses. When the disease is in advanced stage patients are often homebound and lose contact with the hospital specialist teams. If complications occur, and the primary medicine team can not help, patients are taken to the local hospital passing through the A&E department and are seen by doctors and nurses who may not have had any previous contact with them. If they survive the hospital admission they are then discharged at home where they stay until next complication. Specific care facilities do not exist and many patients whose family member cannot cope with their disability and needs, are admitted to care homes where often die.

The role and use of advance directives have not been clarified by Italian politicians as yet. Patients are thus not allowed to decide for their future therapeutic options if they lose their mental capacity or consciousness. If a patient with ALS/MND arrives at the A&E unconscious from an episode of acute respiratory insufficiency, it is the resuscitation specialist who may decide on the use of invasive ventilation, and maybe tracheostomy. If the patient previously stated that he did not want that option, and families testify that, it is the doctor who will decide and he has to take the decision on the patient's best interest. This often results in an unwanted tracheostomy.

It is uncertain if there is a role for a specialist palliative care service (SPCS) in the care of these people and of their families and the literature seems to indicate that the services have a role as, there is a high prevalence of uncontrolled symptoms, unresolved psychosocial and spiritual issues. The SPCS may provide comprehensive care of both patients and families, care in the preferred place of care and help to stay in the preferred place to die, which usually is home, in Italy. A home based palliative care team can achieve better level of care than primary care alone, reducing the number of unwanted hospital admissions, and, consequently, the risk of undesired invasive therapeutic interventions.

SPCS are often involved in the care of people with cancer, facing death and dying and are able to provide better information for patients and families and better chance to plan the care. If a hospice inpatient facility is available respite can be offered to support the home care assistance and this respite is very much appreciated by lay carers and often by patients because they are concerned of the workload that they cause to their familiars. However admission is often feared by those carers who are used to provide a personal high quality assistance to their loved ones, as they have concerns as to whether the same care can be provided in the different setting and are often sceptical about this possibility. Families involved in the care of people severely affected by neurodegenerative disorders may require support, perhaps greater than in cancer, mostly due to the longer end of life trajectories and the progressive continual losses and adaptations that they have to face but this care has not been available in Italy.

Specialist palliative care teams often have an ambiguous approach to the care of patients with a diagnosis other than cancer. Their competence can be helpful in symptom control, psychosocial and spiritual care and end of life care, but they may express concern about difficulties in foreseeing prognosis and lack of competence about the unknown challenges related to the management of this group of patients. Some hospices opened their services to ALS/MND patients, but later expressed concern about the care they had been asked to provide as they felt unprepared when had to manage respiratory machines or when assistance last much longer than their usual cancer patients.

Italian neurologists, from their part, recognised the need to incorporate palliative care in their professional knowledge. Recognising the importance of previously international statements published for the American Academy of Neurologists about the duty of neurologists to provide palliative and end of life care to their patients facing threatening life conditions (AAN 1996, AAN 1998) they started new education courses in their annual meetings or through residential modules for neurologists involved in the care of people affected by long term conditions. Within the Italian Society of Neurologists (SIN) a group of study for bioethics started 20 years ago and in 2000 changed its name in Group of Study for Bioethics and Palliative Care in Neurology (Defanti 2000). The declared aims of this group are to enhance the bioethics reflection on the issues arising in the care for people affected by neurological disorders, promote good research in this field and improve the education of neurologists in palliative care. A document about the equity of provision of palliative care for neurological patients, analysing the bioethical principle of distributive justice, will be published in next months. This approach shows that neurologists are trying to establish connections with palliative care and this collaboration should be encouraged and strengthened..

2.5 Palliative Care in Neurology

2.5.1 Introduction

Since St Christopher's Hospice was opened in 1967 and the new era of palliative care started the involvement in the care of people with neurological disease has increased. Dame Cicely Saunders, founder of St. Christopher's Hospice and pioneer of modern palliative care, wrote in 1981:

“Our Lady's Hospice in Dublin (1879) and St Joseph's Hospice in London (1905) both included patients with long-term illness in their wards and St Christopher's Hospice (1967) followed this tradition and opened with at least 10% of its ward beds for patients with advanced neurological illnesses. The need of patients with motor neuron disease continues and their care has been integrate into the life of the Hospice” (Saunders, Walsh and Smith 1981).

The links between palliative care and neurology were established early in the development of the hospice, but later as hospices were established the main area of care was for patients with cancer.

An appreciation of the role of palliative care in neurological conditions was expressed in 1996 in a document published by the American Academy of Neurologists which suggested that neurologists have to understand, learn and apply the principles of palliative care because many patients affected by neurological disorders die after long illness and the neurologist is often the principal or the only consulting physician (AAN 1996). In the 1999 the AAN published the results of a survey that highlighted a gap between established legal, medical and ethical guidelines for the care of severely ill neurological patients and the practice and beliefs of many American neurologists (Carver *et al.* 1999). A position statement defined the official position of the American neurologists about the assisted suicide and euthanasia, in which the AAN strongly refused these practices, but evidenced the importance of palliative care as an option aimed at relieve sufferance of patients dying of neurological conditions (AAN 1998). In 1999 the AAN published the first evidence-based review on ALS/MND where guidelines and recommendations were provided for the different stages of the disease, with a focus on palliative and terminal care (Miller *et al.* 1999). These practise parameters have been recently reviewed and authors conclude that no controlled studies examined the treatment of pain, dyspnoea, hospice care and recommended design of controlled trials of terminal symptom management, advanced directives, hospice and spiritual care (Miller *et al.* 2009). A Cochrane systematic review studying multidisciplinary care for ALS/MND patients did not find any high quality study on this topic and concluded that further research is needed into appropriate study designs; outcome measurement; caregiver needs; and the evaluation of optimal settings, type, intensity or frequency and cost-effectiveness of care in the ALS/MND population. The authors suggested that future research should focus on observational designs to assess care and outcomes in 'real-life' settings (Ng, Khan and Mathers 2009).

The interface between neurology, rehabilitation and palliative care should be explored to provide long-term support for ALS/MND and for other progressive neurological conditions (Turner-Stokes, Sykes and Silber 2007). These findings must be read knowing that ALS/MND is the neurological disease in which the highest palliative care involvement has been obtained and were most publications have been made (Oliver, Borasio and Walsh 2006), but this disease is by no means the only neurological condition requiring palliative care (Borasio and Voltz 2005).

2.5.2 Neurological conditions for which palliative care can have a role

Neurodegenerative conditions are progressive and incurable conditions that ultimately lead to a state of total functional incapacitation and death. These conditions are "terminal" and, therefore, should be managed with a palliative care approach (Low *et al.* 2003).

Neurologists recognise their need of education and knowledge about end of life care as one of the major barrier to providing care for patients and families at the end of life (Foley and Carver 2001). One concrete initiative to overcome this difficulty was the publication of a specialist book titled "Palliative care in Neurology" (Voltz *et al.* 2004), specifically aimed at improving the knowledge of neurologists wishing to providing excellent care for neurological patients through the final stage of their disease. This book is also a precious source of information for those palliative care teams who are interested in being involved in the care of neurological patients not knowing many

aspects of the natural history, end of life trajectories, symptoms and other psychosocial, spiritual and ethical issues of the neurological disorders. From the table of contents of this publication it is possible to see how many different neurological conditions exist for which death is attended as an expected outcome and for which a palliative intervention can be adopted:

- Stroke
- Demyelinating disease (such as multiple sclerosis)
- Neurological neoplasms
- Movement disorders (such as Parkinson's disease and related disorders)
- Dementias
- Infectious diseases
- Diseases of motor nerves (typically ALS/MND)
- Muscular dystrophies
- Epilepsy
- Pediatric neurological disorders.

In addition specific neurological conditions, usually resulting from complications of neurological diseases, can be of interest for palliative care: Persistent Vegetative State, Quadriplegia and Paraplegia and the Locked-in Syndrome. Common symptoms in these disorders are described and treatment options provided. Ethical issues, like the difficult decisions at the end of life, are explored and the last chapters focus on the other general aspects of the palliative care (psycho-social, spiritual and cultural) and on the need for palliative care education for neurologists.

The disorders listed above can therefore be considered as the ones towards which SPCS should orient their interest when designing new services or when extending pre-existing ones to people affected by neurological conditions.

Some disorders, like the stroke or dementias, have high prevalence rate and represent some of the most frequent cause of death in western countries. If all the people dying for these causes had to be cared for by a SPCS this would have a dramatic impact on the workload. Furthermore most of these patients die in hospital (stroke) (Hamann, Rogers and Addington-Hall 2004) or in nursing homes (dementia) (Volicer 2004) settings where palliative care is not usually provided by SPCS.

As previously reported ALS/MND is the neurological disorders for which most evidence of the involvement of palliative care and specifically of SPCS is reported. Other disorders like multiple sclerosis have received little attention from palliative care, generally patients were considered as chronically ill, with progressive disability, high symptom burden, but not people affected by a life limiting condition for whom palliative care could play a role. A change in this position has started since evidences that MS patients have an increased suicide rate and that up to 5% of them die from euthanasia or physician assisted suicide in those countries where these procedure are legal (Van der Wal and Onwuteaka-Philipsen 1996) and when epidemiological studies evidenced the reduction in the life span of these patients (Bronnum-Hansen *et al.* 2005, Bronnum-Hansen *et al.* 2006). To date MS is the only neurodegenerative condition for whom a specific SPCS was designed, assessed and its outcomes evaluated with a phase 2 Randomized and Controlled Trial (Higginson, I. J. *et al.* 2008).

Movement disorders represent another group of neurological conditions that respect the criteria of incurability, relentless progression and shortened life expectancy. In this case the number of affected patients is very high, 25 fold more than ALS/MND, and some authors described the different phases of the illness trajectories including a palliative care stage where the specific drugs lose their efficacy and the focus of care is just on comfort and symptom control (Bunting-Perry 2006).

Overall, until now, many studies explored the unmet needs of patients experiencing the advanced stages of neurological conditions (Chahine, Malik and Davis 2008, Kristjanson, Aoun and Oldham 2006), as well as the satisfaction or dissatisfaction of the received services (Kristjanson, Aoun and Yates 2006). Literature reviews have described and compared the physical unmet needs of these patients (Saleem, Leigh and Higginson 2007, Higginson, I. J. *et al.* 2006a, Lee *et al.* 2007). There is a common feeling that the palliative care skills of the SPCS could have a positive impact on these unmet needs. What is lacking yet is the evidence that SPCS can make the difference on these outcomes in this population. A Canadian group reviewed the literature exploring the issue of palliative care in neurology and discussed the existing evidence base. The author conclude that current literature reflects primarily expert opinion and describes a growing interest in the early introduction of palliative principles into neurological care. Early initiation of palliative interventions has the potential to improve quality of life, enhance symptom management and assist in advance care planning. Further data is required to determine whether this shift in philosophy has a positive impact on patient care (Gofton, Jog and Schulz 2009). Another systematic literature review evidenced the need to explore more fully the challenges put forward by diseases other than cancer, such as neurological illnesses and heart disease suggesting that the impact of SPCS has not yet been studied extensively above all in these diagnosis (Zimmermann *et al.* 2008). Recently a care pathway for meeting the palliative care needs of people living with neurological conditions has been published. The pathway was designed to support timely referrals and coordination of care through a patient's illness journey incorporating the provision of palliative and end-of-life care (Brown and Sutton 2009). This novel option offers the opportunity to test the referral requirements and should facilitate the interaction among neurology, palliative care and rehabilitation.

In Italy the Study Group of Bioethics and Palliative Care in Neurology of the Italian Neurological Society has published papers focused on the palliative care themes related to neurology and the problems faced by the people dying for neurological diseases. One describes how the neurological patients die, highlighting the mechanisms of death related to most neurodegenerative conditions (Defanti 2005), the clinical and ethical appropriateness of sedation in palliative neurological treatments (Bonito *et al.* 2005), the role of advance directives in neurological patients (Bonito 2005), the discontinuation of life support measures in patients in a permanent vegetative state (Bonito *et al.* 2002) and other ethical issues regarding neurology and palliative care (Defanti *et al.* 2007, Gasparini *et al.* 2006, Gasparini *et al.* 2004). However there are no published experiences of SPCS discussing the involvement in neurological care in the literature. It would seem that neurologists recognised the need to start interaction and relationships with palliative care whereas the palliative care experts did not seem so interested in being involved in the care of people dying for neurological diseases.

From this evidence we can conclude that:

- further studies aimed at better interpret the palliative care needs of patients (and their families) severely affected by neurodegenerative conditions are required
- many people affected by these disorders die without receiving palliative care
- SPCS are to be involved in the terminal stages of the diseases, although their role and the impact on the palliative care needs of this population is still unclear

For all these reasons some of the neurodegenerative conditions that literature and previous experience associated to palliative care will be introduced and common features as well as peculiar characteristics will be described.

2.5.3 Neurodegenerative disorders considered in this research

After having introduced the potential role of palliative care in neurology and having discussed the previously published experiences a brief description of the neurodegenerative conditions that are considered in this research project is now presented.

The three groups of neurological conditions involved are :

- Amyotrophic Lateral Sclerosis, also called Motor Neurone Disease
- Multiple Sclerosis
- Movement Disorders represented by the Parkinson's Disease and by the so called Atypical Parkinsonian Syndromes or Parkinson's Plus Syndromes (Multiple System Atrophy and Progressive Supranuclear Palsy)

These neurological disorders, though being very different for many physiopathological aspects, share some common features:

- All are considered neurodegenerative progressive disorders (Voltz *et al.* 2004)
- None of these conditions is considered to be curable (Elman *et al.* 2007)
- Some symptoms are common to these conditions and can be approached in similar ways (Saleem, Leigh and Higginson 2007)
- All are considered life limiting conditions, even though presenting very different trajectories (Millul *et al.* 2005, Bronnum-Hansen *et al.* 2006, Muller *et al.* 2001)
- National and international directives have suggested a role for palliative care in the management of these diseases (DH-UK 2005, AAN-EHS 1996)

Taken together these conditions are not rare reaching an overall prevalence of 374/100.000 (Doyle *et al.* 2004).

2.5.4 Amyotrophic Lateral Sclerosis or Motor Neurone Disease (ALS/MND)

Motor Neurone Disease (MND) is a term covering a wide range of diseases involving the motor neurone cells. Amyotrophic Lateral Sclerosis (ALS) is one form of these disorders, but usually they are used as synonymous, even though in USA it is generally called ALS (and also Lou Gherig's disease, from the name of a famous baseball player who died of it) and in the United Kingdom and other Commonwealth countries it is known as MND (Shaw, C. 2006). For the purpose of this thesis the term ALS/MND will be adopted to describe this condition.

2.5.4.1 Epidemiology of ALS/MND

ALS/MND is a relatively rare neurodegenerative condition of unknown aetiology (Rowland and Shneider 2001), with an incidence in Europe, Middle East and North America of about 1.4-2,5 new cases each 100.000 inhabitants per year and prevalence rates 2.7- 4.3 points higher (Brooks 1996). Age and gender specific incidence rates, published by the European registers, show that the incidence of ALS increases after the age of 40 years, reaching a peak in the late sixties or early seventies, followed by a rapid decline (Logroscino *et al.* 2008). In Italy the Ministerial Committee for diagnosis, care and assistance of patients with ALS report an incidence of 2-2.5/100.000 inhabitants/year, a prevalence of 6-8/100.000, with a slight male dominance 1.2 – 1.3:1, a mean age onset of 63 years and a mean duration of the disease of 2.5 years (Bendotti *et al.* 2005). 90% of ALS are sporadic, but a 10% are familiar form with autosomic

dominant inherited disease, being passed down through the generations, often linked to a gene mutation on the chromosome 21, the copper/zinc superoxide dismutase (SOD1) (Siddique *et al.* 1991, Rosen 1993).

Diagnosis is generally based on the El Escorial Criteria –EEC– (Brooks 1994, Chaudhuri *et al.* 1995) even though rigid application of the EEC excludes at least 40% of ALS patients from enrolment in clinical trials at presentation, and up to 10% of patients at the time of death (Traynor *et al.* 2000, Logroscino *et al.* 2008).

2.5.4.2 Natural History of ALS/MND

ALSMND is characterized by onset of symptoms and signs of degeneration of primarily upper and lower motor neurons, leading to progressive weakness of bulbar, limb, thoracic and abdominal muscles exiting into rapidly progressive paralysis and respiratory failure. (Rowland and Shneider 2001, Andersen, P. M. *et al.* 2005). ALS/MND cause significant disability early and relentlessly progresses so that most patients lose the ability to walk, feed and toilet themselves, speak and swallow (Shaw, C. 2006). Cognitive functions tend to be spared by the disease, even though signs of fronto-temporal dementia (FTD) have been reported up to 15% of patients (Neary and Snowden 1996, Neary, Snowden and Mann 2000), although higher rates are reported, up to 65% of cognitive dysfunction (Barson *et al.* 2000, Strong *et al.* 2003, Lomen-Hoerth *et al.* 2003). Typical patterns of impairment relate to alterations in mental flexibility, verbal and nonverbal fluency, abstract reasoning and memory, for both verbal and visual material (Strong *et al.* 2003, Abrahams *et al.* 2004) with selective cognitive impairment in the form of verbal fluency deficit accompanied by emotional lability and depressive symptomology (Abrahams, Leigh and Goldstein 2005).

The clinical features can be considered in relation to neurological regions or levels: bulbar, cervical, and lumbar. Bulbar-onset patients present with slurring of speech (dysarthria), difficulty swallowing (dysphagia), or both. Bulbar involvement can be lower motor neuron (bulbar palsy), upper motor neuron (pseudobulbar palsy), or both. Bulbar palsy is associated with upper and lower facial weakness and poverty of palatal movement with wasting, weakness, and fasciculation of the tongue.

- Pseudobulbar palsy is characterised by emotional lability (also known as pathological laughing or crying), brisk jaw jerk, and dysarthria.
- Cervical-onset amyotrophic lateral sclerosis presents with upper-limb symptoms, either bilateral or unilateral. Proximal weakness can present as difficulty with tasks associated with shoulder abduction and distal weakness can manifest with impairment of activities requiring pincer grip. Upper limb signs might also be upper motor neuron, lower motor neuron, or both. The arm can be strikingly wasted with profuse fasciculation and brisk reflexes.
- Lumbar onset patients present with weakness of the legs, often with a mixed picture of wasting and spasticity
- Progressive muscular atrophy may present with weakness of the arms or legs, and the features are of lower motor neurone lesion only, at least initially. The prognosis is longer – often 5 to 10 years

Bulbar onset patients tend to be older, are more likely to be female, have a shorter period of time between symptoms onset and diagnosis and a shorter life span if compared with limb onset patients (Iwasaki *et al.* 2002).

Older ALS patients survive for shorter times. A tendency for older women to suffer bulbar-onset of ALS is well known (Iwasaki *et al.* 2002, Chaudri, Kinnear and Jefferson 2003).

Negative prognostic indicators included site of disease onset, older age and progression rate of respiratory, bulbar and lower limb symptoms (Hoffman 2008). Eventually, ALS/MND results in total dependence for meeting all basic needs, and often the focus is palliative care (Mitsumoto and Rabkin 2007). The disease is usually fatal in 2-4 years (Ringel *et al.* 1993, Borasio and Voltz 1997) although there are variation in life span amongst individuals (Shaw, P. J. 1999). The median time from onset until death has been found to be 23 to 48 months in various series with 5-years survival rates ranging from 9% to 40%, whereas 10-year survival rates are 8% to 16% (Simmons 2005) and patients usually die of respiratory failure (Neudert *et al.* 2001b, Borasio and Voltz 1997).

No known definitive treatment or cure exists for ALS. The only medication which may extend lifespan is riluzole (Rilutek), approved by the Federal Drug Administration (FDA) in 1995 and has been to slow down the deterioration associated with ALS. Riluzole acts to inhibit glutamine release, inactivating sodium channels or interfering with neurotransmitter actions, thus slowing neuronal damage and retarding disease progression (Hoffman 2008).

A population based outcome study in Ireland concluded that Riluzole reduced the mortality rate by 23% and 15% at 6 and 12 months, respectively, and prolonged median survival by 4.2 months (Traynor, Alexander and Corr 2003, Simmons 2005).

2.5.4.3 The Palliative Care Approach

ALS/MND is the first neurological condition to which a specific book exploring the palliative approach has been dedicated. The main thesis of this publication is that being this neurological condition progressive, incurable, and leading to death in some months-few years, palliative care should be involved in all the stages of the disease. The authors suggest that a multidisciplinary approach of care should start from the beginnings, with a proper disclosure of the diagnosis and continue through all the stages of the disease until the death of the patient and the bereavement counselling to the family. The aim of palliative care in ALS/MND is, for the authors of this book, to maximize quality of life of patients and families by relieving symptoms, providing emotional, psychological and spiritual support as needed, removing obstacles to a peaceful death, and supporting the family in bereavement (Oliver, Borasio and Walsh 2006). The palliative care involvement, therefore, should neither be restricted to pure symptomatic treatment nor to the terminal stages of the disease (Borasio, Voltz and Miller 2001).

In 1999 the American Academy of Neurology published an evidence based review about the care of the patient with ALS/MND. Clinical questions addressed in treatment recommendations were breaking the news, symptom management, nutrition management, respiratory management and palliative care. In the research recommendations section for palliative care they list (Miller *et al.* 1999):

1. Study symptom prevalence and quality of life in the terminal phase of ALS.
2. Test the effectiveness of current treatments for palliative care.
3. Develop new therapeutic approaches for terminal ALS symptoms.
4. Study the effect of hospice care on quality of life for patients with ALS and families.
5. Compare hospice home care with hospice inpatient care.
6. Study the role and impact of advance directives on the dying process.
7. Include palliative care of the dying patient in medical education.
8. Examine the psychological impact of ALS on caregivers and health care providers.

Acknowledging the importance of these recommendations 6 years later another working group underlined the need for guidelines specifically for end-of-life care in patients with ALS (Mitsumoto *et al.* 2005). Authors stated that little evidence was available in the published literature that identifies optimal management approaches for caring for the dying patient with ALS. In this review other issues are added to the previous indicated in the AAN Practical Parameters study: attention to caregivers, psychosocial and spiritual aspect of care, access to and cost of the care and quality of life.

Palliative care options are included in another review by the European Task Force on diagnosis and management of ALS/MND (Andersen, P. M. *et al.* 2005). These authors recommend that a palliative care approach should be incorporated into the care plan for patients and carers from the time of diagnosis. Early referral to a specialist palliative care team is often appropriate. Palliative care based in the community or through hospice contacts (e.g. home care teams) can proceed in partnership with clinic-based neurological multidisciplinary care. Clinical guidelines for symptom control are also suggested, confirming the common approach to symptoms like chronic pain and terminal breathlessness with the use of opioids.

2.5.4.4 Specialist Palliative Care Provision

Patients affected by ALS/MND have been cared for by specialist palliative care service since the opening of the first modern Hospice: St Christopher's in London in 1967 by Dame Cicely Saunders (Saunders, Walsh and Smith 1981).

Studies aimed at evaluating the quality of care and the adherence to the recommendations provided in the guidelines discussed in the previous subchapter conclude that patients affected by ALS/MND (and their families) are more likely to receive better care if they are cared for in specialized clinics, also called tertiary clinics, dedicated to the care of this illness (Chio *et al.* 2006, Chio *et al.* 2004b, Bradley *et al.* 2004, Miller *et al.* 2000). Reports from databases of patients cared for by neurological ALS/MND tertiary clinics show that between 36-64% of the patients used hospice facilities (Albert *et al.* 1999b, Mandler *et al.* 2001).

Oliver showed how the specialist palliative care involvement in the care of ALS/MND patients in UK and Ireland varies from unit to unit. Many hospices are involved only in the terminal stages of the disease or in providing respite care and later terminal care. There is a need to develop collaborative relationships with other disciplines and services so to provide more co-ordinated care, in response to and involving patients and their families in the decision of care (Oliver and Webb 2000). These results were confirmed by a European survey among the members of the European ALS Study Group on standards of palliative care in the clinical management of patients with ALS and their families. Authors conclude that, although great efforts are made by the centres to offer the best possible palliative care to ALS patients, discrepancies in the type of care offered are evident and might be resolved by adopting common standards (Borasio *et al.* 2001).

The impact of SPCS on the palliative care outcomes has been described indirectly in seminal papers discussing the role of the hospice care for this population (O'Brien, T., Kelly and Saunders 1992, Hicks and Corcoran 1993) and has then been reinforced by international multicentre studies analyzing the quality of death and the effectiveness of palliative measures in Germany and in the UK (Neudert *et al.* 2001a). These studies

report the usefulness of the palliative intervention, but were all retrospective and without a control group.

Home care received by patients with ALS/MND was described as inadequate and too late to relieve the burden placed on family caregivers (Krivickas, Shockley and Mitsumoto 1997), but recently another study reported that the need for coordinated home care nursing is essential in meeting the patient's multisystem and emotional needs (Hoffman 2008).

In Italy the specialist palliative care involvement in ALS/MND is sparse and anecdotic. Italian Neurologists, adopting the international recommendations of the AAN, where the first to start an approach towards the palliative care and a specific committees was created inside the Italian Neurological Society to study the clinical and ethical issues binding neurology and palliative care (Defanti 2000).

In conclusion the literature strongly advocates the involvement of specialist palliative care in ALS/MND, neurologists recognise the role of palliative care in many aspects of the care of these patients, some experiences of SPCS are described, but prospective randomized controlled studies evaluating the impact of specialist palliative care on the PCO are still lacking, even though several areas of ALS patient care would benefit from controlled studies to establish an evidence base for treatment decisions (Borasio *et al.* 2001).

2.5.5 Multiple Sclerosis

2.5.5.1 Definition

Multiple sclerosis (MS) is a chronic recurrent inflammatory disorder of the central nervous system (CNS). The disease results in injury to the myelin sheaths, the oligodendrocytes and, to a lesser extent, the axons and nerve cells themselves. The cause of MS is unknown although immune mediated mechanisms are almost certainly involved, either primarily or secondarily, and many authors favour a primary autoimmune basis for MS. MS is characterized pathologically by patches of demyelination that are found multifocally within the CNS white matter. Grey matter is relatively spared, as are the nerve axons although recent reports have highlighted the importance of axonal injury (Goodin *et al.* 2002).

2.5.5.2 Epidemiology

Women are affected more often than men. The disease typically becomes clinically apparent between the ages of 20 and 40 years, although, it can begin either earlier or later in life.

- Prevalence of the disease depends on the geographic area being more frequent in countries close to the poles and less in equatorial areas. Prevalence rates in Canada, Europe, and the United States (US) range from 100-200 cases per 100,000 population according to the American Academy of Neurologists and the total estimated prevalence rate of MS in Europe, for the past three decades, is 83 per 100 000 with higher rates in northern countries. Prevalence rates are higher for women for all countries considered with a female:male ratio around 2.0. The highest prevalence rates have been estimated for the age group 35–64 years for both sexes and for all countries (Goodin *et al.* 2002).

- The estimated European mean annual MS incidence rate is 4.3 cases per 100 000 (Pugliatti *et al.* 2006).
- Despite the wealth of epidemiological data on MS, comparing epidemiological indices among European countries is a hard task and often leads only to approximate estimates. This represents a major methodological concern when evaluating the MS burden in Europe and when implementing specific cost-of-illness studies (Pugliatti *et al.* 2006).
- 80% of patients present with relapsing/remitting disease and, typically, the illness passes through phases of relapse with full recovery, relapse with persistent deficit, and secondary progression. In about a quarter of patients, multiple sclerosis never affects activities of daily living; conversely, up to 15% become severely disabled within a short time. Episodes happen at random intervals, but initially average about one per year, decreasing steadily thereafter. In 20% of patients, the disease is progressive from onset, hence termed primary progressive—affecting the spinal cord and, less frequently, the optic nerve, cerebrum, or cerebellum (Compston and Coles 2002)
- The neurology services in the United Kingdom estimate that there are 85.000 people with MS at any time, of whom around 10% have primary progressive MS, and 40% have secondary progressive MS (Higginson, I. J. *et al.* 2006b, Multiple Sclerosis 2003).
- 10% of patients with MS would be expected to have an EDSS of 8 or above at any one time (Weinshenker *et al.* 1991). EDSS (Kurtzke 1983) is a disability scale 0 to 10 with higher scores indicating worse disability. A value of 8 indicates a patient essentially restricted to bed or chair or perambulated in wheelchair, but may be out of bed itself much of the day; retains many self-care functions; generally has effective use of arms. On average patients spend 2 or 3 years in this stage and are considered in advanced stage of the disease (Compston and Coles 2002).

2.5.5.3 Clinical forms of MS

Four clinical evolution of the disease have been described (Lublin and Reingold 1996):

- **Relapsing / Remitting (RRMS)**

This form accounts for approximately 85-90% of MS cases at onset (Goodin *et al.* 2002). The disease takes the form of relapses or attacks interspersed with periods of remission. This is because the CNS can often compensate for areas of damage by repairing myelin or by re-routing messages around the problem area. In a remission, symptoms that were disabling during a relapse can virtually disappear. Occasionally there is some residual damage after a relapse, leading to incremental levels of disability. Exacerbations may be triggered by viral and bacterial infection but not by vaccination or immunisation. Exposure to heat and high humidity may amplify symptoms (Uhtoff's phenomena) as may strenuous exercise and menstruation. Pregnancy protects against exacerbation but the risk of relapse is increased following delivery (Lorenzi and Ford 2002). The treatment for acute relapses is to treat any underlying infection before giving a course of oral steroids. Steroids have been shown to influence the rate of neurological recovery but not the ultimate degree of recovery. For those with relapsing disease who meet the criteria set by the Association of British Neurologists, disease-modifying therapies (DMT) can be prescribed (Goodin *et al.* 2002).

- **Secondary Progressive (SPMS)**
About 75% of people whose disease pattern begins with relapsing/remitting symptoms go on to develop secondary progressive disease. This heralds a gradual increase in disability irrespective of the presence of continuing relapses. For 50% of people this change occurs within the first ten years of diagnosis. This 'transition' can be quite gradual and may only be recognised in retrospect, as the awaited remission never happens. For others progression can be quite marked, and the loss of the improved quality of life previously experienced in remissions can be profound. This transitional period can be emotionally and physically difficult, as people fear the disease has 'finally caught up with them' and often worry for the future (Lublin and Reingold 1996).
- **Primary Progressive (PPMS)**
About 10% of people have this chronic condition from onset. Symptoms gradually worsen over the years, without relapse or remission. This form of MS is not amenable to the disease-modifying treatments that have received so much media coverage (Multiple Sclerosis 2003).
- **Progressive/Relapsing MS (PRMS)**

This form begins with a progressive course although these patients experience occasional attacks, which are superimposed upon their steadily progressive disease course (Goodin *et al.* 2002).

2.5.5.4 Mortality and end of life

Despite being considered a chronic condition causing disability, but not death, we know that overall mortality is three times higher, and life expectancy 10 years less among people with MS than general population (Bronnum-Hansen, Koch-Henriksen and Stenager 2004). The suicidal risk is double respect general population (Bronnum-Hansen *et al.* 2005). General cause of death reported in the Danish MS register are cardiovascular and respiratory diseases, infections, suicide and fatal accidents. Risk of death from cancer in MS seems lower than general population, (Bronnum-Hansen *et al.* 2006).

Severe MS disability, as measured by an EDSS of 7.5 or higher is a major risk factor for death with case fatality ratio for this group of patients approaching 4 times the rate for controls (Sadovnick *et al.* 1991).

2.5.5.5 Specialist Palliative Care needs and involvement

The evidence of symptoms and other unmet needs that can be faced by SPCS for people with MS was reported by many authors.

From these papers it results that in MS six symptoms affect more than 50% of patients: problems using legs, problems using arms, fatigue/lack of energy, spasms, pain, and feeling sleepy. Many symptoms in people severely affected by MS are as highly prevalent and severe as those experienced by patients with advanced cancer. Increased disability is associated with increased severity for some symptoms (Higginson, I. J. *et al.* 2006a).

Neurologic bladder, sexual dysfunction, swallowing and speech impairment, cognitive decline and psychiatric complications can worsen the clinical picture of these people, and symptomatic relief and counselling of patients with MS have a strong impact on quality of life (Ben-Zacharia and Lublin 2001).

Qualitative research provided evidence of disbelief and devastation, losses and forced life choices, tracking down services and information, and sadness and relief. Authors conclude that given the duration, range of symptoms and distress often associated with MS the research raise the important question of the role of palliative services in supporting the person with MS and the family (Wollin, Yates and Kristjanson 2006). Similar findings related to the difficulties to cope with the losses and changes, as well as the discomfort in having to fight to obtain everything, above all to track down the services were reported in other studies (Edmonds *et al.* 2007b, Edmonds *et al.* 2007a, Edmonds 2006). Probably due to the long trajectory of the disease MS patients are more likely to live alone, without caregivers, compared with people severely affected by other neurological disorders (Kristjanson, Aoun and Oldham 2006).

MS is the only neurodegenerative disorder for which the involvement of palliative care has been evaluated with a RCT. The results show that the involvement with the palliative care service appeared to positively affect nausea and problems sleeping to some degree, with the effect strongest after initial contact with the clinical service. It also seemed to have a positive impact on informal carer wellbeing. User satisfaction with the service was high among MS patients, carers and especially other health care professionals. Available time, symptom management, end of life decision making and liaison appear to be important components of the service.

Involvement with the palliative care service did not shorten life in patients severely affected by MS. Findings suggest that short term interventions from specialist palliative care may be beneficial for some patients. The authors conclude that whilst there were merits to the model of service delivery evaluated in their study, further work is required to determine the most appropriate model(s) of service delivery to best meet patients' and carers' needs, building on existing expertise in neurology, rehabilitation and palliative care. (Edmonds *et al.* 2006).

2.5.6 Movement disorders

Movement disorders are a group of neurodegenerative conditions represented by the Parkinson's disease and by the atypical parkinsonian syndromes.

For the purpose of this thesis movement disorders will be presented as a common group formed by Parkinson's disease and the atypical parkinsonisms (multiple system atrophy and progressive supranuclear palsy). This group will be represented with the abbreviation PDs.

In the following paragraph an introduction to the movement disorders considered in this research is provided.

2.5.6.1 Parkinson's disease

Parkinson's disease (PD) is the second most common neurodegenerative disorder after Alzheimer's disease and its prevalence is increasing with the populations aging (Kristjanson, Aoun and Oldham 2006, Lanoix 2009).

PD is a progressive, degenerative disorder of the nervous system characterized by asymmetric onset of bradykinesia and rigidity (Hudson, Toye and Kristjanson 2006). It causes substantial morbidity and may result in a shortened life (O'Brien, T. 2002) Tremor is present in approximately 75% of cases, and muscle weakness and postural

instability are also characteristic. The disease can present differently in individuals, its progression is variable and the degree of disability can fluctuate over the course of a day. Depression is a common complication (Stein and Read 1997).

Although PD is not thought to directly cause death, it does shorten the lifespan and people in the later stages of the disease may have symptoms that exacerbate other fatal illnesses. If the uncertainty of the disease is considered a barrier to effective palliation, some authors see it, rather, as another reason for neurologists to be more involved in the care of PD patients (Liao and Arnold 2007). People with PD face a variety of losses in body image, mobility, social role and independence (Abudi *et al.* 1997).

2.5.6.1.1 Epidemiology

The prevalence of PD in industrialised countries is generally estimated at 0.3% of the entire population and about 1% in people over 60 years of age. PD clearly is an age-related disease: it is rare before age 50 years and the prevalence increases with age up to 4% in the highest age groups. It is unclear if there is a higher frequency of PD in males, as studies have not confirmed this (de Lau and Breteler 2006). An overall prevalence in a general population can be estimated at 160/100.000 inhabitants (Doyle *et al.* 2004) even though this raw data is strongly influenced by the age classes of that population.

Reported standardised incidence rates of PD are 8–18 per 100 000 person-years and increase of the incidence is seen after age 60 years (de Lau and Breteler 2006).

Prognosis, mortality and causes of death will be discussed together with the atypical syndromes.

2.5.6.1.2 Mortality and causes of death

In one of the first papers about the natural history of PD Hoehn & Yahr found an excess mortality in PD patients over the age matched normal population of about threefold and concluded that “The state of parkinsonism severely limits life expectancy”(Hoehn and Yahr 1967). In the post Levodopa era have found reduced excess mortality but mortality ratios were still between 1.5 and 2.5 and average life expectancy has been extended from 7.5 to 11 years though with wide variations depending on the response to the drug.(Shaw, K. M., Lees and Stern 1980). The mean survival for idiopathic PD ranges from 9 to 15 years, age of onset seems to be the best predictor of death within ten years (MacMahon 1999, Hely *et al.* 1999).

The course of the disease, when there is a good compliance and response to Levodopa, is characterized by 5 good years. After 10 years patients experience several drug changes and became progressively disabled, with or without mental problems. Often these events determinate that patients leave their home and are admitted to long term facilities where mortality increase rapidly (Clough and Blockley 2004).

There is little data on the usual cause of death of PD patients. Pneumonia is the leading cause of death in Parkinson’s disease followed by cardiovascular events, stroke and cancer in most surveys. Parkinsonism is also indicated as cause of death up to 38% of patients. (Poewe 2006). Causes of death among PD patients were identified from death certificates and compared to a population-based control group of 4,491 individuals. Mortality was greater in male PD patients (41%) compared to females (28 %) and age, disability scores (Hoehn & Yahr stage) at baseline were greater in the PD patients who had died during follow-up compared to survivors. More PD patients (20%) died from

pneumonia than controls (9%) while more controls (23 %) had died from coronary heart disease compared to patients with PD (13%) (Beyer *et al.* 2001).

Cognitive decline as well as older age at onset have been identified as predictors of decreased survival in several studies and is a substantial contributor to the death of a large number of people. Dementia is a clear marker of poor prognosis and 25-40% of patient eventually develop dementia (Jellinger *et al.* 2002, Buter *et al.* 2008, de Lau and Breteler 2006) while up to 78% in advanced stages have cognitive impairment (Bunting-Perry 2006, Weintraub and Stern 2005)

The symptoms of PD may also exacerbate other illnesses that can be fatal, such as pneumonia and cardiovascular disease.(MacMahon 1999)

2.5.6.1.3 Common symptoms

Analysing the motor complications of PD the most frequent and typical are hypokinesia/akinesia that usually respond well to the L-Dopa therapy, but when it loses its efficacy they evolve towards stiffness and freezing episodes. Dyskinesias and dystonia are caused by the dopaminergic drugs and appear in the advanced stages, when the therapeutic interval between the positive effect and the side effects becomes too limited or totally absent. In this situation motor symptoms are fluctuant and the patients alternate episode of freezing in which are completely blocked, with episodes of dyskinesias when they cannot control their movements (Elman *et al.* 2007). Dyskinesias can cause weight loss and increase frailty. Excessive dyskinesias can cause death from exhaustion. PD in latest stage will cause immobility(Clough and Blockley 2004).

Non motor symptoms are various and often not recognized and undertreated. Studies have reported a prevalence of pain in an estimated 40-85% of people with PD; more than 20% of patients experience more than one pain at the time and this symptom could be PD related or not. Overall, analgesic use was low (Ford 1998, Lee *et al.* 2006c).

Speech, swallowing, sleep, bladder and bowel functions are often impaired (Sandyk 1986). Speech problems may be accompanied by swallowing difficulties and hence weight loss. Dysphagia and cough may indicate inhalation problems which lead to chest infections. Immobilization, falls, chest and urinary infections, and exhaustion weight loss may lead to death (Hely *et al.* 1999, Hudson, Toye and Kristjanson 2006).

As mentioned before one of the trigger conditions that indicate a short life span in PD are the cognitive dysfunctions, leading to dementia, or psychiatric complications like the psychosis induced by the dopaminergic drugs (Elman *et al.* 2007). Other psychiatric complications are anxiety and depression that can affect up to 40% of the patients and that in 50% re not recognized by the neurologists (Shulman *et al.* 2002)

2.5.6.2 Parkinsonisms (PD atypical forms or PD plus syndromes)

Parkinsonisms, PD atypical forms, PD plus syndromes are all synonymous terms to identify movement disorders that share common features with the typical PD, but present other peculiar features that make these syndromes recognizable and different from PD. One common characteristic of the atypical parkinsonisms is the low response to the L-Dopa and to the other dopaminergic drugs, which represent the mainstream of the pharmacological therapy for the extra-pyramidal movement disorders. When the disease progress, palliative needs successively increase (Sjostrom, Holmberg and Strang 2002).

Parkinsonisms that not respond to L-dopa carries a much worst prognosis (Clough and Blockley 2004). Progressive Supranuclear Palsy (PSP) and Multiple System Atrophy(MSA) are the atypical PD forms that reflect the most well developed knowledge base (Sano 2006) and therefore are the most reported in literature.

2.5.6.2.1 Multiple system atrophy

Multiple system atrophy (MSA) is a degenerative disorder of the central and autonomic nervous system. According to seminal researches MSA presents an unremitting downward course with life expectancy of 5-8 years (Quinn 1989). Other authors report a longer median survival, around 9 years from the first symptom to death (Wenning *et al.* 1994). The disease affects slightly more male than females with a ratio of 1.3/1 (Bower *et al.* 1997). Usually there's earlier onset falls associated to bulbar symptoms. Hallucinations and dementia tend not to be specific features (Clough and Blockley 2004). Recent epidemiological surveys reported a prevalence rate of 4.4/100,000 and an incidence rate of 3/100,000/year (Wenning, Geser and Poewe 2005). Italians data report a prevalence of 17-29/100.000 for the population older than 55, but with most prevalence concentrated between 60-79 (81% of the cases), whereas in the 55-59 and >80 groups the prevalence rate is respectively of 9.8% and 8.7% (Chio, Magnani and Schiffer 1998, Morgante *et al.* 1992, Schrag, Ben-Shlomo and Quinn 1999, Vanacore *et al.* 2001). Like PD people affected by MSA presents the typical extrapyramidal symptoms (bradikinesia, tremors and rigidity), complicated by other physical symptoms. In particular the patients experience motor impairment (100%) accompanied by cerebellar signs and non motor symptoms like autonomic signs, as erectile dysfunction in males, bladder disturbance and postural hypotension (82-100%). Other common problems are constipation (57%), dysphagia (30%) and respiratory troubles (42%) that can lead to the death of the patients (Colosimo and Pezzella 2002).

2.5.6.2.2 Progressive supranuclear palsy

Progressive supranuclear palsy (PSP) or Steele-Richardson-Olszewski syndrome is a neurodegenerative disease of middle and late age often mistaken for PD. Again early onset falls, absence of response to dopaminergics characterized this condition. Particular features are severe ocular movement disorders starting with impaired voluntary vertical eye gaze and slow saccadic eye movements. The annual incidence of the disease is 5.3 new cases per 100.000 person years and the prevalence of the disease is estimated between 1.39 per 100 000 population in the US and 4.9 per 100 000 in UK. It is likely that these figures are not precise with a considerable under-estimate, as many patients with PSP are not diagnosed and die with other diagnoses. The incidence of PSP increases with age (1.7 at 50–59 years, 14.7 at 80–99 years). Males are affected more commonly than females (Rehman 2000, Schrag, Ben-Shlomo and Quinn 1999). For PSP Median survival time from symptoms onset is 5.6 years (Bower *et al.* 1997, Litvan *et al.* 1996). At the present no curative therapies exist for this condition and the therapy is only symptomatic (Rehman 2000). The commonest clinical feature is represented by mobility problems associated to visual symptoms that are often severely disabling. Complications of accidental falls, swallowing problems and respiratory infections are the main causes of death (Nath *et al.* 2003). Other common symptoms are constipation, bladder problems, depression, and muscular stiffness – spasms (Saleem, Leigh and Higginson 2007).

2.5.6.2.3 Movement disorders common palliative care issues

The Parkinson's-plus syndromes, multi-system atrophy and progressive supra nuclear palsy, which are relatively rare conditions, are included in those 6% of UK hospice patients having non-malignant diseases (Morris and Gonsalkorale 2004).

Both MSA and PSP share common palliative care problems with PD (Clough and Blockley 2004) and are often misdiagnosed as PD (Schrag, Ben-Shlomo and Quinn 1999). The treatment is similar for all these conditions and focuses on controlling symptoms and on increasing or maintaining quality of life. In this regard, the situation is very similar to the multiprofessional and multidisciplinary team approach used in palliative care (Doyle *et al.* 2004). Some patients respond to levodopa or dopamine agonists such as apomorphine, although to a lesser degree than Parkinson patients (Sjostrom, Holmberg and Strang 2002). Atypical parkinsonian syndromes considered as a whole present an average survival time from diagnosis of 9,8 years (Thomas and MacMahon 2004a).

The relevance of aspiration pneumonia as a contributor to the increased mortality in Parkinson's disease and in the atypical syndromes is also suggested by a post-mortem study showing a tight correlation between the onset of clinically relevant dysphagia and survival time across a group of 77 postmortem confirmed cases of different degenerative parkinsonian disorders including PD, MSA, PSP. Survival time after onset of complaint of dysphagia is similar in PD, MSA and PSP: 15-24 months, and latency to a complaint of dysphagia is highly correlated with total survival time in all disorders (Muller *et al.* 2001).

2.5.6.3 Specialist Palliative Care involvement

Very few experiences of a direct involvement of SPCS in the care of PDs patients were found in the literature.

For the vast majority of those who work in palliative care, the only contact with PDs is when it is incidental to a cancer diagnosis (Morris and Gonsalkorale 2004).

A theoretical model of care have been proposed starting from a classification of the clinical stages of the diseases: Four disease stages for PD patients have been identified: diagnosis, maintenance, complex and palliative. The palliative stage has a mean duration of 2.2 ± 2.2 years for PD, and 1.5 ± 1.2 for atypical (Thomas and MacMahon 2004a, Thomas and MacMahon 2004b). This wide range of time spent in the last stage of the disease, which can be up to 4.4 years for PD and up to 2.7 years for the atypical syndromes, shows how difficult is to predict the prognosis in PDs even in very advanced stages. This is one of the reasons for which SPCS have been so little involved in the care of these patients. Bunting Perry proposes a model describing the trajectory of PD related to the increase of physical disability and the linear relationship existing between the loss of efficacy of treatments aimed at prolonging the patients' life and the increase of the palliative care needs. Hospice care is proposed as terminal care in the very advanced stages extended to the bereavement care for the family after the patients' death.

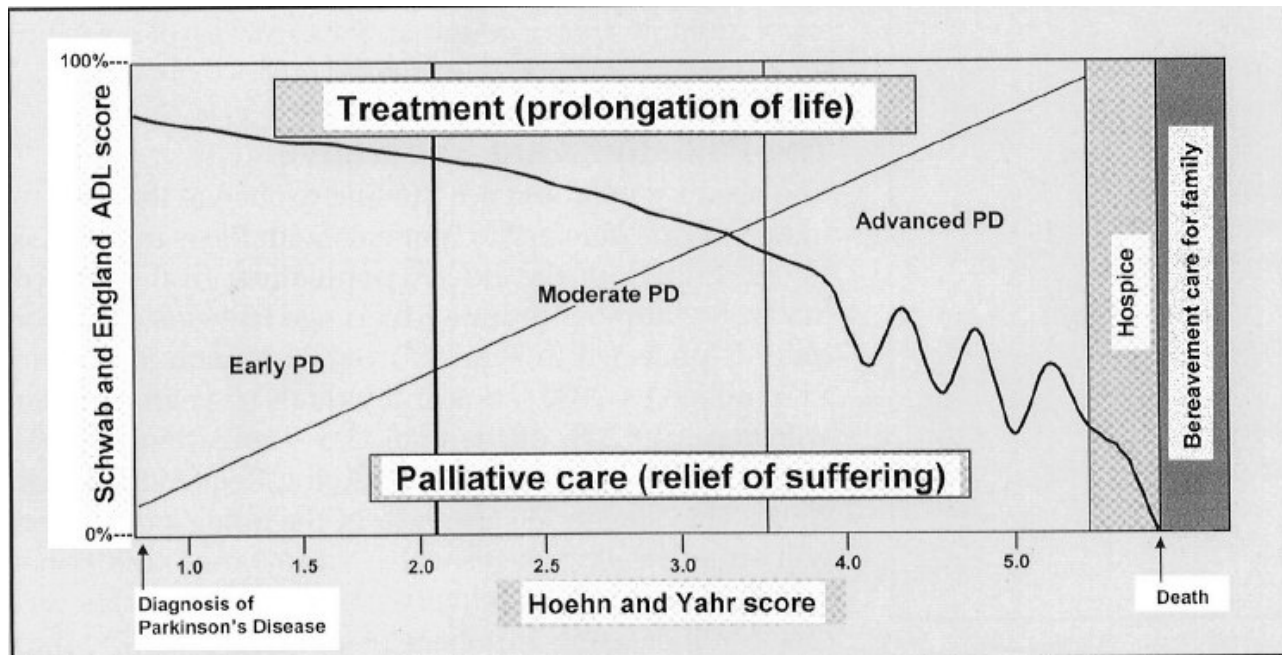


Figure 3: PD trajectory and Palliative Care involvement (Bunting-Perry 2006)

The palliative stage of PDs is characterized for having the aim of relieving symptoms and distress in patients and carers, morbidity relief and maintenance of dignity and remaining functions despite the advanced disease. The role of a SPCS should be to provide advice on administration of medications, progressive dopaminergic withdrawal, analgesia, sedation, control of urinary symptoms, skin care and psychosocial care. The outcomes should be absence of distress, symptoms controlled and maintenance of dignity (Thomas and MacMahon 2004a).

Patients may experience several episodes of physical and cognitive decline to the disease or co morbidity. In this stage the SPCS should take an active role. Advance directives, symptoms control and life prolonging strategies (e.g. PEG) should be discussed and put in act if patients and families wish. A further discussion should focus on the possibility of withholding or withdrawing treatment at the end of life (Bunting-Perry 2006).

In a survey in Oregon family caregivers of deceased PDs and ALS/MND patients were asked about symptoms, treatment preferences, health care usage and psychosocial experiences during the last month of life. Authors conclude that, in the views of caregivers, suffering associated to ALS/MND is no more severe than suffering associated to PDs, and both groups have unmet palliative care needs in the last month of life. PDs patients had significantly shorter hospice care than ALS/MND patients (Goy, Carter and Ganzini 2008).

At the author's knowledge, to date, no evaluation of the impact of SPCS for PDs has been published.

2.5.7 Neurodegenerative conditions summary

In the previous chapters evidence from the published literature were reported describing the clinical and epidemiologic main characteristics of ALS/MND, MS and PDs , the

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common illnesses trajectories, causes of death, main palliative care issues and, where existing, previous experiences of the involvement of specialist palliative care.

In summary we can conclude that all these disorders share common features such as the relentless progression, the incurability, the high level of physical dependency in the advanced stages, the high prevalence of physical symptoms, the psychosocial and spiritual unmet issues. All these disease cause a significant reduction of the patients' lifespan even though this is more evident for ALS/MND and for the atypical parkinsonian syndromes rather than in MS or PD.

Little, no evidence exist about the impact of specialist palliative care on the common palliative care outcomes experienced by this population.

2.6 Patients' needs at the end of life

This section of the thesis follows the description of the neurodegenerative conditions and explore the concept of the human needs. The proposed pathway starts from a theoretical concept of human needs, translate it to the hospice setting exploring the needs of people in the advanced stages of life threatening conditions and then apply these concepts to the neurodegenerative conditions explored in this research.

2.6.1 Maslow's concepts of the human needs

Human needs have been described in Maslow's theory of human motivation (Maslow, A. H. 1943). He stated that human needs arrange themselves in hierarchies of prepotency. This is to say that the appearance of one need usually rests on the prior satisfaction of another, more pre-potent need.

His theory is often represented in a pyramidal shape:

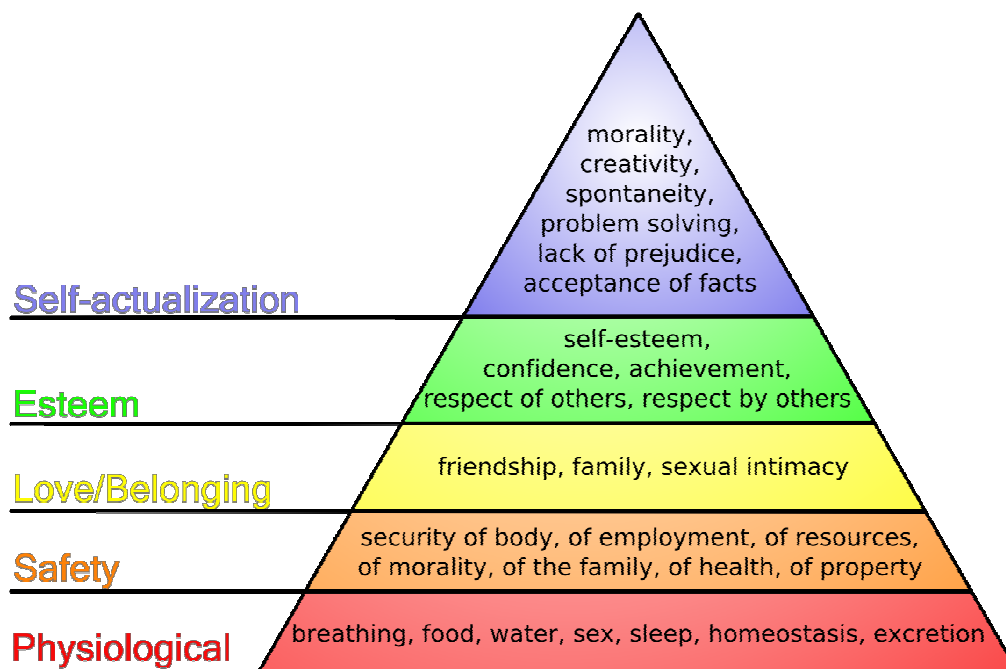


Figure 4: Human needs adapted from (Maslow, A. H. 1943)

Chapter 2. Background: Literature Review

At the base of the pyramid the group of needs with the highest hierarchical position are placed:

- Physiological needs are the most pre-potent of all needs. If these needs are not satisfied the organism is dominated by these, and all the others may become simply non-existent or be pushed into the backgrounds.
- The safety needs follow in order of importance. They can however emerge when physiological needs are relatively satisfied. This group of needs seems related to the infancy and childhood where they are more pronounced and clearly expressed. The author describes this phenomenon justified for the fact that adults tend to inhibit reactions to potential threats due to social habits, whereas children react in a total fashion if they feel they are endangered. The need of protection is generally satisfied in well developed countries where citizens are no longer threatened by wild animals, extremes temperatures or assault and murder.
- The love and belonging needs emerge when and if the 2 previous are relatively satisfied. People feel keenly, as never before, the absence of friends, or a loved one, a wife or children. In our society the thwarting of these needs is commonly at the base of maladjustments and more severe psychopathology.
- The esteem needs is the desire for a stable, firmly based, high evaluation of themselves, for self-respect, or self-esteem and reflects the respect obtained from the others. Satisfaction of these needs leads to self-confidence, worth, strength, capability feeling of being useful to the world, but thwarting of these needs produce feelings of inferiority, weakness and helplessness. These can be the base for neurosis.
- The self-actualization needs are on the top of the pyramid, meaning that can be achieved only if all the previous are at least partially satisfied.. When these needs are satisfied we may claim that a person is fully satisfied and realized. They rely on the specific individual vocation of each person. It can be exemplified by the sentence “ What a man can be, he must be”. A musician must write music, a poet must write, a painter must paint and so on. It is the tendency to become everything that one is capable of becoming (Maslow, A 1970).

The relationship among these categories of needs is certainly hierarchical, but this does not mean that a basic need must be satisfied 100% to let the following need to come out. So most people have these needs satisfied in different proportions. The fundamental meaning of this approach, that will be discussed and applied in this thesis, is to describe the palliative care needs of participants, looking at the relationship between these unsatisfied needs and the priorities of people with advanced neurological disease.

2.6.2 Patient needs and the role of palliative care

The hierarchical concept of needs proposed by Maslow has been widely used in business and social sciences, but it appears only sparsely in the palliative care literature. Zalensky and Raspa applied this model to the hospice and palliative care settings finding high similarity with the Cicely Saunders's concept of "total pain" affecting physical, psychological, social and spiritual components of persons at the end of their lives (Clark 1999).

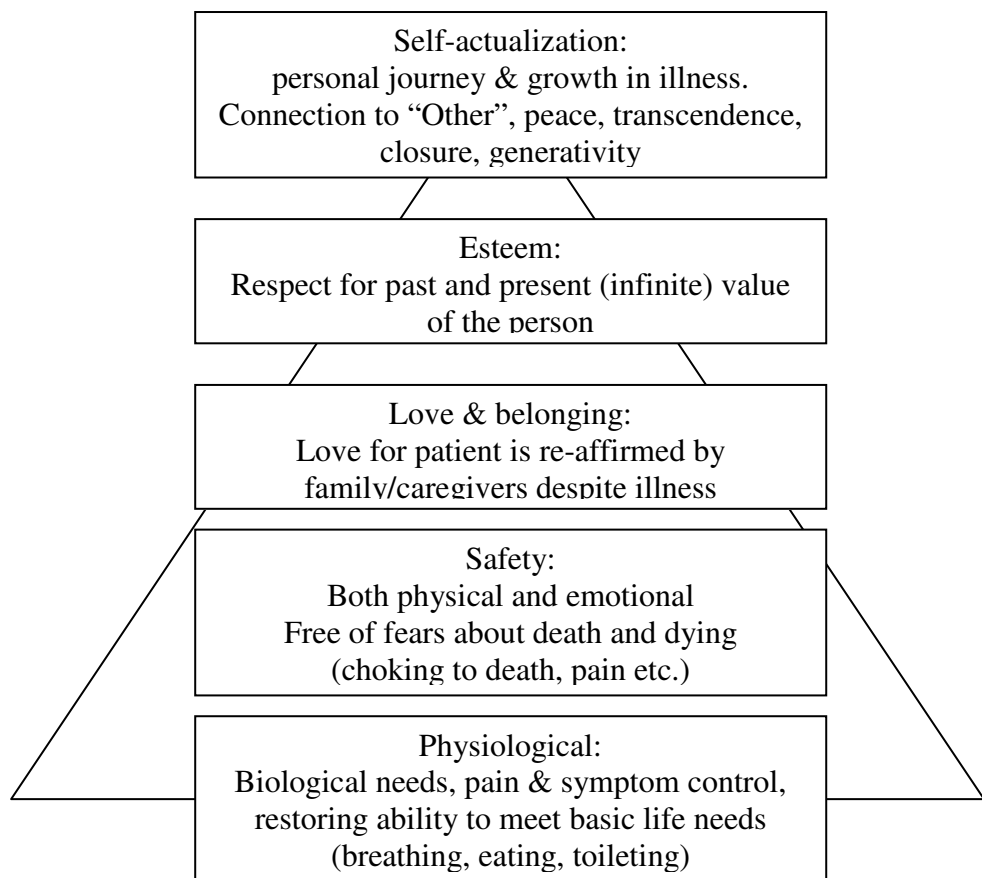


Figure 5: Maslow's hierarchy adapted to hospice and palliative care. The figure diagrams the dependence on lower needs; the apex of the pyramid suggests that higher needs are less frequently realized.

Adapted from: (Zalenski and Rasna 2006)

This adaptation leads to imagine that if physical symptoms are not controlled, e.g. a person is in physical pain or experience severe breathlessness, probably emotional, spiritual and social issues will be a minor concern for him or her. But if impeccable symptom control has been achieved, as suggested in the WHO definition of palliative care (W.H.O. 2002), and probably even if this result is obtain in a lower extent than impeccable, other needs will emerge and are to be somehow fulfilled in order to reach the top of the pyramid where self-actualization is located. We know how the loss of many capabilities experienced by people severely affected by long term conditions can affect esteem and love and belongings too (Edmonds *et al.* 2007b, DH Longterm Conditions NSF 2005).

It is however true that different needs can have different impact on one's personal quality of life as it is shown in many published experiences (Chio *et al.* 2004a, Fegg *et al.* 2005, Kaub-Wittemer *et al.* 2003, Neudert, Wasner and Borasio 2004). Palliative care needs were explored in patients, lay carers, multi-professional palliative care providers and managerial skate holders in Northern Ireland and the main areas of needs identified were social and psychological support; financial concerns, needs for choice and information. Furthermore participants highlighted the inequity of provision of palliative care for patients with cancer and non cancer diseases.(McIlpatrick 2007)

2.6.3 Palliative care needs and neurodegenerative disorders

Palliative care needs (PCN) can be represented by the unsolved problems faced by people severely affected by advanced incurable diseases and by their families. This concept is strictly related to the definitions of palliative care (W.H.O. 2002), where describing what palliative care is the various needs are used to define the specific concept. PCN are listed as the components of the "total pain" (Richmond 2005) and included in the concept of palliative care outcomes, those outcomes that are to be obtained to improve the PCN(Higginson, I. J. 1997b).

PCN can be classified in:

- Patient needs
 - Physical: uncontrolled symptoms, physical limitations due to disability, need for special device to overcome movement limitations, communication impairment, tubes for artificial nutrition or hydration, aids to help breathing like non invasive ventilation, or invasive ventilation, etc.(Addington-Hall, J. *et al.* 1995, Addington-Hall, J. M. and Higginson 2001, Albert *et al.* 2001, Albert *et al.* 2009, Archibald *et al.* 1994, Borasio and Oliver 2006, Borgsteede *et al.* 2009, Chahine, Malik and Davis 2008, Colosimo and Pezzella 2002, Cornish and Mattison 2000, Kristjanson, Aoun and Oldham 2006, Kubler *et al.* 2005, Lee *et al.* 2007, Lisak 2001, Maloni 2000, Miller *et al.* 2009, Miller *et al.* 1999, Mitsumoto *et al.* 2005, Moulin 1989, Moulin 1998, Rabkin *et al.* 2005, Rehman 2000, Rehman 2001, Saleem, Leigh and Higginson 2007, MSCCPG 1998)
 - Psychological: emotional, anxiety and depression, coping with disease and losses, information and communication, abandon, fears (Bolmsjo 2001, Boston, Towers and Barnard 2001).
 - Spiritual: the meaning of the diseases, relation with faith and spirituality, fear of the death and of the dying process, sense of guilt (Taylor and Herr 2004, Lambert 2006).

- Social: isolation, economical concerns, loss of job, change in familiar and social roles (Adelman *et al.* 2004, Aoun and Kristjanson 2004, Chio *et al.* 2005, Clarke, D. M. *et al.* 2005, Gallagher, D. and Monroe 2006, Goldstein *et al.* 1998, Kaub-Wittmer *et al.* 2003, Love *et al.* 2005, Edmonds *et al.* 2007a, Hudson, Toye and Kristjanson 2006, Spliethoff-Kamminga *et al.* 2003, Cotterell 2008)
- Choices: place of care, place of death, advance care planning (Albert *et al.* 1999a, Albert *et al.* 1999b, Bolmsjo and Hermern 2003, Wollin, Yates and Kristjanson 2006, Cotterell 2008, McCluskey 2007)
- Families needs
 - Support: practical help, education, financial and working adaptation, social network, the importance of family atmosphere, help for the children and other weak family components etc (Caap-Ahlgren and Dehlin 2002, Seymour *et al.* 2003).
 - Psychological: coping with losses and preparation for the death of the beloved, prevention of complicated-pathological bereavement and survival after the death of the patient (Dawson and Kristjanson 2003, Hughes *et al.* 2005, Oliver and Gallagher 1998).
 - Spiritual: similar to the patients' ones.

2.6.4 Summary of the patients' needs

In conclusion of this chapter on the palliative care needs faced by patients severely affected by neurodegenerative conditions and their caregivers, it results that a big amount of research was performed and published providing strong evidence about the presence of these unmet needs.

All subgroups of needs categorised by Maslow's approach adapted to palliative care, as well as Cicely Saunders's components of total pain are represented in this population. SPCS aim at meeting the palliative unmet needs of their users, therefore when such services are to be evaluated the domains to be chosen for the assessment must come from these categories of human needs applied to the advanced stages of the diseases towards the end of life.

3. The Qualitative Needs Assessment

3.1 Introduction

After having reviewed the literature about the needs of people severely affected by neurodegenerative conditions a qualitative needs assessment of these needs, in patients and their informal carers living in Turin area, was performed. The point of view of the professional carers was also explored.

This approach is part of the MRC framework (Campbell *et al.* 2000), as shown in Figure 3.1, as the modelling stage (phase 1) of the framework. This, in turn, influences the theory (preclinical phase) and as well because it helps in the strategic design of the project and can predict major confounders.

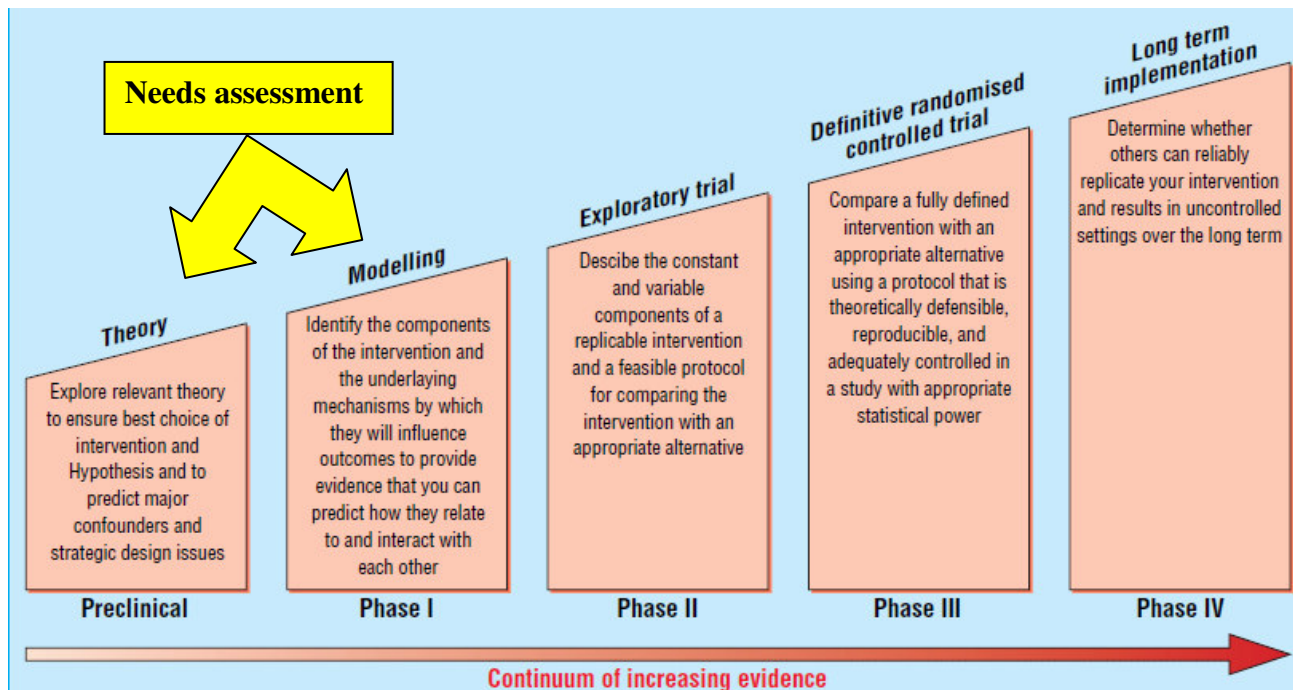


Fig. 6: the modelling phase, phase 1 of the MRC

As previously shown in the literature review of the palliative care un-met needs of people severely affected by neurodegenerative conditions it is now evident that many physical, psychological, spiritual and social issues are not met by the available service for the care and the assistance of this population. These problems can be related to the pathophysiology and the evolution of the diseases (physical needs), emotional and relational aspects (psychological needs), transcendence, faith and meaning of the experience (spiritual needs), integration in the social network, quality of the received care (social needs).

Most of the previously published research is specifically diagnosis related and provides information about different stages of the diseases. Outcomes measured by the rarer quantitative studies focus on specific variables like symptoms or social issues, but never on the full comprehensive of the load of various problems experienced by this heterogeneous group of patients and their families. Recommendations from the published literature suggest that improving awareness of the needs of patients dying

from disorders other than cancer should lead to an increase in referrals to specialist palliative care services (Edmonds 2004)

3.2 Aims of the qualitative study

The purpose of this project was an accurate evaluation of these unmet needs in people severely affected by ALS/MND, MS, PD and related disorders living in Turin city and metropolitan area in order to model a new specific SPCS aimed at providing help for their needs.

Specific aims were:

- explore the lived experience of the diseases told by the directly involved protagonists
- highlight the problems faced by patients and their families ordered and weighted by participants
- analyze the coping strategies adopted
- explore the level of satisfaction about the available services and the participants views about the new palliative care service
- compare the congruence of the problems aroused in the interviews with the previous published experiences.
- collect data from professionals directly involved in the care of these people, their impressions about the quality of the provided care and their point of view towards the forthcoming palliative care service

For this reasons the need to proceed to an in-depth assessment of the potential future user of the service was felt as urgent. Surveys exploring these needs in sample of patients with the same diagnosis (Kristjanson, Aoun and Oldham 2006) revealed how in order to better satisfy the needs tailored service were required. The prevalence of physical problems has been published (Saleem, Leigh and Higginson 2007) and the unique and overlapping aspects of each of these disorders discussed with an emphasis upon the clinical management of symptoms. The authors concluded that effective end-of-life palliative care depends upon an understanding of the clinical aspects of each of these disorders (Elman *et al.* 2007).

The option to proceed with a survey using questionnaires was excluded because no specific tools validated for such an heterogeneous group of diseases was available (for both patients and carers) and, above all, because quantitative methods could not substitute the big amount of data that can be collected by meeting face to face with so highly physically impaired participants and observing their living environment. The meeting with patients, families and professional carers to explore the experience of the disease and allowing them to rank their needs and the impact of the existing service allowed the development of the qualitative study.

3.3 Methodology of the qualitative study

The qualitative study (NeuNeeds: the unmet needs of people severely affected by neurodegenerative conditions) is composed by 2 parts:

- 1) In depth, face to face interviews with patients (and their main carers, if available).
 - a) Patients, specifically adults severely affected by either ALS/MND, MS, PD or related disorders, with mental capacity to give a consent permission to the study

- participation, resident in the area covered by FARO domiciliary home care service (in order to be potentially involved in the future new SPCS).
- b) Their lay carers, usually a familiar, sometimes a paid carer. If the patient did not have any carer or preferred to be interviewed alone this was allowed.
- 2) Focus groups with professionals (involved in their care).
- a) Professional carers directly involved in the patients' care: usually neurologists, but also rehabilitation specialists, physiotherapist and speech and language therapists. Nurses and general practitioners were to be involved, but ultimately their focus groups did not take place for organizational problems.

3.3.1 The in-depth interviews

3.3.1.1 Methods

Interviews were conducted at patients home, if possible, to enable participants to be in their own environment reducing the stress induced by being in an external setting. Other settings were accepted only if the patient had been admitted to the hospital or other facility and was keen to be interviewed in that location (it was not considered acceptable to ask participants so highly disabled to leave their house just to be interviewed). Two researchers led the interviews in order to both control the technical equipment and dedicate time and attention to the interviewees. Each interview was video and audio taped and field notes were taken. This was due to the high communication impairment of most of the patients. In the same video screen the pair formed by the patient and his or her main carer were filmed. Video recordings allowed to catch the non verbal expression of the severely compromised patients, sometimes their face mimic was the only sign to confirm or deny what was told by their carer.

Participants were previously informed by the professional who made the referral (usually the neurologist or rehabilitation specialist) about the aims of the research. Professionals were asked to identify patients and their main lay carer, usually a family member. Before the beginning of the interview the study protocol was provided to the participants and a consent form was delivered. As most patients were so disabled to be unable to sign the written form, a verbal or a clear consent was audio and video recorded. A specific informative form for the patients' general practitioner was given to the family asking them to give it to their doctor at the first occasion.

The in depth interview began with a brief summary of the research aims and participants were informed that this assessment was aimed to help the development of a new service that was not available at that time and that might or might not be offered to them (depending mostly on their will and in part on the future evolution of the research project). This was done not to create unreal illusions and, on the other hand, to avoid that participants felt forced to receive the forthcoming service.

When the scene had been set up researchers asked the patients, if able to communicate verbally or using Augmentative Communication Aids (e.g. a voice amplifiers) or Alternative Communication Aids (alphabet and picture boards, computer keyboards or eye gaze computer systems) and the carer (if present) to describe their experience of disease speaking freely of the themes that they felt more important. Methodologically this possibility offers the advantage to receive information from both the patient and the carer, but also to see the interaction between them. It permits to see what happens when one participant talks about the problems lived by the other one (eg a carer describing the physical symptoms of the patient, or the patient talking about the burden of care experienced by his or her familiar). A limit can be that some issues will not be raised not to hurt their loved one and for this reason some carer added some piece of

information at the end of the interview talking with one researcher while the other was staying with the patient. Two patients e-mailed, after the interview extra comments that they could not say during the interview or that did not want to share with their carer.

The researchers just hinted the main area of interest of the research: physical, psychological, spiritual and social aspects including comments on the service available and experienced. It was then reinforced the fact that it was up to them to cover all these aspects or just focus on one or two. Also the order in which they were to talk about their problems was free.

Participants were informed that all data were going to be managed for research purpose only, that were going to be treated anonymously, and that the content of the interview was not communicated to their professional carers.

Participants were informed that they could stop the interview at any time, if they felt this was too much a burden for them, and that could withdraw their consent to the use of the data even at the end of the interviews if they felt that the content could be not acceptable or not adequate.

If some important piece of information about the care programme, serious physical uncontrolled symptom or therapeutic options emerged from the interviews the researchers offered to get in touch with the professional carers who referred the patient to inform them about these issues.

3.3.1.2 Patients' referrals

Patients referrals were made by neurologists and other professionals involved in their care. Participants were identified from neurological ambulatories for the specific diseases in Molinette Hospital (Turin), neurological department, and in S. Luigi Gonzaga Hospital (Orbassano). The former host an ALS/MND regional centre (a tertiary clinic), an MS ambulatory, and a PD clinic (regional centre for movement disorders and recruitment and selection for Deep Brain Stimulation). The latter host the regional MS centre, a general neurological ward and a respiratory unit where patients affected by respiratory impairment are referred for breathing problems.

3.3.1.3 Inclusion criteria for the patients

- Diagnosis of one of the following neurodegenerative conditions:
 - ALS/MND
 - MS
 - PD or atypical syndromes (Multi System Atrophy MSA, Progressive Supranuclear Palsy PSP).
- State of severe advanced disease. Conditions were considered severe if:
 - They met the criteria of the specific clinical indicators of advanced disease presented in the Gold Standards Framework prognostic guidance for the specific diagnosis (Gold Standards Framework 2006)
 - The neurologists or other professionals directly involved in their care stated that they had serious palliative care needs (Higginson, I. 1997a,pag 220) or were presumed them to be in their last year or months of life.
 - Disability scales were considered only to help professionals to better understand the target of our population. For ALS/MND we

suggested to refer patients with ALSFSR-R ≤ 24 (Cedarbaum *et al.* 1999), for MS EDSS $\geq 8,5$ (Higginson, I. J. *et al.* 2006c) and for movement disorders a H&Y score ≥ 4 (Bunting-Perry 2006).

- Absence of a cognitive impairment at a level not permitting to give the consent to the interview and / or to let their opinion to come out during the interview.
- Consent to the interview
- Resident in the territory covered by FARO SPCS

3.3.1.4 Exclusion criteria for the patients

- Diagnosis other than those listed
- Severe cognitive impairment
- Unable to express their views even with communication aids
- Unable to give consent
- Resident outside of the territory covered by FARO SPCS

3.3.1.5 Sampling

The first criteria for sampling was heterogeneity of the sample, which means that the three groups of different diagnosis (ALS/MND, MS and PD-related disorders) had to be represented.

The second criteria was that different clinical conditions, but all in the advanced stages, were to be represented in the sample, so research team could collect data from a variety of different clinical conditions.

The theoretical sampling method (Coyne 1997) was used - the sampling decision was based on the analysis of the data progressively acquired in the interviews and, therefore, is not established "a priori" but is developed in the course of a study.

To achieve this result a first pool of 10 potential interviewees representative of all conditions and with different clinical features was selected. They were interviewed and data were collected and analyzed. Afterwards other interviews were added until saturation of the themes was achieved.

3.3.2 The focus groups

Focus groups were aimed at exploring the professionals' point of view about the unmet needs of this population, their opinions about the existing services and suggestions for the SPCS in development.

A secondary expected outcome was to see interaction among them and find out differences in opinions and strategies of care.

Finally this was a chance to start the creation of a network with them foreseeing the importance of a future collaboration when their patients were to be referred to the service.

3.3.2.1 Participants

Participants were required to be professionals caring for patients with the characteristics listed for the interviews. It was planned to be a multi-professional and multidisciplinary recruitment. Professionals working in clinics specifically dedicated to the care of ALS/MND, MS, PD and related disorders as well as primary medicine doctors and nurses, were invited.

3.3.2.2 Methods

Events were video and audio taped and field notes were taken.

Focus groups were to happen in professionals' place of work in order to obtain a good participation and consent a relaxed environment. Two researchers were present during the events, again with the tasks of caring at the technical equipment, lead the groups and take the notes.

Participants were asked to introduce themselves, their role and position in their job, their professional experience with patients severely affected by neurodegenerative conditions. Interviewers introduced the focus group presenting the research project, declaring the interest for the participants contribute and listing the main area of interest: physical, psychological, spiritual and social issues experienced by their patients, their professional opinion on how these patients were managed, their level of satisfaction about the offered care and suggestions about the development of the new SPCS.

Rules for the event were previously stated by the researchers: participants had to provide a written consent to the participation (were provided with the study protocol and a consent form to be signed). No judgements or professionally inadequate comments were accepted. Professional secret was established on the content of the focus group. Data was to be used for scientific purpose only and treated anonymously.

3.3.2.3 Sampling

The same methodological approach adopted for the interviews was used. Professionals had to represent all the diagnostic categories listed in the patients' inclusion criteria section. They also had to work in clinics or ambulatory specialized for the care of the selected diseases in one of the two main hospital of Turin's metropolitan area (Molinette hospital and San Luigi Gonzaga). Focus group were supposed to be multidisciplinary and involving professionals working in primary home care also.

3.3.3 Data analysis

After each event (interview or focus group) a transcript verbatim of the audio content was performed by the main researcher (S.V.). Video was used to add information about non verbal communication, to explore facial expressions of those patients with communication impairment and to identify the professionals who participated at the focus groups. Transcript verbatim was in the original Italian language and the events were not transcribed in English as suggested by Strauss and Corbin (Strauss and Corbin 1998, p 285). Data extract from the text coded in the different categories were subsequently translated in English and are presented in the result section of the qualitative study of this thesis.

The methodological approach adopted was a qualitative analysis of the data. The transcripts verbatim were read and re-read and significant themes drawn from the data (microanalysis) (Miles and Huberman 1994).

Interviews and focus groups themes were coded independently by two researchers and then summarized in one document with the researchers' agreement.

An open coding of these main themes was performed by the 2 researchers (S.V. and G.G.) on each interview transcript verbatim.

Computer based software (Microsoft WORD and Microsoft Excel) were used to code the transcriptions.

The analysis of the data was based on the content analysis (Mayring 2000, Mostyn 1985). The methodology of the content analysis is described below:

- First step: the results were grouped into 5 categories of needs: physical, psychological, social, spiritual and issues about services satisfaction. These categories had been previously obtained from the literature search and mirror the various components of the “total pain”, described by Dame Cicely Saunders (Clark 1999), (Clark 1999), and also reflecting the adaptation of the human needs described by Maslow (Maslow, A. H. 1943) and adopted by hospice care generally (Zalenski and Raspa 2006).
- Second step: The main researcher (S.V.) performed an axial coding (Strauss and Corbin 1998) of these preliminary needs in order to reduce the large number of emerging problems into the predetermined 5 categories.
- Third step: a comparative analysis of the needs among the 22 interviews was made by calculating the prevalence of the different needs within the 5 different categories. This has enabled the themes to be quantified (how many times a particular theme was mentioned by all participants during the same interview, or how many times particular needs arise from different interviews) (Strauss and Corbin 1998).
- Inter-coder reliability (Mayring 2000, Strauss and Corbin 1998) was established involving a third independent researcher who did not participate at the interviews, nor at the focus groups.
 - The second and the third researchers (G.G. and C.R.) performed a further axial coding on 5 randomly chosen interviews and the results were matched with the original 5 interviews that were axial coded by the main researcher. A congruence of 50% of the overall axial codes was considered adequate. Tables with results of the coding process were made. Diagrams were drawn to show results, which were then matched by the main researcher (S.V.) in order to see the degree of concurrence.

The content analysis of the results produced a quantification of the different needs which were reported by participants. These data were used to identify a number of domains to be assessed as trial outcomes in the following part of the project (see the explorative randomized trial in section).

Apart from this procedure, which can reflect a quantitative attempt to evaluate qualitative data, a further analysis was conducted on the emerged themes: citations extracted by the transcript verbatim were used to better understand the qualities of the different reported needs.

These could reflect:

- The inner properties of the uncontrolled symptoms, (e.g. the intensity of pain or its characteristics, the effectiveness of therapeutic strategies, the adherence of participants to the prescriptions).
- The meaning of the psychosocial or spiritual issues (e.g. the individual impact of feeling abandoned or socially isolated, the coping strategies related to the disability, the impact of the care giving tasks on the carers’ personal quality of life, the relevance of the spiritual distress on participants)
- The perception about of the available services (e.g. if the degree of satisfaction of the participants about the services was influenced by personal aspects such as

the attitude of the carers towards them, or depending by general organizational issues like the presence or the absence of needed services).

The aim of this further analysis of the data was to understand the meaning of the various reported needs and then use the results to model the forthcoming service (e.g. by providing education to the personnel of the SPCS on specific aspects of the care that participants reported as lacking and important, or creating a network of professionals working in different settings aimed at providing a more effective and comprehensive set of services).

3.4 Ethics

The study was approved by the Ethics Committees (EC) of the two hospitals involved in patients recruitment. The approval documents were then mailed to the University of Kent Ethics Committee, as this study is part of a PhD programme at this University.

Participants' consent was obtained in the forms described in the previous paragraphs: a written consent by the lay carers, professionals and by those patients able to sign. Verbal or non verbal assent for patients was acceptable for people who were unable to sign.

Data were treated anonymously and conserved safely by the main researcher.

Video and audio registrations were made only to better interpret the contents of the event and to create a precise transcript verbatim.

In case of severe suffering during the interviews (both caused by the disease e.g. a physical symptom or induced by the interview itself, e.g. an emotional distress) researchers were bound to stop the interview and help the participant through professional intervention (researchers are physicians and nurses experienced in palliative care) offering a quick referral to their professional main carer (usually the ones who referred the patient for the study).

3.5 Results of the qualitative study

3.5.1 Introduction

In this section results of the Neuneeds qualitative assessment will be presented. Results will be divided in two different sections:

- the interviews to patients and lay carers
- the focus groups with professionals

Data collection started on the 14th of May 2007 with the first interview and terminated on the 27th of August 2007 with the last focus group.

3.5.2 The interviews to patients and lay carers

3.5.2.1 Sample description

22 in-depth interviews were conducted with people severely affected by ALS/MND, MS, PD and atypical syndromes (PDs). Informal carers were also interviewed with patients in order to better understand patients needs when affected by communication impairment and, also, to explore the needs affecting them as caregivers.

Overall participants' and informal carers' characteristics are reported in tab 3.1

interviews	n=22			
Patient's gender	M=18 (82%)	F=4 (18%)		
Patient's age	mean=61,5	range (39-80)		
Main carer	Wife=16	Husband=3	Daughter=1 Mother=1	None=1
Place of interviews	home=20	hospital=1	Nursing home=1	
Paid carers	n=13 (59%)			
Diagnosis	ALS/MND n=9	MS n=7	PD n=5	MSA n=1
Disability level	ALSFRS-R* mean=11,3 range= 3-23	EDSS** mean=9 range=8-9,5	H&Y*** mean=4,5 range=4-5	

Tab 3.1: overall participants characteristics

- ALSFRS-R (Cedarbaum *et al.* 1999) relates to ALS/MND
- **EDSS (Kurtzke 1983) relates to MS
- ***H&Y (Hoehn and Yahr 1967) relates to PDs

3.5.2.2 Description of disease specific groups

Because of the different diagnostic subgroups a further description of the sample within the 3 selected diagnostic groups is provided.

Subgroups were composed by ALS/MND patients, MS patients, and PDs patients.

Within the ALS/MND subgroup

- 5 patients were using non invasive ventilation (NIV), 3 of them for more than 14 hours per day, one for about 8 hours per day and one for 2 hours per day.
- 3 patients were tracheostomized and requiring or receiving invasive ventilation (IV) 24 hours per day
- One patient was not using mechanical ventilation, even though it had been prescribed, because he could not tolerate the mask. This patient used low flow of oxygen during the night hours
- 5 had to be fed by PEG
- All interviews were conducted at patients home, all patients were cared for by a family carer and all but one had also one or more paid carers to help in the home assistance

Within the MS sub group

- 2 patient were in NIV during the night
- 3 patients used aspirators for bronchial secretions and coughing machines for weak cough
- 4 had to be fed by PEG
- All interviews were conducted at patients home, all patients all but one were cared for by a family carer and 3 had one or more paid carers to help in the home assistance.

Within the PDs group

- None was using mechanical ventilation or other respiratory aids
- One patient had a PEG that was used to vehicle continuous infusion of L-DOPA
- One patient was in Deep Brain Stimulation
- Two patients were not interviewed in their home: one because was admitted in hospital for symptom control, one had recently been admitted in a nursing home for respite.
- All patients were cared for by a family carer and one had also a paid carer

The common features of the sample were that all patients had been defined as severely affected by their neurodegenerative condition, no curative option was possible and the level of disability was very high (Kollewe *et al.* 2008, Higginson, I. J. *et al.* 2006c, Thomas and MacMahon 2004a). For these reasons all were defined as being in a palliative stage.

3.5.2.3 Data analysis of the interviews

3.5.2.3.1 Inter-rater reliability

The inter-rater reliability analyzed matching the coding process of 5 (22.7%) events randomly chosen among the 22 interviews showed a good overlapping among the 3 independent researchers involved in the analysis of the data. This was probably due to the clear exposition of the themes by the participants.

In the tab 3.2 results of the inter-rater reliability test are presented.

Analysed Domains	Matching among the 3 reserchers (%)
PHYSICAL	72
PSYCOLOGICAL	54
SOCIAL	72
SPIRITUAL	87.5
SERVICES	76.2
Overall Matching	72.34

Table 3.2: Inter-rater reliability test among the 3 researchers expressed in percentage of matching of coded domains

An overall of 72.3 % of codes where coincident among the 3 independent researcher on 5 randomly chosen interviews. This result means that matching the codes found out independently by the 3 involved researchers in the 5 randomly chosen interviews in 72.3% they were coincident, meaning that the qualitative data had been categorised in the same code.

3.5.2.3.2 The content analysis of the physical needs

A content analysis (Mostyn 1985, Mayring 2000) of the needs that emerged from the interview is now presented.

In chart 3.1 the physical need that came out from the 22 interviews are shown:

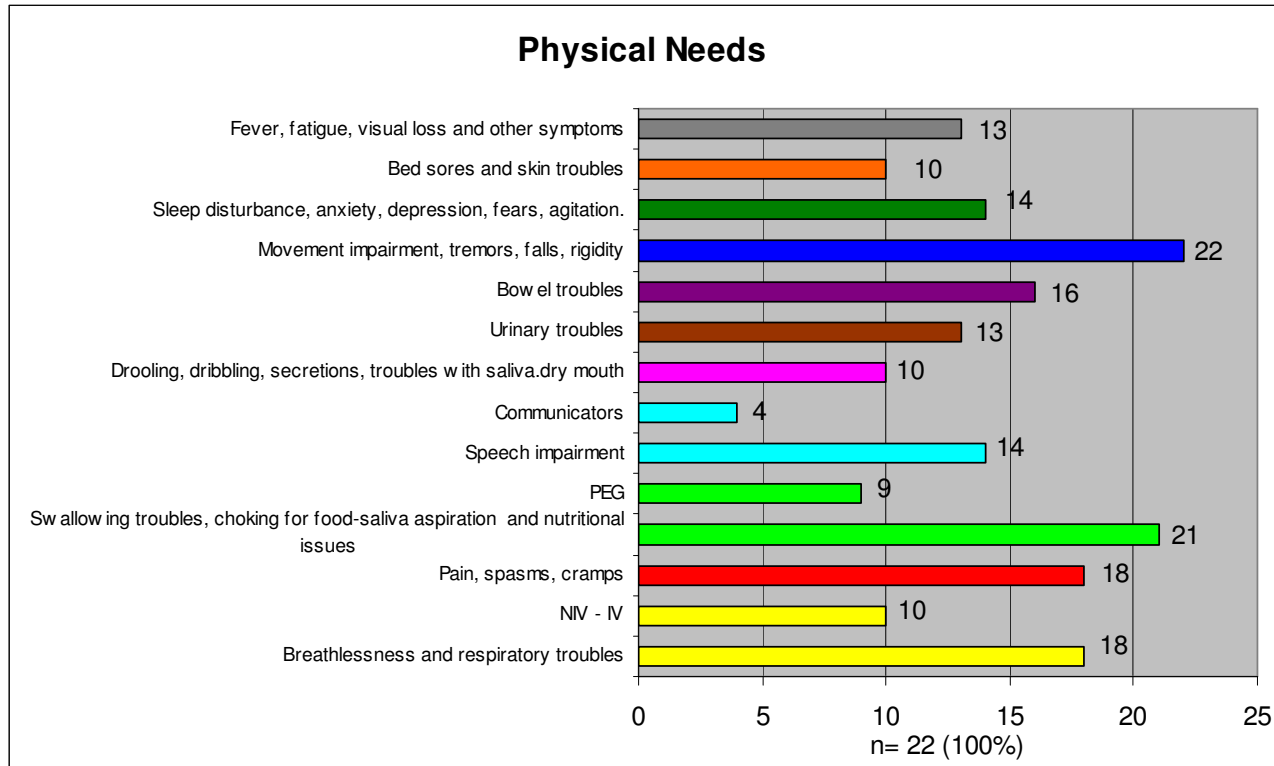


Chart 3.1: the physical needs

6 participants talked about EoL decision to be influenced by physical issues

NIV= Non Invasive Ventilation

IV= Invasive Ventilation

PEG= Percutaneous Endoscopic Gastrostomy

In table 3.3 the physical needs are reported in percentage also with further details.

PHYSICAL NEEDS	Quotes in the 22 interviews	Percentage
Breathlessness and respiratory troubles*	18	81.8%
Pain, spasms, cramps	18	81.8%
Swallowing troubles, choking for food-saliva aspiration and nutritional issues**	21	95.5%
Speech impairment***	14	63.6%
Drooling, dribbling, secretions, troubles with saliva, dry mouth	10	45.4%
Urinary troubles	13	59%
Bowel troubles	16	72.7%
Movement impairment, tremors, falls, rigidity	22	100%
Sleep disturbance, anxiety, depression, fears, agitation.	14	63.6%
Bed sores and skin troubles	10	45.5%
Fever, fatigue, visual loss and other symptoms	13	59%
<p>*NIV= Non Invasive Ventilation \ IV= Invasive Ventilation or oxygen supply were used in 10 patients (45,4%)</p> <p>**PEG= Percutaneous Endoscopic Gastrostomy. 9 patients (40,9%) had a PEG placed</p> <p>*** Alphanumerical tables and/or Electronic Communicators were used by 4 patients (18%)</p>		
<p>table 3.3: the physical needs</p> <p>footnote: in 6 interviews participants talked about End of Life decision to be influenced by physical issues</p>		

3.5.2.3.3 The physical needs detailed analysis

In this section data used to create the various categories of needs emerged from the interviews will be shown

Participants will be identified with the following categories:

- Diagnostic groups: ALS, MS, PD
- Participants' role: P=patient, C=carer, I=interviewer
- Numbers are used to identify participants

Examples:

- PPD2: in this case is a patient (first P), with a movement disorder (PD) and is the number 2 of the diagnostic group.
- CALS5: carer (C) of an ALS patient, number 5 of his or her diagnostic group

Age, and time from the diagnosis and further comments are also displayed when the information is available and useful for the purpose of the analysis.

Not all the data is presented in this section, just those significant citations that led the investigators to the conclusions presented in the discussion section.

3.5.2.3.3.1 Movement impairment

Movement impairments and related symptoms (rigidity, tremors and falls) were reported by all participants 22/22 (100%).

As expected from the high degree of disability of the sample, in all interviews the impact of limitation in movements and the symptoms related to this impairment were discussed by patients and carers. It was often the first physical need reported by participants when prompted to talk about their suffering.

Patients still able to walk underlined the difficulties met in reduced ambulation, fluctuation of symptoms and progression of the disability (Oliver and Borasio 2004, Clough and Blockley 2004, MacLeod 2004, Saleem, Leigh and Higginson 2007):

PPD1(male, 80 years, PD diagnosed 9 years ago and CPD1, his wife)

I= how does he move at home?

C= with a walking stick. We have a wheelchair to go out. We had a walker (walking aid) but we returned it because it wasn't of help.(...) another problem is his muscular rigidity. (...)

I= did he have muscular rigidity at the masticator muscles? P=I had, now it's slightly improved.

Movement impairment caused insecurity, loss of control and dependence (Edmonds *et al.* 2007b, Bunting-Perry 2006):

PPD2 (male 70 years old, diagnosed 6 years ago, CPD2 his wife)

P=I went close to an object that I wanted to grab and when I was at the right distance... I went through. (...)

C=In the night he got up, lost his equilibrium and.. fell down

P=(...) I feel I am blocked.

C= his legs get blocked.

P= once I felt it happening. I felt like it advised me, then it happened but I was prepared for it.

I= did you have enough time to swallow your rescue drug before you got blocked?

P= yes, but the effect arrived later. Now it happens so quickly that I'm aware of it when things are already done.

C= sometimes he stands and at once can't walk. (...) and falls down.

P= this is scaring. I fell several times. Last July I climbed those 3 steps of that stepladder and then I was abruptly blocked. I fell down. I had such a bad back pain!

When the disability evolution is progressive linear falls and consequent complications can be seen as the trigger for physical dependence (Hammerbeck and Garret 2006):

PALS3(male, 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago, CALS3, his wife)

C= in these 5 years he progressively lost the capability to walk. Before he needed a crutch, then 2 and now the wheelchair.

P= yes, because 2 years ago I fell down and fractured a small bone of the pelvis. I had not to move for 90 days, but since then I stopped walking.

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Sometimes disability can create different views within the family. Some carer can think that their loved one is responsible for not fighting against the disease, not collaborate with his therapists (Bolmsjo and Hermeren 2001, Sjostrom, Holmberg and Strang 2002).

PMSA1 (MALE 73, diagnosed 4 years ago. Bedridden, CMSA1 his wife and D his daughter)

I= do you think he cannot walk because he lost his muscular strength or for other reasons?

D= one component is that he didn't want to collaborate with the physiotherapists. (...) we went to meet him, when he was admitted to the rehabilitation clinic, we invited him to go to the physio sessions, but he didn't push his wheelchair. not because he couldn't, because he did not want to.

C= but our GP and our neurologist say this his not his fault, it's due to the disease.

And there are chronic situation where it seems that complications like falls have been accepted by patients and carers as normal occurrence in their life:

PPD4 (Woman, age59 Diagnosed 33 years ago, in deep brain stimulation-DBS- since 1999, totally aphonic, CPD4 her husband)

The patient talks about the good improvement obtained with the DBS, because before she was stuck in the wheel chair, dribbling saliva and dysarthric. She partially recovered in motor symptoms, but never recovered her voice nor the balance:

P= (...), but I fall down.

C= every day she falls down.

P= but they advised me before the operation that I was going to fall.

(...)

P= now I don't have objects falling from my hands anymore, as it happened before the operation.... Now I fall with objects in my hands!

Immobility caused distress and required continuous presence of carers at the patient's bed side, the burden of care increased with the functional impairment (Oliver 2004, Hecht *et al.* 2003)

PALS1 (man, age 48 , bedridden, quadriplegic, totally aphonic, tracheostomized, CALS1 his wife)

C= another physical trouble is that he wants to change his position or move from the bed to the wheelchair, but there's not always someone available for it.

Symptoms related to the movement impairment were also reported as very distressing: In some case the motor symptoms can cause physical pain (Lee *et al.* 2006c):

PPD5 (woman in her 70es, diagnosed 18 years ago, wheelchair bound, many a finalistic movements, spasms and tremors, CPD5 her husband)

C= tremors are bad because she suffers a lot, but she doesn't risk to fall down being blocked. When she has movements she's better because she can walk. But it's more dangerous because of the risk to fall down.

Stiffness sometimes is not controlled even with innovative invasive strategies and can impact on the quality of care and on the burden of the carers (Higginson, I. J. *et al.* 2006a, Ben-Zacharia and Lublin 2001):

PMS7 (man aged 69, diagnosed 25 years ago. Wheelchair- bed bound, anarthric, CMS7 his wife)

C= we had botulinum injections against stiffness. (...) It was 2004 and the neurologist suggested it. They did it in the inner part of the tights.

I= were those the most rigid muscles?

C= yes. Now if I want to bend his legs I have to call my son. I can't do it by myself. In the beginning it looked if it worked, I was able to open easily his legs. Neurologists said it was more helpful for me than for him, because it makes easier to wash him. We repeated it 3 times, after the doctor said it couldn't be repeated anymore.

Movement disorders can be very distressful for both patients and carers and cause confusion about the meaning of the experience. Drugs can be efficient but sometimes side effects force professionals to consider their withdrawal or the medication may lose its efficacy (Calne and Kumar 2003, Clough and Blockley 2004).

PPD3 (man in his 70, diagnosed 27 years ago. Cognitive impairment, admitted in a nursing home for respite, CPD3 his wife)

C= my husband had very awful crisis, where he started trembling and shaking, he had foam to his mouth... he looked as he was in his last minutes...he fell down frequently, hit his back, but luckily had never fractured his bones. This events lasted for about 20-30 minutes, than they passed without any intervention. (...)

C= about 10 years ago they prescribed apomorphine injections. I injected it when he was blocked. He was taking his drugs around the clock and this one if needed. It worked well, but one day the neurologist told me to stop it because it could harm him.

Rigidity or flaccidity can be present in the same patients in different times, depending by external factors or medical complications (Metz, Patten and McGowan 1999, Maloni 2000, Lisak 2001)

PMS3 (Male, age 46, blind because of MS, EDSS 9,5 quadriplegic, almost unable to speak. Fed by PEG, CMS3 his wife)

C=(...) Usually he is very rigid and has clonus. But with the fever he was flaccid and soft.

Subjective distress is caused also by dyskinesias due to the drugs needed to avoid the freezing phases(Clough and Blockley 2004, Chase, Engber and Mouradian 1994, Lee *et al.* 2007)

PPD2 (male 70 years old. Diagnosed 6 years ago.)

I walk very badly. Once I walked better. My hip is aching.

I don't have many tremors. I've never had. I have these dyskinesias. It's a continuous movement, I don't have rest. But not tremors. (...) I can't stop all my

limbs from moving continuously. I dance and can't control it. It's very hard to enter in the car.

3.5.2.3.3.2 Dyspnoea and respiratory troubles

These symptoms were reported in (81,2%) and were particularly prevalent in ALS/MND patients with respiratory impairment (Saleem, Leigh and Higginson 2007).

In the following comments it results, even though NIV can improve these symptoms, this solution can cause other forms of distress and cannot arrest the progression of the symptom (Lyll and Gelinas 2006)

PALS4 (male, 61, quadriplegic, diagnosed 3 years ago, on a wheelchair, cachectic, severely dyspnoeic in NIV)

P= (I feel) great weakness in my legs, tiredness, I can't climb the stairs anymore, and above all, shortness of breath (he says this with a very low voice, sustaining his head with one arm..)

C= breathlessness debilitated him a lot. He couldn't walk because of it more than because of muscle impairment. (...) and this cause great fatigue (...)

C= in the night he use NIV by mask. Now he also use it many hours during the day.

P= the mask is a problem.

C= it tears his skin. Now he's got this pressure sore on his nose. It happened 15 days ago.

Now we have another mask... our lung specialist provided it. But this machine is very sensitive and the alarm starts each time air flows outside the mask. In the night you get crazy for it!

Dyspnoea is present in patients with diagnosis other than ALS/MND too, as shown in this example of shortness of breath episodes for this MS patient (Higginson, I. J. *et al.* 2006a):

PMS1 (Male 53 years old, diagnosed of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):

P=I've got troubles with breathing. They started 3 years ago and I'm using a ventilator to sleep, since then"(...) "I had apnoeas. I spent 3 minutes without breathing.(...) I just couldn't breath, I looked at my wife, but I could do nothing,(..) without breathing. (...) Now, with the mask I can sleep. Before I couldn't sleep. I had many apnoeas and sudden awakenings. I had to get used to it, because it's not comfortable to sleep with this mask: on summer it is worst, it's hot and you sweat a lot; on winter it flows cold air. But at least now I can sleep.

C= Before using the mask we had to run twice to the A&E in the night because he couldn't breath

And in movement disorders (Lee *et al.* 2007) as well as shown below

PPD5 (woman in her 70s, diagnosed 18 years ago, wheelchair bound, many a finalistic movements, spasms and tremors, CPPD5 her husband).

P= this is a very bad disease, because when I'm feeling well... suddenly it starts shaking, trembling (she's trembling while saying this) and it doesn't stop. (....)... I feel something clenching my throat.
I= do you mean shortness of breath?
P= yes.

Respiratory symptoms can be triggered by the frequent and recurrent respiratory tract infections. These events represent a frequent cause of death in neurodegenerative disorders and are among the criteria proposed to be admitted to hospice care (Clough and Blockley 2004, The-Gold-Standards-Framework 2008, Bronnum-Hansen, Koch-Henriksen and Stenager 2004). The following experience about these issues is reported:

PMS3 (Male, age 46, blind because of MS, EDSS 9,5 quadriplegic, almost unable to speak. Fed by PEG, CMS3 his wife)
C=In 2003 he had an aspiration pneumonia. Before that I had to spoon feed him because he couldn't use his hands anymore. But then things went worst and he started choking when he had to swallow both solids or liquids. One day he went in coma and was admitted to the hospital where diagnosed this aspiration pneumonia.
Since then he's fed by PEG.

3.5.2.3.3.3 Pain

Pain syndromes, painful muscular spasms or cramps appeared in 18 interviews (81,3%). Some participants reported pain syndromes spontaneously (Lee *et al.* 2006c, Thomas and MacMahon 2004b):

PPD5 (woman in her 70es, diagnosed 18 years ago, wheelchair bound, experiencing dyskinesia, muscular spasms and tremors, CPD5 her husband)
P= I have pain.(...) my goodness it's so painful!
C= yes, when she trembles so much (....) she doesn't take any pain killer, when tremble stops she's quickly better.

Pain is a common sequel of complication of the disease, like fractures following falls (Lee *et al.* 2006c, Calne and Kumar 2003)

PMSA1 (man, age73,diagnosed 4 years ago. Bedridden, unable to communicate)
C= he has pain. His femur fracture was complicated. The orthopaedic surgeon said he was going to be in pain for all his life. He seemed to want d him to be in pain for a long time, as it would have meant a long survival.
With tramadol he's pain is relieved. Without it his legs are retracted and a long and hard string appears at the back of his thighs. We tried to leave him without the drug, but the pain and contractures reappeared

Pain is not immediately recognised by the participants as a problem related to the disease. It take some time to think and report the symptom which is present even though often not directly caused by the neurodegenerative condition (Maloni 2000, Higginson, I. J. *et al.* 2006a):

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PMS4 (woman, age 69, diagnosed 42 years ago. She lives alone, cared for by paid carers, spends alone most of the day and the night)

P= I don't have pain. No, what am I saying? I have leg pain, a lot. But I think they're age related or due to rheumatisms or arthrosis (...) I don't have cramps, but spasms and inexhaustible clonus at my right leg.

In the following citation the first answer is negative about pain, then the patient quickly realise that she is experiencing pain, even though not directly caused by the disease, but by the immobility and the device used to be moved (Borasio and Oliver 2006)

PALS7, (woman, age 60, diagnosed 12 years ago, quadriplegic)

I= are you in pain?

P= no, well yes, my shoulder, but it depends on positions. (...) I also have a chronic inflammation at this shoulder. Than it gets jammed in the slinging of the hoist. So using it 3-4 times a day... it hurts.

Pain often was reported as skeletal or muscular and often not treated by physicians who fear complications (O'Brien, T. 1993b):

PALS6 (male, 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic. Communicates with a board giving short answers)

I= are you in pain

P= yes, postural.

I= what if you change position, does it improve?

P= yes

I= do you take pain killers?

P= no

C= I talked to the doctor, but he said no.

P= cannabis?

I= does PHT help for postural pain?

P= no.

Pain can often have neuropathic features in MS (Maloni 2000, Metz, Patten and McGowan 1999)

PMS3 (male 46 years old, blind because of MS, EDSS 9,5 quadriplegic, CMS3 his wife)

C= he has spasms and muscular clonus. He's taking baclofen and dantrolene sodium. We put it in PEG. (...) if he doesn't take it his stiffness increase a lot.

C= in the night he wakes up and wants to be turned around, or asks me to stretch his limbs.

C= he has pain. Above all trigeminal pain. He was successfully taking gabapentin, but now they changed it because he should pay for it (the drug is reimbursed by the Italian NHS for chronic pain caused by cancer only). So they're giving him Carbamazepine, but doses are too low yet.

In movement disorders pain was seen as well

PPD1 (male 80 years old PD diagnosed 9 years ago)

C= he had spasms from the base of the neck to the face. They're planning a MNR for it.

P= it was very painful!

I= now is it gone?

P= yesterday evening again. But they're doing intravenous infusion for it.

C= and drops too. Alprazolam 12-15 drops, but after 4-5 hours it comes back again.

3.5.2.3.3.4 Swallowing troubles

Choking when taking food or liquids, saliva aspiration and nutritional issues were reported in 21/22 (95.5%) interviews. 9 patients were receiving feeding and hydration via a PEG.

Swallowing troubles in general have a very bad impact on the carers, forcing them to spend a great deal of time with the patient and changing their habits in food preparation. Often episodes of food aspiration led to aspiration pneumonia with hospitalization. The presence of these problems has been identified as indicators for referral to palliative care (Brown and Sutton 2009)

PMS3 (Male, age 46, blind, EDSS 9.5 quadriplegic, CMS3 his wife)

C= we can't leave him alone. Today is a good day for him, but often he has sudden worsening and requires constant attendance. For instance he can't swallow saliva, so frequently he chokes and we have to aspirate his mouth (...)
One day he went in coma and was admitted to the hospital where diagnosed this aspiration pneumonia.

Since then he's fed by PEG.

Swallowing difficulties can worsen suddenly causing great concern and, eventually respiratory tract infections

PMS7 (Male, 69, diagnosed 25 years ago. Wheelchair bound, anarthric) CSM3 his wife.

C=(...) swallowing troubles began. It was years since he had these problems, but at that time he couldn't swallow at all. He put food in his mouth and spit it out.(...) We waited until the spring of 2004 where he was even worst. He had an appointment in the nutrition clinic for the end of August, but I asked them to anticipate it otherwise he wasn't going to survive so far. We had been informed about PEG for a long time. They admitted him to place it and during this hospitalization he got a Pseudomonas aeruginosa pneumonia with fever, secretions, breathlessness. We were persuaded that he wasn't going to come back home. Well he came back with a PEG

Increased salivation or swallowing difficulties can impact adversely on patient's quality of life and cause important changes in other function too, such as communication, loss of pleasure in eating The fears and changes may lead to the discussion and acceptance of end of life decisions (Miller *et al.* 1999, Hardiman 2000, Heffernan *et al.* 2006) The decision to accept a tracheostomy is well described in the following citation:

PALS7 (woman, 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24\24 by nasal mask that consent her to talk, CALS7 her husband)

C= salivation is increasing. Salivation is increasing a lot.

I= swallowing?

P= as well.. in this period there's an acceleration (...)

I= what about feeding?

P= I eat everything blended. Since two month I have to do like that. But it's tiring, so I can't eat as much as I'd like to. It starts a tachycardia and I feel shattered. So I introduced snacks but certainly I eat less than I used to.

I= anyone told about PEG?

C= a long time ago they talked us about it.

P= let me say.. according to a medical indication I required a tracheostomy last year. But because I can speak they wanted me to decide about it trying to preserve my best quality of life. Now I'm speaking badly and having troubles in feeding, so if they'll do PEG and Tracheo in one time... (...) but it's a recent issue because until one month ago it was better.

Another experience in a recently diagnosed ALS/MND patient whose relatives were struggling to realize the various losses that had developed over a few months.

PALS9 (male, 77 diagnosed 8 months ago, DALS9, his daughter and CALS9 his wife)

I= how long does he have the PEG?

D= since March. They placed it because he lost a lot of weight, he didn't eat anymore.

C= now he eats very little. He eats something each meal, but very little.

P= it's due to a lack of appetite.

I= doesn't it go the wrong way?

C= yes, swallowing is difficult. He's having jelly water. Water goes the wrong way.

P= jelly water also requires some time to be swallowed....

Movement disorders often cause troubles in swallowing as reported below by the elderly husband of this PD patient (Saleem, Leigh and Higginson 2007)

PPD5 (woman in her 70s, diagnosed 18 years ago, wheelchair bound, many a finalistic movements, spasms and tremor, CPD5 her husband)

C= swallowing is another trouble. I have to whirl everything I cook for her. But she complains that everything has the same flavour (...) she's still eating by her self, but it happens that I have to spoon feed her. The bad aspect of this disease is that it changes quickly.

3.5.2.3.3.5 Speech related problems

Communication problems, with intensity from mild to severe with total impairment and loss of speech, were detected in 14/22 interviews. Four patients were using aids to communicate: 2 used alphanumerical boards read by the caregiver, one used both a

board and an electronic device and one a computer with electronic computerized software.

The ALS tracheostomized patients had lost their capability to speak and all needed electronic devices to improve their communication. They reported this need as probably the most impacting their actual quality of life (QoL):

PALS2 (male 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic and cognitively not impaired, and CALS2 his wife)

C= a big discomfort is the impossibility to speak. We use this tab that we saw on the Discovery Channel. It's quicker than the other communicators and electronic devices that we had the chance to use. The only device that really helps and works is MY TOBII package. But it cost 21.000 euros.

Anyway, communication troubles are one of the worst aspects of this disease.

In another experience of a patient in similar clinical conditions the wife clearly states that communication using alphanumerical tabs is difficult and the results affected by the ability of the carer to use it. This ultimately causes a restriction in social activity (Scott and McPhee 2006) because friends or other visitors cannot communicate with the patient if an educated carer is not present and help facilitate communication:

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized. He communicates using an alpha-numerical table, and CALS1 his wife)

C= (...) now we use this table. We learnt it because we had to. They told him to look at the letters and that we had to look where he was looking at. We use this instead of other communicators because this is quicker. The paid carer and I were able to communicate quickly. If others come, we must act as an intermediate.

In other groups of patients loss of speech develops more gradually but this always seem to led to a negative impact on relationships and QoL:

PMSA1 (man, age 73, diagnosed 4 years ago. Bedridden and severely dysarthric, CMSA1 his wife)

C= He lost his speech one month ago. Since then it is really difficult to communicate with him. Today he has fever and it is totally impossible to know how he feels.

Few patients with mild- moderate communication impairment wanted to discuss their feelings about it and the connection with other losses

PALS9 (male, 77 diagnosed 8 months ago)

P= when friends come home to visit me I can't use the mask because it prevents me from speaking. So I can't speak and if I speak a little more than I get breathlessness.

Or

PPD4 (Woman, age 59 Diagnosed 33 years ago. In deep brain stimulation since 1999, totally aphonic had a good recover of motor functions, but no improvement of the voice that is barely perceivable), CPD4 her husband:

C= she's aphonic. That's not due to her throat (...) it's because of the brain damage.

P= it's a long a time ago since I'm like this. I tried speech and language therapy but... (it didn't work at all)

C= there are periods, during the day, that her voice is a bit better and other that you can't even hear it.

3.5.2.3.3.6 Oral symptoms

Mouth symptoms like dry mouth or drooling and issues related to difficulties in managing secretions were reported in 10/22 interviews.

It was often referred as a disabling condition and participants were not very satisfied about the effectiveness of medications. Sometimes it was difficult to distinguish between drooling and the upper respiratory tracts secretions, but both had a bad impact on daily management of the patient:

PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric, fed via PEG, cognitively not impaired, communicates via alpha numerical table) CALS8 his wife:

C= he has drooling of saliva, but only during the day. It's not a problem in the night. he's taking amitriptyline, 10 drops in the morning and 2 in the evening. Usually he doesn't have big amounts of saliva a part for when he laughs or when he has to use a lot his facial expressions.

P= I have a bad sensation in my mouth, like thick secretions.

This MS patient's mother was very confused about the therapy and used an aggressive approach, like stimulating his vomit with the aspirator, to clear dribbling of saliva and respiratory secretion. During the interview one episode was caught on video and the patient looked as if he was really suffering during this procedure. They did not appear to be well informed about the palliative remedies to tackle this symptom and her GP had not been helpful with the prescriptions:

PMS5 (male, 49 years old, diagnosed 24 years ago. EDSS 9,5. patient unable to communicate) CMS5 his mother:

C= he has this bad drooling. I must stimulate his cough with the aspirator. Now he's breathing well, but it's not always like that. Often he has rattle with thick secretions. So I have to stimulate his cough and vomit. But after he feels better. He's not taking drugs for drooling because there aren't any. A neurologist suggested me atropine but my GP didn't want to prescribe it because he thinks it's dangerous for his heart . So he gave us an homeopathic preparation of this drug.

(.....) I heard that this antidepressant (amitriptyline) can be useful for drooling, but how can you give an antidepressant to a person in his condition?

This carer instead followed the neurologist's prescription, but decided to stop the drug because it did not provide the expected result (without asking the doctor for other drugs or change in dosage)

PMS7 (Male, 69, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired) CMS7 his wife
I= Is drooling a problem?
C= yes, since a long time. I aspirate it. In 2005 the neurologist prescribed him amitriptyline. He took 5-7 drop twice a day but it wasn't helpful, so we stopped it.

Other patients reported unsatisfactory treatment of these symptoms and talked about the bad impact on their QoL due to the side effects of the drugs:

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24\24 by nasal mask)
P= (...) also salivation is increasing. Salivation is increasing a lot.(...)
I= did you use drugs to dry secretions?
P= yes, I took amitriptyline, but 2 drops were enough to asleep me for the whole afternoon. A neurologist suggested me to try with an antiparkinson drug (Bornaprina) that should work in low doses. I tried it for two days, but last night it caused me a bradycardia. So I stopped it.
However all these drugs cause saliva to become more sticky. I heard about botulinum injections that reduce salivation for 3-4 months. But doctors don't concur on it.
Now we have the aspirator at home.

This patient wanted to say these words using the alphanumerical table clearly trying to make humour about the symptom. His wife, who works and does not spend the day with him, is not keen on medication for the symptom, meanwhile the patient's mother, who spends the day with the patient and has to aspirate him often, is of a different opinion.

PALS2 (male 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic and cognitively not impaired) and CALS2 his wife:
P= a big physical problem is that I dribble down like a slowcoach or a camel.
C= they prescribed him amitriptyline but I'm not happy being antidepressant. He takes 5 drops a day and doesn't work.
This is a disagreement between us. I think that saliva is normal and it's a side effect of the disease. He and his mother claim that it decreased with the drops.

3.5.2.3.3.7 Urinary troubles

13 patients out of 22 (59%) had some form of bladder dysfunction. Those could be both incontinence or storage impairment (MacLeod 2004). Devices such as incontinence pads, internal or external catheters were widely used. ALS patients tend to be less likely to suffer these issues, but one patient reported this problem:

Incontinence can be overcome using external catheters that are effective in males, even though not always well accepted. In the following

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PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):

P= I've been using an external catheter since 3 years. My wife changes it for me.

I= have you ever had retention problems?

P= no!

C= (..)He has bladder problems in storing urine.

P= I use the catheter only during the day, in the night my wife puts me on a pad.

Another MS patient has the opposite problem and his wife had to learn how to do extemporary catheterizations during the day.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

C=He needs to be catheterised twice a day because he could not pass urine. I do that. In the morning before going to work and in the evening. Now he doesn't urinate a lot since he has swallowing difficulties and drinks very little .

In movement disorders urinary problems can be related to motor symptoms. In this case the patient focus his answer to the improvement experienced after prostatic surgery, but his wife highlights how when the patient is in freezing or when in the night he is out of the drug effect urinary problems appear.

PPD2 (male, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias) CPD2 his wife

I= do you have urinary problems?

P= I was operated at my prostate and recovered well. Before I had to use a catheter for 5 months and was it terrible(...) now, if I can reach the toilet is quiet ok if I can hit the centre of the loo (speaking respectfully!).

C= when he is blocked he needs a container to urinate because he can not reach the toilet. No, he cannot do it when he is blocked.

P= yes, and this is due to the neurological progression of this disease and the limitation of the drugs' efficacy. Once I could do it, now it cost me a lot.

Recurrent urinary tract infections affected some participants:

PPD1 (male, age 80, cognitively slow but able to understand and severely disabled in his movements. During the interview was admitted in the neurological ward for symptom control) CPD1 his wife.

C= he use incontinence pads in the night because of his incontinence. He's been using it for two years.(...) during the day he wears the pads, but can control urine. Sometimes he has urinary tract infections. We had to come to the hospital because he had urinary retention due to his prostate.

I= does sometimes hurts when you have to urinate?

P= yes, it does.

C= this is one of the reasons why we are here in the ward. He was a bit blocked for his prostate.

I= did they apply a catheter?

C= yes, for 10 days. But it is not just the prostate, it is also due to his muscular blocks that prevents him from watering well, it is for his stiffness.

In this MS patient a complicated urinary tract infection nearly caused death. Fever made him unconscious or barely intelligible. His wife testifies how challenging was to obtain a diagnosis and a correct treatment after they had been told by the A&E of the local hospital that he was dying and nothing could be done.

PMS3 (male, age 46, blind, quadriplegic, muscular spasticity, fed by PEG), CMS3 his wife.

C= now he has a catheter life. Before I did 2 catheterization per day. But this caused infections in the urinary tract. Now we do periodic Uri analysis with cultures and our GP prescribes antibiotics. Some months ago he had a sepsis and almost died because of it. I never saw him so bad. (...) When he has fever it's impossible to understand what he wants or says. We spent horrific days when he was in fever ad I called the emergency, they sent an ambulance and we arrived at the A&E. A doctor saw my husband and told me that he was dying, they could not do anything for him and that the hospital was not appropriate for him. I did not know what to do, where to go. We went back home and I called the GP begging him to come home and visit him. He came but then told me that if at the a patient so ill was not to stay at home so prescribed an immediate admission for the hospital, he called the ambulance and we were taken back to the A&E. as soon as we arrived the same doctor asked me what I was doing there again! They told they were not going to do any investigation or treatments and just put him in a dark room at the end of the ward. Luckily a nephew of mine is a physician working in a private clinic and he interested himself in our case. After 40 days we brought him to the private clinic. Fortunately there they found out that he had nephrolitiasis with perinephric abscess. They put a double J catheter in his ureter and placed the catheter life.

3.5.2.3.3.8 Bowel troubles

17 out of 22 (72.7%) patients reported intestinal problems.

Constipation was the most frequent problem and it was present in all the diagnostic groups:

PMS4(71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P = bowels... that's a problem. Because I eat very little, I tend to swallow dry food, I don't move and above all, I don't have the chance to go to the toilet by myself.... Nurses have to do rectal explorations twice a week. That is scheduled on Monday and Thursday. The problem is when I feel a stimulus on Tuesday... I have to pay other people to help me.

Constipation is a typical symptom in PD and often is complicated by the specific treatments.

PPD5 (woman age 78, cared for by her husband -CPD5- at home)

C = her bowel doesn't work at all. She's constipated. Now she taking Tamarine jam. It works quiet well, but not perfect.

Another situation, in this case is an ALS patient where intestinal symptoms are not correctly managed as suggested by the “stepped care approach to care” whereby patients receive dietary advice first, with oral laxatives prescribed later if necessary (Thomas and MacMahon 2004b)

CALS5 is the wife of PALS5, a totally dependent gentleman wheelchair bound and severely dyspnoeic
C= he's stypitic. We do enemas to him, the small ones... 2-3 per day. If a cork happens I have to do 5.

Another trouble is loss of control of the sphincters, above all in MS patients:

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.
P= I use jellified water. (...) it's very useful. It helps me for the bowel also.
C= (...) but I have to help him manually to evacuate the bowels.

Or

CMS6 wife of PMS6 a 53 years old gentleman EDSS 9, fed by PEG, almost totally dependent in the ADL
C= he has his pad on for the whole day. He lost his stimuli so he does everything in it. We have to change it in the night as well, we have to put the alarm clock. For instance last night I forgot to change it and when this morning the home assistants came to wash him he was soaked in the excrements.

3.5.2.3.3.9 Sleep disturbance

14\22 (64%) interviews highlighted how patients are disturbed during the night and this was usually be due to physical problems, anxiety, fears, depression, agitation or other psychiatric symptoms.

Some referred that nights were disturbed for physical problems (Saleem, Leigh and Higginson 2007)

PALS9 (male, age 77, diagnosed 8 months ago, in NIV, CALS9 his wife)
I= do you sleep well?
C= he wakes up 2-3 times per night to urinate. He needs a person who removes his mask and replace it afterwards, he can't do it by himself.
P= I take lorazepam to sleep.

When very high disability limit almost any movement physical problems like gastro oesophageal reflux induced by enteral nutrition via PEG can impact on the quality of sleep

PMS5 (male, 49 years old, diagnosed 24 years ago. EDSS 9,5, CMS5 his mother)
C=in the night he has to sleep almost seated because of reflux. He had food in his mouth, it happened 2-3 times per week.

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Sleep quality can be altered for the loss of control of the tongue and risk of suffocation (Borasio, Voltz and Miller 2001)

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24 hours a day)

P= when I'm in bed I feel like my tongue slipping behind and I fear about choking. For this reason I sleep in a semi seated position.

Non motor symptoms can be mirrored by night disturbances in movement disorders (Chen, Trombetta and Fernandez 2008)

PPD2 (male 70 years old. Diagnosed 6 years ago.

C= since December 2006 his conditions are worst. He can't sleep in the night. Drugs aren't working anymore, even if they changed it(...)

C= he arrived home from the hospital and couldn't sleep anymore. They quit all his drugs because of glaucoma. He just didn't sleep at all(...) he had hallucinations.

P= yes, I saw things like a tank with a stick inside and it looked to me like a person. (...) or tiles that began to fly...

Participants reported they couldn't sleep well because of psychological sufferance and often sedative drugs had to be taken in order to allow them to have a proper rest (Oliver 1998, Mitsumoto *et al.* 2005).

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24/24)

P= I use a benzodiazepine to control anxiety (alprazolam) and an antidepressant. (Elopram). It's been some years ago since I started those drugs. I can't say if they work or not. I've always been a positive person, but in times of troubles they are of help.

And this can impact on their carers quality of sleep, particularly if the patient is tracheostomized requiring continuous attention even in the night hours (Mockford, Jenkinson and Fitzpatrick 2006).

PALS6 (male,age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

C= He takes his drops of anxiolytic to sleeps. He has frequent awakenings, he doesn't sleep continuously. He wakes up because he has to be aspirated and turn in the bed. Some night are quiet, others are not. I sleep in the double bed with him so I feel when he needs me

I= so neither of you sleep.

C= no. and it seems funny, but if I'm relaxed he doesn't sleep, while when he's calm I can't sleep

Even though the role of benzodiazepines and other hypnotic have been questioned in palliative care (Hirst and Sloan 2002) sometimes they can have a positive impact on patients and their families quality of sleep as shown by the following two citations

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PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic)

C= to sleep he needs 20-25 drops of lorazepam. Compared to the early years it's better. Before it was a tragedy

One patient refused to take drugs, even if he could take advantage from it, because he feared the impact of it on his conditions.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.)

I= do you sleep well?

C= well, not so well...

I= do you take drugs to sleep?

C= no, no. all these substances act on the nervous system...

3.5.2.3.3.10 Skin troubles

In almost half of the interviews (10/22, 45.5%) skin troubles were reported

Bed sores in patients with very limited movements. Patients with movement disorders are subjected to these problems above all when they are admitted in hospital facilities where they are more exposed to medical complications (Clough and Blockley 2004):

CPD3 is the wife of PPD3 a gentleman in his 70, diagnosed 27 years ago. Cognitive impairment, Wheelchair bound, living in a long term facility)

C= last year he was admitted to a clinic and after few days the nurses told me that they had to leave him in bed because he was troublesome, always moving, too demanding. They said that he refused physiotherapy, that he didn't want to leave the bed. So I asked the doctor and he told me he was taking a drug that our neurologist suspended months before because it blocked his legs. I said him to stop it, but he refused. So I took my husband back home.

It caused him a bad pressure sore at his back. It has not healed yet. It was due to both the facts that they left him in bed and to the drug they gave him

Skin irritation due to devices needed for reasons other than pressure. In the following case it was a device used to control incontinence to cause skin ulcerations

PMS5 (male, 49 years old, diagnosed 24 years ago. EDSS 9,5. patient unable to communicate), CMS5 his mother

C= he use the urocondom for incontinence. His carer changes it every morning. Unfortunately it sometimes cause pressure sores on his penis.

In ALS/MND also skin problems can occur and again immobility and co morbidity are common cause.

CALS8, wife of PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric)

C= he has a mycosis under his armpits and this cause itching. He suffer of psoriasis too (...) he had bedsores at his back. It was due to a diarrhoea

syndrome that he had on spring. I cured it with creams. Now I still put it because his skin remained very sensitive.

Irritations and painful sores are commonly related to the use of the masks for non invasive ventilation as testified by many participants, here an example:

PALS7 woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24/24 by nasal mask

I= any nasal problem?

P= yes, I had pressure sores inside, but now the body adapted and it made a corn. I also had rhinitis problems and I couldn't assuage it. So an otorhinolaryngologist came and gave me a local therapy. It worked for 1 year, but now it's starting again,

4.5.2.3.3.11 Other symptoms

Other disturbing symptoms, such as fever, visual loss, fatigue, were reported as severe in 13/22 interviews (59%):

Most frequent were recurrent fever and visual loss:

CPD4 is the husband of PPD5 (woman in her 70es, diagnosed 18 years ago, wheelchair bound, many a finalistic movements, spasms and tremors)

C= fever is common with her. Not long ago she had fever. We went to the hospital and they diagnosed a bronchitis. She had 38.5°C. I didn't understand why, but she was trembling the whole day. Then we measured her temperature and there it was.(...) She's almost blind. One eye isn't working at all, at the other she has a bad cataract.

CMS6 is the wife of PMS6 (man 53, diagnosed 16 years ago wheelchair bound, moves with many difficulties his upper limbs)

C= he's almost completely blind because of MS. He can't read nor watch TV. He likes music, so he puts on his I-pod.

Visual problems can be side effects of the therapy as comes out in the following interview of a PD patient who had to be operated in emergency for an acute glaucoma due to L-Dopa.

PPD2 (male 70 years old. Diagnosed 6 years ago), CPD2 his wife

P= they had to operate my eye. They did an iridectomy old style. They said it wasn't enough just change the crystalline lens.

C= he was taking Madopar. On the evening of the 6th of December 2004 he began complaining of a headache. During the night it got worst and in the next morning his eye lost sight.

I= do you remember which eye?

P= the right one, my head felt like if it was blowing out.

C= so we went to the hospital (...) and they treated him with laser but he was not recovering his sight. So they decided to admit him and gave us the diagnosis of acute glaucoma.

P= eye pressure was 70! (...) I read on the drug leaflet that Madopar could cause acute glaucoma, the worst of all

3.5.2.3.3.12 End of life themes

These issues arose in the interviews while participants were talking about physical symptoms, including comments:

CALS1 is the wife of PALS1 a gentleman 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized)

C= he wasn't eating anymore. He weighed 35 Kgs because he had swallowing problems so he wasn't eating nor drinking enough. We went to the hospital because he was not breathing anymore. He was using NIV by mask, but he didn't care about it. When doctor advised him that a PEG was needed he did not consent to it. He was determinate to end his life.

But when the respiratory arrest occurred and we ran to the hospital, everything changed. He was admitted to the emergency department where he had his PEG and the tracheostomy. Since that moment a new life has started.

I= did you make that choice?

P= yes!

PALS5 (male in his 70ies, quadriplegic, aphonic, on a wheelchair. he tries to communicate using a computer keyboard. Very slow. He is a retired doctor), CALS5 is his wife

C= he's firmly contrary to tracheostomy. He prepared a living will stating this.

P= (writes) show them the will. (then, addressing to the interviewer who previously discussed the issue of withdrawing treatments) I agree with your position that withdrawing invasive ventilation is not euthanasia.

CALS8 is the wife of PALS8 (man in his 70s, diagnosed 9 years ago wheelchair bound, totally anarthric, fed via PEG, cognitively not impaired, communicates via alpha numerical table)

C= PEG was placed 3 years ago. (...) one day, it was Saturday, he had a persistent cough. He was already using a thickener for the water because he had problems with swallowing. (...) he refused to go to the hospital because he feared that they'll put him a tracheostomy. (...) in the evening I was very scared because of the incoming night. We called our sons to convince him to go to the hospital but he didn't consent to it. (...)

PMS4 (female, 69 years old, diagnosed 42 years ago. She lives alone, cared for by paid carers, spends alone most of the day and the night. EDSS =8 wheelchair bound)

P=My respiratory tests were totally unsatisfactory, but I refused mechanical ventilators. I've had enough, I lived my life.

3.5.2.3.3.13 Summary

From the content analysis of the physical problems raised by the participants of the interviews eleven groups of symptoms were identified: Movement problems, breathlessness and respiratory troubles, pain, swallowing issues, speech related

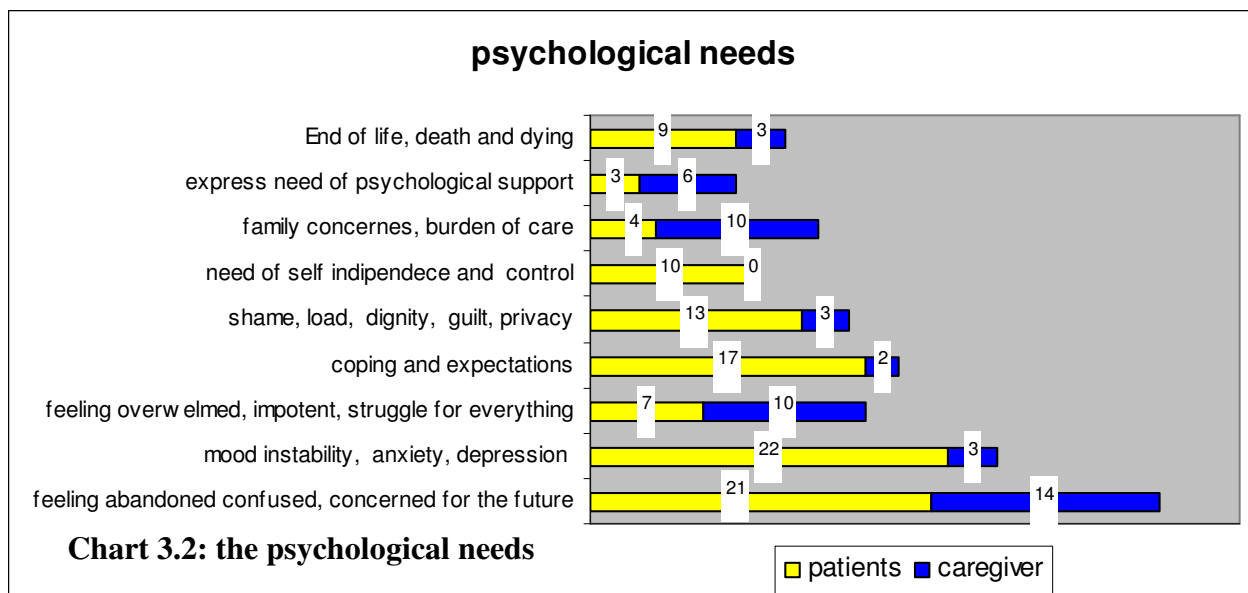
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problems, oral symptoms, urinary problems, bowel troubles, sleep disturbance, skin troubles and other symptoms.

Although some symptoms were more present in specific conditions, like respiratory problems in ALS/MND or urinary troubles in MS, most of the reported physical problems were appeared across all the conditions. Symptoms were reported directly by the patients and sometimes by the carers. Physical issues accompanied the patients throughout all the course of the disease but were more intense and prevalent in the advanced stages. Physical problems were cause of reflections about end of life choices in some cases.

3.5.2.3.4 The content analysis of the psychological needs

In chart 3.2 the content analysis of the psychological needs that emerged from the interviews are shown. Because psychological themes could be related to both patients and carers needs data are presented separately for the two typologies of participants:



In table 3.4 psychological needs are listed in figures and percentage

PSYCHOLOGICAL NEEDS quotes in the 22 interviews	Patients* (%)	Caregivers** (%)
feeling abandoned, confused, concerned for the future	21 (95)	14 (67)
mood instability, rage, fears, anxiety, depression	22 (100)	3 (14)
feeling overwhelmed, impotent, struggle for everything	7 (32)	10 (48)
coping and expectations	17 (77)	2 (10)
shame, load, dignity, guilt, privacy	13 (59)	3 (14)
need of self independence and control	10 (45)	-
family concerns, burden of care	4 (18)	10 (48)
express need of psychological support	3 (13)	6 (29)
End of life, death and dying	9 (41)	3 (14)

Table 3.4: the psychological needs

Footnotes:

*needs related to patients were expressed by patients for themselves or by caregivers on behalf of the patient when he or she was not able to communicate

** % on caregivers were calculate on N=21 because one patient living alone did not have any caregiver participating at the interview

3.5.2.3.5 The psychological needs detailed analysis

In this session data about the psychological issues that came out from the 22 interviews will be shown. Symbols and short forms are the same adopted for the physical needs. Quotes can be from patients or carers or both, but it will be specified who they are related to, in order to identify if the displayed need is a patients' or carers' need.

3.5.2.3.5.1 Feeling abandoned, confused, concerned for the future

This group of needs was very frequently reported by participants:

In all interviews these themes appeared, sometimes directly from patients' voice, in other occasions introduced by the carers and could be related to all participants.

Usually there was a strong agreement between patients and carers about these needs. In the next citation both participants at the same interviews talked about feeling abandoned by friends and relatives:

PMS1 (Male 53 years old, diagnose of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife)

P= "we would need a magic wand!"

I= why do you say so?

P= " to solve everything! Because we are abandoned.

C=The disease takes away people, relatives and friends. It's also our fault. We create barriers. Maybe we have too many expectations on them.

Indifference and isolation are used as synonymous and relate to the sense of abandonment that is the opposite of the need of love and belongings described by Maslow (Maslow, A 1970)

PMS2 (male, age 43, EDSS 9, quadriplegic) CMS2 his wife

P= mine is one of those cases where people say: " If you need a hand call me" but they think " if you don't call it's better. And if you call, well do it only once."

C= yes, there's a lot of indifference.

I= do you feel like a sense of isolation, void?

C= yes, but you learn to let it slide over.

P= you don't have to think about it, because if you do it...

Loss of friends is seen by participants as an example of abandonment. Those persons who used to have plenty of friends and visitors try to explain the reasons for this loss. In the following example the carer talked on behalf of the patient who couldn't speak, but clearly agreed with his wife position:

PALS1 (man, 48 years old, bedridden, quadriplegic, totally aphonic, tracheostomized. Fully aware of his situation. He communicates using an alpha-numerical table). Caregiver CALS1, his wife.

C= we are isolated. We lost many friends. He used to have so many friends and people who came to see him. But that is because he was a town councillor and they needed him. Since he's ill they disappeared. Few of them helped us when he was in the hospital, but when we came home they vanished. We looked for them,

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but the plea is always the same: “ we can’t see him like this” or “we are too sorry for him”.

The same carer continues complaining about the fact that because they seem coping very well with the disability and the organization of care, friends, professionals, social assistants abandoned them thinking that they do not have unmet needs.

C= I want to underline that you see us like this (e.g. well organized) for a series of lucky circumstances. That’s because he devoted himself. He didn’t lose his heart. (...) But you have to know that it’s not easy at all to live in our condition, this situation is slightly heavy to carry on.

But this, somehow, plays against our interests. You see people think that as long as we are well organized and independent we don’t have needs. They tend to leave us in a corner. But you can’t always wait until last minute to obtain what you need. Help is needed when it’s useful, not when nothing can be done anymore.

This MND/ALS patients’ wife describe the frustration felt because friends got lost due to the communication impairment, and how in a first stage it was the patient who turned away his visitors because he felt ashamed of his condition

PALS2 (male 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic and cognitively not impaired) and his caregiver (wife) CALS2. The patient communicates through an alphanumeric table with his wife and mother, the only family component able to use it.

C= the problem with the use of the tab to communicate are outsiders. Hardly people coming here want to try. Maybe friend are worried to mistake. You exhort them, but they don’t want. At the end you isolate yourself because people comes once and don’t comes back because feel embarrassed(...)

C= friends are very few.

P= you can count them on the tips of one’s fingers.

C= we are isolated.

I= aren’t you a cause of it?

C= not now. In early years yes, because he was ashamed of his condition.

Another example of abandonment from friends and families as well

PMS6 (male 53, diagnosed 16 years ago wheelchair bound, moves with many difficulties his upper limbs, drinks using a feeding bottle. He is cognitively not impaired, but he looks like if he were living in a dream) and his wife CMS6

C= we are isolated. Many of the things we used to do before we had to leave it. We went out with friends, we had to renounce to it. Friends and relatives don’t come to visit him. I think it’s because of his drooling, or because he lost his urine from the catheter... it can be disgusting (...)

C= even with his family we had problems. His mother didn’t want to come here and spend sometime with him. They put him aside. They do many things for his sister who’s healthy and has got a child, but nothing for him.

Participants felt abandoned by the professional carers as well. Following some example of these experiences are displayed.

CMS3, wife of PMS3 (Male 46 years old, blind because of MS, EDSS 9.5 quadriplegic, almost unable to speak. Fed by PEG). She is talking about their experience with the GP who did not want to help them at home and rejected the patient after he was discharged from the hospital:

C= (..) I called his GP. I was really angry. He answered me:- There's nothing I can do. It's not useful that I come and visit him. He's just coming from the hospital, if they weren't able to do something for him there...- (and added) - It's up to you to convince yourself that your husband's conditions are very serious. You can't keep him at home. You have to take him in a place, you can't manage him at home.- This is what they told in hospital too, but I wasn't convinced.

(...)

C= (after her GP rejected them as patients): Goodness me! What if we won't find another family doctor! I came home really disconsolate.(...) one thing is being helped by supportive persons, another is being alone and feeling abandoned. (...) when you're left alone sometimes you can't be rational and objective enough to manage him.

In this case the wife of this PD patient in really bad conditions admitted in a nursing home for respite care complains about the neurologist's behaviour

PPD3 (male in his 70, diagnosed 27 years ago. Cognitive impairment. Patient able to consent to the interview, but participate very little. Continuously shaking his limbs and constantly moaning. Sometimes he cries. Wheelchair bound.) and his wife caregiver, CPD3. The patient is staying in a nursing home

C= I'm really angry with the neurologist that has always cared for him. I expected him to come here and visit him because he had always followed his situation, so once (...) he should have come to see how he is going on. (..) If you need an help there is no-one available!

Another cause of abandonment can be the disease itself. This MS patient reports what happened to her marriage after the diagnosis

PMS4 (woman, 69 years old, diagnosed 42 years ago. She lives alone, cared for by paid carers, spends alone most of the day and the night. Wheelchair bound, she can move, with plenty of difficulties, her right hand. Left hemiplegic, left amaurosis)

P= 10 years after diagnosis and 15 after our wedding my husband left me. We had a 14 years old son. He left me alone with my mother saying that he didn't want to be my nurse anymore. Later we divorced.

Carers can fear to abandon their loved ones even when a respite admission has been carefully planned. This carer describes her feelings when she had to leave the patient in a specialized facility for rehabilitation and respite

PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired) CMS 7 his wife.

C= the second respite care admission at Piancavallo was the most difficult. We came back home earlier than planned because I couldn't leave him there alone

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(...) he wasn't able to speak, to move and on that time he was losing his hearing. He was isolated, how could I leave him alone?

Another cause of suffering is confusion about what is happening that is related to the difficulties to cope with the disease and concern for the future. This makes carers and patients life very hard to organize.

PMS2, (man, 43 years old, married, with a 10 years old daughter, quadriplegic) and CMS2 his wife.

C= when we had the diagnosis we were 8 months far from our wedding. We were confused, we didn't know what to expect from it (the disease). Personally I've never thought to abandon him. There was love (and there's still), so we went on.

Confusion can be generated by the therapeutic options as well. Sometimes advance directives (not legally binding in Italy) are not adhered to and this can be a cause of confusion and regret

PMS5 (man, 49 years old, diagnosed 24 years ago, unable to communicate, legally unable to make decisions his mother is his legal guardian. The patient seems favourable to the interview. Wheelchair bound, fed by PEG, several gags for his dribbling.) CMS5 is his mother.

C= when he was admitted for high fever and choking, they suspected an aspiration pneumonia, we had a visit from a psychologist. They came to obtain is consent to do the PEG.

I= was he contrary to that?

C= yes, definitely! We ask the neurologist If this was to be considered therapeutic heroism (overtreatment) and he said no. He added that PEG would have prolonged his life of 2-3 years (...) but he was firmly contrary. At that time he had moderate speech impairment, but he could express himself very well. But at once, one of the doctors came and told me: " Mrs X this is not good. Even if he's contrary we have to do it. We need his consent because he can't go on like that. He must do it. So they convinced him that after PEG he could continue to eat normally. But that was false! Nothing, he can't even swallow a sip of water. Now 4 years have passed!

Confusion and abandonment can lead to fear for the future. In the next citations examples of these feelings are reported.

After being discharged from the A&E because the doctors said her husband was dying and that was not the right place of care for him and after being rejected by their GP who was not keen on caring for such a problematic patient, his wife says:

CMS3, wife of PMS3

C= obviously now, anytime something wrong happens to his health conditions to me is a nightmare! What if I should call the emergency again? I take him back to the A&E and the doctor will tell me again: - are you here again?- well, I start thinking a little and say: - I don't know-.

Fear of bad symptoms can lead to fear for the future

Patient PPD2 (male 70 years old. Diagnosed 6 years ago.)

P= I have the psychosis and the fear of swallowing problems. I am aware that this will happen and I'll have to suffer also for this problem.

When patients are dependent on life sustaining devices like mechanical ventilators patients can dread technical failure and this can have a bad impact on their quality of life

PALS3(male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home) and his wife CALS3.

P= I fear that the ventilator could break, maybe in the night. I'm terrified. Or between Saturday and Sunday...

(...)

P= I'm really worried about the future.. I think at tomorrow, how it will finish.

C= lately yes, very much. He wasn't like this. After diagnosis I was really bad and he helped me with that. But now, as he realize that his strength is decreasing, he is really suffering on that side.

3.5.2.3.5.2 Mood instability, rage, fears, anxiety and depression

Issues related to these domains were present in all interviews and generally related to patients. Only three carers reported these problems related to themselves.

The most frequently reported theme was patients' mood instability usually detected and reported by the carer.

CMS1 is the wife of PMS1 (man 53 years old, MS diagnosed 18 years ago)

C=His mood changes daily. It depends on the day.

Emotional incontinence is another form of mood instability as well described by this pair

PPD2 (male 70 years old. Diagnosed 6 years ago). His wife CPD2.

C= he' very emotional.

P= yes, but there are moments in life where it's easier to control emotions...

Carers as well reported difficulties related to their mood due to their role of caregiver of a very disabled patient

PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired) CMS 7 his wife.

C= (..) I should go to a psychologist to understand the way I am. (...) I realize that I have strange reactions like laughing or crying, that are odd for an equilibrate person as I am. I'm aware I'm in troubles.

Rage is another emotion who came out from the interviews. Again it could be directly reported by the patients

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PALS3(male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home) and his wife CALS3.

C= but the neurologist told you that there are many new experimentations..

P= come on, how long will they take! Look at me now, look at the way I am now! I can't stay all this time here, set on the chair, with nothing to do. I can't concentrate on the things I can do. If I have to use tools such a computer, I don't have patience. After 2-3 minutes I quit. What's the meaning in living like this?

Suggested by the carer when talking about the patient

CMS3 is the wife of PMS3 (male 46 years old, blind because of MS, quadriplegic, almost unable to speak. Fed by PEG)

C=after two car accidents due to his visual loss he had his drive licence suspended and had quit driving, he couldn't go to work and began shutting himself up in his grief and rage.

Rage was also described as the result of unsatisfactory relationships with health professionals supposed to care for their beloved ones.

CPD3= I'm really angry with the neurologist that has always cared for him. I expected him to come here and visit him because he had always followed his situation, so at least once (...) he should have come to see how he is going on.

CMS3= (...) I called his GP.I was really angry. He answered me:- There's nothing I can do. It's not useful that I come and visit him. He's just coming from the hospital, if they weren't able do something for him there...

Another expression of psychological suffering were fears.

Patients and carers talked about their fears of the disease and its consequences

PALS7(woman 60 years old, diagnosed 12 years ago, in NIV 24 hours a day and considering about the PEG and the tracheostomy)

P= mine is a very long story, 12 years. I'm a long lasting, am I not? But talking about this there's a scaring aspect: slow is the disease, slow is the agony. Everything is longer.

(...)what scares me more is the slowness of the agony. I'm afraid of the day where I won't be able to talk and I won't be able to open my eyes. That's scaring. Because you feel you don't have any chance to do something. (...) I know of a woman from Brindisi that couldn't shut her eyes. Her husband closed them for her using patches for the night and in the morning he removed it. Poor lady!

PPD2 (male 70 years old. Diagnosed 6 years ago.) his wife CPD2.

P= I'm very scared. I'm always terrified to fall.

CALS8 is the wife of PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric, fed via PEG, cognitively not impaired, communicates via alpha numerical table)

C=we both participated to the informative meetings held by the ALS group. They were interesting, but the session where the neurologist spoke of the disease was terrifying for us. The world collapsed on us. We arrived home... if I think at that I still cry...

And other causes of fears were uncertainty about the future and death and dying

PALS3(male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home) and his wife CALS3.

P= I fear that the ventilator could break, maybe in the night. I'm terrified. Or between Saturday and Sunday...

PALS5 (male in his 70ies, quadriplegic, aphonic, on a wheelchair.) his wife caregiver CALS5.

C= he has so many fears... he fears to die.

Anxiety and depression were explicitly mentioned in some interviews.

This happened when discussing about therapies or when participants were asked about psychological problems

PPD1, (male 80 years old PD diagnosed 9 years ago) and his wife CPD1

C= Often he cries and is sad. A neurologist prescribed him Elopram, but now they stopped it. Anyway, sometimes his mood is very low.

C= he has moments where he's anxious. He wasn't so before this disease. Now he's in therapy with Alprazolam for insomnia, but...

PMS6 (male 53, diagnosed 16 years ago, wheelchair bound) and his wife CMS6.

C= he was depressed. He's in treatment with seropram. He doesn't speak a lot about the disease. I try to stimulate him, I scold him because he let himself go. He acts like if he said: "I'm ill and that's it!" I don't like it. Other patients are more reactive than he is.

3.5.2.3.5.3 Feeling overwhelmed, impotent, struggle for everything

These needs emerged from numerous interviews and were equally distributed between patient and carers.

Participants described their sense of impotence regarding the evolution of the disease, their feeling of being spectators of the disease without having any chance to modify the impact of the disability on them.

PMS4(71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P= I don't want to do nothing! No more therapies. I refuse it because if you say I don't do nothing and then you accept... when you see that they don't work...

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it's a terrible disappointment. And delusion kills. This is the MS reality. Don't hope, accept it.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL)

P= when you have to go in a one way street.. you can't turn or do a U-turn. (looking at his wife) you must accept it. (...)if my nose is itchy I need someone to scratch it.

Impotence can be experienced when patients' physical troubles impede the normal daily activities, by affecting those skills that patients were used to have and that suddenly become overwhelming.

PPD2 (male 70 years old. Diagnosed 6 years ago) his wife CPD2

P=Once I was queuing in my GP's ambulatory and there was another patient waiting. He was older than me, but I was rigid as... my legs were chained. The sensation is awful, like having lead blocks at your feet and you can't raise them from the floor. As if they were stuck.

Impotence is also when carers cannot help patients because they are not able to understand what is happening

CMS3 is the wife of PMS3 (male 46 years old, blind because of MS, quadriplegic, almost unable to speak. Fed by PEG)

C= he had fever, I tried to talk to him, but it was not possible. When he's like that it's absolutely impossible to communicate. This is another terrible aspect, it's devastating because he tries to speak, but it's impossible to understand what he means. So you stay there and don't know what to do.

There is also the fear of being overwhelmed by the experience of the disease and become unable to control emotions

CMS1 is the wife of PMS1 (man 53 years old, MS diagnosed 18 years ago)

I= what about a psychological help?

C= I don't know..... I'm concerned that emotions would overwhelm me

Struggle for everything was denounced by carers and, although in less number, patients talked about the difficulties related to having to fight to obtain help and consideration from others.

This could be related to other family members as described by the wife of this MS patient:

CMS1 is the wife of PMS1 (man 53 years old, MS diagnosed 18 years ago)

Anything you need you have to ask and ask and ask...

(...)Relatives are like new shoes, after a while they hurt, they are tight. However you always have to ask, they don't see your needs. Once I had to do a hole in the wall. They knew it but no one offered to do it. They were expecting me to ask them for it. When I argued they said: - why didn't you ask?-. they should be more sensitive about our needs.

Others described how difficulties came from professionals, both health and social workers. In the following citations carers complain about the social assistants who are accused not to be of help when asked to provide help, causing them to struggle to obtain economic support or technical devices

CALS1 is the wife of PALS1 a man 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.. He communicates using an alpha-numerical table. He's waiting an electronic communicator (MyTobii) that will allow him to have a voice decoder, e-mail access. This device is very expensive but could have a dramatically positive impact on his QoL

C= to obtain what we now have got, we had to pester the life out of them. Ask and ask and ask. And the strange thing is that they came here to interview us asking us about what things could be done to improve the quality of life of the patient, his family and all this staff... these are good initiatives, but then when you have to go to the NHS office to obtain the authorization for devices or subsidies or aids.. well nobody cares about you!

PMS2 is a 43 years old man, quadriplegic, spending his days stuck in the wheelchair. He lost his job and his wife had to reduce hers to 4 hours per day only in order to have time to care for him and for their 10 years daughter.

P= (when the social assistant came home to evaluate their economic status to decide if they could receive a social allowance) She didn't even have a look at the person on the wheelchair, who was the real problem! (meaning that she was only interested in the house to evaluate their economic status rather than the real problem that was his physical inability)

This carer is really frustrated and negative about the possibility to receive any kind of help after years spent struggling to obtain what she felt were her unmet needs

CPD3 is the wife of PPD3 (male in his 70, diagnosed 27 years ago, wheelchair bound., totally dependent in the ADL)

C=If you need an help there is now one available!

3.5.2.3.5.4 Having to cope with losses, having unmet expectations

These issues were frequently raised by patients or by carers when talking about patients' feelings. This group of needs, very consistently with other published experiences, involves difficulties in accepting the physical disability caused by the disease, the relentless progression of the symptoms, the loss of social activities, the loss of loved people and goes towards the uncertainty of what will happen seeking for cure through improbable and costly journeys to far east countries and looking for strange therapies found on the internet.

This MS patient is talking about the difficulty to accept the wheelchair. His wife refers that their coping strategy is based upon making fun of the disability and they act like if she is trying to harm him in order to make jokes and laugh of it. They both state that crying is not a good coping strategy and does not induce other people to provide help.

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PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

P= when you have to go into a one way street. you cannot turn right or left or do a U-turn.(looking at his wife) you must accept it. (...)

C= we try to make some fun of it. Sometimes people who don't know us well says:-Gosh, are you killing him?- and sometimes he says to his doctors:- Look that if you can't explain my death.. it was her who killed me, it's an homicide!- I know this does not seem appropriate, but if you don't act like that... what can you do? You can cry once, twice.. and then what?

P= if you cry once people can tell you they are sorry.. poor man. The second time they grow weary, they're fed up with you.

C= crying is not useful. Doesn't solve any problem.

P= and doesn't even patch it.

Difficulties to accept disability and problems related to the disease progression are well described by this PD patient, fully aware about the disease and what can happen in the future because he is an active member of the PD patient society and read many books about movement disorders

PPD2 (male, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias) CPD2 his wife

P= with the progression of the disease, which is degenerative, being limited in the therapy because of side effects what happens to me? Things that once I gladly did, now I do reluctantly, they cost me fatigue. I perceive my slowness in movements, it's a feature of the disease (...)

C= we can't play cards with our friends anymore, he falls asleep.

P= because of the drugs. It's not my fault.

C= we can't go sightseeing too, as we were used to.

P= it's due to the disease progression. I'm limited because of the drugs. (He shows us his book about PD related problems and adds):" if I had written this manual I would have done it identical".

Disability can drive to other losses like job loss that can cause economical problems, but moreover impact really badly on patients' expectations and self esteem

CMS3 is the wife of PMS3 (male, age 46, blind, quadriplegic, muscular spasticity, fed by PEG)

C= after the hospital admission (when he was diagnosed) I hoped he could come back home fully recovered. Unfortunately it wasn't like that because his is a Secondary Progressive MS, so it worsened continuously.(...)

C= losses were continuous, month after month. He found a new job as telephone operator, he could do it being blind, but it didn't last for long because he lost the sensibility of the skin of his fingers and couldn't feel the keys.

The following example shows how difficulties in coping with symptom progression can lead to trying many alternative approaches in foreign countries, hard therapies like abstinence from food or being eager to participate to any new drug trial.

PALS2 (male, age 39, tracheostomized, fed by PEG, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2

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P= the most tragic period was when I fell

C= he fell continuously. He had 2 devastating years. That was an horrible period where psychologically you don't accept the disease. You don't believe your doctors.. and we went abroad looking for alternative solutions.

We tried everything: Germany for injection of stem cells, France for macrobiotics, Perugia for fast. For the official biomedicine we participated to a trial with IGF (...) and Rilutek. But then attending at medical conferences you see that when there are benefits they are limited in time and disillusioning and so you wonder if it's worthy to continue.

P= I refused the PEG in the beginning because I didn't want to lose the pleasure of eating.

C= but they told you that you could continue eating something, I think it was also a psychological refusal.

P= food was the only thing that was left to me. It so true that now I look at TV programmes about cooking.

Alternative therapies can become an obsession in those families where the disease progression is not accepted as shown below

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

C= we went twice to Beijing where he had 2 stem cells transplantations. Our neurologist tried to discourage us, but he wanted to try. Fortunately our insurance covered the costs.

P= the question is always the same: What I do have to lose?

C= since they couldn't offer him any alternative!

P= it can work or not... but if it works.. than it's just money, until we have it! (...)

C= this disease is so quick that you don't have time to obtain what you need that you have to face another loss.

Patients realise that research is far from being of help for them and this causes disappointment as appears in the following citation

PALS3(male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home) and his wife CALS3.

C= he was a very active and sporting man. Immobility destroys him psychologically. Being forced in a wheelchair is something unnatural.

P=yes, it's terrible.(...)

P= last respiratory control they told me I was improved...

C= maybe because ventilator is doing its job

P= I can't be improved.

C= well the disease is progressing.(...), but the neurologist told you that there are many new experimentations..

P= come on, how long will they take! Look at me now, look at the way I am now! I can't stay all this time here, set on the chair, with nothing to do. I can't concentrate on the things I can do. If I have to use tools such a computer, I don't have patience. After 2-3 minutes I quit. What's the meaning in living like this?

Patients who are aware of their conditions because diagnosis was not told or because they don't want to discuss it tend to be unhappy about their health carers

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This recently diagnosed patient is severely dyspnoeic and thinks that his breathing problems are not well treated by his lung specialist.

PALS9 (male, age 77, diagnosed 8 months ago, in NIV, he was not informed about his diagnosis) and CALS9 his wife

P= I need a doctor able to cure my breath! (...) I don't say that doctor that is caring for me are not good, but my breath doesn't improve.

(...)

P= (abruptly, without forewarning) Isn't it possible to be cured from this illness?

C= this is what he's interested in! please talk to him about it.

In the following experience the patient lost his speech and was very closed in himself so it was not possible to know how much he knew about the disease progression. The only clue that we obtained was that his coping strategy was the hope in new research as his wife referred.

CMSA1 wife of PMSA1 a gentleman 73 years old, diagnosed 4 years ago. Bedridden with muscular rigidity and serious communication impairment

I= your husband talked to the neurologist about the disease, about prognosis..?

C= he didn't talk a lot of his disease. We talked a lot about research. He knew perfectly about his diagnosis, he read the examinations reports, probably he commented about it with his doctors, but not with us.

(...) I restate that he was aware of his disease, but didn't talk with us. And he knew that we were aware too. (...)

When the disease progression shows patterns different from what are normally expected, patients and carers can be deceived that this is the effect of alternative treatments. This can create false expectations and delusions.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized) and CALS1, his wife

C= our GP is a homoeopath, and cures him with her therapies. Because of it we eliminated almost all of his drugs. Now we don't know if these homoeopathic drugs works, but actually we must say that his disease is still. He's one o the few cases where this disease is not progressing.

This evidence shows the many losses experienced by an MS patient with an extremely long illness experience. From her words shines the different physical, emotional, relational losses that she had to cope with

PMS4 (71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers. She is divorced after being abandoned by her husband who said he could not cope with her disease)

P= it's a continuous of losses: job, satisfactions, divorce, disabilities (...)

(after I divorced) I found another man, with MS as well. We fell in love. He was divorced as well, a former football player. He was an excellent company. (...)

*than something unexpected happened. Liver cancer, the worst type. He died in 1995. (...) so an extra suffering. Each time a worst relapse on me (...)
I accepted the wheelchair only when my partner died. On one side it's an advantage, but on the other side you quit doing those little movements you used to do before.(...) ... what can I do? I can cry and I'll soon have liver trouble, or I can crush my head against the wall, throw myself out of the balcony, but I can't because I'm unable to stand anymore, poison myself... but I don't have drugs with me.. so or you accept it or otherwise... it's the same!
I've had enough, I lived my life. If you only knew how many people and friends I saw dying because of this disease. The last one was a friend of mine, a woman with MS: she was only 44!*

Carers also experience difficulties when they have to cope with their loved one losses. These result in a growing workload for them and satisfactions are not easy to find out as this participant testifies

CPD3 is the wife of PPD3 a gentleman in his 70, diagnosed 27 years ago. Cognitive impairment, Wheelchair bound, living in a long term facility)
C= I'm sorry if I said weird things. This is my experience: suffering and sacrifices. No satisfaction.

3.5.2.3.5.5 Feeling a burden for others

The sensation of feeling a burden for people living around is both common and very negative for participants

An MS patient explain this feeling with the following sentence

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):
"after so many years you start feeling a like load for the others. A deathly load. Because I have a must condition and alter other people's choices. You always need to ask. I don't like it."

This ALS/MND patient, even though recently diagnosed, report how the need to rely on other member of his family affects his independence and force his relatives to stay behind him

PALS9 (male, age 77, diagnosed 8 months ago, in NIV, CALS9 his wife)
P= if I could drive I'd go wherever I like.... But like this...I have to ask them to drive for me. I lost my independence.

When physical dependence becomes total, like in the following case, even simple tasks like scratching one's own nose is not possible and someone has to do it on patient's behalf

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL)
P=I literally need someone always behind me.... if my nose is itchy I need someone to scratch it.

In this dramatic testify the extreme consequence of feeling a burden is told. This patient tried to commit suicide in order to relieve his loved wife from the burden of his own care

CALS8, wife of PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric)

C= this winter he tried to take his life. (...)I went to see him in the night and he had stuffed a handkerchief in his mouth. (the patient smiles). I took it off and asked him why... he said he was doing this for me, because I'm full of troubles and infirmities. I answered him that my infirmities would continue anyway and is would have been a further problem to me. He promised not to repeat it, but four nights later he stuffed 2 handkerchiefs in his mouth. So I got angry and told him that if he succeeded police would have charge me for murder. (...)

In some case carers report that caring for patients totally disabled for many years is really troublesome. This MS patient's wife said this sentence when the patient was not listening to her, but obviously these feelings cannot stay hidden in a long relationship like theirs is.

PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired) CMS 7 his wife.

C=I don't want him to listen to.. he has never understood what it really meant to me... I can't tell you these things right now! He has never understood the troubles that we had...if he knew he'd live too bad

3.5.2.3.5.6 Shame, dignity, privacy

These words were also used to express feelings that can be grouped in the previous set of needs. These are all uncomfortable sensations related to the lack of self esteem, respect from the others, achievement.

These uncomfortable feelings concur to patients isolation. In the following example a patient talks about shame and closure

PMS4 (71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P= I'm not participating the patients meetings organized for the patients association. My partner didn't like it. When you go and see, after 1 year, how other patients worsened you mirror your worsening. So I don't go to these dinners. Cheese and bread on my own are better!

(...)

P= I don't go out because of a sense of shame. I'm ashamed of being ill, of the wheelchair, I fear to fall. It happened once, just downstairs on the sidewalk in front of the house door. There was a little gap among tiles and it blocked the wheels. I fell and hit my face. I felt worst for the sense of shame than for the injury.

Disability itself can cause sense of shame in some participant. The typical signs of a movement disorder are unbearable to be shown for this PD patient

PPD2 (male, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias)

P= I'm a proud man. I feel ashamed as if I were a thief. (...) I'm ashamed when people see me like this, blocked. I know that it's not my fault, but it's disgraceful being like this. Shame is the most invalidating problem of the disease to me. It's humiliating. I'm hurt when I read sorrow in other people's eyes.

And when a young man is completely dependent in the daily activities privacy can be an issue. This patient had to accept his parents help for his own care while his wife is out to work. He clearly states how he does not have time to stay alone by himself because, being tracheostomized, he need continuous control of his life sustaining machines.

PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.

P= another unsatisfied need is privacy! I'd like to stay alone for a while.

C= (...) he had to accept a carer out of his parents because they are getting old. He yielded only when he understood that it was necessary

3.5.2.3.5.7 Need of self independence and control

Terms to express this group of psychological needs were the following:

Need to feel useful, to be able to do things on their own, to communicate with others when speech is impaired, to cope without external help.

These issues represent the opposite face of physical dependence and often were proposed by those participants who were concerned for being a burden for the others.

The next citation is from a carer who has been involved directly in the care of her ill husband for 25 years and, in the mean time, she had to care for their son and daughter and, eventually lost her job not to leave the patient in other people's hands

CMS 7, wife of PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired)

C= I've always refused help from others, like volunteers and now I'm regretful for this. I didn't want to involve my son and my daughter because I didn't want to affect their lives

The same carer (talking on behalf of her husband who could not talk because his speech was completely affected by the disease, but who agreed not verbally to these sentences) describes how important is self independence for a man who has always lived an active life and that progressively lose these capabilities

C=Imagine that one day he called me to show how he was still able to tighten a screw. Is it possible that a man need to show something like that?

One coping strategy, shown below, adopted by this couple formed of the ill husband and his active wife to maintain independence and control is to fight together against the disease trying to stay compact and leaving the rest of the family out

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic) CALS6 his wife

P= we are serene because we are foolhardy!

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C= we had our crisis, but perhaps it's a matter of temperament. He has a strong determination to fight against his disease, I follow him... so we plucked up courage. But this doesn't mean that we didn't have problems or obstacles. We had it.

We don't involve our relatives in our problems. We gave us a commitment, we have to manage alone. Our son, well he'll have his own life so we don't want to involve him too much.

Patients, above all in early stages of the disease tend to refuse external help because accepting it would mean accept the progression of the disability. This elderly man, completely aware of the nature of his illness, refused to accept a necessary aid like the wheelchair because he wanted to show that he could cope with the increasing disability without external help. At the time of interview he was very disabled, he completely lost his speech and was stuck on the wheelchair, but when his wife told this story he smiled confirming that these were his real intentions.

CALS8, wife of PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric)

C= he has always been a tough man. When we went to the legal visit for the invalidity pension he refused to go with the wheelchair despite the fact that some suggested him to do it. He wanted to go with his cane.

Aids can be of extreme help for those patients who want to fight against disability. Loss of communication skills affects deeply one owns quality of life. When patients obtain and accept external devices like electronic communicators their regain some degrees of freedom and really appreciate it.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL)

P= the laptop? It's a Saint, a blessing! Because at least with it I feel useful having something to do.

These two patients, both tracheostomized, cognitively not impaired and showing a high level of life attachment, confirm the importance of electronic communicators as tools to enhance their independence, to stay in touch with the world and to improve their self esteem

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic), CALS his wife

C= we are looking forward to obtain a new vocal communicator. It would be so important for him. He likes very much when people come here to meet him. A lot of people don't want to come afraid to disturb us. Marco would be so happy!

I= what if volunteers came here to spend sometime with you?

P=Please, send them!

(...)

P= (communicating with the eyes through an alpha numerical table) to me the possibility to communicate means a lot. It would improve my quality of life. Even my psycho- physic balance would be enhanced.

PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.

P= as long as I could I surfed on the internet and I was full of friends with my disease. We could communicate, share suggestions.

C= this gave him a good support both moral and psychological.

3.5.2.3.5.8 Family concerns, burden of care

Family is one of the most important factor for the quality of life of people living with advanced neurodegenerative disorders.

It is both an essential part of their care, because it allows the patient to be cared for in their homes, because it provides practical help, love and social relationships, but it can be a cause of concern. Family carers, from their side feel the importance of their role, but they complain also about the hardness of their task, lack of external help and, sometimes they ca not cope with their loved ones disability and decide to abandon the patient.

This PD patient shows his disappointment for not being able to provide support to his wife who has recently been hit by an acute stroke and is slowly recovering

PPD2 (male, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias) CPD2 his wife

P= another cause for my suffering is that I can't help her (crying), and she has her physical problems too.

But patients suffer a lot when they feel they have been abandoned because they were affected by their neurological conditions

PMS4(71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P= 10 years after diagnosis and 15 after our wedding my husband left me. We had a 14 year old son. He left me to my mother saying that he didn't want to be my nurse anymore. Later we divorced.

In some families the disease is not an obstacle to the normal relationship as the carer of this totally impaired patient testify

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized) and CALS1, his wife

C=despite the disease if we have to quarrel we do it easily. Our relationship didn't change.. if things were not going well before, well they aren't going well neither now (and smiles) right dear?

P= (using the alpha numerical table) yes! (smiling)

Tiredness and burn out of the carers is really an issue as testified by the following experiences.

PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired) CMS 7 his wife, SMS7 is their son

C= about myself I must say that that I'm burned out.

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S= it's what me and my sister are trying to explain to her.(...) it's not just a question of money, it's she that doesn't want to accept a concrete help. (...) it might be a relief to know that there's a paid carer able to stay with dad and give you 2 hours to do what you like.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

C= personally I'm very distressed! I'm tired, I don't even have 5 minutes to do what I like. I can't go out, I mean anything. (...) since he has been tracheostomized we can't leave him alone not even for 1 minute!

Carers can feel preoccupied of what could happen if they had accidents and could not provide care to their loved ones

CMS1 is the wife of PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night)

C= The disease is present and we have to live with it. I never ask anything to anyone, but life would be much easier if I knew that someone were available. I'm anxious when I have to leave and drive: what if something happens to me?

CALS6 is the wife of PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

C= I'm terrified to fall ill. If it had to happen... who'd care for my husband!

Inside families there is not always concordance about the strategies to adopt for patients' care

DMSA1 is the daughter of PMSA1 a gentleman 73 years old, diagnosed 4 years ago. Bedridden with muscular rigidity and serious communication impairment. She talks about her mother who is providing care to the patient and does not follow their physicians prescriptions about therapies

D= my mother is like this. She continuously cuts his drugs... it's not easy.

I= do you think she has unattainable hopes about the disease progression? Is she aware of what will happen?

D= I think she knows everything.

Other family members can represent a further cause of concern above all when they are fragile and cannot cope with what is happening

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

C= me, my daughter and my son we all are in psychotherapy. Above all my son he is very shut off, is difficult to communicate with him. He suffered a lot for his father condition. He is depressed now. He's 26, probably he is more frail than we are... he wasn't able to study anymore and this was frustrating to him. He saw his father's conditions worsening and became intractable.

3.5.2.3.5.9 End of life issues

Psychological unmet needs contribute to end of life decisions. Some participants talked about their position about life sustaining devices or decision related to take their lives when were talking about their psychological status

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):

P= Sometimes I say I'll go to The Netherland's, so I'll avoid problems. Sometimes I think... well, maybe if they change a little our laws.. they should change our laws. When one arrives to an unbearable level. I don't say it for me. It's for people around me. It's not good to sacrifice people.

I= so you say you'd purchase euthanasia because you feel like a load for your family?

P= " as well. (the dog starts barking)... but without me even the dog would die.

PALS3(male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home) and his wife CALS3.

P= I really wonder if taking these drugs is worthwhile...

I= can you tell me something about the psychological difficulties in coping with this disease?

P= I don't want to think about it (...) if I think at what will happen to me... I don't want to arrive at the tube solution (tracheostomy). When.it'll be necessary, well to be bedridden, eyes fixed on the ceiling... I can't accept it. And when I think about Welby (an Italian ALS tracheostomized patients who ask for the withdrawing of his invasive ventilation), he just wants to die. But if you do it than the doctor will be persecuted. Can not a man just decide on his own?

(...) I'm very sorry for my family, my wife, my son, but this is not life. If there were other exits, but like this...

Uncertainness about end of life choice (that in Italy are not clearly regulated by law and advance directives are not legally bounding the doctors choices) can be a cause of concern and worry for both patients and carers

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

P= I don't want to be tracheostomized. But I can't be sure that this advance directive will be respected because in Italy they aren't recognized by the law.

C= a person we know was tracheostomized in a hospital against his will.

P= you know the world is full of dick-heads!

C= I fear to face a supporting interventionism doctor, of those ones supporting life against everything. I believe in self determination. If it were for me.. hurrah for euthanasia, hurrah for the Netherland's!

CALS5 is the wife of PALS5, a totally dependent gentleman wheelchair bound and severely dyspnoeic

C= he doesn't want an invasive ventilation. He's very determinate in that. He prepared a living will stating this. I agree with him.

3.5.2.3.5.10 Need of psychological support

Psychological support was discussed in several interviews and positions about this help were disputed.

Good experiences were reported by some participants

PPD5 (woman age 78, cared for by her husband -CPD5- at home)

C= 2 years ago the Italian Parkinson's disease patients association organized a respite care admission for both of us in a nursing home. We spent 15 days with 3 other couples with the same disease. It was very good, we also had the chance to have talks with psychologists. It was very helpful for us.

I= would you like to have someone to talk to, to give vent your spleen to?

P= I feel this need!

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24 hours a day) and her husband CSLA7.

P= psychological support is so important. It's not just talking to someone, but it's receiving help. I learnt that in the morning I have to create a list of the things that must be done. Anytime I complete one I cancel it from the list. It helps a lot by keeping your mind busy and don't let worries enter.

Another point was the importance for relatives to save some spare time for them. This helps them to clear their mind from my needs and to me because when they come back they are motivated and fresh. You hardly understand these things by yourself. The psychologist helped us a lot.

Other participants were contrary or not satisfied about the help received from psychologists

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

C= he attended for a long period the psychologist of the ALS centre. Now we have a psychotherapist coming here. She comes each 7-10 days. We pay for this service.

P= now it's more a friendly relationship than a professional one! I don't have enough strength to talk, so she talks to me about movies, news etc. it's no more a therapy

PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.

C= we denied the psychological support from the ALS group. We weren't interested in it. I know they have groups of patients and relatives that meet there, but I don't want to go. It's too heavy to me. Many of them are at the beginning of this disease, I don't want to revive those moments. (...) if I see them I can just cry and can not stop.(...)

When we were in Perugia for this alternative approach of the fast he had to meet the psychologist, it was scheduled. He hated it because he was told that the disease was his fault, that was a kind of answer of the body because he didn't want to accept certain situations.

In some cases the position was not clear and participants were undecided about to try a support or not

PMSA1 (MALE 73, bedridden, caregivers are his wife and one of his daughter CSMA1 and DSMA1)

I= ever tried a psychological support?

D= no.

D= (crying, when her mother isn't in the room) she's the problem! Anytime one of us says something, she replies that we don't understand, and that we cannot understand!

3.5.2.3.5.11 Summary

A huge range of psychological themes emerged from the content analysis of the interviews. The raised issues regarded many different aspects of the psychological human needs. Some were about the personal feeling of being abandoned by the professional carers or by friends, relatives because of their conditions. This imply confusion and concerns for the future.

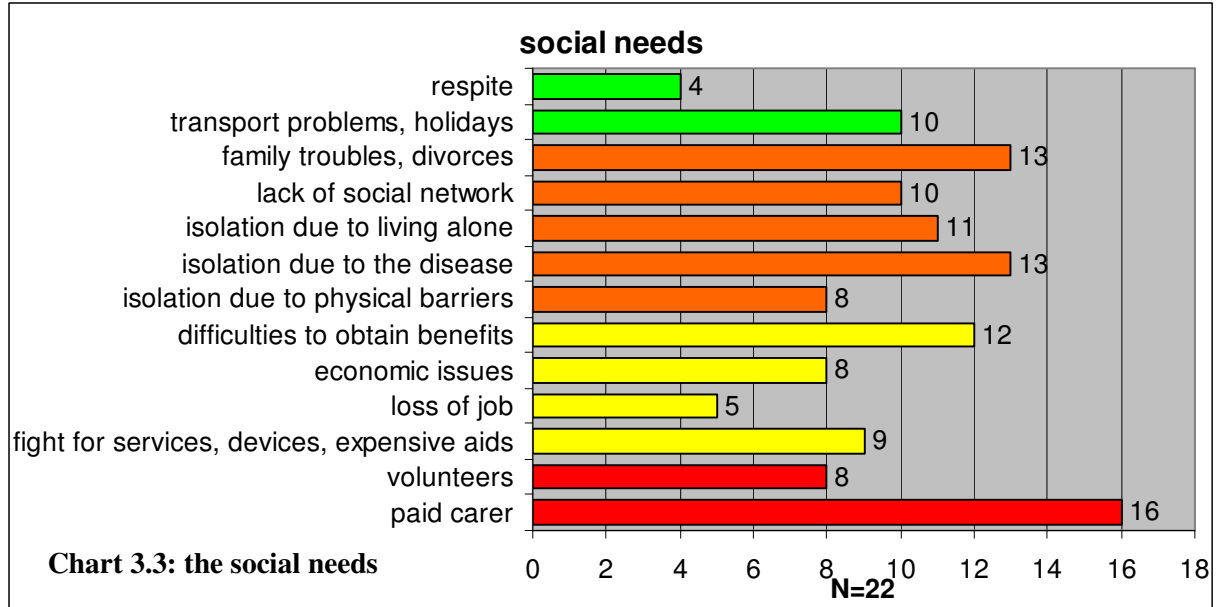
Mood instability, leading towards anxiety and depression were sometimes declared by the participants, but in many interviews could be detected from non explicit statements or comments. Difficulties in coping with the disabling disorders arouse frequently often accompanied by unrealistic expectations of clinical improvements. Other issued reported were the unpleasant feeling of losing control, feeling impotent or overwhelmed by the disease, worries about being a burden for the others. Less frequently, but highly significant for the participants who talked about this feelings, were conditions like loss of dignity, need of more privacy and sense of shame.

The need for a specific psychological support was discussed by participants who had already experienced it with different opinions and satisfaction. Psychological issues triggered some comments from participants about end of life decisions.

Family carers took the chance of the interviews to testify how difficult and exhausting is the long term care of their loved ones and the worries that they feel about what could happen to the patients if something happened to them.

3.5.2.3.6 The content analysis of the social needs

The content analysis of the social needs which emerged from the 22 interviews is summarized in the chart 3.3 and table 3.5.



	Quotes in the 22 interviews	Percentage
Need of respite care	4	18.8%
Transport, holidays	10	45.5%
Family problems, divorce	13	59%
Isolation (lack of social network)	10	45.5%
Isolation (living alone)	10	45.5%
Isolation (disease)	13	59%
Isolation (physical barriers)	8	36.4%
Financial issues	8	36.4%
Difficulties to obtain social benefits	12	54.5%
Loss of job	5	22.3%
Difficulties for services, devices	9	41%
Need of paid carers	16	72.8%
Need of volunteers	8	36.4%

Table 3.5: the social needs

3.5.2.3.7 The social needs detailed analysis

Results related to the social needs of participants to the interviews will be displayed in this section. Abbreviation forms will be used as described for the physical needs section.

Social needs were grouped in four main categories as shown with the different colours in chart 3.3.

Each group will be presented and described with citations extract from the interviews used as data.

3.5.2.3.7.1 Isolation

This group of unmet social needs was the most represented in all the content analysis. A total of

41 citations in the 22 interviews were related to these themes.

Lack of social network was one of the main cause of isolation of both patients and carers. Friends and relatives often do not collaborate and tend to vanish over time like this carer reports

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

C= friends were very helpful when he was at the hospital, but since we came home they aren't so collaborative. Some of them disappeared and we lost them before he was admitted to the hospital. Some were very close to him, we wouldn't expect them to vanish like this.

This carer was talking about the decision to admit her husband to a long term facility because she was not able to manage him at home. Even though they have relatives they cannot be of help because of other serious health problems

CPD3 is the wife of PPD3 a gentleman in his 70, diagnosed 27 years ago. Cognitive impairment, Wheelchair bound, living in a long term facility

C= our relatives aren't much of help for us because they have their own problems. My niece has a congenital malformation and so she's often in the hospital and my daughter has to stay with her.

Some families are really alone because they do not have relatives living close to them and no other help from neighbours or from the community

PPD1 (male, age 80, cognitively slow but able to understand and severely disabled in his movements.) CPD1 his wife.

C= we have 2 sons, but they're married and live far from (...). At home we are alone and I have to do everything by myself.

I= do leave him at home alone?

C= yes, he stays at home alone. I don't go away half a day, but a couple of hour to go shopping .. yes.

Another situation where the elderly couple is really isolated, not having anyone able to provide help. They do not have sons or daughters nor close relatives.

PPD5 (woman age 78, cared for by her husband -CPD5- at home)

I= do you have sons or daughters?

C= no, we are alone!

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(...)

I= do you think that a priest could help you to cope with your disease related distress?

C= well, we have been living here for 4 years and.. well volunteers from the church say that priests are few and have so many people to visit..

We had 1 priest who came in the past, but now he disappeared

(....)

C= I have to go shopping, on errands and to manage the house. She stays at home with the paid carer we have. She's staying here for 3 hours in the morning and sometimes in the afternoon if I have to go out.

In this situation the patient's wife complains about their state of loneliness in which even their close families do not provide help

CMS6 wife of PMS6 a 53 years old gentleman, fed by PEG, almost totally dependent in the ADL

C= even with his family we had problems. His mother didn't want to come here and spend sometime with him. They put him aside. They do many thing for his sister who's healthy and has got a child, but nothing for him.

This carer suggests how things change over time. In 25 years of caregiving to her husband she felt her needs becoming higher and the help provided by friends being less concrete and satisfactory.

CMS7 is the wife of PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired).

C= we are isolated because I have always refused to be helped.

I am plenty of needs, but this is the way I am. (...)I'd feel to go and leave him only if I knew he's very well cared for. (...)

C= friends have always been supportive to us. When I worked they spent their mornings with my husband(...). But now thing are worse. They come here and find him cleaned and prepared and don't do anything. I know this discourse is unpleasant, it look like if I didn't need help from people. I have plenty of needs. They were so helpful when my son and daughter went to school. I changed my shifts to manage our lives. Then since I retired, my life is finished. Before I had some time for me, I went out for a pizza with my colleagues.. now I'm shut in this house and I know it. I reached a point where I can't have a chat with others.

This carer recognize to be the only one able to care for her wife, even though he has to care for is mother too, who is living with them and is very ill having a cancer in advanced stage. In this case visits from friends and other people of the community seems not being much of help

PPD4 (woman, age 60, care for by her husband –CPD4- at home, in DBS)

C=I am the only one who takes care of her. I'm retired now and tale care of home management and errands..

(...)

C= we know that AIP (PD patient's association) organize meeting, parties etc, but we don't participate. We don't even have contacts with our Parish church or other social centres. We just have friends and neighbours coming for some visits.

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In the following citation we have both physical problems who concur to limit the visits of friends and the loneliness of the patients' wife who does not receive enough help and has to leave the patient alone

PALS9 male, age 77, diagnosed 8 months ago, in NIV, CALS9 his wife, DAL9 one daughter

C= he's very happy when his friends and the priest come to visit him they talk about their past, things they did together (...) unfortunately his breath gets short quickly when he talks and he needs the mask. With it he can't talk anymore.

(...)

C= knowing that there's a person who can spend 2-3 hours one afternoon per week would very important to my mother. She could plan her shopping without feeling in a rush.

D= it'd be important for dad too, he likes so much talking with people!

Another cause of isolation is the disease itself. Participants describe how symptoms and signs of an advanced disease can scare other people and cause sense of shame resulting in closure and isolation.

PPD2 (male, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias) CPD2 his wife

C= we were used to go out: we went out to play cards with some friends of us, but now we can't anymore because he falls asleep.

P= my immobility is self-evident. I can't walk, I have many difficulties to enter in the car because I can't open the door and because I continuously move like dancing.

(...)

C= we can't go out for tourism, as we were used to, because he's ashamed and so, in conclusion, we are always blocked at home.

Here there is a further example of how patients feel the disease can take people away

CMS6 wife of PMS6 a 53 years old gentleman, fed by PEG, almost totally dependent in the ADL

C= we are isolated. Many of the things we used to do before we had to leave it. We went out with friends, we had to renounce to it. Friends and relatives don't come to visit him. I think it's because of his drooling, or because he lost his urine from the catheter... it can be disgusting.

Physical barriers are another cause of social isolation. Carers and patients complained about their difficulties when they have to leave their homes and often have to renounce and stay at home when going out could be very important for their lives

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):

P=My daughter had her degree on May, but I couldn't participate to the ceremony because of the stairs.

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C=It's difficult to go out with the wheelchair because of obstacles. Even where there are ramps, often they are dangerous; I feel like the chair slips away from me.

PPD1 (male, age 80, cognitively slow but able to understand and severely disabled in his movements.) CPD1 his wife.

C= we have the problem to go out because of the stairs we have in our apartment building. We should place a lift, but we don't know if the other tenants will agree.

In the following two reports carers explain how physical barriers and lack of external help cause patients to spend lots of time alone in their homes when they have to go out on errands and cannot take their loved ones with them

PALS3 (male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. On NIV at home) and his wife CALS3.

C= leaving home is a problem: there are stairs, then we have to book an equipped taxi and it takes 24 hours to book it. For this reason I quit going out, it is ages since I left home last time.

(....)

C= unfortunately I'm alone and I have to do all the shopping, errands etc. so I have to leave him home alone.

CMSA1 wife of PMSA1 a gentleman 73 years old, diagnosed 4 years ago. Bedridden with muscular rigidity and serious communication impairment, DMSA1 his daughter

D= how can we go out with dad? He doesn't leave the house. There are so many physical barriers to leave the house that we can't push him out.

(...)

C= I'm always so busy. I've got to go to the GP's ambulatory for prescriptions, or to the district office for authorizations for medications, to go shopping and other errands... I don't have anyone who can help me in this daily tasks.

Barriers are not only physical, sometimes human component contribute to the physical isolation as this carer explained

CMS6 wife of PMS6 a 53 years old gentleman, fed by PEG, almost totally dependent in the ADL

C=Unfortunately, even if we had a good aid to go down from the stairs, when we were in the cortile we couldn't exit because the other inhabitants of the building left their car everywhere precluding our way out.

Families sometimes had to move to better places to overcome physical barriers. In the following citation the carer talks about these issues and adds how being alone in the task of caring for her highly disabled husband means having to involve their only son in his daily needs. Patient and carer tried not to be a burden for their son, but obviously an external help would be of help.

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic). They're considered a "social family" and live in a council flat

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C= we had to move to this flat because where we were living before we didn't have a lift and to go out we went through a slide.

(...)

C= I've always tried not to involve our son too much in his father's care. I don't want him to be oppressed by this hardship. But he has always helped me a lot.

(...)

C= when we are alone we try to manage ourselves our problems. We ask for help only if strictly necessary. If he needs to go to the toilet and we are alone he wants me to call our son at work. So he asks me when he'll be out of his turn of duty. But I say to him: "let's do it ourselves... and we do it!"

Isolation is also the consequence of being abandoned. The following two quotes are related to two patients who have been abandoned by their spouses because they said they could not cope with the disease

PMS4(71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P= after the divorce (...) now I live alone.

P= I spent 15 days as a prisoner in my house because the transport footboard that enables me to descend last steps of the stair to go out, went out of work. It has been working there for 40 years. I did maintenance interventions when required. Now I applied for a new one. The city council will contribute with 2500 euros that will be reimbursed to me in 2009. the rest will be charged to me. Total cost is 7500 euros plus manpower.

PMS5 (male, 49 years old, diagnosed 24 years ago. Quadriplegic, fed by PEG, cognitively impaired, barely able to consent to the interview) and his mother CMS5

C= he is married and has got a son. His wife left him when he was seriously ill. His son comes to see him sometimes, or call to have news, but in fact, he is alone and I am the only one who takes care of him

A consequence of being isolated can be the fear of falling ill and having to place the patient into hospital or in a long term facility. This carer highlights how her work is not recognized by the community as helpful and cost saving. She underlines how external help could prevent problems to her health status and the costs of hospitalization for both of them.

PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric, cognitively not impaired, communicates via alpha numerical table) and his wife CALS8

C= I'm afraid of what could happen if I fell ill. I don't know how much would it cost to the community if they had to hospitalize both of us.

(...)

C= I can't manage this situation alone because I'm old and ill.

3.5.2.3.7.2 Economic issues

In this group of unmet needs the following categories were gathered: financial issues, loss of job, difficulties to obtain economic benefits, equipment, services and expensive aids.

About economic related troubles we had the following information:

- 5 participants said they lost their job because of the disease,

- 8 declared to be in serious financial troubles,
- 12 stated their difficulties to obtain sick benefits
- 9 reported difficulties in obtaining equipment, services or important expensive aids, such as electronic communicators.

Job loss, financial issues and benefits

The loss of a job has economical and motivation implications. It is another loss that, added with physical losses, has a very bad impact on patients' quality of life, self esteem and self actualization. (Maslow, A. H. 1943)

Following two different examples of how this loss contributed to amplify patients dissatisfaction.

PMS4(71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P= I lost my first job because of the disease. It was very important to me. I created the company where I worked, together with my boss. I left that place because of the several hospital admissions I had in that period. I had to quit because I couldn't go on like that anymore. It was a great loss. I entered there when I was 18 and left married with a son... that was a family to me. (...). After the divorce my sister in law found me another place of work as a secretary in a school. I had to quit that too because I had to climb some stairs and I reached a point where I couldn't afford it anymore.

This MS patient complains about their economic restrain after his wife decision to leave her job in order to care for him

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):

P=My wife lost her job in order to have time to care for me. I expected them to give her some money for it. Nothing. She had to retire earlier from her job, and so she's having a smaller pension. We are living with my disability pension and the disability cheque..

And this carer describes how hard is to care for his very ill husband having health troubles herself. She states that their incomes are not enough to engage paid carers.

PPD1 (male, age 80, cognitively slow but able to understand and severely disabled in his movements) CPD1 his wife.

C= his pension is about €1000 per month. If I had to pay for caregivers day and night... I couldn't afford it. So if I had a problem like a back pain... I don't know what I could do (...) I had 2 hips replacement, shoulder surgery, I must take care of myself.

This MS patient wife testify how her decision to reduce her working time resulted in financial straits for their family

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

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C= I had to cut my salary changing my full time job to a part time one. Now I work 4 hours a day to care for him and our daughter (she's affected by juvenile rheumatoid arthritis).

We have to pay a mortgage for our flat, a loan at interest for the van. But they said that as long I work and he has his pension we are wealthy and they won't give us nothing.

Paid carers are scarce resources and family have to pay for them. This woman didn't know that she could apply for a sick benefit, a monthly cheque provided by the town council specifically aimed at supporting family who are caring for seriously disabled persons in their homes.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

I= do you have a sick benefit for his condition?

C= no, he receive a pension and the accompaniment subsidy. We don't even know what the sick benefit is (...) the lady who is caring for him, I should ask her to stay 1 week-end per month, day and night, in spite of the fact that she has her daughter to look after. But she's staying here 5 days a week and then it costs. If we were extremely rich I could pay for a carer 24\24 and my life would continue exactly as it was before.

This wife obtained a sick benefit enough to provide paid carers for her disabled husband, but she was not reassured about its continuation into the following year. This is a great cause of concern for her who is still working in a high responsibility position

CMS3 is the wife of PMS3 (male, age 46, blind, quadriplegic, fed by PEG).

C= I don't ask for nothing. I just want to care for my husband at home. I think that, as a citizen, I've my rights, not only duties. I have the duty to work, to care for my husband and I think I'm socially useful because if I put him in a structure it'd be much more expensive for the society(...)

C= (...) now things are worse. He needs assistance 24 hours a day. Now, after a long bureaucracy, we obtained a sick-benefit. It allows us to pay a caregiver 8 hours a day, 5 days a week, while I'm at work. It was approved for this year, I hope they'll renew it and maybe increase a little so I can have this carer helping me on Saturday morning when I have to bath him.

Here similar examples of how social supports are not tailored on families' needs are shown.

CALS6 wife of PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

C= my son has just started his first job. The welfare assistant told me they'll cut our subsidies because of this. But his intake is e very low and he just uses it for his leisure...

(....)

C= I left my job 6 years ago, when they conceded him the accompaniment cheque. I was working part time and it wasn't worthwhile to work and pay a carer for doing what I could do. My husband now receive the invalidity subsidy, the accompaniment cheque and since the beginning of this year a sick benefit that allows me to pay for this carer 2 hours a day. It's not enough at all, I'll go

and discuss with the welfare assistant to increase its amount. The social office send us also another person for 2 hours per week. He wasn't useful at home, so we agreed that he goes shopping or goes on errand for us.

CPD3 is the wife of PPD3 a gentleman in his 70, diagnosed 27 years ago. Cognitive impairment, Wheelchair bound.

C=. *I don't receive any taxi coupon and I have to pay for ambulances.*

(....)

C= money is a problem: he receive 1100 euros for his pension and 400 euros for the accompaniment check. Here the charge for us is 1200 euros per month. But as I take only 230 euros for my pensions I have to live with 530 euros. It's not much, barely enough to survive.

3.5.2.3.7.3 Material, medications, bureaucracy

Four participants talked about social difficult related to aids delivery, bureaucracy. Having to assist disabled patients for very long time caused carers to report difficulties related to the procedures that are to be renewed cyclically and cause waste of time in NHS offices.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

P= why anytime I need something I have to do that long bureaucracy?

C= I'm like one of the family in that counter. I go for the bedpans, catheters, jelly water.. I spend so much time there and I have to subtract it from my work or from the family.

PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.

C= we have some problems with the provision of material from the NHS and the pharmacy and the company delivering these things.

Patients and carers recognize that domiciliary aids like wheelchairs, orthopaedic beds, lifters are very useful and necessary, often difficult to accept because when they are required they mean a progression of the disease. In the following example this patient's wife suggest that neurologists, rehabilitation specialists and providers should be more efficient and prescribe these facilities earlier in the course of the disease.

PALS3(male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home) and his wife CALS3

C= for this kind of patients aids should be provided quickly. Maybe anticipating their needs. Now he has a good electric bed, but we in the beginning they gave us another mechanical one and we had to change it after a short time. Why didn't they prescribe this one at the beginning? They know the way the disease progress. The same with the wheelchair.

Even palliative drugs are not always reimbursed from the NHS and their cost can be a real burden for families in long periods.

CPD3 is the wife of PPD3 a gentleman in his 70, diagnosed 27 years ago. Cognitive impairment, Wheelchair bound, living in a long term facility)
C= our neurologist prescribed to my husband a drug to substitute apomorphine. I had to go to buy it in a Pharmacy far from our city. It wasn't reimbursed by the NHS and was very expensive. I had to pay about 230 euros each box. It was effective, but we had to stop it.

Expensive devices

Participants highlighted the difficulties they met when they needed expensive aids. For people severely disabled lack of electronic communicators or cars adapted for disability can be fundamental facilities can cause social isolation. In Italy there has been big discussion about the possibility to provide these aids to people severely affected by neurodegenerative conditions.

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic)
P=If you need aids they give them to you only if they are disease related, I mean wheelchairs, hospital bed, air mattress etc, but as I need expensive remotes to control my electronic devices, or I had to buy a special van where I can enter with my chair using an elevator... well they didn't help me at all.

This family pioneered the request to obtain computer based communicators. The patient is a former local politician and raised his problem to the public opinion. He produced a video and broadcasted it on the internet to produce awareness about this issue.

CALS1 is the wife of PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized. Fully aware of his situation. He communicates using an alpha-numerical table. He's waiting an electronic communicator that will allow him to have a voice decoder, e-mail access etc.)
C= this new device, the communicator,(MY TOBII software package) is too important for him. It'll arrive next week, we had the chance to try it in a session at the hospital. It's great. But it's very expensive (€21.000). we applied for it at the NHS office, but they denied it. As it is fundamental for his Quality of life we decided to have it in any case. So he created an association called "A voice for Michele". They are doing fund raising for it. On a parallel plane we continued asking to the NHS, he wrote to the town council, to the member of the regional committee in charge for public health and an the end they approved the grant. Now they say that they are providing it to him, but it was us who had to fight and ask and pester them!

The following testimonial shows the similar problems in a tracheostomized ALS/MND patient

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic) and his wife CALS6
*I= what would you think could be helpful to you now?
C= communicator, we are anxiously waiting for it!
(....)
P= (in the letter to the director of district NHS office to obtain the communicator) I need 24\24 assistance and for this, fortunately there is my wife*

who does this impeccably. I really need this communicator. I tried it on June at the hospital. It'd allow me to navigate on the internet and write and send e-mails with its eye controlled device. On the screen there's the fundamental alarm key that can be activated with only a glance! It's an extraordinary tool for people in my conditions. If you won't authorize this tool I won't be able to afford it, because I'm a social case...

3.5.2.3.7.4 Transport, holidays and respite

Overall 14 participants raised unmet needs that could fit into these categories.

Transport

7 participants referred problem related to difficulties in transport. Adaptation of private cars, high cost of ambulance or taxis adapted for disability were the main topics of this category.

This carer was explaining the reasons that induced her to admit her husband to a nursing home. One of the causes was lack of transport for his needs, when he had to go to his rehabilitation sessions, ambulatory controls etc.

*CPD3 is the wife of PPD3 a gentleman in his 70, diagnosed 27 years ago. Cognitive impairment, Wheelchair bound, living in a long term facility)
C= I have to pay for all transports. I don't receive any taxi coupon and I have to pay for ambulances.*

But it is not only a financial problem since even those participants whose transports were free of charge were not satisfied by public transports.

*PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.
C= to transport him to the hospitals for his routine controls is a problem. We don't pay for the ambulance because it is offered by the district domiciliary service. But we must wait for them hours, above all when it's time to come back home.*

To move persons with high disability means moving equipments as well. Families report difficulties for this reason

*PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.
C= we had to buy a Doblò (van that can host the patient on the wheelchair) to move. The problem is that if I drive I can't look after him and his arm falls down... so, when we have to go for his visits, pump titrations etc, we call the AISM (patients association). Their volunteers come with their van, but you have to book their service about 8 days before, so sometimes we're late at the visits.*

Patients struggle to go out and to leave their rooms in which they are often confined for long time.

CALS6 is the wife of PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

C= it's so important to him to go to his home village in summer. When he's there he recuperates. He has his friends, his land.. It is located 800 kilometres far from here.

But to go there we have to rent an ambulance where we can plug the ventilator and the in-exsufflator. It's very expensive. We travel when they come in these whereabouts to transport another patient, so we can save some money travelling in there way back.

Holidays

When asked about important issues 10 patients and families participants raised their desire to go on holiday. This was not just for leisure, but really to have a break far from the disease, from the setting where they have to face disability, isolation and losses. It is an attempt to reinforce normal family relationships, play with children and feel “normal”. Unfortunately it seems that it is not so easy to satisfy this need.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife

P=It's a long time since I can't go on holiday (...) it's not just a question of where, but also of how I can get there

C= if I drive our car I have to choose if carry him or his belongings, his baggage, 2 wheelchairs.... AISM (patients' association) doesn't offer this service. You feel discouraged when have to move so many aids like pads, devices etc .

Respite

Some carers talked about their tiredness due to the burden of the care they have to provide.

This carer describes how heavy is to care for her tracheostomized MND/ALS husband

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

C= personally I'm very distressed! I'm tired, I don't even have 5 minutes to do what I like. I can't go out, I mean anything. (...) since he has been tracheostomized we can't leave him alone not even for 1 minute!

Similar feelings appear in this interview with the wife of a very ill MSA patient

CMSA1 wife of PMSA1 a gentleman 73 years old, diagnosed 4 years ago. Bedridden with muscular rigidity and serious communication impairment

C= I'm always so busy. I've got to go to the GP's ambulatory for prescriptions, go to the district office for authorizations, to go shopping and other errands... I don't have anyone who can help me in these daily tasks.

This MS patient' wife testify how hard is to accept to be helped. Even though she states to be really tired and in need for respite she is not sure about a hospice admission

because she would feel guilty and is not confident about the quality of care that her husband could receive in her absence.

PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired) CMS 7 his wife.

C= I have always refused to be helped. I am plenty of needs, but this is the way I am. (...)I'd feel to go and leave him only if I knew he's very well cared for.(...) A respite care admission would really be of help. (...)

C=Before I had some time for me, I went out for a pizza with my colleagues.. now I'm shut in this house and I knew it. I reached a point where I can't have a chat with others.(...)

C= about myself I must say that that I'm burned out. (...)

C=I need to go away for some days, to walk off, to sleep... also in the day, even if he's quite without troubles I've always my ear pointed on him. I want to stay alone!

I think a respite care could be helpful, but I have to see if I'd be able to leave him.

It's so hard to leave. I left him only twice, always to go to visit my daughter who's living abroad. I need a strong motivation and the certainty that he's well cared for.

3.5.2.3.7.5 Need for paid carers and volunteers

In 16 interviews out of 22 need for paid carers able to provide help in the care of the patient came out. 8 participants stated that they would be happy to receive visits from volunteers.

This is a striking need showing different problems: it can be due to financial issues because paid carers often are totally at families charge, but it can be a problem of acceptance since many participants testified how hard is to trust unknown people giving them hard responsibilities from which their loved ones lives can depend.

Paid Carers

Some carers urge for practical help in the house

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

C= we called them (the social service managers) because we need help. A person who can spend the morning with him. I don't need a nurse, because I do everything of it. Just someone who stays with him

P= that's our main need!

This carer reinforce this concept stating how help is required for many different tasks

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

C= in terms of needs, we need help for everything! Family management, housekeeping are very challenging. You are keen to help as much as possible, but you're so busy! So if there were another person who could help in the day this would be so useful!

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(...)people totally unable to do necessary things should receive constant assistance!

Similar findings came out in this interview where the patient' wife and their two daughters were discussing about their needs at home

PALS9 (male, 77 diagnosed 8 months ago) and his daughter CALS9, his wife WALS9, and a second daughter DALS9

W= he needs assistance for the whole night. It's a problem because he has wires and tubes all around him.

C= he sleeps in his bed and one of us has to spend the night close to him. We work on shifts, one night each.

D= so, every now and then, we have to pay for a carer who gives us a rest.

This patient recognises that his wife needs help. He complains about the absence of help and about financial support

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife)

The first need we have is a help for my wife! My wife had to be admitted to the hospital for surgery. She is my only caregiver and I'm totally dependent on her. I didn't have any form of assistance. I had to pay a private carer days and nights. And when I ask to detract its cost from the taxes they didn't allow me. Not even an Euro. The more you are down the more they push you down. If you have money you can afford it, but if you don't, well..

This PD patient' wife talks about the impact of her poor health status on her difficulties to care for her husband. She is really worried about what could happen if she was not able to care for him for an acute disease

PPD1 (male, age 80), CPD1 his wife

C= his pension is about 1000 euros per month. If I had to pay for caregiver day and night... I couldn't afford it. So if I had a problem like a back pain... I don't know what I could do. (...) I had 2 hips replacement, shoulder surgery, I must take care of myself.

This other carer is already helped by paid carers, but she complains because with the worsening of her husband disability she needs more help.

PMS3 (male, age 46, blind, quadriplegic, muscular spasticity, fed by PEG), CMS3 his wife.

C= I just want to be helped a little more than I was in the past years. What I need is not to be left alone (in the management of my husband), because otherwise sometimes you are not rational enough. (...). Now things are worse. He needs an assistance 24 hours a day. Now, after a long bureaucracy, we obtained a sick-benefit. It allows us to pay a caregiver 8 hours a day, 5 days week, while I'm at work. It was approved for this year, I hope they'll renew it and maybe increase a little so I can have this carer helping me on Saturday morning when I have to bath him

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Other similar data appear in the following citations were participants, both carers and patients, recognise how the need for good paid carer is not satisfied in their situations.

CALS5 is the wife of PALS5, a totally dependent gentleman wheelchair bound and severely dyspnoeic. PALS5 (male in his 70ies, quadriplegic, aphonic, on a wheelchair. He is a retired doctor).

C= I have to pay for a private assistant. He's living with my husband 15 hours a day. He's very good, we couldn't stay without him. I can't manage to toilet him or move him anymore. In the hours when we he is off duty I'm alone and I'd need a help. I'm very tired.

This MSA patient' daughter adds how in their experience paid carers do not collaborate with their programme of care and they cannot rely on them.

PMSA1 (MALE 73, bedridden. caregivers are his wife and one of his daughter CMSA1

C= it's not easy to find a good paid carer. We changed a couple of them. They don't do what you ask them to. My father has to some physiotherapy exercises during the day. But if you don't control them they don't do it.

This couple reports other problems with their paid carers

PPD5 (woman age 78, cared for by her husband -CPD5- at home)

C= I have to go shopping, on errands and to manage the house. She stays at home with the paid carer we have. She's staying here for 3 hours in the morning and sometimes in the afternoon if I have to go out. (...)

C= in the morning she stays home with the carer. We have several problems with these persons (...) we had to change many of them because they are not Italian, they are from eastern Europe and are irregulars. Many of them they steal in our flat. We had their names from a volunteer of our church. She said she knew a friend of her...

P= this one is even worse than the others.

C= the problem is that I have to leave her at home with them.

In this situation paid carers are absolutely necessary because the patient lives alone and without this help she could not stay in her preferred place of care

PMS4 (71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

I live alone. I'm receiving a paid caregiver since 16 years. She's paid for the social assistant for 2 hours and IN2 in the morning. Than there's a boy who helps me when I have to go to bed or get up.. sometimes he goes on errand for me.

I= is the municipality paying for him?

P= yes.

I= is he paid for an amount of hours?

P=yes, about 60 hours per month. We manage it depending on my needs.

The following data is from an interview with an MS patient wife during which their son arrived at home and provided some comments about his mother attitudes in the care of the patient.

PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired) CMS 7 his wife and SMS7 is their son.

S= it's not an economic problem. It's her difficulty to accept help now (...). We would need to be addressed to a service that could help us.

S= it's what me and my sister are trying to explain to her.(...) it's not just a question of money, it's she that doesn't want to accept a concrete help. (...) it might be a relief to know that there's a paid carer able to stay with dad and give you 2 hours to do what you like.

Volunteers

Issues about the need and the role of volunteers appeared in the data with different meanings. In some case they were available, but their role was not clear at participants eyes.

PPD5 (woman age 78, cared for by her husband -CPD5- at home)

I= do you have volunteers who come here and stay with you?

C= yes, volunteers from the parish church. They come to bring us the Communion. (...) volunteers, if we really need them, come and give us a hand..

P= (Angrily!) What do you say? I've never seen a volunteer here washing a plate!

C=(..) there are 2-3 volunteers from the church.

P= But what do they do?

C= nothing... they just come and visit us.

I= are you happy that they come?

P+C= yes.

This carer feels that volunteers could be a solution for their needs, but she says that she could not find any and she fears that they could not cope with her husbands' problems

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

C= here voluntary service don't exist. We would have really benefit from it. Maybe to ease a little our carer during the day. She has a child and sometimes would require some rest. And then it would be money saving for us! We went asking to our parish church, but they don't have anyone. And then I fear that when they see such machines they'd flee.

This patient identified clearly how a volunteer could help her for.

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24 hours a day)

P= I'm looking for a volunteer to help me by turning my books pages. It would be very important to me because real books are better than electronic ones.

This patient looked enthusiastic at the idea to receive volunteers

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

I= wouldn't you feel invaded or oppressed if other people came here?

P= no!

C= many don't come saying they don't want to disturb... My husband on contrary, would be so happy to have them here!

I= what about volunteers, would you be happy to have someone coming here to chat with you?

P= please send them here!

This carer says volunteers can be a good idea for the next future, even though they had never thought about it.

PPD2 (male, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias) CPD2 his wife

C= we have never looked for volunteers because we can manage things by ourselves. But tomorrows who knows. I am of his age and have my problems.. I hade a mild stroke and still have problems at my left side.

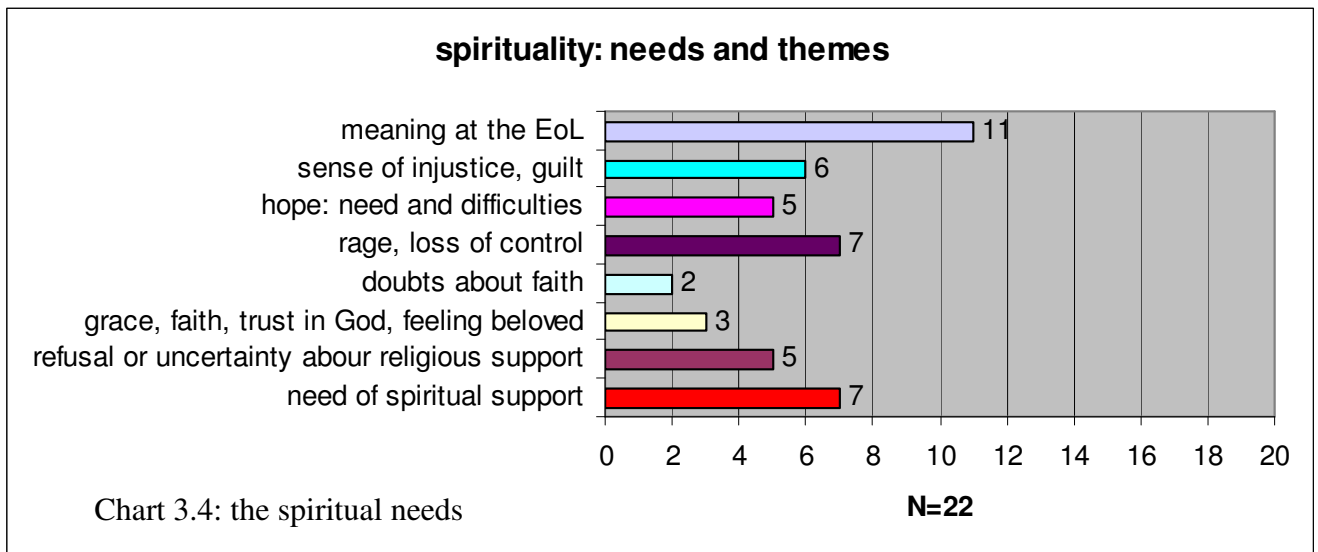
3.5.2.3.7.6 Summary

Among the different categories of social needs reported in the interviews the sense of social isolation for both the patients and the carers was the most frequent and burdensome. Economic and financial problems were also discussed together with the need of home care assistants and volunteers required to provide help, but difficult to obtain from the existing services. The long duration of the diseases together with the high disability of the patients raised other peculiar social issues like the problematic of the transports and the need of holiday and respite.

3.5.2.3.8 The content analysis of the spiritual needs

Spiritual issues were less frequently reported by participants than other categories of unmet needs. Often researchers had to probe this group of existential needs with direct questions. In only a few interviews participants talked about these themes at the beginning of their stories. Usually this group of needs was discussed at the end of the events and, in some cases, participant recognised that spirituality was very important for their QoL.

In the following chart 3.4 and table 3.6 the results of content analysis related to the spiritual themes emerged from the interviews are summarized



Spiritual needs and themes		
	Number of citations	Percentage
Meaning	11	50%
Justice	6	27%
Hope	5	23%
Rage, loss of control	7	32%
Faith, religion, love	5	23%
Spiritual support	12	55%

Tab 3.6 the spiritual needs

3.5.2.3.9 Spiritual needs detailed analysis

In this chapter a detailed analysis of the spiritual content of the interviews is presented. Participants will be identified with the same short forms used for the physical needs.

3.5.2.3.9.1 Meaning

Searching for a meaning of the lived experience is a typical spiritual need (quote). Some participants reported to have found a meaning in their experience of disability and life threatening condition, but this does not mean that they accept it easily. Others stated to believe they can find that meaning in the future.

This patient recognises her difficulties in accepting her condition

PALS7 (woman 60 years old, diagnosed 12 years ago, quadriplegic, disharthric, dyspnoeic, NIV 24\24 by nasal mask that enable her to talk).

P=This doesn't mean that I accept this disease. I deal with it. I'm sure that I'll discover the reason for this when I'll be up there. Because down here I don't see any reason for this.

This patient was experiencing a strong revision of her life and she felt that the impact of the disease had been so bad for her and her husband that she would have changed her mind about her choices in life if she had known it beforehand.

PPD4 (woman, age 60, cared for by her husband –CPD4- at home, in DBS)

P= if I knew that I was going to fall ill one year after our wedding I wouldn't have married.

This carer, after having described the psychological difficulties of their younger son in coping with his father disease resulting in deep changes in his behaviour talks about the meaning of such devastating experience for the other familiars and the role of external help

CALS4 is the wife of PALS4 (male, 61, quadriplegic, in a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago)

C=We believe in psychotherapy. For our family it is very important. It is helping my son that is very depressed (...) and it is not easy to have him like that wandering in the house in that condition. It's also a matter of spirituality in a sense

Decisions and choices at the end of life can be affected by having found a meaning for what is happening in that experience.

This carers explains that being aware that her husband is likely to die changed her plans about the place of care for her loved one

CMS3 is the wife of PMS3 (male, age 46, blind, quadriplegic, muscular spasticity, fed by PEG),

C= doctors told me that my husband arrived at the end of his life. For this reason they wanted me to place him in a nursing home because I wasn't going to

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be able to manage him at that stage, at home. I said:” What? I’ve cared for him for years alone and now that you say that he’s at the end I should park him somewhere else? Absolutely not! We’ve never been divided except for the short time when I had surgery. I’ll take care of him in our home.

There were contrasting findings between similar situations that caused completely different decisions. This was particularly clear about end of life decisions. Patients who found a meaning in life even with high disability coped well with choices who prolonged their life in spite of higher disability. Some would do the same choices again, some do not want to consider it. All these choices hide different meanings deeply found by participants in their lives when physical conditions dramatically change.

This citation is from a ALS/MND patient who was tracheostomized in emergency for a respiratory arrest even though he had previously refused this option. Following this decision he is happy and would do it again. His wife is very supportive of his choice despite the high burden of care that she has to face.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife
*P= well, I think I’m lucky because my disease is stable.
I= would you consent again to the tracheostomy?
P= yes.
I= do you mean it’s worthwhile living like this?
P= yes
C= my idea is that living is always worthwhile. I can’t accept the idea of dying.
(...) my opinion is that life must be lived in any condition. This is true only if you’re conscious, because if you’re not others will have to decide for you.*

In contrast another patient with the same diagnosis decided not prolong his life because he could not find a meaning in his life in that condition.

PALS3 (male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home) and his wife CALS3.
*P= I don’t want tracheostomy. That is not life.
(...)
P= everyone can think what prefer. I don’t want to suffer for these things.
Please, let’s give a blow, I don’t want to suffer more.*

Another patient reports the same decision and his wife talks about their concern not to be respected in their decision.

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4
*P= now I state that I don’t want to be tracheostomized, but the neurologist told that even if I prepare a living will it might not be respected by a doctor if I were took to an intensive care unit, in case of emergency.
C= I think I’d like that every person could decide freely about his end of life decisions.*

This patient gave his consent to tracheostomy and mechanical ventilation, but he says that sometimes he thinks to withdraw it. Having found a meaning in his actual life he endures in his decision to stay alive

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

P= Sometimes I wonder, without finding any answer: “ there is more courage in living or in dying?” if euthanasia were legal perhaps... but then not, I can't and I don't want to quit.

This very religious woman thought a lot about the decisions at the end of life. She was considering tracheostomy and PEG at the time of the interview because her breathing and swallowing were both deteriorating quickly. Asked about these decisions and the implication of the meaning in her choices she said:

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24 hours a day)

P= I read the letter written by Cardinal Martini and I found it wonderful because, talking about heroic treatment, he says that:” don't allow a person to reach the end of his life means refuse his or her meeting with the Father that is waiting for him or her.” And that's true. Everyone of us has its own time

She wanted to comment about the discussion that was on the news on those days in Italy about the request of a tracheostomized ALS/MND patient to withdraw his mechanical ventilation and to die peacefully. She commented about the meaning of that decision stating that it was morally correct because that man fought with all his energy against the disease, but at the end his suffering became meaningless:

I think that Welby's case, the ALS patient who died when his doctor sedated him and switched off his ventilator and is under a trial for euthanasia, shouldn't be considered euthanasia because a man who lived 20 years communicating with the eyes, tracheostomized, with a PEG, showed a strong attachment to life. If then, in spite of his invasive ventilation, he had apnoeas and suffered for breathlessness this means that his body couldn't go on anymore.... So let's unfurl the sails!

Finding a meaning can be described as the sense of completeness of one owns life as testified by this patient

PMS4 (71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P= What I cannot accept is therapeutic heroism, overtreatment. If necessary I'll state it on a written form. It's useless, it just prolong absurd suffering, delusions. This is what I think.

(...)

My respiratory tests were totally unsatisfactory, but I refused mechanical ventilators. I've had enough, I lived my life.

3.5.2.3.9.2 Injustice

When the sense of meaning cannot be found a sense of injustice about what is happening to affect patients and families.

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Sometimes it appears under the form of a recrimination towards God or the fate when participants feel not to deserve their conditions

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night)

P=I've never hurt anyone and I'm not even able to swear... and that's the nice. I never swear, I store all inside, also pain, and the is the way they pay me back.

This patient has been affected by MS for decades and is actively involved in the local MS patients association. She stated that she accepted her conditions, but she feels unfair to see younger people facing worsening disability without having the possibility to stop it.

PMS4 (woman, 69 years old, diagnosed 42 years ago, quadriplegic, living alone)

P= we have so many young people affected by this disease in our association. Boys that didn't receive anything in this life.

Injustice is also described as the lack of reward for having lived a socially correct life, with the expectation to enjoy serene years after retirement.

PALS3 (male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home)

P= I worked 37 years and never asked for sick-leave. (...) can you imagine 37 years without a single day of sick-leave? Then I retire, I was so well.. and now this. I cannot accept this, no way!

Having to struggle to obtain what participants thought to have the right for evoke another sense of injustice. This patient feels he has the right to obtain an electronic communicator and wrote a letter to the Health District manager complaining about the refusal that he had received. He uses a metaphoric expression to state his sense of injustice for not having had his need satisfied because he politely avoided to quarrel with the involved office.

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

P=Unfortunately I learnt that in some people's life I'd rather enter and exit on the tip of the feet.

A further cause of injustice can be caused by the sense of guilt that is well explained in the interaction between this patient' wife and her daughter

CMSA1 wife of PMSA1 a gentleman 73 years old, diagnosed 4 years ago. Bedridden with muscular rigidity and serious communication impairment.

DSMA1 their daughter

C= when you have to face with experience like this one .. life changes so much. You don't have time for yourself.

D= she could have time if she let us help her. But she doesn't permit to be helped because if she saves some time for her then she feels guilty and she feels like if this was unjust.

3.5.2.3.9.3 Hope

The spiritual meaning of hope can be seen as having the hope to improve ones own physical condition.

PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.

P= the only thing that really support me is hope. I hope to recover. Is the hope that one day they'll find a cure for me. Is this a visionary hope? Well it helps to boost my morale!

Hope can also be an attempt to justify strange decisions, such as in the following example where this couple went twice to China for a stem cells transplantation (that did not provide any improvement) against the neurologist's advice

PALS4 (male, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4.

P= what did I have to lose?

C= no one could offer him alternatives.

Lack of hope can lead to desperation and confusion

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night)

P=After so many years of my disease I don't know what to hope, what to think, what to say, what to do.

Hope can be seen as the opposite of acceptance

PMS4(71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P= these are MS realities: It's useless to have hope. You just have to accept.

Carers can experience frustration when they feel that their loved one lost his or her hope and do not fight against the disease.

CMS6 wife of PMS6 a 53 years old gentleman EDSS 9, fed by PEG, almost totally dependent in the ADL

C=He really speaks very little. It's due to the disease. I try to scold him, to stimulate him because he let himself go. He doesn't react as others do. It's like if he said:" I'm ill and it's fine, let me stay". I don't like it, it's like if he lost his hope.

3.5.2.3.9.4 Rage and Control

When asked about their spiritual feelings evoked by the experience of disease interviewees talked about the rage and the loss of control caused by the disease.

PPD2 (male, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias)

P=I become angry with the Chief Clerk who stays over there (meaning God) and I tell him: " Please look on another side, is it possible that you always have to look here!"

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night) *P=It looks like I'm the only crucified here! I've always been correct with the other people, polite with everyone. This is the way I'm paid back.*

Relentlessness of the disease progression cause rage and affects the coping strategies of the families.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

P=It's because of all that happens to you. When you say, come on perhaps we can breathe a little, we can have a break, suddenly something worst happens!

Rage can be the strength enabling patients' to go on in life as testified by this patient who gave us part of a letter that he wrote with the computer to the NHS manager who should approve his application for a new computer based communicator

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

P= "I know I'm a load for everyone, but what can I do? My disease takes away everything: strength, voice and dignity. I can't recover, but help me to survive. According to statistics I should have been dead a long ago. But I have a strength and a rage inside that I could smash the world."

Rage can also be a symptom of frustration for being stuck in a wheelchair and totally dependent on the others.

PMS4 (female, 69 years old, diagnosed 42 years ago)

P= for the family you're just a heavy load. This is the dark thing, this is the thing that makes me curse, swear.

The importance of being in control of what will happen and to maintain autonomy in decisions can be associated with the sense of frustration reported by participants about the potential loss of this aspect of life.

PALS7 (woman 60 years old, diagnosed 12 years ago)

P= about living will I agree but: it must be done when I am very close to a decision. I can't state now about issues that will happen in 10 years. Then it's an absurd that to do this you have to go to a notary.

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

P= now I state that I don't want to be tracheostomized, but the neurologist told that even if I prepare a living will it might not be respected by a doctor if I were took to an intensive care unit, in case of emergency.

C= I think I'd like that every person could decide freely about his end of life decisions.

3.5.2.3.9.5 Faith, religion and love

Spirituality and religiousness are not synonymous. Love is often described as the main characteristic of God and has a profound religious meaning. Participants talked about these themes highlighting their different feelings and position about these issues.

This patient totally impaired in his speech, showed us this prayer that he wrote on his computer to express his faith

PALS5, male in his 70ies, quadriplegic, aphonic, on a wheelchair. he tries to communicate using a computer keyboard. Very slow. He is a retired doctor)

P= " Faith to me is the pursuit in the fatigue, in the dark of doubt. I don't believe in sudden flashes, in the graces meant as miracles. Doubt, discount, light. Grace is for very few on Damascus road. My Lord, this is not the time to search the reasons for faith. Highest Grace is abandon myself in You. Total trust in your hands, my Lord. To live in daily precariousness with my angels and in Your Love"

And this woman talks about the importance to have faith and how she can justify what happens in life, find a meaning, because she believes in God's plans

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24 hours a day)

P= faith helps in the important things of life. This was our lives journey. I've never thought to be immune from illnesses. I had friends who died of cancer, 2 sons of our friends died for car accidents... nobody was born with guarantees.

(and talking about withdrawing vital support treatment)

P=I think that is a way to say: let me go up there, because up above someone is waiting for me.

This couple is very religious and found a deep support in their faith. The patient was not able to talk, but he strongly wanted to testify his faith and agreed with his wife statement

PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric)
CMS7 his wife.

C= after diagnosis my husband put his life in Our Lord's hands.

(...)

C= I try to explain my husband's point of view about the meaning of this disease and relationship between spirituality and this experience, I'll ask him to confirm or not if this is correct: in the beginning of the disease he said – I'll never ask to

Our Lord to heal me, because if he wants to he does it anyway. If he gave this disease it must be of use for something or someone

Faith is not always seen as having a positive impact on participants' experiences.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

C= We are believing, but... sometimes you ask yourself if you have to believe or not...

CMS6 wife of PMS6 a 53 years old gentleman EDSS 9, fed by PEG, almost totally dependent in the ADL

C= we used to attend the church. Now he doesn't feel like going anymore. Once two priest of our parish came for dinner, it was nice. But then they changed and we didn't have the chance to meet the new ones.

3.5.2.3.9.6 Religious support

Support provided by priests or other religious assistance was very well accepted and recognized as positive by a part of our participants and refused or not seen as useful by others.

This can be affected by having been religious persons (or not) before the experience of the disease, but also by the possibility of being supported by someone who can or is supposed to provide religious support. The following citations testify how good relationships with religious professionals result in a good spiritual support during the disease.

PPD1 (male, age 80, cognitively slow but able to understand and severely disabled in his movements. During the interview was admitted in the neurological ward for symptom control) CPD1 his wife.

C= he has always attended our parish church. He went to the Mass. (...) now the priest should come to meet him. I always say that I should go and tell him to come!

P= I follow the Mass in television.

C= (...) it helps him, because he's so sad and often cries.

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night)

P=Once a month a priest comes here to meet me. He gives me the Holy Communion. We exchange 2 words, we talk about some things. It's a small relief.

PMS4 (71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

P= as religious support I've a good relationship with the parson. (...) he knows that I curse like a Turkish, but he says that the Lord understands everyone's intentions. He's a person with whom I can talk about everything. He's a real support. According to our Church dispositions I shouldn't receive the Holy Communion, being divorced. He and his predecessor have always given it to me.

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Once he went to the hospital where my partner was admitted for his cancer. He went to meet him and told him he was there by chance, just because he went to see a parishioner admitted in the same hospital. Later on I happened to know that it wasn't true. He went there just to meet him. We found responsible people.

PALS9 (male, age 77, diagnosed 8 months ago, in NIV, CALS9 his wife)
C=He's very happy when the priest and his friends come and talk about their past, the things they did together...

Sometimes a religious support would be well accepted, but this need cannot be satisfied because it is not available

PPD5 (woman age 78, cared for by her husband –CPD5- at home)
I= do you think that a priest could help you to cope with your disease related distress?
C= well, we have been living here for 4 years and.. well volunteers from the church say that priests are few and have so many people to visit..
We had one priest who came in the past, but now he has disappeared.

Lack of contacts with the local church can be due to little no interest of patients or families about it

PPD4 (woman, age 60, cared for by her husband –CPD4- at home, in DBS)
C= we don't have any contact with our church. We just have friends or neighbours who come and visit us.

Some participants wanted to state clearly that they were not religious persons and, consequently did not want to receive any form of spiritual support

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4
C= we are not religious at all and we don't believe in religious support.

Religious support was sometimes tolerated, though not really felt as important or not specifically tailored on peoples' needs

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized) and CALS1, his wife
C= there's his uncle, he's a retired priest, sometimes he comes to visit him.
I= is he helpful?
P= (he smiles ironically)
C= I think that these issues are depending on the person. Or they help you and you start believing or... personally I live the sense of this experience day by day.

3.5.2.3.9.7 Summary

Spiritual needs themes that could be extract from the interviews focused mainly on the concept of meaning of the experience of the disease and ambivalence about the role of faith, religiousness and spirituality at these stages of the disease.

Other profound spiritual issues reported by participants were the sense of justice or injustice related to what was happening to them, the need and difficulty to hope, the sense of rage and fear to lose control of their existential values.

3.5.2.3.10 Service satisfaction

Patients and their informal carers provided comments about the degree of satisfaction related to the available services that they experienced in the advanced stages of the diseases.

Results from the content analysis of the interviews are presented in this section of the thesis.

Comments could be either positive or negative in terms of effective help received when needs occurred. The primary aim of this enquires was to determine what could be done by the experimental new SPCS to integrate existing services by:

- providing effective help in those fields were existing services were lacking or not tailored on patients needs
- on the other hand, not to double existing services when they were fully satisfying participants needs.

3.5.2.3.11 Content analysis of the service satisfactions' themes

The content analysis of the transcript verbatim of the interview revealed the following fields of interest to be explored:

- Hospital based services
- Primary home care and emergency services
- Other services like respite facilities, physiotherapy, speech and language therapy
- Social services, technical support

Results revealed various levels of satisfaction in the different analyzed services.

In table 3.7 these results are summarized and presented categorized in terms of satisfactory and not satisfactory outcomes from the participants perspective.

Service	Satisfied	Not satisfied
Hospital based services	16/22	8/22
Primary home care and emergency services	9/22	22/22
Other services like respite facilities, physiotherapy, speech and language therapy	11/22	17/22
Social services, technical support	7/22	14\22

Table 3.7: satisfaction or not satisfaction of participants about different specific services. In each interview more than one comment could happen about the same group of service, for this reason the sum can exceed the number of interviews

Some comments were also captured from participants' reports about their point of view about a new experimental SPCS aimed at providing help in their conditions. 15 participants provided a positive comment about this possibility, none were contrary.

3.5.2.3.12 Detailed analysis of the service satisfactions' themes

Data from the interviews related to the degree of satisfaction about the available services are now presented.

Participants were invited to describe the service that they had experienced, were experiencing or that were required. Some data was extracted indirectly from the transcript verbatim when occasionally comments about services appeared.

Hospital based services

All interviewees experienced hospital based services during the course of the disease. Generally they were in charge to a specific neurological clinic or cared for by the respiratory team. Some also experienced nutritional services or were admitted for sometime in a general hospital ward.

Overall both patients and carers were satisfied about the support received by hospital services. Neurologists and respiratory services were generally reported as satisfying participants' needs.

Patients and carers appreciate the effort of professionals to provide a comprehensive care even when it is out of their duty or specialty

*PMS4, 69 years old woman, wheelchair bound, diagnosed 42 years ago
P= the neurologist who diagnosed my MS, 42 years ago, Prof M, is like a brother for me, a relative, a real friend. For all my needs (work related, familiar, personal etc) I've always relied on him. He has always been available with me... to me he's a person to be put on a pedestal.*

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PPD2 (man 70 years old. Diagnosed 6 years ago. Severe dyskinesias and freezing)

P= the neurologist that is caring for us is one of the best available, we're very satisfied of him

They recognise the positive attitude of those professional carers that try to compensate the lack of existing home services by visiting them when necessary. They seem also happy to be involved for scientific and educational purposes as described in the following citation where this tracheostomized patient was filmed by the respiratory team and the video displayed in scientific meetings to show their home organization and good coping with the disability.

PALS1 (man 48 years old, tracheostomized, bedridden, quadriplegic, totally aphonic) caregiver CALS1, his wife.

C= We are cared for by Dr Z. He's the best for us. They have always been very supportive with us, even when we came back home.

I= who trained you on how to manage respiratory devices?

C= they did it, in hospital. But when we came home we modified some of them... when we arrived home we were a bit unprepared. You know in hospital if something doesn't work you simply call for help, but at home... he needed some time to learn how to recognize symptoms and troubles and to realize if we had to worry or not. But to achieve this some time was needed.

I= what about the respiratory service of the hospital: do they come here?

C= no. well if they come they do it when they're not on duty. Dr Z. comes, the others as well. They are happy to come and see the way we are organized. They filmed our place and showed it in a congress.

When participants complained about hospital based services it was usually because they felt that their needs were not being met by the service or because they felt abandoned by their carers when the disease progressed and professionals could not be of help anymore.

In the following sentence appears how this MS patient did not feel safe during routinely respiratory controls feeling in danger for his health rather than helped by the tests and visits

PMS1 (Man 53 years old, diagnosis of MS 18 years ago, quadriplegic, using NIV for respiratory insufficiency)

P=I stopped going to follow up visits for the ventilator. I used to go each 6 month. They visited me, controlled my oxygen saturation with a sample of arterial blood. Once they were doing an X-ray scan to my back. I could not control my chest and neck and I was falling down; if it were not for my wife, who quickly caught me, I would have broken my neck and that would have been the end of the story. This happened 2 years ago. Since then I decided not to go to these control visits anymore. Another point is that they analyzed my respiratory functions with those tests where I had to inspire and expire in a tube. What if the patient before me had a chest infection? I've enough problems like this, I don't want to risk. Yes, they changed the nosepiece, but the tube? That is the same for all patients!

Next is an example of not satisfaction about the neurologist who diagnosed and cared for this patient for a long time. His wife complains because she felt like he discharged

them when he became very ill and now he had to be admitted to a nursing home because she was not able to care after him at home anymore.

PPD3 (man in his 70es, diagnosed 27 years ago) and his wife CPD3.

C= we have always had a good service from our neurologist. Lately he washed his hands about our situations. (...) when we were in need there were not available places for us. We had to book for the visit but the waiting list was of about 5-6 months. Even if it was urgent we had to wait. (...)

C= the day I was carrying him back home from the clinic (where he had been admitted for symptom control and were his condition worsened and they caused him a bedsore) a neurologist of the clinic took me aside and whispered to me: "I want to suggest you one thing, if you own a flat sell it, and with the money you obtain put him somewhere. Your husband is not to be cared at home."

I answered him: "Do you tell me this? Do you know how many stairs I had to clean, how many toilets I had to wash to have these four walls? I'll do anything but selling the flat to park my husband."

Primary home care and emergency services

Available home care services were reported as less satisfactory from participants. These services are provided by the Italian National Health Service (NHS) and are free of charge for patients. They are coordinated by the general practitioner (GP) and are based on the intervention of district nurses (DN). Usually the primary home care intervention consist of scheduled visits of both GP and DN that manage infusions, medications, wound dressing.

GENERAL PRACTITIONERS

Of the 14 participants who talked about their GPs results were ambivalent. 6 reported good satisfaction, 8 not.

The following is a list of citation from participants who stated to be satisfied about their GPs involvement. On average those declaring satisfaction seem not to rely very much on GPs' clinical interventions, but they rather appreciate collaboration, availability of domiciliary visits when required, easiness in prescription and bureaucracy.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

C= our GP she's very collaborative. She doesn't come regularly to visit him here. Sometimes I call here and she comes, takes his blood pressure etc.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

C= our GP, well, she is very good. When she is in service. She's so much busy that she's often missing. There are her substitutes and she explains them very well about our problem when she's off, so it's ok

PALS2 (man, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.

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C= The GP is collaborative. She has to come each month on schedule to prepare prescriptions. We opened ADI (district domiciliary service), but we only use it because of ambulance free transport

On the contrary those patients and carers not satisfied about their GPs complain about their doctors' lack of keen to visits patients at home or difficulties to contact them out of hours.

CPD1, wife of PPD1 (man 80 years old PD, diagnosed 9 years ago, unable to leave his house for muscular stiffness and gait problems)

C= (our GP) doesn't come frequently to visit him at home. Once I told him: - Come to visit my husband-, but he is not so keen to come!

CMSA1 is the daughter of PMSA1 a 73 years old gentleman bedridden totally unable to move, communicate and completely dependent in the ADL. The patient' main caregiver is his old wife

C= my mother is continuously moving between GP and district office. I cannot believe that. I think that the GP should come to visit my father at home at least once each 2 weeks and prepare his prescriptions here. But he doesn't come at all!

Another source of not satisfaction is related to the lack of knowledge of GPs and their substitutes about the specific neurological conditions. For this reason participants tend not to call them knowing that they are of little, no help.

PPD4 (woman, age 60, cared for by her husband –CPD4- at home, in DBS)

P=Our GP is not always on duty, and even when he is, he's not able to manage most of the problems related to the disease.

C= often he's out of service and his substitutes don't even know her. I think that a continuity of care is lacking.

PALS7, (woman 60 years old, diagnosed 12 years ago)

P= unfortunately GPs often cannot give correct information about this disease, and are not always attentive to patient's needs. This is my experience.

Finally sometimes the relationship of care between the GP and the patient can become unbearable due to the high intensity of the required assistance. In the following situation the carer of this very ill patient told the story of an intense personal relationship between her husband and his GP which ended dramatically because the doctor was really too much involved in the assistance for many years and decided to refuse to care for the patient leaving him and her wife in despair.

CMS3 is the wife of PMS3 (man, age 46, blind, quadriplegic, muscular spasticity, fed by PEG).

C=(...) he was bad again, flaccid, with fever. I called our GP, he said he was not coming to see him because: " If they didn't help him at the hospital, what can I do?" I insisted. He came, did not visit him, just prescribed an immediate admission to the hospital and called the ambulance (...)

The patient goes back to the hospital where he is immediately discharged home because the hospital doctor said to the carer that he was not to be admitted in the hospital for high risk of complications.

C= (as soon as we arrived back home) I called his GP. I was really angry. He answered me like this :” There’s nothing I can do. It’s not useful that I come and visit him. He’s just coming from the hospital, if they weren’t able do something for him there... (and added) It’s up to you to convince yourself that your husband conditions are very serious. You can’t keep him at home. You have to take him in a place, you can’t manage him a home.”

In a few days they received a letter from their GP in which he rejected the patient and his wife

C= (after the GP rejected them as patients): Goodness me! What if we won’t find another family doctor! I came home really disconsolate.(...) one thing is being helped by supportive persons, another is being alone and feeling abandoned. (...) when you’re left alone sometimes you can’t be rational and objective enough to manage him.

DISTRICT NURSES SERVICE

Public home nursing service is called ADI (domiciliary integrated assistance). Usually this service provides technical nursing support managing iv infusions, bed sores dressings, and other specific nursing tasks that cannot be done by informal carers. This service was not designed to provide continuous care for chronically ill patients. Among the advantages of this service are the direct provision of medications and equipment without bureaucratic burden for families and direct access to free transports with ambulance towards hospitals and ambulatories. District nurses see many patients during their daily activity and their telephonic accessibility is normally limited at the early morning hours. They work in strict collaboration with GPs. DNs do not receive a specific education in palliative care and are not specialist in symptom control or counselling. They work on shifts and for this reason there is not continuity of care for their patients resulting in daily visits everyday from different professionals.

Due to the characteristics of this service only few of the interviewees experienced this service and could provide comments about it. Generally those who had been discharged from the hospital with fluid infusion or need for PEG management had the chance to be cared for by the DNs service.

Comments from those participants who received the service were collected and are now presented in terms of satisfaction or not satisfaction. Some participants did not have the service, but talked about its potential need.

Overall in 10 interviews issues about DNs was arisen. In 2 cases this was reported as a good and helpful, 8 did not report this as helpful or stated that they felt not to require it.

One carer said she was happy of the DNs service because nurses enabled her to become confident with the management of her husband PEG and tracheostomy.

CALS6 is the wife of PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic)

C= ADI’s nurses came for a period o time to medicate his PEG and tracheostomy. Now they taught me how to do and I do it by myself.

This MS patient lives alone and needs extemporary bladder catheterisations during the day. This was possible because DNs come regularly to perform it at her home. She is very happy with this service, but she recognises that in the future this service could be discontinued and she is worried about this.

PSM4 (female, 69 years old, diagnosed 42 years ago)

P= I've district nurses coming 3 times a day for extemporary catheterisations, Monday to Friday and twice a day Saturdays and Sundays.

(...)

I'm concerned about the district nursing service. I don't know how long they'll be able to continue to come 3 times a day for my catheterisations. I heard they're planning a reorganization of their service and chronic patients will be discharged.

Some carers felt that the DNs service is only temporary and that the aim of it is ultimately to discharge them. Probably the process of education to the management of the equipment that the DNs carry on aims at enabling the family to deal with the management of the tracheostomy and PEG, but this is seen by families as an attempt to leave them alone.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

C= we have the ADI service now (district nurses service). They came to medicate his tracheo wound, but they want us to learn how to do it ourselves, we're learning it. They used to come daily, I mean Monday to Friday. Now they just pass 3 times a week. Probably they will stop coming very soon.

(...)The same happens with the domiciliary assistants that come to wash him, and change his postures. They came everyday, then 5 times a week, now only 3... I feel they will leave us early. Yes, they informed us they can always come back if you call them ... but...I feel they want to get rid of us.

Some families experienced bad service from DNs and reported it during the interviews

CMS6 wife of PMS6 a 53 years old gentleman, fed by PEG, almost totally dependent in the ADL

C= district nurses came a couple of times to dress his PEG wound. I'd rather they not came! They weren't of help at all and their dressing wasn't good for him. I got rid of them quickly!

PPD5 (woman age 78, cared for by her husband -CPD5- at home)

C= I do the dressing of her PEG wound. District nurses came for the first times, then I learnt how to do it. Now it's clean and neat. I renewed it this morning. Material was provided by district nurses when they came. We still have some and I use it. When it will be finished off I will go to my GP and she will prescribe it, I guess. Now, if I were in trouble, I would go to the neurological ambulatory of the hospital where they know us well and can be of help. I do not want to have the DNs around anymore.

Another cause of frustration is bureaucracy required to obtain the service. In the following rows this is well explained: by the time needed to be seen by the DNs family

themselves achieved their outcome and when they arrived the problem had already been solved.

CALS8, wife of PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric.

C= when he had his bedsores our GP advised me that we had to activate the district nurses service (ADI). We did the papers and when they called me back to know what I needed It one month had passed. I cured him in the meanwhile and he had fully recovered. If they came earlier I'd have saved a lot of money as they give materials without any charge. On the contrary I paid everything.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

P= an informative office is needed. They should advise about your rights, bureaucracy... where you go if you have particularly needs and they answer .

C= I didn't know the jelly water. It was the speech therapist who advised me of it. Why the district nurses didn't say nothing about it?

They should improve their organisation and the homecare service office should quickly give information and support.

Some declared to use just some benefit from the homecare public service like transports

PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.

C= We opened the ADI (district domiciliary service), but we only use it because of ambulance free transport.

P= (writing on his electronic communicator) We don't need district nurses to come here.

EMERGENCY SERVICES

Emergency services in Italy are organized in a quick intervention ambulance team with a national phone number (118) that can be called by anyone who is facing an health emergency. After a first telephonic triage a nurse decides which colour code to assign to the call. This can be:

- white code (inappropriate call, refer the patient to his or her GP),
- green code (not life threatening condition, send a simple ambulance to transport the patient to the local hospital),
- yellow code (not immediate risk of death, but serious condition, send an ambulance with a specialized nurse to treat first problems and then take the patient to the hospital),
- red code (immediate risk of death, send an ambulance with an emergency team with physician and nurse)

In the local hospital patients are admitted in the Emergency and Acceptation department (A&E). in that setting if the patients conditions are life threatening they are seen by an anaesthesiologist-resuscitator that decides the appropriate treatment. Because in Italy advanced directives are not recognised by the law if a patient arrives at the A&E unconscious it is up to the physician to decide if undertake cardiopulmonary resuscitation or tracheal intubations. This is a major cause of concern for those patient (usually with motor neurone disease) who decided not to be resuscitated in case of acute respiratory insufficiency.

The following quotations from the interviews testify difficulties faced by the families and patients who participated at our interviews about this issue.

CALS8, wife of PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric)

C= my husband doesn't want to be tracheostomized at all. But he's scared that once in the hospital they could do it against his will. For this reason we had to discuss one full day because he didn't want to go to the hospital when he had an aspiration pneumonia(...) One day, it was Saturday, he had a persistent cough, fever and was breathing badly. He refused to go to the hospital because he feared that they'll put him a tracheostomy (...) in the evening I was very scared because of the incoming night. I called our sons to convince him to go to the hospital but he didn't consent to it. (...) he did not even want to call the doctor on duty for the general practise, he was really terrified. His cough was continuous and shaking. Fortunately we called this doctor and he was very good. He said he needed to go to the hospital, but reassured him that being conscious now one could ever do something against his will. at the end we went to the hospital.

Another similar situation was testified by this ALS/MND patient completely impaired in his speech, but strongly determined not to be tracheostomized.

PALS5, (man in his 70s, quadriplegic, aphonic, on a wheelchair. He tries to communicate using a computer keyboard, but is very slow. He is a retired doctor. He wants us to read his living will form that had been filled with a notary. He asks us to use its content for our research)

P= with this Living Will I affirm that I am aware to be affected by the motor neurone disease. I know that I can have a respiratory arrest at any time. If this happens I do not want to be resuscitated and I do not want no invasive ventilation, nor a tracheostomy. I ask to the emergency team that could be called in case of an acute respiratory insufficiency to avoid any life sustaining treatment on my person. This is my will and I am determined to obtain what I desire for myself even though this is not recognised by the actual Italian legislation (...)

Another cause of complain in our participants sample was due to the organization of the emergency departments.

When a totally paralyzed patient arrives for an emergency there is the risk to be treated like a not physically impaired person. This implies the risk of being let on a stretcher for a long time with a high risk of skin damage and symptoms due to immobilization.

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):

I= what happened when you had to go to the A&E?

C= it was because of respiratory troubles in the night: I called the emergency because he was not breathing (...) they came quickly and took him to our local hospital where they are not used to treat neurological patients (...), that was my fault, I talked to the people of the ambulance, but I could not convince them to drive him to the S. Luigi hospital, where he's well known.

P= it was Christmas Day and I had to spend the whole day on the stretcher! It was terrible and I can not describe what happened to my skin...

Organization can be critical when a physically impaired patient is to be cared for in the emergency department also because they are structured to face acute events, but there is a lack of coordination with the domiciliary services. Emergency doctors are invited to keep these very ill patients at home, but if therapeutic options do not work and patients must go to the A&E there is the risk of being seen as an inappropriate admission in the wrong setting. This can cause problems as reported below

CMS3, wife of PMS3 (male, age 46, blind, quadriplegic, muscular spasticity, fed by PEG)

C= (...) I lived again the drama that happened when he was in coma for his pneumonia. The paid carer was at home with him; he called me at work telling me that my husband was breathing badly, with secretions out of his mouth. I told him to call 118 (emergency). I arrived home while the doctor was visiting him. He said that he had high fever and prescribed an antibiotic. He added that fever would have gone and that it was not useful to go to the hospital (...) After 5 days of fever (I've never seen him so bad), one night he started vomiting. He vomited his soul! What a shambles. At that time in the night I could only call the on call service of the NHS. He had fever, his temperature was 40.6°C. the doctor on call did not come to visit him at home, he just told to wait the effect of the antipyretic. After half an hour he called me back. Fever was 40.5°C. He said it was going better, just to put ice on his forehead. (...) I really felt alone! I had to clean vomit, care for him, I spent the whole night with thermometer in my hands...(...). The day after we went to the hospital with our GP's prescription for an emergency admission . Another terrible quarrel. The A&E doctor looks at me and says: "What are we going to do with your husband? Anytime he doesn't feel well you bring him here?" I show him our GP admission letter. He answered me that I had to convince myself that my husband was terminally ill and so I did not have to take him to A&E, it would only be worst for him for high infection risks. Anyway, they put him in a small dark room. Time passes and nothing happens. I wanted them to do some exam to understand the origin of fever. I was scared of pneumonia. They told me I wasn't supposed to teach them their job. (...) they didn't have any available bed, he had to stay on the stretcher. They told me to look for another accommodation outside the hospital. The day after I called S. Luigi hospital, the social assistant... nothing. No one could help me (...).

Again lack of coordination can be a cause of bad service for patients who need the emergency service being affected by a chronic condition

PMS7 (Male, age 73, diagnosed 25 years ago. Wheelchair bound, anarthric, cognitively slightly impaired) CMS 7 his wife.

C= we had many problems with the hospital and emergencies services because of lack of coordination. We had an urologist who came home for catheter substitution who claimed that we had to be cared for by the urologists working in the hospital where we're in charged. Of course the latter said that we already had the former... during that quarrel we spent two years with my husband who yelled in the nights because of bladder pain. We called the emergency and told us that this was not a situation that they could manage. What are they there for?

Sometimes informal carers are more experienced than professionals working in A&E in the management of particular devices. In the following report this old carer was used to manage his wife's DuoDopa infuser connected to the PEG, but when they had to go to the A&E he was not allowed to stay with his wife and nurses did not know how to use that pump.

PPD5 (woman age 78, cared for by her husband -CPD5- at home)

C= Emergency department of our local hospital now is fine. It was not since like this few years ago. (...) We are usual customers (laughing) We had to go there twice in last month: once for a bronchitis, the second time for a stomach pain. The problem was that they didn't allow me to stay close to my wife and I'm the only one able to manage her duodenal pump, so the alarm bipped and... they should let me stay, I know there's a law for it, but they said that relatives were not allowed...

Finally for patients living alone even the call to the emergency can be a problem, like in this case where the patient is physically impaired and spends the night alone. A paid carer in the evening put her in bed and then comes back in the morning.

PMS4 (71 years old woman diagnosed 42 years ago. Quadriplegic, wheel chair bound, lives alone helped by paid carers)

I= what if something happen when you are alone at home. Do you have someone you can call?

P= yes, my cat! (joking).

I= have you ever called emergency?

P= once I did it, but they didn't come. I was alone in the night and had an acute diarrhoea due to a mistreatment that I received during a physiotherapy session. So I called the emergency and the doctor on call replied me that it was a kind of antibiotics overdose. He suggested me to wait until next morning and then to go to the hospital for controls. So I did. But I spent the night alone, with constant diarrhoea (...)

Other services: physiotherapy, speech and language therapy, respite facilities

PHYSIOTHERAPY AND SPEECH AND LANGUAGE THERAPY

The physiotherapy (PHT) and speech and language therapy (SLT) services are provided by a private non profit organization (Don Gnocchi) in collaboration with the NHS. These services are activated by both GPs and hospital specialists and offer packages of interventions at home as courses of 15-20 treatments that are to be renewed time to time. Being these services very important for both symptom control, prevention of complications and maintaining of physical mobility they are strongly required by patients. Unfortunately many participants complained about the lack of continuity of PHT and SLT claiming that, when many months pass between two courses they lose the advantages of the previous intervention.

Some participants recognised the benefits received from the PHT and SLT provided by the Don Gnocchi

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CPD1 wife of PPD1, man 80 years old, diagnosed 9 years ago.

C= we had a physiotherapist who came home for 15 sessions from Don Gnocchi service. He also called the SLT, she's very good. It helps him swallowing water.

PALS3 (male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. On NIV at home) and his wife CALS3.

P= I have a PHT coming once a week for physiotherapy sessions. She comes from Don Gnocchi service. Well she a nice girl, she makes those nice massages, lovely.

C= well, the neurologist said he has to do passive PHT. She has to work on his limbs. It's a good service. The physiatrist comes, prescribes a course of sessions and the physio comes once a week. Renewal is almost automatic.

PALS8 (male in his 70, diagnosed 9 years ago) wife CALS8

C= we are very satisfied of the Don Gnocchi physiotherapist service. They come here regularly and courses are renewed automatically. They could provide speech therapy as well, but he doesn't want it anymore

This patient lives in a village in a rural area outside the city of Turin where the NHS district directly provides PHT and SLT service. She was very satisfied with it

PALS7, (woman 60 years old, diagnosed 12 years ago)

P= we are very lucky in this area. Our local NHS district provides everything we need, very quickly. We have a superb FKT service

Other participants though reported less satisfaction about this kind of service.

Main reason of not satisfaction was lack of continuity of the home PHT service

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night)

P=Physiotherapy: they should do it at home, maybe just a couple of time per week ,but continuously.

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

C= (about PHT and SLT) we have both the services from Don Gnocchi. The problem is that they come and they are very useful, only for a course of about 15 sessions. After that we have to renew the request and obtain the authorisation. It takes months.

P= it's a plague! It should be automatic.

C= if you know that a young man needs passive physiotherapy... you can't think that at the end of the course he'll walk again

P= I'd love to have PHT in a swimming pool, they say it would be very helpful for my symptoms!

C= yes, and they have the pool at the Don Gnocchi centre. But you know what? If he goes there, there are physiotherapists to treat him in the water, but no one will take his napkin off! And because I must go to work I can not accompany him there. I can't lose other working days.

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P= I need a person with me 200% of the time, I can not do anything by myself being so paralyzed . For this reason I had to refuse admissions in places where they could do water gym.

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

C= physiotherapy is a problem. It's very useful, but Don Gnocchi provides only one course of 15 sessions and then there's a hole of 4 months. We have to pay for another therapist to have a continuous service.

PPD5 (woman in her 70es wheelchair bound.) and her husband CPD5

C= physiotherapy: we have just finished one course from Don Gnocchi domiciliary service. Now, to renew it I need a visit from the physiatrist. I went to reserve it and they gave an appointment for January 2008 (the interview happened on June 2007). And after the visit I'll need the authorization from the NHS district. And after that there will be the Don Gnocchi waiting list... Furthermore she needs more speech therapy than physio. She requires it to improve her swallowing. But she should receive it when needed it, not one year later!

Other participants complained about the efficacy of physiotherapy itself because they did not obtain the expected results or because they were not satisfied by the relationship with the therapists.

CSM5, mother of PSM5 (male, 49 years old, diagnosed 24 years ago.)

C= he had a physiotherapist who came to treat him from Don Gnocchi, but it wasn't helpful, it didn't change his conditions at all, so... There's nothing that works.

CMS6 wife of PMS6 a 53 years old gentleman EDSS 9, fed by PEG, almost totally dependent in the ADL

C= physiotherapy was provided by Don Gnocchi. First courses were fine, but this year they sent to us a physiotherapist who wasn't reliable at all. He was always late, his treatment were very light, he just moved up and down his forearm and his leg.

P= he called it physiotherapy! (laughing)

C= so I complained to the service, but they said they couldn't change him. Since then he's not doing any physiotherapy.

Other participants did not receive the home physiotherapist service and discussed about other bad experience faced when they used territory based facilities

CPD3, wife of PPD3 (male in his 70, diagnosed 27 years ago)

C= he did physiotherapy, but not at home. I had to accompany him to the PHT centre and I had to pay a taxi for it. I applied for taxi free coupons three years ago. We are still waiting!

PPD4 (woman, age 60, cared for by her husband –CPD4- at home, in DBS)

C= We went to do speech therapy to the local centre here, but it didn't work.(...) We would require domiciliary physiotherapy and speech therapy.

P= I think that a domiciliary physiotherapy would be very useful. If I have to go to do sessions to the physio-centre I feel down before leaving my house. When I arrive there I'm not able to do anything. It becomes useless. It's also a mental difficulty. When you're not expecting it, this disease takes you in. (...) you feel frail and you fear the things you have to do.

RESPITE FACILITIES

Patients living with long term neurological conditions spend long periods of time at home. Respite admissions in non hospital settings can be required to alleviate the caregivers burden and sometimes are used for physiotherapy intensive treatments or complementary therapies courses.

No participants experienced an hospice admission because hospices in our region do not offer admission for non cancer patients.

Interviewees reported comments - both positive or negative - about previous experiences in respite settings. 13 of them stated to be keen on a respite admission or that they had had a previous good experience with this kind of service. Four were contrary to this possibility for previous bad experiences or because did not want to leave their homes.

Some carer recognise openly their need of a period of relief from their task of caregiving, above all when they face the risk or the need to be admitted in hospital for some health trouble.

CMS1, wife of PMS1 a 53 years old gentleman, diagnosed 18 years ago, quadriplegic, using NIV in the night, totally dependent in the ADL

C=About assistance well, being retired I can manage him at home: but a respite care admission is necessary! When I had to went to the hospital for surgery they told me that the only possibility to care after him when I was there was to admit him in the hospital also, that's very sad..”

PPD2 (male, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias) CPD2 his wife

C= last week I asked if there were structures where he could stay just in case I had to be admitted for a stroke.(...) Yes, because I had a mild one recently and if it should happen again? He can not live alone. We would need a place where he could stay for 1-2 weeks if I had to be admitted.

Others consider the option of an admission period in specialized rehabilitation centres in order to have time to deal with other problems like the wife of this patient testifies

PMS2 (male, age 43, quadriplegic, can only move his neck, totally dependent in ADL), CMS2 his wife.

C= I could let him to be admitted at Don Gnocchi for 20 days.. he'd have his water gym, but who'd stay next to him? I can't, I must take care of my daughter too. She suffers of a juvenile form of rheumatoid arthritis and, you know, I have to go to Milan twice a week for her injections!

P= yes, in cases like ours 1+1 is not 2, but 3 or 4! (meaning the burden of care on his wife that has to care for him and their daughter)

Some participants had already experienced respite admissions and discuss pros and cons.

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This patient was very enthusiastic about her respite admissions in two centres far from Turin city, but specifically adapted for patients with advanced neurological diseases

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24 hours a day) and her husband CALS7

C= in Dr Mora centre in Pavia there's the chance to stay with her in the same room. That's important. Pavia is very well organized.

P= yes, before we stayed in Veruno. There's the neurology department now directed by Dr Pasetti. Pavia somehow is better because they have the social assistant inside that indicates all bureaucracy pathways to receive aids and services and, if necessary, directly contact your local social assistant. In Pavia you receive PHT daily, there's the SLT, nutritionists, and psychologists(...)

P= Veruno is a private, but operating with the NHS, centre where I spent a period of time. My husband was not with me. I had my electrical wheelchair (my Ferrari!) and I went around on my own. There were volunteers who spoon fed me at dinner and the nurse who did it at lunch. Personal care was provided by the centre personnel.

This family experienced a respite admission in China where they went to try an experimental therapy with stem cells. Although the therapy was not successful because the disease was not relented they had a very positive impression of the care received.

ALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

C= respite care is fundamental. We received it in Beijing in China. He had 2 sessions of physiotherapy per day, speech and language therapy, massage (...). We, as family members, had the chance to stay with him, have meals with him, sleep in his room, but we could also go out and have a rest as well because he was very well cared for by the clinic personnel. It was like an hospice!

One carer talked about the costs that she has to pay to have respite periods for his very disabled husband in a nursing home

CPD3 is the wife of PPD3 a gentleman in his 70, diagnosed 27 years ago. Cognitive impairment, Wheelchair bound, staying in a long term facility

I= how did you find this nursing home?

C= he was in the waiting list. It's his third re-entry here. Before he came for 1 month, I paid for it, and then he came back home. It's not free. The respite care admission has to be paid.

Now he's been here for 10 months. I pay a part of the charge, the rest is paid for the municipality.

Experiences were not all positive. This MS patient with four decades of diseases on her shoulders is one of the founders of the Turin MS patients association section and collaborated to the creation of respite admissions between the association and Don Gnocchi centre. Despite this she reports not to be satisfied of her periods spent in that facility

PMS4(71 years old woman diagnosis 42 years ago. Paraplegic, wheel chair bound, lives alone helped by paid carers)

Chapter 3. The Qualitative Needs Assessment

P= sometimes I spend 20 days in Don Gnocchi centre, it's a kind of respite admission. They know me very well and so give me this chance. I don't like it very much because I have to share the room with elderly women praying all the time, or crying, and after few days you became more depressed than before. Than somehow, you lose your independence. I won't go this year for these reasons

There are also situations where carer and patients do not agree about this issue. Carers feel the need of a respite period, but patients can feel this as an attempt to place them out of their homes

CMS6 wife of PMS6 a 53 years old gentleman EDSS 9, fed by PEG, almost totally dependent in the ADL

C= once per year he spends 3 weeks in Piancavallo, a rehabilitation centre where he goes for respite care.

P= I don't like it because it's a hospital. I like my home, I want to stay in my place (...)

C=Another key point are respite care admissions in the hospice. They are strongly required.

Some facility specifically created to provide respite care for neurological patients can not satisfy the needs of very advanced guests who are too ill to participate to social activities resulting frustrated and abandoned.

PMS3 (male, age 46, blind, quadriplegic, muscular spasticity, fed by PEG), CMS3 his wife.

C= he spent a period of time at Villa Fiorita, a nursing home specialized in MS care. I had to put him there because I was going to be operated and then I needed a period for recovery. I went to meet him before my operation and it wrung my heart. He was alone in a corner of the living room. While other patients were consuming their meals he couldn't because he is fed by PEG. Nobody was caring for him. His nose was dirty... once his PEG tube lost some liquid, but nobody saw it and I discovered it when he was all mucky.. He was the one in worst conditions.. the others talked to each others, had meals together.. he couldn't because he didn't speak or eat.

Some setting like general nursing homes can even worsen physical conditions of patients admitted for respite, this was certainly the case of this old couple

PPD5 (woman age 78, cared for by her husband -CPD5- at home)

C= we had a respite care admission in a private nursing home paid for the NHS. We both went there because I was very tired, having to care for her alone, and she required some physical therapy. It was our GP who filled the application form for us. She had daily sessions of physiotherapy and speech therapy. It wasn't very good. We had a double room where we could stay together, but food wasn't good at all. I had worsening of my rheumatic pain. When I came back home I wasn't able to walk anymore. Imagine that I entered walking on my legs and when I came back home I needed a wheelchair! Fortunately I recovered after 20 days.

This couple, even if previously declared to be tired and in need of help, clearly decline the possibility of a respite admission

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

P=(communicating through an alpha numerical table) Respite care? I won't go anywhere out of here (he wants her to show us the rest of the house and the garden)

C= me too, if I have to be helped I prefer someone coming here. I don't want him to risk further infections or other complications

Social services and technical support

Patients severely disabled for a progressive neurological disease are likely to need some help from the welfare social service. In Italy social assistants are professionals working for the local offices of the ministry of the welfare. This service can be accessed by anyone with economical hardship, physical disability or social impairment. The main help provided by these professionals to families in which a severely disabled person lives is based on financial benefits like the sick benefit, accompaniment check or other grants to be used to pay for home assistants that care for disabled patients. They also provides information about available services to the person and can refer applicants to transport companies to obtain ambulances or equipped taxis to move patients toward hospitals or ambulatories for routine visits, physical therapies and so on. Finally these offices are those providing technical equipment like electronic communicators, wheelchairs, orthopaedic beds, aids and devices.

EQUIPMENT DELIVERY, FINANCIAL BENEFITS AND TRANSPORT

Interviewees reported their experience when they met these services.

Some were very satisfied for the received benefits. Often this is the case of ALS/MND patients because this has been recognised as a rare condition in Italy and therefore neurologists can directly prescribe the necessary equipment. More difficulties were met by families of patients with MS or movement disorders that do not have the same facilitations

PALS3 (male, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. On NIV at home) and his wife CALS3.

C= for aids provision bureaucracy is simple. It's enough a prescription from the neurologist and NHS district office provides everything.

PALS7, (woman 60 years old, diagnosed 12 years ago, quadriplegic, dysarthric, dyspnoeic, NIV 24 hours a day)

P= I have 2 ventilators. Now they approved the delivery of a new battery. In case of an electric black out I have about 20 hours of autonomy (...). We are very lucky in this area. Our local NHS district provides everything we need, very quickly. We have a superb PHT service, we had all the aids: wheelchair, bed, mattress, hoist, electrical wheelchair.

Even though the social system seems reasonably efficient in the normal equipment provision problems can arise when very expensive devices are required

Chapter 3. The Qualitative Needs Assessment

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized) and CALS1, his wife

C= we obtained our aids: wheelchairs, hospital bed, air mattress, ventilators, aspirator, in-exsufflator etc. But when we asked for other needs like the computer based communicator, which is very expensive but can dramatically change my husband's quality of life) well that was a tragedy!

Some families experience bureaucratic difficulties due to numerous steps to go across cyclically

PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.

C= aids delivery is a long procedure. Each month we need a prescription from our GP, an authorisations from the NHS district office, than we should go to the pharmacy to retire. But now they contract private companies to deliver this material. The problem is we don't have a person like the pharmacist to talk face to face when a problem occurs. And the service from one of this company is fully unsatisfactory. With other things go well.

PALS4 (male, 61, quadriplegic, on a wheelchair, cachectic, severely dyspnoeic in NIV diagnosed 3 years ago) and his wife CALS4

C= our lung specialist prescribed a new mask because he had this skin sore due to the previous mask. If we had to wait for the normal procedure of the NHS we were to wait 20 days...luckily he had one and gave it us (...)Procedures to have aids authorizations and delivery are tricky. We fortunately found a good clerk at the call centre. Many times he helped us when we were in trouble because companies delivered wrong material or we run out of it.

C=. Even if I immediately go to our GP for prescriptions aids provision procedures are long.

Transports can be critical and expensive for patients with movement disability and families require some financial support as testified below

PPD3 (male in his 70, diagnosed 27 years ago. his wife caregiver, CPD3

C= a free transport service would really be needed to go to the hospital visits. They wanted me to bring him there with an ambulance, even if he could go by taxi. I had to pay for this and it was very expensive. (..)

When participants were not satisfied of the social service it could be simply because they had never experienced it being not confident about the possible help

PMS1 (male, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):

I= what about the social assistant: did you ask this service some help for you wife who has to care after you day and night?

P= we have never been to the social assistant. We heard that they can send us someone at home. I think it would only be a trouble, we have our routine and don't want someone else wandering around the house.

Financial support is very important to help families to pay private home carers to help in daily assistance to their loved ones. Interviewees reported their experience about this crucial point.

PPD4 (woman, age 60, cared for by her husband –CPD4- at home, in DBS)
C= nowadays she has her invalidity pension and the accompaniment check. We don't have any other sick benefits nor paid carers helping us at home. No domiciliary services at all.

HOME ASSISTANCE

When social assistances provide paid carers to families caring for very disabled persons often packages of services are tailored around the needs of the family. Our participants reported some criticisms about their perception of the results of this intervention. Sometimes the help provided by the social service is seen as not helpful like if they had to accept prepared package of assistance not fitting their needs

CMS6 wife of PMS6 a 53 years old gentleman EDSS 9, fed by PEG, almost totally dependent in the ADL
C= the welfare assistants provide for us a girl who come 3 hours a day, but honestly it's not enough. They give us a podologist once a month and the barber. He doesn't want this barber, he wants to go to the one he likes.

Paid carers need specific education to be of help. Sometimes their presence is not enough to provide relief

PALS2 (male, age 39, tracheostomized, quadriplegic, diagnosed 14 years ago, totally aphonic) and his wife CALS2.
C= we have a paid carer who comes everyday and a second one who comes some afternoons and Saturday in the morning. But this organization seems me stupid and a strong limit to the service. They offered us a domiciliary assistant, but she can't aspirate him. So she's not of use at all. They said their nurses are on call, but I can't call them each 20 minutes to aspirate him. They should train the assistants to aspirate!

In some case the time spent by the paid carers is not enough to provide adequate help

PALS6 (male, age 50, diagnosed 13 years ago, tracheostomized, bedridden – wheelchair, quadriplegic, aphonic), CALS6 is his wife
C= now there's a domiciliary assistant paid for the welfare office. She's very good and collaborative. But she's paid only for 2 hours a day. In the beginning this was enough, but now it's not. She often has to stay longer to help me with him.

Many participants advocated the need of paid carers to provide help for patients' necessities.

PPD1 (male, age 80, cognitively slow but able to understand and severely disabled in his movements. During the interview was admitted in the neurological ward for symptom control) CPD1 his wife.

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C= if they could come in the morning to help me in his toilette, or shower him... because it's getting hard to me.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

C= home service: we have domiciliary help provided by the welfare assistant. 3 mornings and 2 afternoon they come to help the carer to mobilize him. But frequently they told us that as long as the carer is able to care for him they weren't keen to do it. But even if the lady who cares for him does it, she doesn't have to. It's their task!

TECHNICAL ASSISTANCE

Comments were also captured about the technical assistance provided by those companies which deliver technical equipment such mechanical ventilators that require revision and technical assistance

PALS3 (man, age 60, on a wheelchair, severely dyspnoeic, diagnosed 5 years ago. In NIV at home) and his wife CALS3.

C= ventilator has a free assistance guaranteed 24/24. we have the free telephone number. But we never needed.

PSLA9 (man, age 77 diagnosed 8 months ago) and his daughter CSLA9.

D= we have 2 ventilators. The only problem is that batteries doesn't last for long. Technical assistance is very good.

SPECIALIST PALLIATIVE CARE SERVICE

After participants reported the lived experience of the disease, discussed the unmet needs and provided comments on the received services, researchers asked their opinions about the experimental new SPCS that was under construction. They explained the aims, the organization, the philosophy of palliative care and the resources available asking the interviewees their opinions about its usefulness.

15 participants stated to be keen to receive a service similar to what a SPCS can provide, (sometimes just partial like volunteers or physiotherapy) none of them declared to be contrary to such a service

Some comments are now presented

PPD2 (man, age 70, fluctuating motor symptoms with freezing episodes alternate to severe dyskinesias) CPD2 his wife

C= I hope that if in the future he'll have further problems in movement a domiciliary service with the characteristics that you explained to us will be available.

I= do you think that now a SPCS could be useful to you now?

P= sincerely I've never felt this need till now. For the future it will be welcome.

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PMS1 (man, age 53, diagnosis of MS 18 years ago, quadriplegic, using NIV in the night, CMS1 his wife):

I= Would you accept a SPCS at home?

C= I was very happy when our GP started to do regular visits each month: if there was something else it would be welcome. Our house door is always open.

P+C= very useful would be a service on call.

PALS1 (male 48 years old, bedridden quadriplegic, totally aphonic, tracheostomized.) and CALS1, his wife

C= we would appreciate a home care service, available at weekends too. Someone who can substitute me or the carers for some periods of time.

CALS8, wife of PALS8 (male in his 70, diagnosed 9 years ago wheelchair bound, totally anarthric)

C= we both would be definitely pleased to receive a SPCS service like yours at home. If you start, please contact us.

PPD4 (woman, age 60, cared for by her husband –CPD4- at home, in DBS)

C= we'd really need a service like the one you are presenting. We would require domiciliary physiotherapy and speech therapy.

P= I think that a domiciliary physiotherapy would be very useful.

Summary

Participants provided a number of precious information about the existing services for people severely affected by neurodegenerative conditions living in Turin area. Specialist clinics services were generally reported as positively satisfying the patients' needs, although this is not equally shared among the different conditions. Primary care home services seemed lacking because they are not organized to meet the chronic needs of very disabled people. Those patients requiring technical equipment like mechanical ventilators were satisfied by the technical assistance available. Specialist rehabilitation domiciliary services, although present, did not provide homogeneous and continuous support and participants reported difficulties of access to the services. Finally social and welfare services seemed far from being of help to the social needs for this population. A service with characteristics similar to those provided by a SPCS was strongly advocated by the majority of the interviewees.

3.5.3 Focus Groups results

In this section the results of the 3 focus groups with professionals involved in the care of patients severely affected by neurodegenerative conditions will be shown.

3.5.3.1 Participants

From May 2007 to August 2007, 3 focus groups with a total of 11 professionals involved in the care of the patients who were interviewed and whose results have been reported in the previous sessions were run.

All the participants of the focus groups were professionals working in the 2 hospitals of Turin involved in this study: Molinette hospital and S. Luigi Gonzaga hospital. They were recruited from the specialist clinics of these hospitals directly involved in the care of ALS/MND, MS and PD and movement disorders.

A list of participants is displayed in table 3.8

focus number	participants	gender	professional role	specific interest
1	EPD1	f	neurologist	PD
1	EMS1	f	neurologist	MS
1	EMS2	f	neurologist	MS
1	EMS3	m	neurologist	MS
1	EPC	f	neurologist	PC
2	EALS1	m	neurologist	ALS
2	EPD2	m	neurologist	PD
2	EMS4	m	neurologist	MS
3	EALS2	m	physiatrist	ALS
3	PHT	f	physiotherapist	ALS
3	SLT	f	speech&language therapist	ALS

Tab 3.8: Participants of the focus groups

EMS= Multiple sclerosis expert

EPD= Parkinson's disease expert

EALS= Amyotrophic lateral sclerosis expert

EPC= Palliative Care expert

Nine participants were physicians:

- 8 specialist in neurology. Of these 3 were MS specialists, 2 PD specialists, one was an ALS/MND specialist and one had interests in palliative care.
- 1 was a rehabilitation specialist working in a respiratory centre where mechanical ventilation and cough assisted devices were prescribed.

The remaining two professionals were one physiotherapist and one speech and language therapist both collaborating with the rehabilitation specialist.

3.5.3.2 Focus groups general results

The 3 events were run in the selected hospitals. The focus groups were advertised at the participant services through the direct involvement of the directors of the centres. Professionals interested in the participation were volunteers and did not receive any payment for their presence.

They signed the participants' module of the informed consent as required by the local Ethics Committees. Each event was conducted by 2 researchers, were audio and videotaped and transcript verbatim was performed. The duration of the groups was of one hour of time on average. All participants talked about their professional experience in the care of people severely affected by neurodegenerative conditions.

The general framework of each event was based upon a personal introduction of each participants. They were asked to declare their experience in this field, where they were working, their annual mean case load and the characteristics of their patients.

General rules about the secrecy of the contents of the event, privacy and professional behaviour were discussed. Interaction among participants was encouraged. The facilitator introduced the aims of the study and declared the interest in the following aspects:

- Physical, psychological, social and spiritual unmet needs of the people severely affected by neurodegenerative conditions
- Impact of these needs on the families, caregivers burden
- Their knowledge about the service available for this population in Turin city and Turin area, quality of the home care of their patients
- End of life issues: how and where do their patients die, choices and policies about end of life decisions
- Opinions about the involvement of a SPCS for the home care and the hospice care.

A content analysis of the transcript verbatim was then performed by 2 researchers and then main contents were grouped in a common file.

Main results coming out from the three focus groups are presented in the next session

3.5.3.3 Focus groups content analysis

A content analysis of the main themes arose from the three focus groups are now presented.

Data are grouped in four main areas:

- General experience
- Palliative care unmet needs of the patients (including caregivers burden of care and end of life issues)

3.5.3.3.1 General experience

In this section the following the analysis of the following aspects will be presented:

- number of patients severely affected by neurodegenerative conditions potentially eligible for a SPCS
- available services
- comments about a new SPCS

Number of potential users

The potential number of people severely affected by ALS/MND, MS and movement disorders living in Turin area can be estimated by prevalence data of the specific

conditions, even though there is a lack of information about the advanced stages of the specific diseases. Participants were asked to report the number of patients with the diagnosis above indicated and to define how many of these could be considered in advanced stage and, therefore, potentially eligible for a new SPCS.

Data were different depending on the dimension of the centre where participants were working for and for the different diagnosis. Data are not meant to provide information about the prevalence of the disorders, nor to establish how many people severely affected by these conditions were living in Turin or its metropolitan area, but to consider the proportion of people in advanced stages compared with the overall caseload of the involved centres.

MS specialists reported about 10% of their patients to be in advanced stages

EMS3: "I'm the director of the MS centre in this hospital. We can estimate in 80-100 severely affected MS patients cared for by our centre. We must define what we mean for it, but it is about 10% of all our patients"

EMS1: "we can add 15-20 more MS severely affected patients of our service"

EMS4: "it's hard to estimate for the MS. I do not know if we lose the most disabled ones, usually we see them one or twice per year. I think that bed bound patients seen in our service are about 25-30".

ALS/MND report a higher proportion of patients being in an advanced stage and not able to go to the ambulatory

EALS1: "An estimate of ALS patients cared for by our tertiary clinic can be of 70-100. About 25 are the ones that would require a domiciliary service"

Professionals working in the respiratory centre report higher numbers of ALS\MND patients with respiratory problems cared for by their hospital service

EALS2: "if we consider both patients in non invasive and invasive ventilation we have about 100, only ALS. We have a high turnover"

PHT: "in relation to the dimension of our service it's a big load"

Movement disorders are much more prevalent and very difficult to predict in terms of potential SPCS users. This neurologist working in the tertiary clinic for Parkinson's disease reports:

EPD2: it's difficult to say because the worst ones are lost at follow up. As long as they can't come to the ambulatory we don't know what happens to them. But compared to the numbers that my colleagues are citing now I fear that for all extra pyramidal syndromes, considering also demented patients, we should imagine a number 100 times higher. I'd estimate hundreds of patients eligible for a home care service in Turin area"

Available services

When asked about the availability of specific services for people severely affected by neurodegenerative conditions in Turin city and metropolitan area, professionals described an heterogeneous picture.

ALS/MND though being a rare condition seems to have a better network of specific neurological services above all aimed at supporting advanced patients at home. In spite of this transports and social benefits are difficult to obtain

EALS1: "For home bound patients we can provide a domiciliary service with neurologists than go and visit patients in their homes more or less each 2 months. No transport services for ALS patients is provided. Social benefits are difficult to be obtained. Our neurological service is free of charge for them, but they need paid carers and can receive social benefits only if very poor and lonesome. They are charged for everything they need".

This is not true for movement disorders where neurologists are concerned about the quality of the assistance provided by primary care alone

EPD1: "I don't know what happens when patients are at home: how are they managed, is the carer able to recognize complications? Are GP's able to manage it? It happens that our patients go to A&E of other hospitals for bad dyskinesias, maybe because they swallowed 5 tablets instead of one and their therapy is completely changed when it would have been enough to wait the drug effect.. and consequences are worst!"

EPD2: "I think that a good domiciliary service can improve many outcomes for PD advanced patients. One aspect can be PEG management for those patients using DuoDopa continuous infusion via PEG. We had local complications 100-1000 times more frequent than other experiences in Northern Europe. We think this can be due to a better home care service provided there... this is a palliative intervention because we don't have any therapy that can improve survival in PD"

For MS there is no specific home care service except for the transport service provided by the local patients association that is seen very positively by professionals

ESM4: "AISM (patients association) provides a transport free service to MS patients. This allows also very impaired patients to come to our centre to be visited. This doesn't happen for other categories of patients.

When asked about their feelings about the quality of services available for their patients MS specialists show frustration and impotence

ESM2:" we have structural limits, but specific projects aimed at this aspect should be enhanced. A good domiciliary service ease our job. Patients are more satisfied, better managed and more compliant with therapy (...)I feel impotent. There are structural limitations. We do our best, but...."

Even respiratory team report frustration about the impossibility to provide telephonic support to domiciliary patients

EALS2: "most problematic patients have my personal cellular number (I must say that in our service this is not always seen as positive). But usually they have to call here the service and the secretary office isn't always on call, just during the normal office time."

Some participant discussed about lack of instruments and adequate education in their hospital based teams about specific issues very common in palliative care like, emotional support, supervision meetings or team work strategies

PHT: “we don't have any clinical multidisciplinary supervision, we just have a project for us physiotherapists”

SLT:” we shouldn't answer to our patients’ emotions and deep questions just with our feelings or emotions. But here any operator has its own background and education, we don't have common pathways or guideline. It's not easy”

EALS2” these are hard experience that we have to keep inside. We don't have any form of meeting among ourselves because of lack of time”

3.5.3.3.2 Professionals points of view about a new SPCS

Participants seemed very interested about the creation of a new SPCS for neurological patients.

EALS2: “We were just waiting for a service like the one you're organizing.”

SLT: “It would be really important for patients and families because they wouldn't feel alone. They complain about ADI service not only because they're not efficient, but because they don't visit them regularly, above all in the first days after discharge from the hospital they have to manage 24\24 a patient, perhaps with a tracheo.. it's not easy.”

Home care for advanced stages is strongly advocate to avoid hospital complications for very ill and frail patients

EMS3: for advanced stages we should think about strategies to reduce infections rates. Enhancing home care is fundamental. Hospital admissions should be avoided in far advanced MS, but PD also, above all in winter time, because it expose these frail patients to aggressive bacteria, with high risk of death and bad agony due to prolonged antibiotic treatments.”

EMS4: “a service able to treat complications at home would really be important for this population. Manage fever occurrences, prevent urinary infections etc.”

Professionals are confident that a SPCS could reduce inappropriate hospital admissions or unwanted treatments:

EALS1: (do you think a specific SPCS for this population could be of use?) ” Yes, definitely, above all in these situations where a close follow up could prevent inappropriate treatments.”

EALS2: “absolutely yes. Often patients come here because they know that we are here. Rarely I go to visit them at home. A SPCS could really prevent inappropriate admissions. I think mostly at NIV patients that don't want a tracheostomy. They need a strong and supported domiciliary service.”

SLT: “I think that about the place of death many relatives call the emergency when they feel that the patient is choking to death. I think that must be

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overwhelming, so it's an instinct to run to the emergency. If a palliative care team could be present at home and help.. that could be helpful.... but being alone must be scaring.

EPD2: "I think it would help very much for caregivers education. This is so important. They should be able to recognize early signs of aspiration, cough etc. families aren't easy to foretell. Sometimes they would like to stop treatments very early, in other situation they want heroic treatments in far advanced stages.. Domiciliary treatment of infection would prevent hospital admissions in wards were unprepared professionals aren't able to recognize simple problems: any simple infection can precipitate general conditions of PD patients."

Another specific role of a SPCS identifies by professionals could be to provide support and education to those carers who face difficulties in the management of complex devices like mechanical ventilators. They recognise that the training period that is provided in the day hospital is not always sufficient to enable the carers to be self sufficient at home

SLT: "I think that a home training to continue education on what they learnt here would be precious for our patients and carers"

EALS2: "this is a problem: we educate carers to use respiratory devices only for 2-3 hours here in day hospital, then we see them after a 3 months period of time. Personally, if I foresee problems due to a difficult in understanding I tend to call the family after 1 week. but this is not the normal procedure and not everyone here agree. It is not uncommon to recognise that after three months the ventilator is still at home packed in the original box because they did not feel able to use it"

Even rehabilitation centres in our area are not satisfactory for all patients, above all for the very advanced one, because they require continuous assistance and these facilities are not always prepared to care for totally disabled people, and because they tend to be placed far from the city.

EMS1: " they really need respite, I mean to be relieved a bit. But then patients admitted in facilities feel abandoned. There's another centre (Peveragno) where MS patients can be admitted for respite, but they feel abandoned when are very disabled because they are placed on their wheelchair, fed by PEG, they cannot communicate with the other guests and do not have their families with them.

EPD2: "we can admit PD patient in a clinic where they are treated with physiotherapy and assessed and followed up annually after Deep Brain Stimulation. The problem is the waiting list that is very long. Frequently we have to declare that the patient needs physiotherapy, when it's just a respite care, otherwise it wouldn't be reimbursed. If there was a place of care for respite admission it would be really useful in our area.

Terminal care could be another interesting option for our participants

EALS1: “terminal sedation for acute respiratory failure in ALS/MND patients usually happen at home or in our ward, theoretically it could happen in an hospice if a bed would be quickly available when needed”.

Last role discussed about palliative care facilities was the role of the hospice for respite care or terminal care.

Professionals revealed the difficulties to find appropriate places to admit very disabled patients for limited periods of time, recognizing that hospital wards are not a good solution

EMS1: “Respite care wards able to admit these patients when far advanced are lacking. Hospital wards are not the best place for them. In last 3 months we had 3 MS patients admitted in the hospital for routine assessment and procedures who had hospital acquired infections. 2 of them died. If infections could be managed at home it would be better for patients. 1 patients had 6 episodes of pneumonia at home and always recovered, when he had the first in hospital died for it.”

When patients are young nursing homes are not seen as good solutions because of the high age of the other guests

EMS2: “MS patients tend to die at home, even because there are not so many facilities for them. The only ones available are geriatric nursing homes. So a man in his 30ies, not cognitively impaired can't stay in a nursing home! There is a clinic in Mappano held by nuns where we can admit some young patient with high disability”

3.5.3.3 Palliative care unmet needs

Professionals discussed about the unmet needs faced by their patients. They underlined some difference between this population unmet needs and cancer patients in general. Problems were grouped in physical, psychological, social and spiritual ones as previously presented in the results of the interviews. End of life issues and caregivers burden of care where also explored and are presented in the following paragraphs associated to the specific related patients problems.

Physical needs

Each specialist introduced the symptoms and other physical features typical of the different diagnosis. These conditions show peculiar and common symptoms due to the natural evolution of the illnesses. Some neurologist declared that their perception of patients' symptoms is not always correspondent to what their patients report:

EPD1: for PD advanced patients physical needs are evident from the observer point of view. Patients tend to accentuate them during the visits. But they're also objective.

EMS2: “MS patients with EDSS > 7,5-8 they all are wheelchair or bed bound. Physical needs are so evident. All our patients are treated for symptom control

(...) And then a strong rate of them have cognitive impairments, so their symptom perception is different from the professional perception. .”

The overall symptom load in these conditions seems to be very high as described by the experts who participated to our groups:

EALS1: in ALS physical suffering is very high. Not very much pain, because pain is mostly secondary, it's not due to the disease, but.. There are certainly symptoms to be controlled that can justify a medical intervention at home. Other palliative intervention required are postural education (very useful for postural pain), respiratory management and skin care. For skeletal pain physiotherapy and repositioning are fundamental and effective.

EPD2: many aspects in PDs patients are similar to those descript for ALS ones, high symptom load and physical suffering. I cannot say if monitoring these patients at home can improve their symptom control.

In the following paragraphs some specific physical symptoms will be explored

1. Pain

Pain control is considered as one of the most important goal to be achieved in palliative cancer care. Neurologists and the other professionals involved in the care of people severely affected by neurodegenerative conditions express different opinions about the prevalence and the impact on patients' QoL in their patients.

Some recognise that pain is an issue, but they feel impotent about its control:

EPD2: Pain is definitely misdiagnosed in PD and related disorders. It's frequent and we are impotent. Pain killers are used, but generally it is an undertreated symptom.

EPD1: Pain and gait problems and are the most affecting on quality of life. For pain control we do what we can do. At a certain point it seems unavoidable.

On the other hand some participants think that pain is not a very important symptom for their patients or tend to consider this symptom not as caused by the disease, but a possible complication. From their words it seems that this symptom is not on the top of the list of their worries about their patients' care.

EMS4: in MS pain isn't so frequent. There are painful symptoms due to complication of chronic immobility. Those patients are often referred to pain clinics. But it's not so frequent. Results aren't so clear. Many patients are cognitively impaired so we don't understand very well.

EALS1: In ALS pain is not so relevant. In advanced stages, when the patient is tracheostomized and can live for a long time postural pain can be a problem.

Painful syndromes are frequent in these conditions and are labelled with different names like cramps, spasms or painful fasciculations.

FKT: Spasticity in MS, muscular hypotrophy, cramps or fasciculations in ALS and rigidity in PD impact very badly on patients QoL. Not only because of limitations in their job, but also because they can be painful and impact on family relationships. They can't have meals with their loved if they can't swallow, they lose many important moment of normal family life.

EMS2: Spasticity is an important symptom not always controlled with oral drugs such as baclofen, so we obtain good results by positioning baclofen intrathecal pumps.

When asked about the use of opioids professionals report a general lack of confidence with these drugs for many reasons ranging from fears of side effects to lack of experience.

EPD2: in PD and related disorders these drugs are scarcely used. There is the problem of chronic constipation the always affect our patients and that can be worsened by opioids (like tramadol). for this reason in my personal experience they're not so frequently used. Patients complain about leg pain gait bound and usually complicated by osteoarthritis, back pain etc. we refer them to the pain clinic where they're treated with local injections of steroids, or prescriptions of acetaminophen, and steroids if necessary. For this reason I must say that my experience with opioids is like zero.

EALS1: Generally these pain aren't very responsible to opioids, they respond better to frequent changes of their positions. On average they're used at low doses, but some patient can need high doses. For analgesic purposes also Fentanyl patches are used, with dosages of 25-50 µg/hr.

Other drugs, generally used as adjuvant for pain therapy in chronic cancer pain, seem to make neurologists more confident about their use, even though in the Italian National Health Service they are not reimbursed if prescribed for pain not cancer related. ALS/MND being labelled as a rare condition is an exception:

EPD2: adjuvants such as anti epileptic drugs are effective for neuropathic pain. The problem is that for our patients these drugs aren't reimbursed by the NHS and so patients can't afford it. It reduce our therapeutic choices.

EMS4: we also used a lot gabapentin or pregabalin, but now we can't use it anymore because of their cost.

ALS1: we use gabapentin and pregabalin for fasciculations and cramps. Pregabalin is helpful for anxiety also. Fortunately ALS is considered a rare disease and so all drugs are refunded by NHS

2. Dyspnoea and respiratory symptoms:

Shortness of breath is commonly faced by ALS/MND patients, but palliative options seem to be scarcely known in our reality.

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EALS2: dyspnoea is the main symptom. Due to the respiratory pump deficit. With anxiety it create a self sustaining loop. This is particularly true in ALS where patients are aware of prognosis and psychological reaction doesn't help.

Experts in respiratory management are strongly committed in this symptom relieve, but they focus their interventions on mechanical ventilation.

EALS2: Physiotherapy is ventilation. Even when patients are not yet in respiratory insufficiency a mechanical ventilation (maybe only for 20-30 minutes per day) can expand pulmonary parenchyma acting as a PHT. It can help also to mobilize bronchial secretions

Other specialist do not report dyspnoea as a major concern, they focus on pulmonary infections as a possible cause of death rather than a cause of suffering:

EMS3: in MS respiratory troubles are less frequent than in ALS. In advanced stages we have seizures and recurrent respiratory and urinary tract infections. These problems can cause patients death.

Inadequate cough is seen as a common problem for neuromuscular patients:

EALS2: another symptom is weak cough. It's more frequent in MS than in ALS. It can occur in patients with normal ventilation without respiratory impairment. It cause bronchial secretions, choking etc. This can cause the difficult decision if tracheostomise these persons or not. Most of them are tracheostomized at the end. we have a patient who was tracheostomized for week cough. he has a ventilator at home, but he never use it, he just need to be aspirated. other drugs to reduce secrections aren't so useful and then just delay the tracheo decision. you can prescribe antibiotics, if they work infection will come again. it's a tough task for us. often we feel impotent because of this.

Treatment of respiratory symptoms is generally based on mechanical ventilation that can be invasive or not. Compliance to this vital support is reported as problematic by professionals. There is the feeling that it can be related to the attitude of patients to accept and cope with their illnesses and disabilities:

EALS2 : control of breathlessness is a very subjective issue. Some patient is very satisfied,(sometimes it looks a little big exaggerated like if he was trying to be rewarding us), others are more negative and tell you that it didn't help at all. we may say that, on average, there's a good satisfaction in our patients. only one refused it completely.

FKT: I think that NIV acceptance depends on the psychological acceptance of the disease. Patients with little acceptance of the disease tend to have more difficulty to accept respiratory help. Usually they can appreciate the positive effect after a period of NIV. Patients with better social situation cope better with NIV. The most informed ones surf the internet and it can arise questions about the NIV. Very important is the degree of information they received and their level of acceptance of the bad news.

Opioids are not commonly used to relieve shortness of breath. Some believe their use is limited to terminal sedation, others confess not to be experienced to their use:

EALS1: to control dyspnoea as symptom we don't use morphine because the symptom is very progressive and its use's not suggested by international guidelines, so we use it for terminal sedation only. Opioids have always been used with caution, like benzodiazepines. recently they have become of common use, but experience is not yet so wide.

EALS2: Opioids for dyspnoea? I heard about it, but we're not experienced at all

3. Drooling and mouth problems

Participants mentioned this group of physical problems as frequent and difficult to treat

EPD2: Drooling is a symptom difficult to treat even if you visit the patient daily.

EALS2: Drooling is a big problem. Some anticholinergic drug like scopolamine works, but..

EALS1: Than there are difficult symptoms in general like drooling. it's a very distressing symptom, very hard to control. basic research is required to better understand and manage this symptom. Amitriptyline and scopolamine are used, but not so efficient.

4. Participants listed other physical uncontrolled symptoms experienced by their patients

EPD1: our patients experience gait problems, tremors, rigidity, non motor related troubles, urinary problems, gastrointestinal problems, dysphagia, many other problems.

EMS2: out of spasticity and pain, urologic troubles are very frequent in MS and very advanced patients are mostly catheterized.

EMS2: Most invalidating symptoms in cognitively not impaired patients are mobility troubles, dysphagia, speech impairment and fatigue.

Psychological needs

The psychological problems faced by patients severely affected by neurological conditions appeared in the discussions proposed by professional carers.

Depression, dementia and cognitive changes, difficulties in coping with the disease and the continuous losses, problems related to providing adequate psychological support emerged as important themes seen by the professional point of view.

In this paragraph data related to these issues are presented.

1. Anxiety and Depression

Anxiety and depression are common findings in people affected by advanced diseases. For some specific condition like PD and related disorder this depression can be a specific feature of the disease, sometimes preceding the diagnosis

EPD1: in PD the impact of these symptoms is equal to motor symptoms. Depression, in the end, affects everyone. Often it precedes diagnosis.

Depression can appear in all diagnosis as the consequence of disability and loss of self control. Therapies prescribed to limit the progression of the disease can worsen depression as well

EMS1: depression is present at every stage of multiple sclerosis. There's always fear of something worst that could happen. In advanced stages they recognize that they won't recover. We must add that some of psychological symptoms are worsened by specific treatments. For instance interferon is contraindicated in patients with psychiatric co-morbidity.

Anxiety is often associated to depression expressing the fear for the future and the uncertainty of the present.

EMS2: I confirm that depression is common in all the stages of MS. Being the course of the disease unpredictable patients have anxious conditions even in periods of stability. High risk of relapsing cause anxiety and depression. In progressive forms of MS drugs aren't effective and so patients must assist the progression of their disability without any chance of improving.

EALS1: anxiety and depression are obviously common in ALS/MND as well. These symptoms are challenge our assistances in parallel with the clinic activity (physical symptom control. Above all long surviving patients tend to arise these problems.

Therapies offered to relieve these symptoms are available and effective, but patients' compliance is not always optimal and difficulties in the follow up of very disabled patients can affect the result.

EMS3: anti depressants are effective, but it depends if patients want to receive it.

EPD1: SSRI (inhibitors of the reuptake of serotonin) are effective, even if a compulsive obsessive disturb occurs, but they must be titrated and patients are to be followed up carefully. This is not always the case of our physically impaired patients who are hardly transportable.

The importance of a psychological support was strongly recognized by the participants and its strengths and weaknesses will be discussed later on in this subchapter.

2. Dementia and cognitive changes

With the disease progression cognitive disabilities can appear in all conditions. This is particularly frequent in movement disorders and multiple sclerosis where frank dementia or psychiatric symptoms are common, but in motor neurone disease as well fronto-temporal dementia can occur causing behavioural changes and problems related to decision making.

EPC: thinking about Parkinsonisms and ALS I'd highlight speech impairment without cognitive impairment that cause bad aphasias and have an awful impact on QoL. Dementia and other cognitive impairment impact badly on families. Physicians tend to focus on memory disturbances, but families are more concerned about other psychiatric features like aggression.

Patients with high physical disability are difficult to assist at home and when admissions are required their cognitive status can challenge this decision. Nursing homes, facilities usually admitting elderly demented people, cannot be the best place for young not cognitively impaired patients

EMS2: MS patients tend to die at home, even because there are not so many facilities for them. The only ones available are geriatric nursing homes. So a man in his 30ies, not cognitively impaired can't stay in a nursing home! There is a clinic in Mappano (a village near by Turin) held by nuns where we can admit some young patient with high disability. This is the only facility that provides admissions for very disabled MS patients, cognitively impaired or not. When they are demented probably they do not understand where they are, but when they are competent it is very hard for them to accept to stay and eventually die in that setting.

3. Coping with the disease progressions and continuous losses

Professionals are aware of the difficulties faced by people experiencing relentless disability.

They report their difficulties in providing help and the resistance of patients to accept aids that could ease their invalidity because when they propose these devices patients understand that the disease is progressing and they do not want to accept it.

EMS3: decisions about PEG or baclofen pumps cause high psychological suffering to patients. (...)MS patients hardly accept efficient therapies even when they are very effective on symptomatic outcomes. Patients tend to refuse because to accept it they should accept the progression of their disease. Once they accept it they're happy with their decision. So it's not enough to offer an aid, often we receive a refusal. This is frustrating because you, as a professional, know that they need that help, but you recognize their feelings and often have to wait until they cannot cope anymore to prescribe these aids.

Physicians report to be challenged by questions about relational aspects like sexuality, arguments in which they do not feel to be sufficiently prepared to provide help

Chapter 3. The Qualitative Needs Assessment

EPD1: they always ask me about this! When they enter in the ambulatory and I ask How they are... they say they want to talk about their sexual courses. Often is the first thing they mention. It is a little bit embarrassing for me because I do not know what to say..

Decision making and the acceptance of the disease progression are strongly related. Life sustaining treatments, such as mechanical ventilation, are effective on symptom control and can significantly extend life, although can not relent the disability progression. Professionals working in the respiratory clinics recognize the importance of the psychological support on the compliance of patients and families with these devices, and, as a consequence, on the outcomes of the treatments.

EALS2: psychological distress in our patients is the most distressing to me as a professional, and often we work with the psychologist on this theme. Some patient cope extremely well. We have some that being tracheostomized and quadriplegic can surf the web and send us e-mail. Unfortunately others do not cope at all. Usually these are the ones who do not accept the disease and its complications

SLT: Many patients and their families aren't prepared at all when they have to decide. I don't mean they should decide serenely, but at least they should be prepared. Three out of four patients attending our department are not prepared to take the decision about tracheo when required. The prepared ones had previously received an adequate support.

PHT: We saw patients with a tattoo on their throat reporting that they didn't want to be tracheostomized...

SLT: yes, but we also have to say that some patient refused tracheostomy until the moment they became breathlessness. At that time they changed their mind. When they felt they were choking they asked for tracheo.

Patients who cannot cope with their invalidating conditions often arise issues about end of life decisions. This aspect of caring is a main concern for professionals working in the hospital setting.

Discussing about euthanasia, suicide and decisions related to patients will to end their lives evoked an interesting debate among our participants

EPC: I remember a 60 years old ALS patients who asked his neurologist if he could end his life. He had just been diagnosed. He discussed with me and the other colleague about euthanasia. He was a cultured man who knew how he could die because of the disease and explicitly asked to die. Another patient with MSA, after an evident worsening of her conditions asked about how to terminate her life, she asked why in Italy it's not possible to be helped to die. I've never had a patient who took his life, but with these diagnosis the problem is real.

EMS2: fortunately I didn't have any experience like this. In MS the problem is due to the disease progression. ALS patients are vigilant until the end of their life. MS patients, luckily, have cognitive impairment in far advanced stages and this helps. They are not so aware of their conditions. Carers, well, they are more

aware of this. Diagnosis is a difficult moment, than generally patients accept it because there are remissions, there is a lot of research...

EMS3: these situations (suicides, request of euthanasia) are more common than in cancer because it's a longer disease. Even advanced stages can survive for years. We had patients who attempted suicide, but never succeeded. Some of them are at risk. Above all the early and the far advanced stages. They do what they can, with drugs overdose or trying with their hands.. above all young patients in relatively good conditions that don't accept it.

EPD1: in literature these problems are reported. I don't have this experience. It may be due to the fact that PD patients tend to be older than MS, or because of their particular character.. If they talk about this probably they're trying to keep your attention on them, it's more a provocation. Sometimes it happened with younger patients who said:" What do you think doctor, wouldn't it be better to end up this situation? But if my wife does it I'm happier!" Probably younger patients are more at risk because of the long life they have to live with the disease, but fortunately they respond well to therapies.

4. Role of the psychological support

when asked to describe which kind of psychological support their services were able to provide to these population professionals reported different experiences. Some groups, such as the ALS/MND one, are better organized and have a proper psychological support service available for both patients and relatives, even though it is basically hospital based and so difficult to reach homebound patients

ALS1: We have a psychological support service that works very well. Patients, carers and friends are supported in a very good way. The problem is that our psychologist can't go out of the hospital and so when the patient is bedbound often cannot be of help anymore.

Specialists recognise that this support is very important throughout the course of the disease

SLT: I think that a psychological support should start soon after diagnosis. They have to focus on end of life decisions, like if they are to decide for a tracheo, yes or not. It would be sensible if psychologists were included in the information and support pathways. In facts, in our reality, they are not.

In conditions other than ALS/MND the available psychological support is not well established and no home service is available. In MS ambulatories a psychologist is available and provides counselling for both patients and families and contributes to help in therapeutic decisions

EMS4: we have a psychologist who helps us in caring for our patients. She is of help above all for new diagnosed patients that have to cope with such a bad news. She works on treatments compliance too. Certainly in advanced stages a support would also be required, but we're not so experienced on that. Being a long term disease patients have their time to use adaptation for their disease.

In movement disorders psychologist are involved mainly for therapeutic decisions. Patients are tested for selection about deep brain stimulation (DBS), in fact only those without signs of cognitive deterioration can be selected for this treatment. This service is of little, no use for psychological support

EPD2: our patients are continuously tested by our psychological group, but they're tested in order to monitor their cognitive performance as it is important to decide if they can be candidates to DBS or in follow up after its placement. In ordinary visits we try to support them in informal ways, but we don't have any specific team dedicated to this point. There are big necessities for this kind of support as nothing is available for our population

Social needs

Professionals who participated in the three focus groups dedicated some comments to the social unmet needs of their patients. They declared that their opinions were largely based on what patients and families described during the clinics, rather than a direct experience based on having seen their homes or their social environments because only a few of them could visit patients at home.

Social isolation, financial concern and caregivers burden were the most prevalent themes which emerged from the discussions.

1. Sense of social isolation and lack of social support

Isolation can happen because patients are abandoned by their partners who are scared by the diagnosis or by the disease progression

EMS1: a frequent problem is patients abandoned by their spouse due to the disease. It's awful, above all when young mothers with MS are abandoned with young children. So divorces and abandonment are more frequent than in general population. MS is a scaring disease and many husbands can't cope with it. What surprise us is that it happens just after diagnosis, not after a long time when disabilities impact badly on QoL.

But patients can be isolated even in their own families because they lose physical skills like eating and therefore do not participate in familiar meetings and meals

SLT: This happen when a member of the family thinks that if the patient stays around the table with the others he'll suffer because he or she is incapable to eat like the others. But this cause isolation and loss of the familiar role. When you don't feel like a person anymore you stop your interactions with the others. In the end you don't communicate with your family. Some patients just lie on a wheelchair with the PEG, ventilator.. sometimes someone pass in front of you, tell you a couple of words, but you can't answer... that's isolation.

Isolation is also worsened by lack of help provided in the place of care (home) and in the required way.

EALS1: home care should provide domiciliary assistants to mobilize patients and to enable them to go out when possible. Many tracheostomized patients can

go out with the wheelchair and a portable respiratory machine. Nowadays it is rare that a severely disabled neurological patient can obtain a good home care service

Professionals believe that little practical help and suggestions could have a strong social impact for patients and caregivers if available

EPD2: very easy suggestions at their home could really impact on their QoL. They could really be helpful for carers. Often carers ask me how to achieve services and I don't have any idea of it. Patients tend to be isolated because of lack of transport services. I suggested a number of possible interventions to a patient who came in our ambulatory, but his carer told me that it wasn't feasible because the real trouble was to transport him, and if this problem can not be overcome we in the hospital cannot provide any help.

2. financial and job related problems

When disability impairs patients they lose their capability to work. This is recognized as a further loss by professionals, being both cause of diminished financial income and cause of social isolation

EMS2: another issues are job related problems. Patients tend to hide diagnosis not to be discriminated on their place of work. Or if they work on their own there's the problem of how to manage it when they'll be disabled.

EALS: when patients are homebound, above all if they are totally dependent by a mechanical ventilator, they require continuous assistance. If they are young and have already lost their employment the problem becomes worst because often their spouse has to leave their job to care for them.

3. caregivers burden

This aspect was described above when discussing the need for respite care admissions and can be implemented by the previous comments about the need of social support in terms of home care assistants and financial support.

Specifically professionals recognize the high burden of care induced by having to care for very disabled patients for months and sometimes years with little no time to rest or to take some time for their own lives.

EMS1: they really need respite, I mean to be relieved a bit.

EALS1: respite admission is very important for caregivers. It would be so important to have facilities available for respite periods here in Turin. I think that for our patients respite admissions can be very useful to support their carers. Maybe they could use 15 days just to sleep!

Spiritual needs

Existential questions are reported by participants to the focus group as inevitable in the advanced stages of neurological diseases. Often these are triggered by end of life decisions or spontaneously discussed by patients during routinely clinics.

Professionals declare that they do not feel at ease when these issues come out in their practise.

EPDI: typically patients report something like this: You see dear doctor the main question is “Why me?” And above all “Why now?” I worked all my life and now that I could enjoy it... the matter is that we don't know how to answer, there's non one we can refer them to.

Some participant recognise their potential role in enabling their patients to raise these issues

EPC: it depends very much on how much place you dedicate to communication. If you let these questions arise or not. I think that every patient feels these needs. Support provided is still very low. There are psychological support dedicated services with social workers too, but not about spirituality. Often these questions are asked to physicians because they don't feel a professional judgement. If you leave enough space these questions will arrive.

Spiritual themes can be addressed by relatives after the patients' death, or by patients themselves when the future appears scaring and full of doubts, as this participant testify

EALS1: about spiritual issues they tend to arise in familiars after the patient' death because the disease is so quick... but sometimes in those long term tracheostomized patients (that I personally consider over-treatment or heroic treatment) the existential problems become more important than the practical ones. We receive questions about anguish of the future, fear of what could happen..

Spiritual questions can challenge ones own beliefs and involve professionals in their private intimate life. Sometimes this can be a cause of difficulties in maintaining the professional role and therefore specific spiritual support seems required

SLT: patients with advanced respiratory troubles talk of spiritual questions and these should be addressed to a spiritual assistant or chaplain. We became aware that these heavy questions are asked to us. So we try to answer without giving personal beliefs but... what kind of answers can we provide? (..) this is a further reason to provide a psychological and spiritual support from the time they are diagnosed.

3.5.3.3.4 Summary

The professionals that participated to the focus groups provided deep insight about the various aspects of the care of patients severely affected by neurodegenerative conditions. They confirmed the difficulties to determine the number of potential people in advanced stages that could require specialist palliative care. Professionals denounced their difficulties to provide supportive care to patients very disabled unable to go to the clinics. The end of life phases are not easy to be identified for the unpredictability of the trajectories of the disorders. They are aware of the high burden of physical, psychological, spiritual and social unmet needs faced by their patients and their family carers and some recognize their lack of expertise in managing the peculiar problems caused by the advanced stages. The potential role of specialist palliative care, both for the home assistance and for the hospice respite admissions, was widely recognized and participants confirmed their keenness to help the development of a new SPCS for their advanced patients.

3.6 Discussion of the qualitative study

In this chapter the results emerging from the qualitative study are discussed.

The general framework of the discussion is based upon the following key points:

1. General discussion of the results
2. The physical, psychological, spiritual and social components of the unmet needs
3. Potential role of a new SPCS aimed at meeting the unmet needs
4. Limitations of the study
5. Summary

3.6.1 General discussion of the results

Results of the NeuNeeds qualitative study can be summarized as follows:

- 22 in depth interviews with patients severely affected by ALS/MND, MS, PD and related disorders and their informal carers and 3 focus groups with professionals involved in their care were performed.
- Audio and videotaping and field notes of the events were collected, transcript verbatim performed and a multi step coding procedure allowed to highlight various categories of needs and issues related to patients' quality of life, caregivers' burden and comments on the available existing services dedicated to this population.
- The inter-rater reliability among the 3 independent researchers was higher than 70% in the coding procedure. This represents a clear indication of how explicitly participants identified the needs and themes in a way that independent observer can detect the underlying meaning with little no difficulties.
- The large amount of data results from this study: interviews and focus groups had a mean duration of about 60 minutes each. Transcript verbatim of the 25 events resulted in about 108000 words with an average of 4300 word for each event. Inevitably some information got lost in the coding process, even though a conspicuous amount of data was analyzed.
- Participants seemed interested in the study and keen to collaborate. None declined to participate to the interviews. Focus groups were more complicated to organize and manage as described in details in the limitation section. This can be interpreted as a general interest of patients and families about new initiatives that can have a positive impact on the assistance, while it confirms that for many professionals palliative care is not on the top of the list of their agenda.

3.6.2 The physical, psychological, spiritual and social components of the unmet needs

The content analysis of the results coming out from the 22 interviews and the 3 focus groups reveal a huge number of problems that patients severely affected by neurodegenerative disorders living in Turin city and its metropolitan area (and their families) have to face and that are not fully satisfied by the existing dedicated services. All the components of the so called "total pain" (Clark 1999) are represented in the transcript verbatim of the various events. Professionals involved in the care of this

population agree with their patients about the high number of distressful symptoms and other non physical components of their suffering.

Even though the qualitative design of the study was chosen to provide a deep insight of this population's daily troubles, rather than to quantify the number of the unmet needs, the content analysis showed numerically the big load of troubles present in the sample and can be confronted with previously published data.

In the following sections the various categories of needs are discussed

3.6.2.1 The physical needs

The published systematic review of the symptom prevalence among people affected by advanced and progressive neurological conditions (Saleem, Leigh and Higginson 2007) allows a comparison between the findings of the study and what is known about these issues.

Overall 169 physical symptoms were discussed in the 22 interviews with an average of 13 physical problems each event. Professionals focused more on the major symptoms such as pain, respiratory troubles or typical neurological features like movement impairment, and were less likely to not mention other problems usually well known in the palliative care settings such as the quality of sleep, oral symptoms, or problems of the skin. Voiding functions were also rarely reported by professionals as important problems, whereas patients and families talked frequently about it. Professionals were more concerned about medications, prevention of medical complications and respite admissions.

Pain

Pain is reported both frequent and severe as evidenced in recent literature reviews in this population (Ganzini, Johnston and Silveira 2002, Lee *et al.* 2006c, Higginson, I. J. *et al.* 2006a, Saleem, Leigh and Higginson 2007). In our study more than 80% of participants had some kind of pain or painful conditions. These were not necessarily related to their main diagnosis, but were cause of suffering for patients affected by chronic disability and in their last months/years of life. Some patient reported pain spontaneously, others had to be given hints and support to discuss this issue. Some participants seemed ambiguous about this symptom because they felt that their painful condition was not directly caused by the neurodegenerative condition. They felt relieved when researchers underlined the importance of recognising the impact of any kind of pain in the palliative philosophy of care. Several different definitions were used by participants to talk about painful symptoms and this finding is similar to what described by Saleem *et al.* in his review about the physical symptoms in neurodegenerative conditions (Saleem, Leigh and Higginson 2007).

It appeared that pain was undertreated and participants stated that their physicians feared to use pain killers because of potential side effects like respiratory depression and this confirms what has been described in patients with neurological conditions since many years ago (O'Brien, T. 1993b).

Pain is certainly one of the most known physical symptoms in the palliative care setting and in the past was seen as an inevitable condition for people dying for cancer. The development of the modern palliative care and hospice philosophy allowed millions of cancer patients to be relieved of this awful symptom (Hanks and Forbes 2005). The

assumption is that painful syndromes caused directly by the neurodegenerative conditions or complicating the clinical conditions as co-morbidities can be better controlled if a SPCS can intervene offering its skills as shown for ALS/MND in some hospices (Oliver 1998, Oliver 1996, Oliver and Webb 2000, O'Brien, T., Kelly and Saunders 1992).

Breathlessness and respiratory problems

Respiratory symptoms, particularly causing dyspnoea, appeared in more than 80% of the interviews. This is a very well known condition for ALS/MND advanced patients (Saleem, Leigh and Higginson 2007, Neudert *et al.* 2001b, Lyall and Gelinas 2006), but in our study it was reported by patients affected by MS and movement disorders as well. It is known that for patients with neurodegenerative conditions and severe disability the main cause of death is infection, particularly a chest infection (Sadovnick *et al.* 1991, Koch-Henriksen, Bronnum-Hansen and Stenager 1998, Litvan *et al.* 1996, Beyer *et al.* 2001). Shortness of breath was also found to be correlated with QoL in hospice setting (Steele *et al.* 2005). Participants in this study talked of numerous episodes of respiratory tract infections during the advanced stages of these diseases, causing shortness of breath and scaring both patients and carers. These occurrences often cause the hospitalization of the patients exposing them at the risk of further hospital acquired infections, as reported by the professionals who participated at the focus groups.

Another important finding of this study was that the management of respiratory troubles in patients with severely advanced neuromuscular problems is focused on the possibility to provide an efficient respiratory support, preferably through non invasive ventilation, which has been shown to improve the QoL of the patients and have apposite impact on survival, although is not a curative treatment and cannot arrest the progression of the muscular paralysis (Eng 2006, Mitsumoto *et al.* 2005, Miller *et al.* 1999). Invasive ventilation is a possible option that is offered when non invasive respiratory support no longer control the symptoms or the arterial gas analysis shows an excess of CO₂. Another group of patients that often require invasive ventilation are the bulbar patients, especially for ALS/MND, for whom the ventilation through the mask is often not applicable because the patients cannot control the upper airways (Albert *et al.* 1999b, Albert *et al.* 2009).

In Piedmont, the Italian north western region, where these study was set up, there is a prospective regional register for ALS/MND (Chio *et al.* 2009) containing data of all the diagnosed cases of this disorders since 1995. Recently the neurologists working in the tertiary clinic for ALS/MND in Turin published a retrospective analysis of this register aimed at evaluating the frequency, the clinical characteristics and the outcome of tracheostomy in these patients. This study evidence that the prevalence of tracheostomy is of about 10% and of those patients who had this option they were more likely to be men and young. The median survival time was of 253 days after the tracheostomy and longer survival was related to young age, presence of PEG, being married and cared for by a tertiary clinic (Calvo *et al.* 2009). In this study 3 patients with ALS/MND were tracheostomized and ventilated 24 hours per day. They were all long survivors because they had been tracheostomized respectively 4, 3 and 2 years before and were clinically stable. All of them reported to be happy with this choice, even though one of them had it in emergency and had not been not keen on the intervention beforehand. None of them discussed the possibility of being disconnected from the ventilator. This subject appeared in other interviews with patients with non invasive ventilation that feared this

option because they had been told that if they chose it they could not be withdrawn with the Italian laws. This is certainly a difficult decision even though in most western country it is both legally and ethically accepted (Oliver 2004, Ankrom *et al.* 2001, Moss *et al.* 1996, Borasio, Gelinas and Yanagisawa 1998, Borasio and Voltz 1998).

What appeared to be lacking in the care of these patients was the palliative management of the respiratory symptoms. Experts involved in the focus groups confessed their lack of knowledge in the use of opioids to control breathlessness. Physicians involved in the respiratory management declared to feel impotent when the patients refuse or do not tolerate a non invasive ventilation. They reported that when patients are brought to the hospital for respiratory insufficiency they are more likely to be tracheostomized even if they would not want it because there are not an alternative to control breathlessness. Neurologist stated that morphine was to be used for pain control or for terminal sedation, but that there were not an indication for this drug to control dyspnoea even though this option, commonly used in hospice and palliative care settings (Jennings *et al.* 2002, Lyall and Gelinas 2006, O'Brien, T., Kelly and Saunders 1992, Oliver 1998). Furthermore the indication for using morphine to control breathlessness has been published in guidelines for the management of the ALS/MND patients by neurologists in many occasions (Miller *et al.* 1999, Mitsumoto *et al.* 2005, Eng 2006).

For these reasons it was thought that the skills of a SPCS could have been of use to enable patients who refuse mechanical ventilation and that suffer for shortness of breath to receive adequate palliation of their symptoms and to have their advanced directive respected.

Sleep related problems

The quality of sleep in very ill patients is often poor and these symptoms represent a common feature in different advanced conditions (Hockley, Dunlop and Davies 1988, Solano, Gomes and Higginson 2006, Saleem, Leigh and Higginson 2007).

In this study 64% of participants reported problems causing sleep disturbance. Often the poor quality of the sleep is due to uncontrolled physical symptoms or fear and anguish that may emerge during the night. It is known that insomnia is one of the symptoms that can indicate poor oxygenation of the blood at night time due to respiratory failure – a condition frequent in ALS/MND (Borasio, Voltz and Miller 2001); can be caused by a restless legs syndrome that affect particularly PD's patients (Lee *et al.* 2007); or by painful neuropathic syndromes in MS (Henze, Rieckmann and Toyka 2006).

Being a common condition that impacts badly on the QoL of both patients and their carers this domain was included among the physical symptoms to be assessed in the evaluation of the new SPCS.

Intestinal symptoms

Constipation, diarrhoea, problems due to dietary changes caused by dysphagia and the consequent need to alter the consistence of the food, the placement of a PEG and the feeding through enteral nutrition, the side effects of drugs like the L-Dopa are common findings in patients with advanced neurodegenerative conditions.

In the present study more than 70% of the interviewed patients reported problems related to their bowels.

The intestinal symptoms were also described by the professionals involved in the focus groups that reported these problems as both frequent and difficult to manage.

SPCS have to deal with intestinal problems due to the use of opioids, partial or total intestinal obstruction in malignancy, and cancer related diarrhoea among the others. Constipation is also used as an excuse to avoid the use of opioids and this was been reported by neurologists, patients and carers involved in this study. One of the tasks of a SPCS is to convince the service users that constipation can be improved by applying protocols that are part of the normal routine activity of the hospice teams.

Urinary symptoms

Urinary problems frequently affect neurological patients at the end of life. Voiding dysfunctions are associated with common neurological disorders involving difficulties with emptying and storage. Typically MS patients suffer from detrusor external sphincter dyssynergia when they have a spinal cord involvement. This can lead to urological complications such as hydronephrosis, vescico-uretral reflux, sepsis and urilithiasis (Andersen, J. and Bradley 1976). In PD's patients up to 72% have symptoms of bladder dysfunction, usually a detrusor hyperreflexia (Shah and Badlani 2004, p. 264.) ALS/MND should spare patients from sensitive and autonomic deficits, but when very advanced difficulties related to incontinence or need to use pads, external catheters or sometimes indwelled bladder catheter can appear. When patients lose the ability to eliminate completely the urine from the bladder and some remains stored extemporary catheterization may be necessary. This task is often performed by the patient if he or she maintain a sufficient ability in their upper limbs, or more frequently in the advanced stages by their carers. Patients must not stay wet to avoid the risk of skin ulcers and this cause a high burden of care for their families.

In this study about 60% of participants reported symptoms and problems related to the urinary tract. This group of symptoms is again of common management for a SPCS.

Oral symptoms

In this group of symptoms were grouped all the difficulties related to the mouth discomfort, like drooling and dribbling of saliva, dry mouth, problems with secretions and oral pain. If the swallowing issues such as aspirations, choking for food and dysphagia problems are added we can see that almost all the interviewed patients reported at least of these issues .

It is known that ALS/MND, especially for the bulbar forms, may cause drooling and dribbling of saliva, not from an excess of production, but for the difficulties in swallowing it (Oliver *et al.* 2007, Mitchell, Temperley and Duff 1995, Miller *et al.* 2009) and this happens in MS (Rousseaux and Perennou 2004) and in PD's as well (Bunting-Perry 2006, Calne and Kumar 2003, Clough and Blockley 2004, MacMahon 1999, Wenning *et al.* 2004).

Drugs can cause dry mouth, especially anticholinergics, amitriptyline and opioids. All these categories of drugs are commonly used in palliative medicine and therefore SPCS know well the therapeutic balance between positive and side effects.

Other physical symptoms

Among the coded groups of physical needs other categories found were movement difficulties that obviously affected all the interviewed patients and this can be easily understood from the high disability degree of the sample. Skin troubles and other less common symptoms like fever, visual loss and speech impairment were also reported and patients testified how they impacted on their individual QoL. These conditions are

described in the content analysis of the physical needs, in the results section of the qualitative study. For the purpose of this project the physical symptoms that were chosen, among the ones who emerged from the interviews and the focus groups, were those for which it was thought that the input of new SPCS could make the difference inducing positive change due to its specific knowledge and competence. Of course this cannot be possible for the movement disability, visual loss or loss of the speech.

3.6.2.2 The psychological needs

Abandonment

The sensation of being abandoned by the persons on whom patients and families rely upon, is a very unpleasant feeling leading toward the sense of insecurity. This is what was proposed by Maslow as a fundamental need for the human beings when he described the safety needs:

Practically everything looks less important than safety,(even sometimes the physiological needs which being satisfied, are now underestimated). A man, in this state, if it is extreme enough and chronic enough, may be characterized as living almost for safety alone. (Maslow, A. H. 1943).

Participants reported to feel abandoned by their relatives, friends, doctors and other healthcare specialists. The causes of this abandonment were defined as caused by the patients' difficulties to ask for help, the lack of sensitiveness of the people that should perceive the difficulties of the physically impaired patients and offer themselves to help, the lost of interest of professionals that lose interest in patients when are very ill and can not be cured. The consequences of this awful sensation can be the anxiety, confusion, fears of the future and closure. Recommendations have been suggested by other authors so that attention is given to emotional support, which specifically addresses three main areas of dealing with loss and change for people that are severely affected--physical issues, independence and relationships (Edmonds *et al.* 2007b). Similar conclusions were reached by other researchers for PD patients and carers, highlighting, among the others, the difficulties faced by their interviewees in the advanced stages (Hudson, Toye and Kristjanson 2006). Many patients and carers feel abandoned by their medical attendants when they reach the end stage and are often told that "nothing more can be done"(Gonsalkorale and Morris). Abandonment is strictly related to the quality of the relationships and a key point for palliative care professionals is to put the patients' and the families' needs in the centre of the attentions, fighting the negative common practice in medicine of tending to ignore the dying and treating them differently.

From the professionals point of view this was confirmed by the sense of frustration reported by the focus groups participants. They reported how they feel impotent when patients become too ill to be able to go to the hospital or the ambulatory clinics and they are lost at follow up. Some declared not to know what happen to the patients once they do not come to the appointments. Others reported lack of confidence in the existing domiciliary services provided by the NHS.

Coping with the disease

Difficulties in coping with the neurodegenerative disorders and the coping strategies adopted by the participants were a very interesting finding of this study. It is known that advanced neurological disorders can cause desperation and lead to concerns at the end of life, leading towards suicide (Sadovnick *et al.* 1991) or medical decisions to shorten one's own life (Ganzini *et al.* 1998, Veldink *et al.* 2002, Van der Wal and Onwuteaka-Philipsen 1996). Depression is not the only cause of these decisions, but other conditions such as hopelessness, fear to die with suffering and difficulties to accept multiple losses can induce people to take decisions like these (Albert *et al.* 2005, Kumpfel *et al.* 2007, AAN 1998, McCluskey 2007).

One of the tasks of a SPCS is to help patients and families to cope as better as possible with the diseases and the losses and changes caused by the progressive course of the illnesses (Edmonds *et al.* 2007b, Adelman *et al.* 2004).

From the interviews of this study it results how many causes of losses can affect the patients and their carers. It was possible to detect issues related to difficulties to cope with losses or having unmet expectations (which can be considered as a loss) in 17 out of the 22 interviewed patients. Most reported difficulties were about physical troubles such as having to accept limitations in movements and autonomy, but other losses were reported such as losing a job or the possibility to communicate. Rage, fears, anxiety and depression were again consequences of these difficulties. Some participants spent their time and money seeking for alternative therapies, often having to pay a great deal of money taking journeys of hope in far countries like China.

Family carers reported difficulties defined as having to struggle for everything they needed (Edmonds *et al.* 2007a) because doctors and social assistance were not of help for them, and they had to go through difficult bureaucratic passages to satisfy the patients needs. Some reported that they preferred to quit asking because they lost the hope to be helped confirming that specialised psychosocial care is necessary for carers of very ill and disabled patients (Oliver and Gallagher 1998).

Another psychological need found out in the study was the unpleasant sensation of being a burden reported by the patients (Chio *et al.* 2005). This was usually denied by the carers, even though some declared to be very tired and in burnout syndrome. This sensations confirms what published in the literature about the different perceptions between patients and carers of one another's quality of life and existential issues (Adelman *et al.* 2004, Bolmsjo and Hermeren 2001, Goy, Carter and Ganzini 2008, Kristjanson, Aoun and Oldham 2006).

Anxiety and depression

The methodological design of this study was not meant to detect signs or symptoms of these specific psychiatric disorders, but when direct references to these issues were made the researchers coded it into these categories. This happened also when indirect citations were related to these mood disorders. Even though direct sentences using the terms anxiety or depression to describe the psychological condition of the patients were

seldom used, in all interviews appeared some mentions to the alteration of the patients' mood.

The prevalence of mood disorders in patients with severe neurodegenerative disorders is widely discussed. Depression ranges from 25-50% in MS (Minden and Schiffer 1990, Patten *et al.* 2003, Sadovnick *et al.* 1996), whereas anxiety affects about 36% of these patients (Galeazzi *et al.* 2005, Korostil and Feinstein 2007). In ALS/MND advanced patients depression was found in 19% of patients (Rabkin *et al.* 2005), but it can affect up to 50% and it is strictly correlated with hopelessness impacting deeply on the individual QoL (McLeod, J. E. and Clarke 2007). Anxiety has been described between 19- 24% of patients in an Australian and an Irish survey respectively (Kristjanson, Aoun and Oldham 2006, Clarke, S. *et al.* 2001). For PD's patients anxiety is reported between 43-62% (Chaudhuri and Martinez-Martin 2008, Lee *et al.* 2007) and depression between the 20-40% (Kristjanson, Aoun and Oldham 2006, Chaudhuri and Martinez-Martin 2008), although using other instruments it has been found in lower percentages (3-16%)(Lee *et al.* 2007).

To obtain these prevalence data authors used validated tools that were not part of this study design. From the interviews it emerged clearly how patients and their families' mood were proved by the experience of the disease and participants were facing worries that impacted badly on their QoL. Some experienced fears and concerns for the future, others reported loss of hope for their own future and for their families. Another used term was impotence as to describe the impossibility to do something to improve their conditions. Frequently patients' mood was described as instable, probably a feature difficult to be detected by traditional tools, but recognizable by their carers.

All these conditions impact negatively on the quality of life of both patients and family members and a psychological support should be of help, aside of medical treatment, trying to reduce the negative effects of mood disorders.

Other psychological themes

The need of psychological support was discussed by some participants. The opinions were divided between those who had already successfully experienced this service and those who had a negative impact or that were prejudiced against it. Probably this is not very different from what normally happen in cancer care when often patients and families are not keen at receiving psychologically help, but then realize how helpful it can be to be accompanied during a painful and hard experience so that it may prevent further suffering to the ill persons and pathological bereavement in the survivors.

Less frequently, were other psychological issues raised by the participants: sense of shame for being in such a poor conditions, need of privacy that is completely lost when patients completely paralyzed need someone to wash them, change their clothes, feed them and so on. Someone reported a sense of loss of dignity which is striking for palliative care professionals that have the dignity of the dying person as a guide line for their job.

Need of self independence and control was expressed in some interview and this reflects others' findings in the advanced stages of incurable diseases (Cotterell 2008).

3.6.2.3 The spiritual needs

Among the spiritual themes that were discussed by the participants of this study the most frequent were related to the meaning of the experience of the disease (50%) and those related to the faith, religiousness and spiritual issues like the role of the spiritual support (78%). Other less frequent arguments were the debate about justice/injustice for what is happening to them (27%), the concept of hope (Centers 2001) (23%), usually referred to the hope to find a cure for their conditions (miracles, alternative therapies) and the concept of control (32%) where interviewees were divided between those who said that were losing control and rage usually supervenes taking its place, and those for whom staying in control of their situation is an important defence against the uncertainty of the future.

In the following paragraphs the two most represented groups will be discussed: the sense of meaning and the role of faith, spirituality and religiousness.

The meaning of the experience of the disease

Half of the sample discussed about the following question: Did you find a meaning in the experience of disease that you are doing? Does this make sense in your life? These questions were not part of the scheduled interview scheme, but were among the hints that researchers provided when and if participants asked what did we mean for spiritual needs.

Previously research showed how patients severely affected by neurological disorders or malignancy change their personal values shifting from self-enhancement values towards self-transcendence ones and this could be seen as a coping strategy at the end of life (Fegg *et al.* 2005). What is not clear is the relationship of these domains with the individual QoL. Maintaining a meaningful quality of life centres on psychological, supportive, and spiritual factors, as opposed to physical status (Stromberg and Weiss 2006). From the data of this study it appears that participants with ALS/MND who had been tracheostomized were keener on talking of these items and that they had to find new different meanings in their lives after that important decision to prolong their lives, even if their physical disability did not improve, but continued to worsen. On the other hand those who chose not to be tracheostomized reported more difficulties to find meaning in their lives, as if they were just waiting for the end. Some said that having found a meaning does not mean to accept the situation (Albert *et al.* 2005). So patients who clearly decided not to prolong their lives shift the meaning of their life towards the importance of having their choices respected (Bolmsjo 2001) (in Italy advanced care planning or advanced directives are not yet legally binding and therefore it can be up to the resuscitation specialist in A&E to decide if to resuscitate a patient who lost consciousness for a respiratory arrest, even if he or she had previously stated that did not want a CPR). For others the problem of meaning is widened to their previous choices, for instance one said that if she had known that she was going to fall ill she would not have been married, probably she could not find a meaning in her role of wife being totally dependent by her husband.

Now an important question could be if the input of a SPCS could have an impact on the sense of meaning of the lived experience in these patients. In the published literature are

reported the importance of the personal existential values (Bolmsjo 2001, Mockford, Jenkinson and Fitzpatrick 2006), but whether they can be affected positively or negatively by being cared for by professionals working in an end of life setting remains unclear.

Faith, spirituality and religiousness

Are faith, religiousness, spirituality positive resources that can help to cope with severe disability and life limiting conditions? Or at the opposite persons with a strong spiritual belief are challenged in their faith when such a disastrous disease bursts into their lives? Can a SPCS modify in part these aspects improving the relationship between a dying person and his or her spirituality?

These open questions can summarize the discussion of this point. Information about spirituality, relationship with God, faith and religiousness were collected from those participants who wanted to talk about these themes.

Overall in almost 80% of the interviews something related to these issues came out. For some participants the faith in a God that is love and that has the answer for their unbearable conditions is the meaning in life. Someone discovered his or her religiousness during the phases of the disease, others strengthened their liaison with God after having fallen ill.

For others faith brings with doubts and the experience of the disease challenged their relationship with the church. Plahuta et al studying psychosocial factors in ALS/MND patients, showed that the extend degree to which spiritual beliefs help to cope with ALS is not a significant predictors of hopelessness (Plahuta *et al.* 2002). This finding seems in contrast with what reported by the interviewees of this study for whom, when spirituality was lived as a positive resource, it appeared easier to cope with the disease and to be more positive about the hope.

Spiritual and religious support, that are not synonymous (Murray *et al.* 2004, Strang, Strang and Ternstedt 2002, Clark 1999, O'Neill and Kenny 1998), were also discussed by participants. The sample was divided in a group who stated the importance of receiving spiritual support and the consequent positive impact on the existential aspects of their lives, and a group that was not interested and did not feel that this support could improve their quality of life. Someone sadly reported that a spiritual support would have been welcome, but due to their impossibility to leave their homes they were not receiving an adequate spiritual care.

SPCS have professional spiritual assistance in the staff and often the other “non spiritual” members develop attitudes and skills towards the spiritual aspects of the end of life, chronic disability and other spiritual issues. Professionals who participated at the focus groups recognised the importance of the spiritual needs of their patients even though the definition of the various spiritual components of the spiritual care was not so clear. This is congruent with other studies about this topic (Strang, Strang and Ternstedt 2002).

Do SPCS make the difference for these items? No evidence in the literature was found related to the impact on the spiritual issues of a SPCS for people with neurodegenerative disorders.

3.6.2.4 The social needs

Numerous social unmet needs were discussed by the participants of this study (see table 3 and chart 3 in the qualitative results section). Some were expected by the researchers because had previously been described in the literature, such as the economic and financial problems due to the loss of the job sometimes for both the patient, that losses the ability to work and for the carer (often a spouse carer) that has to dedicate most of the time for the caregiving tasks (Mockford, Jenkinson and Fitzpatrick 2006). Social, and financial issues, should be addressed early in the course of the disease and family carers are to be supported because so often play a central role in management and care of the patients (Mitsumoto and Rabkin 2007). Enduring financial hardship was one of the themes developed in the data analysis of a qualitative study with caregivers of PD patients aimed at exploring the palliative care needs in patients and families affected by this neurodegenerative disorder (Hudson, Toye and Kristjanson 2006). Financial support is one of the variables that can help families to keep at home patients who choose to be cared and die at their homes (Grande, Addington-Hall and Todd 1998). A Dutch study quantified the monthly cost of caring for patients with ALS/MND at home as higher than 1000 Euros per month (Van der Steen *et al.* 2009) independently by the kind of service received (specialist or general care).

The fact that SPCS can reduce the cost of care for terminally ill patients has recently been challenged by a literature review (Zimmermann *et al.* 2008) even though for the families receiving a comprehensive package of services including physicians, nurses, physiotherapists, psychologists and other professionals for free is certainly a precious benefit. The reported financial problems were not only related to the reduction of the incomes or the excess of money spent to pay for paid carers or home assistants to care for the ill patient, but also the difficulties to obtain social benefits like the monthly checks that in Italy are provided to those families who decide to care for relatives with chronic illness in their homes, or difficulties to obtain costly technical devices such as the electronical communicators that allow the patients with severe aphonia or aphasia to stay in touch with the external world by communicating with the others. Adequate physiotherapy and speech and language therapy at home was not always possible without having to pay for it.

Other unexpected social problems were the high rate of divorce and family troubles that were reported as caused by the disease, difficulties in transport and the need of respite admissions and holiday periods for both the patients and the exhausted carers. These themes were collected and examined by the researchers trying to see where a SPCS could be of help by improving some aspects of the social unmet needs.

Social isolation

It appeared straightforward that the main group of needs that can be faced and improved by a SPCS are those related to the social isolation of the patients and the carers. The lives of many neurological patients are significantly limited in social interaction (McLeod, J. E. and Clarke 2007).

Many causes of this condition appeared in the interviews and all were cause of suffering and frustration for all the family members. The disease itself is the main cause of social isolation. The fact that patients experience high disability limits their mobility and

causes their conditions of being homebound. Furthermore friends, relatives and other components of the normal social network around the patient and the family tend to disappear when the disability progress and the condition becomes chronic. Some patients specifically talked of their wish to be visited by volunteers that could just spend some time with them to offer a break from the long hours of solitude. The availability of internet connection, possible even for totally paralyzed people with only spared ocular movements, was described as wonderful by some participants because it allows them to create virtual communities with other persons around the world. Communicators allowed group of ALS/MND tracheostomized and quadriplegic patients to write a book just using their eyes to control a piece of software similar to a word processor on their desktops (Brownlee and Palovcak 2007). The problem was that other patients could not enjoy these facilities because were older, unable to deal with informatics or with severe visual losses that withdrew them from the use of these facilities.

Various studies highlight the need of social support for families and patients where neurodegenerative conditions caused this social isolation. According to a qualitative study in PD for patients and carers there is a need for more specialised and individually adjusted support (Birgersson and Edberg 2004). Other authors underline how action to reduce the burden of care for caregivers better attention to the patients' functional status and to the caregivers depressive symptoms is important (Caap-Ahlgren and Dehlin 2002) and this task could be done by a SPCS. Social isolation is one of the contributors to the reduction of the QoL of PD patients followed overtime (Karlsen *et al.* 2000) as well as for patients with respiratory disorders (Skilbeck, J. *et al.* 1998).

Patients with a chronic illness die with many unmet needs and care teams are frustrated by the lack of resources available to them and admit to be ill-equipped to provide for the individual's holistic needs. Some clinicians described difficulty in talking openly with the patient and family regarding the palliative nature of their treatment. This last statement was among the conclusion of mixed methods study exploring the unmet palliative care needs of end-stage heart failure, renal failure or respiratory disease patients (Fitzsimons *et al.* 2007) and overlaps with some statements of the professionals involved in the focus groups in this study as well. People who found it difficult to talk openly with the patients justified this with the explanation that is was due to the impossibility to offer a continuity of care outside the hospital and therefore the professionals felt that they have lost the patients, when they become too ill to go to the scheduled visits. Remarkably, some patients describe their most stable form of social interaction as being their visit to the doctor or clinic (McLeod, J. E. and Clarke 2007). This is lived as a cause of isolation from the other side, the patient and the family, that experience this distance occurring with the physician as a cause of social isolation.

One question rising from these considerations is the following: it is clear that patients severely affected by neurodegenerative conditions and their carers experience a high burden of social isolation, but in which extent a SPCS can reduce these problems? Theoretically by providing regular home visits, periods of respite care in hospice, providing volunteers and helping the family to engage experienced paid carers known by the service the SPCS could improve the person's experience. It can also facilitate the connections with other services by networking with the specialists working in the hospital that no longer provide active care to the patients, but that can collaborate with the SPCS professionals for specific issues, symptoms or prescriptions. This last consideration covers one of the issues described in the qualitative study of the unmet

palliative care needs of advanced MS patients that was the lack of continuity of care and communication between professionals (Edmonds *et al.* 2006).

The problem is that no prospective studies are available in the published literature that demonstrated that these social outcomes have really been achieved by SPCS in the care of neurological patients. For these reasons the social isolation, in both patients and their informal carers, was chosen among the items to be assessed in the evaluation of the forthcoming new SPCS for people severely affected by neurodegenerative disorders and their caregivers.

Service satisfaction

Services aimed at meeting the needs of people affected by neurological progressive disorders need to be flexible and tailored on the patients and family carers needs (Kristjanson, Aoun and Yates 2006).

Being cared for by tertiary clinics with multidisciplinary teams has been described as having positive impact on clinical outcomes, such as survival and lower hospitalization rates in neurodegenerative disorders (Chio *et al.* 2006) although recently other authors suggested that further prospective studies are needed to confirm the positive impact on outcomes that can really impact on patients' and caregivers QoL (Ng, Khan and Mathers 2009).

From a different perspective the involvement of SPCS in the care of people affected by chronic illnesses that span long periods of time, such as PD, entails negotiating the chronic-palliative interface and requires continued multi-disciplinary professional involvement as well as the integration and recognition of the care provided by family caregivers (Lanoix 2009). Furthermore the interaction among neurology, rehabilitation and palliative care evidence some specific areas of competence for each specialty, but also highlights common areas in which the different specialities, and therefore the various services operating in each sector, must collaborate to fulfil the unmet needs of the patients (Turner-Stokes *et al.* 2007).

In this study participants spontaneously discussed their satisfaction or dissatisfaction of the experienced services. All patients experienced hospital based services, mainly neurology clinics, rehabilitation services, dietitian services or respiratory units. Some needed the intervention of emergency services as well, both at home after calling the ambulance and in the A&E departments of the hospitals. Some participant also experienced home care services, usually provided by the primary medicine facilities, home physiotherapy or SLT. Respite admissions in different inpatients units and social – welfare services were also used by some of the interviewees and comments were collected.

None of the interviewees had experienced a SPCS, nor hospice inpatients admission before the interviews were conducted but some participants wanted to know what these service are and then provided some personal judgements about these facilities.

Those families in which the patient needed particular technical equipment, like mechanical ventilation, electronic communicators, nutritional pumps or orthopaedic aids, technical support services discussed their level of satisfaction or dissatisfaction.

Participants commented on the perceived quality of the received services, if they wished. The aim of this part of the interview was to find out which services were operating in the area where the forthcoming new SPCS was going to operate, their mode

of operation and their impact in terms of user satisfaction. The ultimate objective was to avoid to duplication of existing services, and potentially reinforce, integrate or substitute the less satisfactory services and care they had received.

Overall patients and family carers seemed satisfied of the specialist hospital based services, confirming that specialised units, often multidisciplinary, satisfy the needs of these people (Chio *et al.* 2006). Some criticisms of those services were about the difficult in accessing help rapidly in case of urgent needs. Less satisfaction were reported about the emergency services that seem not prepared to face the difficulties experienced by persons with chronic conditions and high disability. Primary care satisfaction depended mostly on the relationship that participants had with their GPs, the willingness of the latter to be involved in the home assistance of the patient and the specific expertise of the family doctors related to the medical problems faced by the patients. District nurses home care assistance was usually not seen as an appropriate service for these chronic conditions. Technical equipment services were generally defined as fine for those who needed it.

Social – welfare services were the most criticized together with the emergency services. These two services, in the area where the study was conducted, are not tailored on the needs of people with progressive, degenerative and life threatening conditions. Negative reports were directed against the welfare system for its delay in providing the necessary benefits and for the lack of sensitiveness of the operators towards the needs of the families. Carers testified how they had to struggle to obtain their rights and this is coherent with other published evidences (Edmonds *et al.* 2007a). Emergency services are thought and designed to meet the urgent needs of acute patients and their main goal is to save lives rather than improve the QoL. Patients were scared that inappropriate interventions from these services could result in unwanted aggressive treatments, such as intubation and invasive ventilation in neuromuscular patients who did not want any life prolonging measures. Unfortunately advanced care planning is not yet recognized by the Italian law and the risk to be subject to inappropriate treatment exist in Italy; lately the final decision if intervene or withhold is up to the emergency doctor, if the patient lost his or her capacity to communicate. This situation was so frightening for some patients that they decided not to call the emergency in problematic situations so not to be over treated.

In addition professionals working in A&E demonstrated a lack of communication skills about the end of life conditions and this was cause of further confusion for the patients and carers who needed to go to the hospital for acute care that complicated the chronic course of the neurological disorders.

Specialist services at home like the physiotherapy, SLT and the neurology visits (the latter provided for ALS/MND patients only) were very appreciated except for the fact that often they were intermittent. Physiotherapy and SLT, were provided in courses of 15-20 sections and often many months had to pass before receiving a second course. This inconvenience caused the loss of most of the therapeutic advantages achieved with the previous treatment.

Professionals interviewed in the focus groups reported frustration about the lack of continuity of the available services, regretted their sense of impotence due to the impossibility to provide care out-side the clinics to the very ill patients and recognized the potential positive role of a SPCS for both the home palliative care and for the potential use of respite admissions into the hospice.

The comments of those patients and carers who decided to discuss the potential role of a SPCS with respect to the potential help in their conditions were generally positive, even though some participants wanted to state that their conditions are different from cancer and (mostly MS patients) did not see themselves as terminally ill, but chronically ill.

3.6.3 Potential role of a new SPCS aimed at meeting the unmet needs

After the discussion of the various categories of unmet needs in this study participants researchers had to choose those domains, among those who emerged in this study, to be evaluated in the following quantitative study.

Among the various potential outcomes it was decided to choose those that were potentially modifiable by a SPCS.

In the physical symptoms group were identified the following:

- pain (and painful syndromes)
- breathlessness (and respiratory troubles)
- sleep disturbances
- intestinal symptoms
- urinary symptoms
- oral symptoms

In the psychological group were chosen:

- depression
- anxiety
- abandonment
- coping with the disease

In the social categories:

- Sense of isolation of both patients and carers
- Perceived quality of the experienced service for both patients and carers

In the spiritual group:

- The “meaning” of the lived experience of disease
- The help received from faith/religiousness

In addition the Quality of life of the patients and the Caregivers burden of care were chosen as main outcomes for the next study.

The forthcoming new SPCS is supposed to have a positive impact on some of the listed domains that are all part of the direct experience of patients, carers and professionals who face these situations every day.

3.6.4 Limitations of this study

1. The prevalence of problems revealed by the study is not to be considered as extendable to the general population, but it reflects the experience of the sample involved in the present study. Qualitative, by definition, this study was not aimed at calculating any epidemiological information about symptoms or other non physical problems related to the considered disorders. Information was in fact intended to explore the lived experiences of participants and to provide a direct in depth contact between potential users of a new SPCS and operators experienced in the palliative care assistance of people with cancer.

2. Sampling: participants were referred to the study by professionals involved in their care in hospitals (usually their neurologists). This was considered as a good source of potential participants with homogeneous problems and similar conditions. Professionals were asked to refer neurological patients with advanced disease and with unmet palliative care needs. The limit is, by definition, a potential selection bias due to the referers' vision about what the advanced stage was and their personal involvement with both patients and their carers. A further limit was the necessity to interview patients not completely cognitively impaired. This was necessary to obtain their consent to the interview and to receive some information by them, even if just a non verbal assent or deny to their carers declarations about the issues. Unfortunately this caused the potential patients in worst conditions, the ones with severely cognitive impairment, to be excluded by the study and, therefore, some important piece of information (mainly from their caregivers, of course) can have been lost in the current study.
3. Gender: 18 out of 22 patients were males. This was due to chance because gender equality was not considered as a referral goal of this study. Professionals justified this difference as the real situation of their follow up list of patients. In fact it is known that ALS/MND and PD tend to affect more males than females and that even though MS tend to affect more women than men, being male represents a higher risk for a worst prognosis and higher disability progression. Gender is not considered a discriminate for palliative care problems, therefore results can be claimed as reliable even if in presence of such an imbalance between the sexes in the sample.
4. Heterogeneous problems: needs and problems which came out from the interviews and the focus groups were very many and heterogeneous. This is expected in qualitative research when using in depth interviews led by participants when large amount of data are generated from relatively few sources (Payne 2007, p 149). Codes were generated to overcome this difficulty by using a systematic comparison of similar findings in the various events (Strauss and Corbin 1998, p 95)
5. Focus groups participants: Professionals involved in the 3 focus groups were all working in hospital. This happened because a fourth focus group with GP's and DN's working in a primary medicine district was scheduled, but participants decided not to participate at the last moment. The official justification was the very little experience that they have with the home care of patients severely affected by the neurological diseases studied in the present research project. A further limitation is the absence of nurses in the focus groups. When invited to participate nurses of the different clinics agreed to join the study, but they were present in any of the groups. Neurologists reported that this fact could be due to the difficulties of the nurses working in their departments to feel at ease in a group with physicians, specialists in neurology and rehabilitation. To overcome this a further focus groups for nurses only was set up in one of the hospital involved in the project, but again, at the last moment, potential participants denied to participate justifying their absence with problems related to their shifts in the ward or ambulatories.

3.6.5 Summary

From the content analysis and the detailed analysis of the interviews with patients severely affected by severe and progressive neurodegenerative disorders and from the focus groups with professionals involved in their care, many categories of potential palliative care unmet needs were found.

These needs were classified into the following categories: physical, psychological, social and spiritual unmet needs. Comments regarding the quality of the available services operating in the geographic area where the study was conducted were also captured and analyzed.

The main aim of this study was to identify, with a qualitative approach, a number of potential outcomes to be subsequently assessed in a phase II RCT trial, aimed at evaluate the impact of a new SPCS on the palliative care outcomes of this population.

These parameters were chosen from the unmet needs reported in the qualitative study. The chosen domains were six physical symptoms, four psychological issues, two social aspects, two spiritual themes in addition to the patients' Quality of life and the Caregivers burden of care, that are to be considered as traditional palliative care outcomes.

4. The quantitative study

4.1 Introduction

In this chapter of the thesis the quantitative component of the research project is reported.

This explorative randomized and controlled study was called Ne-Pal from the initials of Neurology and Palliative Care.

It represents the last experimental part of the research project and can be seen as a phase 2 study of the MRC framework (Campbell *et al.* 2000) as shown in the figure 7.

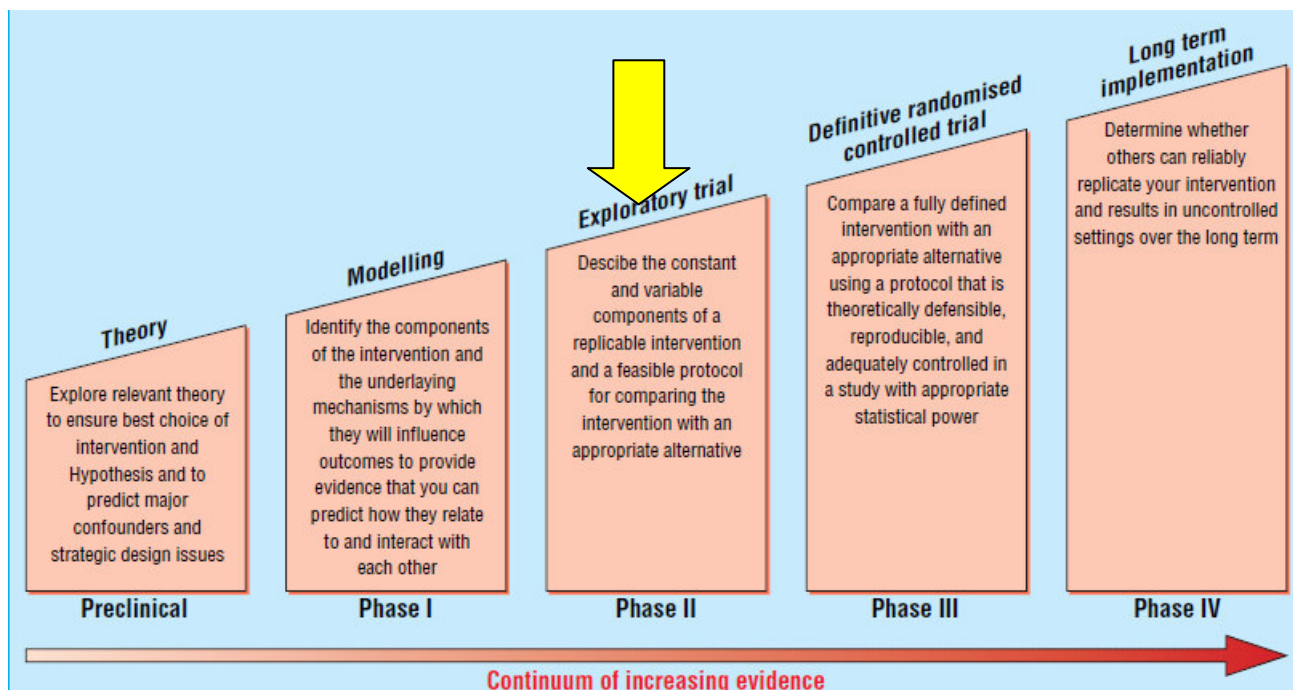


Fig 7: the MRC framework phase 2; the exploratory trial (Campbell *et al.* 2000)

4.1.1 Rationale of the study

Palliative care is no longer a luxury, but can now be defined as a necessity. Recently two major authors, both very well known for their important contributions to the development and the evaluation of palliative care services, stated that: “*The time has come to recognize palliative care as a necessity, and to invest in services and care and in the knowledge to develop them effectively and efficiently*” (Higginson, I. and Foley 2009). In the published literature several studies, book chapters (Borasio, Voltz and Bausewein 2003, Higginson, I. 1997a, Addington-Hall, J. M. and Higginson 2001) and some specific specialist books (Voltz *et al.* 2004, Oliver, Borasio and Walsh 2006, Byrne *et al.* 2009) have reported aspects related to the palliative care issues affecting patients with neurodegenerative disorders, and their caregivers. The majority of research has been based on specific diagnostic groups such as PD (Hudson, Toye and

Kristjanson 2006), ALS/MND (Mitsumoto and Rabkin 2007), or MS (McLeod, S 2001) and often focus on precise objectives such as the physical symptoms (Oliver 1998, Ganzini, Johnston and Silveira 2002, Goy, Carter and Ganzini 2008, Mancardi 2006, Saleem, Leigh and Higginson 2007, Lee *et al.* 2007, Elman *et al.* 2007, Higginson, I. J. *et al.* 2006a), the psychosocial aspects (Rabkin *et al.* 2005, Chio *et al.* 2005, Dawson and Kristjanson 2003, Gallagher, D. and Monroe 2006, Kristjanson, Toye and Dawson 2003, Love *et al.* 2005, Wasner *et al.* 2004, O'Brien, M. T. 1993a, Koopman 2003, Birgersson and Edberg 2004, Pasetti *et al.* 2003) or the spiritual issues (Lambert 2006, Murphy *et al.* 2000, Taylor and Herr 2004). Some discuss the role of the available services (Goodyear-Smith 2005, Kristjanson, Aoun and Oldham 2006, Wollin, Yates and Kristjanson 2006, Higginson, I. J. *et al.* 2006b, Chahine, Malik and Davis 2008, Kristjanson, Aoun and Yates 2006, Addington-Hall, J., Fakhoury and McCarthy 1998, Giles and Miyasaki 2009), but very few aimed at quantifying the impact of SPCS for this patient group (Edmonds *et al.* 2006, Higginson, I. J. *et al.* 2009). No studies were found in the literature exploring the impact of a SPCS for people severely affected by ALS/MND, MS, PDs (considered as a whole group of very ill neurological patients) on the main palliative care outcomes (Higginson, I. 1997a, p. 220, Doyle *et al.* 1995).

The issue of assessment of palliative care has been widely debated for cancer care and a recent review claims that the evidence for benefit from specialized palliative care is sparse and limited by methodological shortcomings (Zimmermann *et al.* 2008). Carefully planned trials, using a standardized palliative care intervention and measures constructed specifically for this population, are needed. In contrast with this position the project ENABLE 2 (Bakitas *et al.* 2009) showed with a RCT that QoL and depression were improved by the SPCS, even though the physical symptoms were not. A literature review stated that SPCS provide significant benefits in advanced cancer (Higginson, I. J. *et al.* 2003).

The qualitative assessment, described in the previous section of this thesis, highlighted various unmet palliative care needs in both patients and their informal carers, and provided a list of potential outcomes to be measured in this quantitative phase.

4.1.2 Brief precis of the study

To assess the potential impact of a new SPCS on some Palliative Care Outcomes (PCO) of people severely affected by ALS/MND, MS and PDs an explorative, randomized, phase 2 study, was set up by FARO Foundation in collaboration with the neurological clinics of two main hospital of Turin and its province.

The main aims were to quantify the impact of the new service on patients' QoL and caregivers' burden of care. Physical symptoms, psychosocial and spiritual issues were considered as part of participants' QoL and were also included among the outcomes. These domains were chosen among those which came out from the qualitative part of the research project (Neu-Needs).

Other objectives were to observe recruitment, drop out rate, mortality and, in general, the feasibility of a prospective randomized study in this population.

Methods, results, discussion and recommendations are presented and discussed in the following subchapters.

4.2 Aims of the study

4.2.1 General aim

To assess the impact of a new SPCS on some PCO in patients severely affected by ALS/MND, MS, PDs and in their informal carers

4.2.2 Specific aims

To compare the effects of a four months provision of a SPCS versus standard care on:

1. primary outcomes:
 - a. Patients QoL
 - b. Caregivers burden of care
2. secondary outcomes
 - a. Patients' symptom control, psychosocial needs and spiritual needs
 - b. Caregivers social needs
 - c. Overall service satisfaction
3. further outcomes:
 - a. recruitment process
 - b. attrition rate
 - c. mortality in the groups
 - d. feasibility of the study

4.3 Methodology of the quantitative study

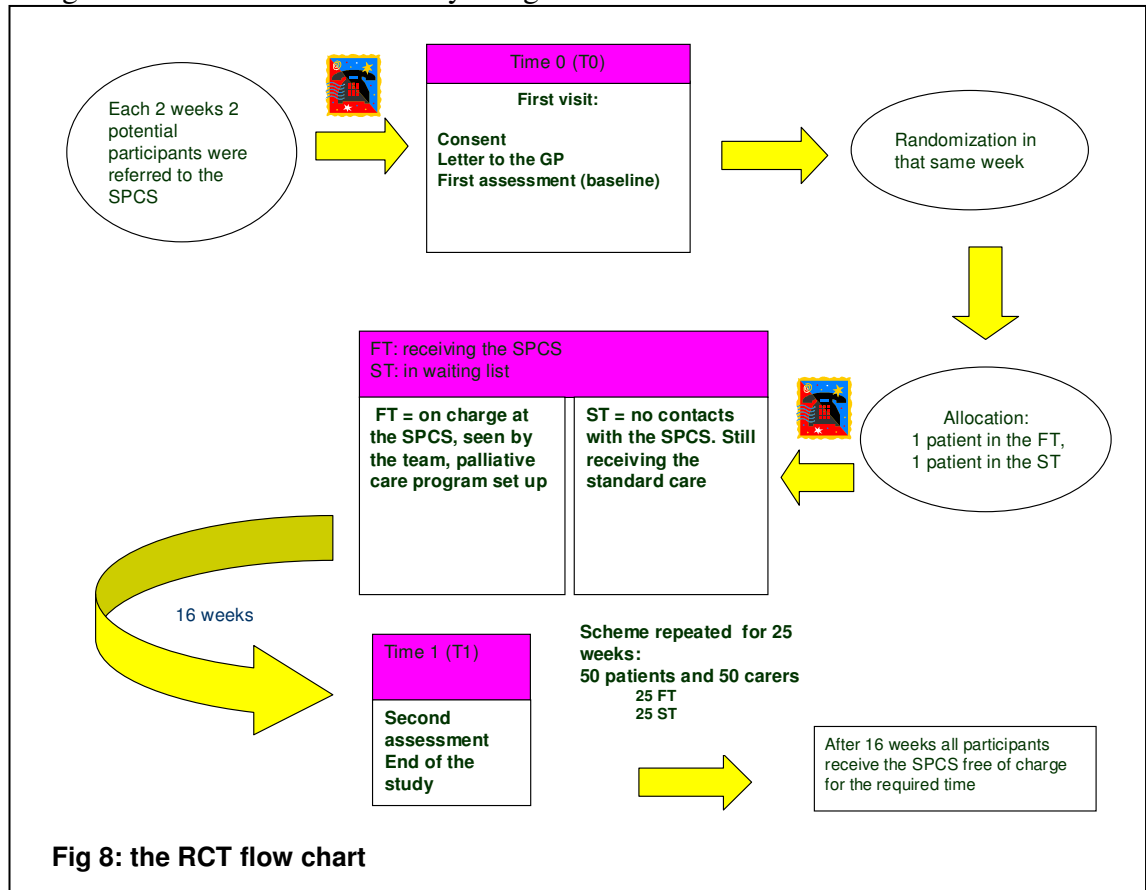
The study methodology was a Randomized Controlled Trial adopting the Fast Track (FT) versus Standard Track (ST) design (also called waiting list study).

This prospective randomized controlled study was previously tested in a similar study on MS patients and their carers by the Palliative and Supportive Care Research Group of the King's College, London (Higginson, I. J. *et al.* 2008). The authors concluded that this methodology was both feasible and reliable in a study of palliative care in which the prognostic features of participants would allow a wait similar to the length of the usual waiting list.

In this experimental design two arms of the study are created:

- a FT group in which the allocated participants receive immediately the intervention, in our case the SPCS.
- a ST group where the allocated participants wait for a period of time established by the study protocol (16 weeks), before receiving the intervention. In this period of time (the waiting list) participants of the ST receive the conventional available standard service (Best Standard Practice) that in this study was represented by the primary medicine service and the specific neurological hospital based services.

In figure 8 a flow chart of the study design is shown.



Potential participants were referred to the study by their professional carers, neurologists or other specialists working in hospital services (e.g. respiratory or rehabilitation clinics). Professional carers involved in participants' recruitment were part of the network of hospital based services involved in the overall service development and evaluation procedure. They had previously been informed of the inclusion and exclusion criteria (described below). They were asked to identify and refer patients in pairs each week, so that each pair was made of two patients with the same diagnosis and, possibly, similar clinical features (e.g. both with no respiratory equipment or both with non invasive ventilation or both in invasive ventilation). Professionals were also taught to introduce the general principle of the study in order to exclude those patients and their families not keen on participate at an experimental study.

Having obtained a preliminary informal consent to the participation in this study, the hospital specialists referred the potential participants to the research team. The researcher contacted patient or their family by telephone to explain the aims and the methodology of the study. The presence of the informal carer was encouraged at the first visit during this contact. If consent was obtained the research team set up a first visit at the participants' home, or in another setting if this was wished. During the first visit the study protocol was explained in details and a written copy of it was left for the participants, together with an informative letter for the GP, and a formal written consent form to be completed and signed by participants.

As difficulties in the signing and completion of the forms was expected for some patients with severe disability, an oral or a clear consent expressed in the presence of the researchers and the informal carer was considered valid.

After this formal phase had been completed the two researchers started the baseline assessment (T0). One researcher interviewed the patient while the second interviewed the caregiver. Usually this happened in two different rooms giving the chance to talk in depth of the participants' experiences. At the end of the visit the questionnaires and test sheets were collected and stored by the main researchers.

After the two potential participants were interviewed, the randomization occurred. This happened at the end of the same week of the interviews. Participants were immediately informed by telephone about the group they had been allocated in. The FT one was informed that they were going to be contacted by the SPCS at the beginning of the following week, while the ST was told that he or she would have been contacted in 16 weeks. The professionals who had referred the patients were then informed about the result of randomization.

During the 16 weeks of intervention participants in the FT group received the SPCS (described in details in the following subchapters), and could also receive the standard care if they wished. The ST group received the standard care and did not have any input from the SPCS.

After 16 weeks the 2 participants were reassessed by the research team, if still alive and not lost at the follow up.

This T1 interview represented the end of the study for both the FT and the ST participants, but it was also the starting point for the ST to receive the SPCS, while the FT could continue to be cared for by the SPCS if wished.

This sequence repeated for 50 weeks allowed the enrolment of 50 patients and their carers.

This simple methodology allowed a comparison between two groups of patients and their carers. Both were assessed at T0 (baseline) before being randomized. After 16 weeks FT had already received the SPCS, while the ST not, therefore a comparison among the groups reveals which differences was due to the SPCS input – at T1.

4.3.1 The Specialist Palliative Care Service

The SPCS represents the intervention in this study. It was a newly formed service set up by FARO Foundation in Turin in 2006. It was an extension of the existing specialist care palliative home service for cancer patients, operating in Turin city and its province since 1999, and the hospice inpatient facility with 14 beds, operating in Turin city since 2001.

The domiciliary team comprises physicians expert in palliative care and symptom control, nurses with specific training in palliative care, psychologists and physiotherapists. A team of volunteers collaborates with professionals providing help to the families.

When patients are cared for by the SPCS they receive regular home visits by both the doctor and the nurse (called the “mini-team”) and can be seen by the psychologist and/or by the physiotherapist if the mini-team members recognise the needs, which would be appropriate for further input. A bereavement service is offered after the death of the patients for the family components.

Visits are scheduled depending on patients' needs: in cancer patients they are seen on average twice per week by the doctor and twice by the nurse. The mini-team is on call for their patients Monday to Friday 08.00-20.00. In week-ends, or bank holidays, two doctors and four nurses are on call for all patients under the care of the SPCS. During the nights the service does not work, but there is an agreement with the emergency service to provide home assistance in case of serious problems occurring in the night hours that families could not solve with the indications previously provided by the mini-team. Medical notes are left in the patients' home and can be used by other professionals to ascertain the on-going therapy and the programme of care.

The Hospice facility is located inside an hospital of the city of Turin. The personnel work in shifts, covering 24 hours per day providing continuous assistance to the in-patients. Physicians working at the hospice are members of the home care team, visit patient daily and are on call in the night if emergencies occur.

Overall the FARO SPCS is composed by 60 professionals. All of them participated to the education programme included in the new SPCS for neurological patients. The professionals participated in lectures about the main palliative care needs of this population, attended field experiences in hospital wards and ambulatory clinics to learn about the use of respiratory devices and other technical equipment and were encouraged to participate to specific courses and congresses regarding palliative care in non cancer conditions.

4.3.2 The standard care

For standard care are meant all those social and health services provided by the Italian NHS in the area of the study (Turin city and its province).

Standard care included:

- the patients' General Practitioners.
- the District Nursing service.
- Neurologists, dieticians, rehabilitation specialists, respiratory specialists working in hospital ambulatory clinics .
- Emergency services.
- Social services, including paid carers and benefits advice .

4.3.3 Specific measured domains

According to the results of the qualitative analysis a number of relevant domains potentially modifiable by the intervention of the SPCS were identified:

1. Patients Qol
2. Caregivers burden of care
3. Physical symptoms:
 - a. pain
 - b. shortness of breath
 - c. quality of sleep
 - d. urinary problems
 - e. intestinal problems
 - f. oral symptoms
4. Psychological issues
 - a. feeling abandoned
 - b. coping with the disease

- c. anxiety
- d. depression
5. Social issues
 - a. social isolation
 - b. service satisfaction
6. Spiritual issues
 - a. meaning of the experience
 - b. help from faith

4.3.4 Measurement tools

The following tools were used to assess the previously listed domains:

1. Patients QoL: The Schedule For The Evaluation Of Individual Quality Of Life-Direct Weighting (SEIQoL-DW) (Mountain *et al.* 2004, O'Boyle, C. *et al.* 1996a, O'Boyle, C. *et al.* 1996b, Hickey *et al.* 1996)
2. Caregivers' burden of care: The Caregiver Burden Inventory (Novak and Guest 1989).
3. Physical symptoms (Pain, Breathlessness, Sleep disorders, Urinary symptoms, Intestinal symptoms and Oral symptoms): Visual Analogue Scales (VAS) or Numerical Rating Scales (NRS) 0-10
4. Psychological issues:
 - a. For Anxiety and Depression the Hospital Anxiety and Depression Scale (HADS)(Zigmond and Snaith 1983)
 - b. For the other domains (Feeling abandoned and Coping with the disease): Visual Analogue Scales (VAS) or Numerical Rating Scales (NRS) 0-10
5. Social issues (Social Isolation and Service Satisfaction rate)
 - a. Visual Analogue Scales (VAS) or Numerical Rating Scales (NRS) 0-10
6. Spiritual issues (Finding a meaning in the experience of disease, Help received from faith)
 - a. Visual Analogue Scales (VAS) or Numerical Rating Scales (NRS) 0-10

Patients were also assessed at baseline (T0) for:

- cognitive status: using a short form of the mini mental state test called abbreviated mental test score (AMTS) (Hodkinson 1972) to exclude the effect of a cognitive impairment on the results in the 2 groups
- physical disability using:
 1. general tools to evaluate the daily activity performances: the Activity of Daily Living (ADL) and Instrumental Activity of Daily Living (IADL) tests (Kempen and Suurmeijer 1990).
 2. specific disability scales for the three diagnostic groups
 - a. ALSFRS-R for ALS/MND patients (Cedarbaum *et al.* 1999)
 - b. Expanded Disability Status Scale (EDSS) for MS patients (Kurtzke 1983)
 - c. Hoehn and Yahr (H&Y) for PDs patients (Hoehn and Yahr 1967)

- the pressure sores risk and other skin problems related to disability using the Braden scale (Bergstrom *et al.* 1987)

All the used tools were translated in Italian and the participants records were gathered in personal folders (Case Record Forms- CRF) by the main researcher (SV).

An English version of the used forms is attached in the appendix section (Appendix 2, chapter 9.2).

The measurement tools were selected applying the following criteria:

- evidence from the literature of previous utilization and validation in studies involving patients with these diagnosis in a palliative care setting.
- advice from palliative care specialists involved in the clinical practice of this group of patients.

According to these principles some tools were validated and previously used to measure similar outcomes in patients with neurodegenerative disorders.

The Schedule For The Evaluation Of Individual Quality Of Life- Direct Weighting (SEIQoL-DW)

The SEIQOL-DW (Lee *et al.* 2006b, Chio *et al.* 2004a, Pollmann, Busch and Voltz 2005) had been used to measure the QoL in all the diagnostic groups. It has demonstrated reasonable construct validity for use in assessing individual quality of life in a group of individuals with other neurological disorders (LeVasseur, Green and Talman 2005) even though in patients with cardiac diseases seems to be a good instrument to determine patients determinants of QoL rather than a quality of life instrument (Moons *et al.* 2004).

The schedule for the evaluation of individual quality of life (SEIQoL) was developed to assess quality of life from the individual's perspective (McGee *et al.* 1991, O'Boyle, C. A. *et al.* 1992). It is an interview based instrument. The investigator can assess the level of functioning in, and relative importance of, those areas of life nominated by the respondent. This tool has been defined as a complex measure of a complex process and its use in routine clinical situations may prove impractical (Hickey *et al.* 1996). To overcome this difficulty a shorter form has been created: the SEIQoL-direct weighting (SEIQoL-DW) (O'Boyle, C. *et al.* 1996a, O'Boyle, C. *et al.* 1996b).

The direct weighting instrument is a simple apparatus consisting of five interlocking, coloured laminated circular disks that can be rotated around a central point to form a type of piechart. The laminated disks are mounted on a larger backing disk, which displays a scale from 0 to 100, and from which the relative size of each coloured segment can be read.

Each segment is labelled with a life area nominated by the respondent as being important to his or her overall quality of life. The respondent adjusts the disks until the size of each coloured segment corresponds to the relative importance of the life area represented by that segment. These segments may be adjusted and readjusted until respondents are satisfied that the proportion of the pie chart given to each life area accurately reflects the relative weights they attach to those life areas. The weighting procedure is quick to administer, colourful, tactile, and easy to understand. The SEIQoL-DW was developed and validated and found to be a valid and reliable measure of explicit weighting policies for quality of life domains (Hickey *et al.* 1996).

The SEIQOL-DW index is the overall measure of individual QoL. It is the result of a formula involving the satisfaction and the weight of each elicited area and is a number ranging from 0-100 - the higher meaning a higher QoL.

The Caregiver Burden Inventory (CBI)

The Caregiver Burden Inventory (CBI) is a 24-item, five-subscale multidimensional measure of caregiver burden giving a sensitive reading of caregivers' feelings and a sophisticated picture of caregivers' responses to the demands of caring (Novak and Guest 1989). In a previous study with caregivers of ALS/MND patients authors demonstrated a correlation between the level of quality of life of the informal carers and the worsening of patients' disability (Chio *et al.* 2005). The five sub-scales explore time dedicated to:

- the care
- impact on life
- physical fatigue
- social factors
- emotional issues.

For each item, carers are asked to indicate how often they have felt that way (0-4). The final index is a composite measure, combining the different aspects of caregivers' reactions to their involvement (Higginson, I. and Harding 2007, p 107). The sum of the scores of the subscales (ranging from 0-96) provides the overall final index. The higher results indicate a higher burden of care and, consequently, a lower QoL of the carers.

Visual Analogue Scales (VAS) and Numerical Rating Scales (NRS)

VAS and NRS are common measurement tools used in palliative care (Caraceni *et al.* 2002). They are simple instruments used in clinical and research settings generally used to measure the intensity of a phenomenon (Hauser and Walsh 2008).

VAS are represented by a linear row, usually 10 cm long, in which at both endings the measured object is defined. One extreme of the line is defined as the absence of the symptom or problem, the other is the maximum intensity imagined by the participant. Participants are asked to define one point of the line representing the intensity of the studied problem. Researchers, using a ruler, report the intensity of the symptom as the closest number 1-10 from the point indicated by the participant.

NRS are similar to VAS, but in this case numbers are shown 0-10 and the responder has to elicit the intensity of the problem from the same interval.

These tools are defined as interchangeable showing overall good feasibility, reliability (internal consistency), and convergent validity and can be chosen depending from the physical ability of participants to draw a point on the line or respond to the numerical scale choosing the identified number (Hollen *et al.* 2005).

In the present study VAS and NRS were chosen to assess those domains identified from the qualitative study for which no validated measurement tool could be found in the literature. General and specific outcome measurement tools widely applied in the palliative care settings, such as the Palliative Outcome Score, recently extended and used in a study on MS (Higginson, I. J. *et al.* 2009), measure many of the listed problems, but are not validated for ALS/MND or PD. For this reason, and considering the explorative purpose of the present trial, the research team decided to measure the impact of the SPCS on the physical, psychological, social and spiritual issues highlighted in the previous qualitative study, singularly, by using these simple tools in which participants could rate the problem intensity before and after the intervention or the standard care.

The Hospital Anxiety and Depression Scale (HADS)

This tool has been used to assess the presence of anxiety and depression in palliative care (Doyle *et al.* 1995).

It consists of two scales, one for anxiety and the second for depression both formed of 7 items. Each item has a four point (0–3) response category so the possible scores range from 0 to 21 for anxiety and 0 to 21 for depression. An analysis of scores on the two subscales could be regarded as being in the normal range if the score is of 0 to 7 for either subscale, a score of 11 or higher indicating probable presence ('caseness') of the mood disorder and a score of 8 to 10 being just suggestive of the presence of the respective state (Snaith 2003).

The Abbreviated Mental Test Score (AMTS)

This ten question test is the short form of the Mini-Mental State Examination-MMSE (Holsinger *et al.* 2007, Folstein, Folstein and McHugh 1975). Score 0-6 indicate a strong suspect of dementia, 7-8 can relate to a certain degree of mental confusion, 9 or 10 mean normal cognitive status (Hodkinson 1972).

The Activity of Daily Living (ADL) and Instrumental Activity of Daily Living (IADL) test

This general tool is formed of two subscales measuring the capability of patients to be independent or dependent in the daily living activities, such as feeding, bathing, clothing, moving independently and the voiding functions (ADL) or their possibility to use instruments like the telephone, money, drugs, transports, home cleaning, cloth washing, cooking, and shopping (IADL). The two subscales range respectively from 0-6 (ADL) and 0-8 (IADL), the higher scores indicating better performances (Kempen and Suurmeijer 1990). This instrument was used to have an inter diagnostic parameter of disability in order to evaluate the degree of general physical impairment in the group of study.

The Amyotrophic lateral sclerosis functional rating scale revised (ALSFERS-R)

The ALSFRS-R is a well-established and widely distributed score for the functional status of patients with ALS (Cedarbaum *et al.* 1999). It is based on 12 items, each of which is rated on a 0–4 point scale. The rate of total functional disability thus ranges from 0 (maximum disability) to 48 (normal) points.

The Amyotrophic lateral sclerosis functional rating scale revised (ALSFERS-R) score and its progression rate have been debated as survival predictors (Kollewe *et al.* 2008).

The Expanded Disability Status Scale (EDSS)

The Expanded Disability Status Scale (EDSS) is a disability scale used in MS. It ranges from 0 (no physical impairment, normal neurological exam) to 10 (death due to MS) with steps of 0.5 points. EDSS steps 1.0 to 4.5 refer to patients who are fully ambulatory, 5.0 to 7.5 present progressive physical impairment to ambulation. 8.0 or more are patients essentially restricted to bed or chair, 9.0 are defined helpless patients still able to eat and talk. 9.5 of EDSS is a patient unable to communicate effectively or eat/swallow (Kurtzke 1983).

The Hoehn and Yahr score (H&Y)

The Hoehn and Yahr score is a disability scale for people affected by movement disorders. It is formed of five stages 1-5. Stage 1 describes minimum no physical impairment due to the neurological disorder. Stage 4 is described as severely debilitating and stage 5 is for bed bound patients (Hoehn and Yahr 1967).

H&Y 3,5 or more is defined by Bunting-Perry as an advanced stage of PD (Bunting-Perry 2006), probably this overlap with the definition of palliative care stage proposed by Thomas and Mc Mahon (Thomas and MacMahon 2004a).

The Braden Scale

The Braden scale is a risk assessment tool for pressure ulcers. It is a scale ranging from 4-23 points that considers 6 indicators: humidity, sensitivity, nutritional status, movement, friction and slip. Lower scores relate with higher pressure sore risk. 16 is considered a cut off point below which prevention strategies are to be started to avoid the risk of pressure sore appearance (Bergstrom *et al.* 1998, Bergstrom *et al.* 1987).

4.3.5 Participants

Participants of the Ne-Pal study were patients severely affected by neurodegenerative conditions and their main informal caregivers. Inclusion and exclusion criteria are now described.

4.3.5.1 Inclusion criteria

Participants of the pilot RCT were adult patients of both genders severely affected by ALS/MND, MS and PDs and their informal caregivers.

Patients had to be resident in Turin city or in the metropolitan area (territory of provision of the FARO SPCS).

The severity of the disease was defined according to the Gold Standards Framework prognostic indicators specific for the different conditions as shown below (The-Gold-Standards-Framework 2008):

For ALS/MND: At least one of the following conditions were to be satisfied:

- Evidence of disturbed sleep related to respiratory muscle weakness in addition to signs of dyspnoea at rest
- Barely intelligible speech
- Difficulty swallowing
- Poor nutritional status
- Needing assistance with ADL's
- Medical complications eg pneumonia, sepsis
- A low vital capacity (below 50% of predicted, hypercapnia)

For Multiple Sclerosis:

1. EDSS \geq 8,5
2. presence of at least one of the following conditions:
 - Significant complex symptoms and medical complications

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- Dysphagia (swallowing difficulties)
- Admissions with sepsis and poor nutritional status
- Communication difficulties e.g. Dysarthria + fatigue
- Mild cognitive impairment
- Breathlessness

For PD, MSA o PSP presence of the following criteria:

1. Hoehn and Jahre stage ≥ 4
2. No indication for neurosurgical procedures

The presence of 2 or more of the following criteria:

- Drug treatment is no longer as effective / an increasingly complex regime of drug treatments
- Reduced independence, need for help with daily living
- Recognition that the condition has become less controlled and less predictable with “off” periods
- Dyskinesias, mobility problems and falls
- Swallowing problems
- Psychiatric signs (depression, anxiety, hallucinations, psychosis)

4.3.5.2 Exclusion criteria

- Diagnosis other than those listed
- Severe cognitive impairment
- Unable to express their views even with communication aids
- Unable to give consent
- Resident outside of the territory covered by FARO SPCS

4.3.6 Referrals

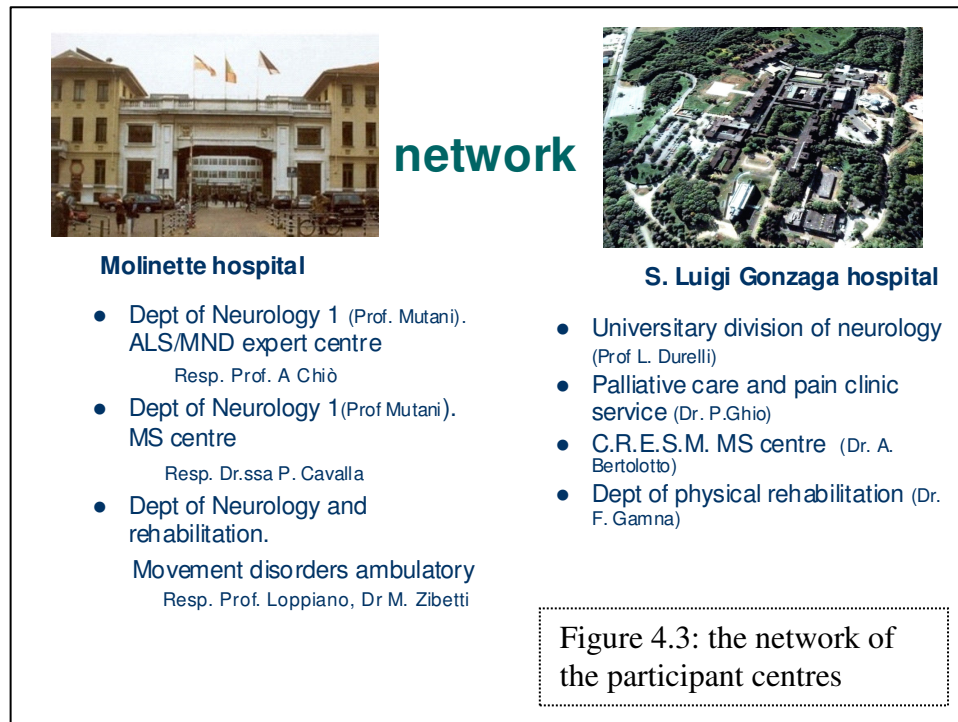
Participants were referred to the study by their specialists, generally neurologists, respiratory specialists or rehabilitation specialists working in the participant centres.

The study protocol was sent to the specialist neurological services of the two hospital involved in the project: Molinette Hospital of Turin city and San Luigi Gonzaga Hospital of Orbassano.

Professionals involved in patients recruitment selected the potential participants according to the previously listed inclusion criteria, asked a preliminary consent to their patients and then communicated the patients contact details to the research team.

4.3.7 Participant Centres

Participant centres are listed in figure 4.3



4.3.8 Research team

The research team was composed of five researchers: two palliative care physicians, two nurses and one neurologist.

The role of the team members was:

- To receive referrals from the participant centres
- To assess participants at T0 and T1
- To randomize and allocate patients in the two study groups
- To collect patients forms and to analyse data

One of the two palliative care physician (SV) was the main researcher and is the author of this thesis.

4.3.9 Data Collection

Each participant was identified with an alpha numerical code. Each patient had a paper form where demographics, clinical evaluation and consent form were collected.

The main researcher was responsible for the data collection and the privacy of the content.

Assessments were conducted by a team of two researchers, one doctor and one nurse. During the interviews usually the doctor interviewed the patient while the nurse assessed the carer.

Participants' forms were filled at T0 and reused at T1 in order to have a single participant form to be analyzed at the end of the data collection process.

4.3.10 Sample size

Being an explorative pilot study the sample calculation was not made on statistical methods, but it was determined on the increase of the work load of the FARO SPCS expected during the enrolment and follow up phases.

FARO regular patients load is about 100 patients at any time. It was considered as acceptable an increase of 50%, represented by the new neurological assistances. This allowed a sample size of 50 participants divided in 25 in the FT and 25 in the ST.

4.3.11 Data Analysis

Data analysis plan is composed of two main aspects: a clinical and a statistical analysis.

4.3.11.1 Clinical analysis

The clinical analysis consider the differences in the two study groups (FT and ST) in terms of clinical significance. Clinical significance yields information on whether a treatment is effective enough to change a patient's diagnostic label or to make a significant change in patients' condition. For pain and sleep disturbance this change has been described as bigger than 13% and 10% respectively (Gallagher, E. John, Liebman and Bijur 2001, Nava and Tali 2003). Published works have demonstrated that a change of 50% of the standard deviation of any QoL tool can be considered a clinically significant change. This translates to a change of 8 to 10 points on a 100-points scale (Sloan 2002).

Changes in participants' tests can be either positive or negative from baseline (T0) and the second assessment (T1). The clinical interpretation of the results in this study has been categorised in three possible groups:

- No clinical significance if the difference between T1 and T0 is lower than 10% of the scale used to measure that domain
- Moderate clinical significance if that difference is between 10 - 19% of the same scale
- Relevant clinical significance when the difference is 20% or higher.

4.3.11.2 Statistical analysis

The statistical analysis plan includes:

- descriptive statistics to evaluate the comparability of the two study groups.
- use of group comparison test to detect differences between the two study groups that can be caused by the intervention. The aim of this procedure is to determine the efficacy analysis. Depending on the characteristics of the variables the Analysis of Covariance (ANCOVA), independent t-test, Chi Square test or non parametric tests have been used to highlight statistical changes between the two groups.

Statistical analysis will be conducted at three different moments:

1. at baseline (T0) to describe the characteristics of the sample (demographics, disability etc), calculate the prevalence of the explored needs and compare the two groups for the outcome measures. The mean scores at baseline will be used as covariate for the ANCOVA test.
2. at T1, after that the FT group will have received the SPCS. This groups

comparison will highlight those variables whose scores will be significantly different between the groups. The latter will be used as dependent variables in the ANCOVA test.

3. comparing mean T1 and T0 scores. This procedure will be performed by:
 - subtracting the mean differences of the outcome measures at T1 – T0 and then calculating the differences obtained between the two groups. This allows to calculate the eventual clinical significant difference between the two groups that can be due to the intervention (having received the SPCS).
 - Performing the ANCOVA test. This allows to determine the eventual statically significant difference between the group due to the intervention. For those variables resulting statistically different at this test the null hypothesis (see below) can be refused.

In the data analysis comparing the two groups (T1-T0) only those participants who completed both the baseline and the post intervention assessment will be included. The death rate will be compared between the two groups to evaluate if the intervention could affect mortality. Patients lost at follow up or that voluntarily ask to leave the study are considered as “drop out”. The dataset was saved both in Microsoft Excell and SPSS software packages to be analyzed.

The null hypothesis

The null hypothesis of this study is that no changes are to be found between the treated and the control groups after the intervention represented by the provision of the SPCS in the treated group versus the best standard care in the control group.

Alpha value

The alpha value, determining the statistical significance found in this study, is set at 0,05. Bonferroni adjustments can be applied in case of multiple variable analysis. In particular this adjustment will be applied for the ANCOVA between the two groups using the T1 mean scores as dependent variables, the two groups (FT/ST) as independent variable and the T0 mean scores as covariate. This test will be applied for the variables that will be significantly different between the groups at T1 and the Bonferroni adjustment will be applied by dividing the pre-set α value (0.05) for the number of the considered variables. The new value obtained will be considered as the new α value determining the statistical significance between the two groups.

4.3.12 Randomization process

The randomization process is very simple and straightforward. It is repeated each week after the recruitment of each pair of participants. The participants CRFs are wrapped in two identical white folders, anonymous and without any possible sign of recognition. The main researcher put both folders on one of the FARO Foundation secretary's workdesk asking him or her to choose one. The selected one became the FT participant, while the other is allocated in the ST group.

4.4 Ethics

The study protocol was submitted and approved by the Ethics Committee (EC) of FARO Foundation. The approval was later sent to the two involved hospital EC's and both declared to be satisfied by the FARO EC approval and did not proceed with a further analysis. The University of Kent Ethics Committee was later informed of the ethics approval of the protocol. A copy of the FARO EC approval is appended in the appendix section of this thesis (Appendix 2, Chapter 9.4.3)

4.4.1 Informed consent

Before the enrolment in the study researchers provide the explanatory sheet and the consent forms (approved by the EC) to the patient and to his or her carer. This allows participants to be informed of the study aims and methodology according to the Declaration of Helsinki developed by the World Medical Association (WMA 2008). A copy of the information letter to the patient's general practitioner is also given to the family asking them to deliver it to their family doctor at the first occasion.

4.4.2 Case record form (CRF)

The main researcher is responsible for all the information included in the CRF. The latter is available for revision in case of control during the trial and afterwards. Data are stored safely by the main researcher both in paper and electronic format.

4.5 The quantitative study results

In this section the results of the Ne-Pal (quantitative randomised control study) study are shown.

4.5.1 Participants

4.5.1.1 Referred patients

From March 31st 2008 to February 28th 2009 52 potential participants were referred to the Ne-Pal study for recruitment by professionals working in the participating centres listed in the methods section.

Participants referred had the following diagnosis:

- Multiple Sclerosis (MS) n=20
- Amyotrophic Lateral Sclerosis (ALS\MND) n=16
- Movement disorders (PDs) n= 16 (PD n=12, MSA n=2, PSP n=2)

4.5.1.2 Potential participants who denied consent

Two out of the 52 referred patients (3,8%) did not consent to participate in the study - both with a diagnosis of MS:

- One said that he did not feel seriously ill and for this reason did not want to be cared for by a SPCT.
- One had recently been diagnosed a breast cancer and was going to start chemotherapy and for this reason decided not to enter into the study protocol.

4.5.1.3 Enrolled participants

The remaining 50 patients and their 50 informal caregivers gave their informed consent to the participation and were therefore enrolled in the Ne-Pal explorative pilot randomized controlled study.

In table 4.1 participants demographics are shown.

4.5.2 General considerations

Patients	N= (%)
Enrolled	50
Randomized	25 FastTrack (50%), 25 StandardTrack (50%)
Gender	M=30 (60%) F=20 (40%)
Diagnostic groups	ALS/MND=16 (32%) MS=18 (36%) PDs=16 (32%)
Age classes (in years)	<44 =8 (16%) 45-64 =17 (34%) >65 =25 (50%)
Informal caregivers n=50 (100%)	Wife=25(50%), Husband=12(24%), Daughter=4(8%), Son=1(2%), Mother=1(2%), Sister=1(2%), Brother=1(2%), Friend=1(2%), Paid Carer=4(8%)
Table 4.1 Ne-Pal participants' demographics and general characteristics	

All the 50 participants enrolled in the study met the inclusion criteria. Two patients were not able to complete the SeiQoL-DW interview and therefore their QoL index at baseline is missing. All patients were able to complete the assessment of symptoms. The psychosocial and spiritual assessment was completed by all participants but two.

Patients were asked to identify their main informal caregiver and decide whether he or she could answer to the tools used for carers. Five patients did not wish their carer to be involved so 5 carers scores have not been included.

Of these five patients who did not want their carers to be involved:

- four were cared for by paid carers
- one was cared for by his wife.

4.5.3 The results at baseline (T0)

4.5.3.1 Outcome measures results at baseline (T0) of the overall sample

In this sub chapter the results of the overall sample are displayed.

The analysis of the tests performed at baseline (T0) revealed the following results

The patients' Quality of Life

The SeiQoL-DW was the first test performed during the baseline assessment. On average it took about 30 minutes to be completed. 48 of the 50 participants could answer to the interview, even though very few could manipulate the disk and for this reason researchers moved it for them until the desired result was achieved.

SeiQoL-DW Elicited cues

The 48 patients who completed the SeiQoL-DW at baseline elicited a total of 239 cues: 47 chose 5 cues each, while one only elicited 4.

In the following table (4.2) a list of the elicited cues at baseline is reported and results are compared to the 203 clues elicited at T1 by the remaining 41 patients who completed the test. Results are displayed in absolute figures and percentage of the sample size.

Cues categories	T0 elicited cues. N=48, (%) *first 5 ranks	T1 elicited cues N=41, (%) * first 5 ranks
Family	48 (100%) *1	41 (100%) *1
Spiritual life	22 (46%)	18 (44%) *5
Entertainment	30 (62.5%) *2	40 (97.5%) *2
Independence	23 (48%) *5	9 (22%)
Symptoms	16 (33%)	9 (22%)
Health status	26 (54%) *3	23 (56%) *3
Friends- relationships	23 (48%) *4	21 (51%) *4
Finance-economic concerns	11 (23%)	8 (19.5%)
Social concerns	14 (29%)	9 (22%)
Information – communication	7 (14.5%)	3 (7.3%)
Others	20 (41.6%)	22 (53.5%)
Total	239	203

Tab 4.2: List of the SeiQoL-DW cues elicited by the patients at baseline (T0) and after the intervention (T1)

The elicited cues, components of the individual QoL of the patients, showed that family is the most frequent cue being present in 100% of the interviews both at baseline and after the follow up period. Entertainment related cues occupy the second position in the choices of the patients with a higher frequency at T1. Health status cues are in the third position at T0 and T1, even though symptoms were considered as a separate group. If these 2 categories were grouped they would represent the 87% of the elicited cues at baseline and 78% at T1. Friends and relationships were in the 4th position in both assessments, whereas at the 5th standings there were independence at baseline and spiritual life at T1.

SeiQoL-DW index

The mean QoL level at baseline of the overall sample is described by the SeiQoL-DW index resulting from the following formula:

$$SEIQoL\ Index = \Sigma(levels \times weights)/100$$

Higher SEIQoL-DW index values indicate higher Quality of life.

SEIQoL index at baseline N=48 mean=62,8 (SD=19,45)

Table 4.3: the overall sample SEIQoL-DW index mean value (SD)

The mean score of QoL in the enrolled patients at baseline was 62,8 on a scale 0-100 (see Table 4.3).

The Caregivers’ Burden of Care

The burden of care was assessed using the CBI inventory. This tool was completed by 45 informal carers at time 0.

The CBI inventory results on the overall sample of carers are reported in table 4.4 divided in the 5 sub-domains (time dedicated to the care, impact on life, physical fatigue, social factors and emotional issues) and the final index. Higher values indicate a higher burden of care.

Domain	N= Number of respondents Mean= Mean value within the sample SD= Standard Deviation		
Time dedicated to the care (Range 0-20)	N=45	mean=16,51	SD=4,346
Impact on life (Range 0-20)	N=45	mean=13,78	SD=5,965
Physical fatigue (Range 0-16)	N=45	mean=8,69	SD=5,464
Social factors (Range 0-20)	N=45	mean=4,38	SD=4,019
Emotional issues (Range 0-20)	N=45	mean=1,42	SD=1,971
OverallCBI index (Range 0-96)	N=45	mean=44,78	SD=14,953

Table 4.4: CBI results at baseline

The Physical symptoms

The explored physical symptoms were pain, breathlessness, quality of sleep, urinary symptoms, bowel symptoms and oral symptoms. Symptoms were assessed using Visual Analogue Scales (0-10 cm) or Numerical Rating Scales (NRS 0-10) depending on the movement possibility of the patients. The value 0 meant absence of the symptoms, the value 10 meant the worst possible symptom experience.

The physical symptoms prevalence and intensity at baseline, for the overall sample, are reported in the tab 4.5.

Symptom	Prevalence	Categorical intensity: Mild=1-3 Moderate= 4-7 Severe= 8-10	Continuous Intensity VAS (0-10 cm) or NRS (0-10 points)
Pain (N=50)	35/50 (70%)	Mild=19 (38%) Moderate=8(16%) Severe= 8(16%)	N= 50 mean=3,38 SD=3,17
Breathlessness (N=50)	36/50 (72%)	Mild=19 (38%) Moderate=15(30%) Severe= 2(4%)	N=50 mean=3,24 SD=2,71
Sleep disorders (N=50)	34/50 (68%)	Mild=14 (28%) Moderate=12(24%) Severe= 8(16%)	N=50 mean=3,34 SD=3,16
Urinary symptoms (N=50)	35/50 (70%)	Mild=11 (22%) Moderate=13(26%) Severe= 4(22%)	N=50 mean=4,14 SD=3,53
Bowel symptoms (N=50)	40/50 (80%)	Mild=9 (18%) Moderate=19(38%) Severe= 12(24%)	N=50 mean=4,96 SD=3,46
Oral symptoms (N=50)	42/50 (84%)	Mild=15 (30%) Moderate=19(38%) Severe= 8(16%)	N=50 mean=4,32 SD=2,93
Tab 4.5 Symptom prevalence and intensity at baseline for the overall sample			

The Psychological symptoms

The prevalence and intensity of Anxiety and Depression at baseline, for the overall sample, are reported in the tab 4.6.

Symptom	Prevalence= Borderline + Caseness	Categorical classification Normal=0-7 Borderline= 8-10 Caseness= 11+	Continuous Intensity VAS (0-10 cm) or NRS (0-10 points)
Anxiety HADS-A N=48	18 (37,5%)	Normal=30(62.5%) Borderline=9(18.75%) Caseness=9(18.75%)	N=48 mean=6,98 SD=4,34
Depression HADS D N=48	18 (37,5%)	Normal=30(62.5%) Borderline=8(16.7%) Caseness=10(20,8%)	N=48 mean=6,75 SD=4,04

Tab 4.6: Anxiety and Depression prevalence and intensity at baseline for the overall sample

The other two psychological domains were assessed using Visual Analogue Scales (0-10 cm) or Numerical Rating Scales (NRS 0-10) depending on the ability to move of the patient. The value 0 meant that the patient felt totally abandoned or could not cope with the disease at all, whereas the value 10 meant that the patient was not feeling abandoned at all or could cope perfectly with the disease.

The mean intensity of the two measured psychological domains, Feeling abandoned and Coping with the disease, are reported in the tab 4.7.

Psychological domains	Continuous Intensity VAS (0-10 cm) or NRS (0-10 points)
Feeling abandoned	N=49 mean=7,45 SD=2,82
Coping with the disease	N=49 mean=5,02 SD=3,16

Tab 4.7 Feeling abandoned and Coping with the disease mean intensity at baseline for the overall sample

The Spiritual issues

The spiritual issues measured were the “meaning of the experience” related to the disease and the “help received from faith”.

These domains were assessed using Visual Analogue Scales (0-10 cm) or Numerical Rating Scales (NRS 0-10) depending on the movement possibility of the patients. The value 0 meant that the patient did not find any meaning in the lived experience of the disease or did not feel helped by the faith at all, whereas the value 10 meant that the

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patient found a full meaning of the lived experience of the disease or felt totally helped by the faith. Results are summarized in table 4.8

Spiritual domains	Continuous Intensity VAS (0-10 cm) or NRS (0-10 points)		
Meaning	N=49	mean=3,76	SD=3,77
Help from faith	N=49	mean=5,08	SD=3,70

Tab 4.8 Spiritual domains mean intensity at baseline for the overall sample

The Social issues

The social domains assessed were the sense of social isolation due to the disease and the satisfaction of the services received. These social issues were assessed both in the patients and the carers.

The sense of social isolation was assessed using Visual Analogue Scales (0-10 cm) or Numerical Rating Scales (NRS 0-10). The value 0 meant that the participants felt totally isolated because of the disease, whereas 10 represented that participants did not find isolated at all.

The service satisfaction was assessed using the same uni-dimensional scale were 0 meant that participants were totally unsatisfied by the available services, while 10 meant total satisfaction.

The results of these two domains assessed both in the patients and the informal caregivers are shown in tab 4.9.

Social issues	Continuous Intensity VAS (0-10 cm) or NRS (0-10 points)		
Sense of social isolation for the patients	N=48	mean=5,27	SD=3,23
Sense of social isolation for the carers	N=46	mean=5,76	SD=2,98
Service satisfaction for the patients	N=48	mean=6,83	SD=2,22
Service satisfaction for the carers	N=46	mean=6,41	SD=2,41

Tab 4.9 Social issues mean values at baseline for the overall sample

4.5.3.2 Baseline results and groups comparison

A description of the sample with comparison between the two groups of the study (Fast Track and Standard Track) is now shown.

	Overall (%)	Fast Track (%)	Standard Track (%)	Statistical comparison
Gender	M=30 (60%) F=20 (40%)	M=16 (64%) F=9 (36%)	M=14 (56%) F=11 (44%)	no statistical difference, $\chi^2=.083$, $p=.77$ Phi= .082
Diagnosis	ALS/MND=16(32%) MS= 18 (36%) PDs= 16 (32%)	ALS/MND=7(28%) MS=10 (40%) PDs 8 (32%)	ALS/MND=9(36%) MS=8 (32%) PDs=8 (32%)	no statistical difference, $\chi^2=.472$, $p=.790$ Phi= .097
Age Groups	<44=8 (16%) 45-64=17 (34%) >65= 25 (50%)	<44=2 (8%) 45-64=12 (48%) >65=11 (44%)	<44=6 (24%) 45-64= 5 (20%) >65= 14 (56%)	no statistical difference, $\chi^2=5.242$, $p=.073$ Phi= .324
Disability specific scales	ALSFRS-r=10,2 EDSS=8,5 H%Y=4,2	ALSFRS-r=10,7 EDSS=8,6 H%Y=4,25	ALSFRS-r=9,8 EDSS=8,4 H%Y=4,12	Independent samples t-test ALSFRS=p.83 EDSS=p.58 H&Y=p.554
ADL	N=50 mean=1,08 SD=1,589	N=25 mean=1,04 SD=1,513	N=25 mean=1,12 SD=1,691	Independent samples t-test no statistical difference, $p=.861$
IADL	N=50 mean=0,98 SD=1,36	N=25 mean=1,20 SD=1,5	N=25 mean=0,76 SD=1,20	Independent samples t-test no statistical difference, $p=.258$
BRADEN SCALE	N=50 mean=15,86 SD=3,289	N=25 mean=16,28 SD=3,298	N=25 mean=15,44 SD=3,292	Independent samples t-test no statistical difference, $p=.372$
AMTS	N=46 mean=9,30 SD=1,33	N=25 mean=9,52 SD=1,0	N=21 mean=9,05 SD=1,6	Independent samples t-test no statistical difference, $p=.235$

Tab 4.10: Patients demographics and main characteristics at the baseline assessment, groups comparison and statistical analysis

These results show that the two groups were similar in their characteristics and dependency.

There were no statistically significant differences in gender, diagnostic groups and age groups between the two groups of the study.

The disability levels were high for all the three diagnostic groups and similar for the two groups of the study. This is confirmed both by the specific disability scales and by the general ADL/IADL scales. As described in the methods for ALS/MND the ALSFRS-R scale ranges from 0 to 48 , where 48 means no impairment in all the considered domains and 0 is a totally dependent person. On the contrary the EDSS for the SM group ranges 0-10 and the H&Y for the PDs from 0-5. In these cases lower values indicate better performance and, therefore lower disability, whereas higher scores indicates higher disability. For these reasons we can conclude that patients enrolled in the Ne-Pal study were very disabled and their disease progression was to a advanced stage, causing severe physical impairment . No statistically significant differences were found between the two groups after randomization. The Braden scale, which is a tool used to assess the risk of pressure ulcers for patients with physical disability, confirms this evidence indirectly because the mean value of the overall sample and of the two groups was between 15 and 18 indicating that patients were at risk of bedsores.

The other domain presented in table 4.10 was the Abbreviated Mental Test Score. This test was used because it was important to know if the participants were likely to answer about the trial outcomes tests. Mean values were in both the overall sample and in the 2 subgroup, all above 9 indicating that participants did not have pronounced mental dysfunction.

4.5.3.3 Outcome measure results at baseline compared between the two groups

In the following tab 4.11 the results of the assessed items at baseline, compared between the two groups are shown

Domain	Overall sample N= number of respondents Mean= mean value SD= Standard Deviation	Fast Track N= number of respondents Mean= mean value SD= Standard Deviation	Standard Track N= number of respondents Mean= mean value SD= Standard Deviation	Statistical comparison between groups Independent t-test
SEIQoL-DW index (0-100)	N=48 mean=62,837 SD=19,45	N=25 mean=66,172 SD=18,13	N=23 mean=59,2 SD=20,57	no statistical difference, p=.222
PAIN (0-10)	N=50 mean=3,38 SD=3,17	N=25 mean=3,00 SD=3,29	N=25 mean=3,76 SD=3,07	no statistical difference, p=.403
BREATHLESSNESS (0-10)	N=50 mean=3,24 SD=2,71	N=25 mean=2,96 SD=2,74	N=25 mean=3,52 SD=2,71	no statistical difference, p=.471
SLEEP DISTURBANCE (0-10)	N=50 mean=3,34 SD=3,16	N=25 mean=3,4 SD=3,46	N=25 mean=3,28 SD=2,894	no statistical difference, p=.895

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URINARY SYMPTOMS (0-10)	N=50 mean=4,14 SD=3,53	N=25 mean=3,68 SD=3,497	N=25 mean=4,6 SD=3,582	no statistical difference, p=.363
BOWEL SYMPTOMS (0-10)	N=50 mean=4,96 SD=3,46	N=25 mean=4,52 SD=3,97	N=25 mean=5,4 SD=2,887	no statistical difference, p=.375
ORAL SYMPTOMS (0-10)	N=50 mean=4,32 SD=2,93	N=25 mean=3,76 SD=2,758	N=25 mean=4,88 SD=3,059	no statistical difference, p=.180
ANXIETY (HADS-A 0-21)	N=48 mean=6,98 SD=4,34	N=25 mean=6,56 SD=4,0	N=23 mean=7,43 SD=4,65	no statistical difference, p=.492
DEPRESSION (HADS-D 0-21)	N=48 mean=6,75 SD=4,04	N=25 mean=6,32 SD=3,8	N=23 mean=7,22 SD=4,3	no statistical difference, p=.448
FEELING ABANDONED (0-10)	N=49 mean=7,45 SD=2,82	N=25 mean=7,44 SD=3,07	N=25 mean=7,46 SD=2,604	no statistical difference, p=.982
COPING WITH THE DISEASE (0-10)	N=49 mean=5,02 SD=3,16	N=25 mean=5,00 SD=3,512	N=25 mean=5,04 SD=2,836	no statistical difference, p=.964
MEANING (0-10)	N=49 mean=3,76 SD=3,77	N=25 mean=3,16 SD=3,965	N=25 mean=4,38 SD=3,536	no statistical difference, p=.264
HELP FROM FAITH (0-10)	N=49 mean=5,08 SD=3,70	N=25 mean=4,8 SD=3,99	N=24 mean=5,38 SD=3,449	no statistical difference, p=.593
SENSE OF SOCIAL ISOLATION (PATIENT 0-10)	N=48 mean=5,27 SD=3,23	N=25 mean=4,88 SD=3,308	N=23 mean=5,7 SD=3,169	no statistical difference, p=.388
SERVICE SATISFACTION (PATIENT 0-10)	N=48 mean=6,83 SD=2,22	N=25 mean=6,48 SD=2,502	N=23 mean=7,22 SD=1,858	no statistical difference, p=.256
SENSE OF SOCIAL ISOLATION (CARERS 0-10)	N=46 mean=5,76 SD=2,98	N=23 mean=6,17 SD=2,605	N=23 mean=5,35 SD=3,325	no statistical difference, p=.353
SERVICE SATISFACTION (CARERS 0-10)	N=46 mean=6,41 SD=2,41	N=23 mean=6,65 SD=2,479	N=23 mean=6,17 SD=2,387	no statistical difference, p=.509
CAREGIVER BURDEN OF CARE OverallCBI index (0-96)	N=45 mean=44,78 SD=14,953	N=24 mean=46,67 SD=17,574	N=21 mean=42,62 SD=11,294	no statistical difference, p=.371
Tab 4.11: baseline groups comparison				

As shown in the tab 4.11 no statistical differences appeared between the two groups of the present study at baseline.

In the following chart (4.1) the mean differences in the two groups at baseline of the measured outcomes will be displayed graphically.

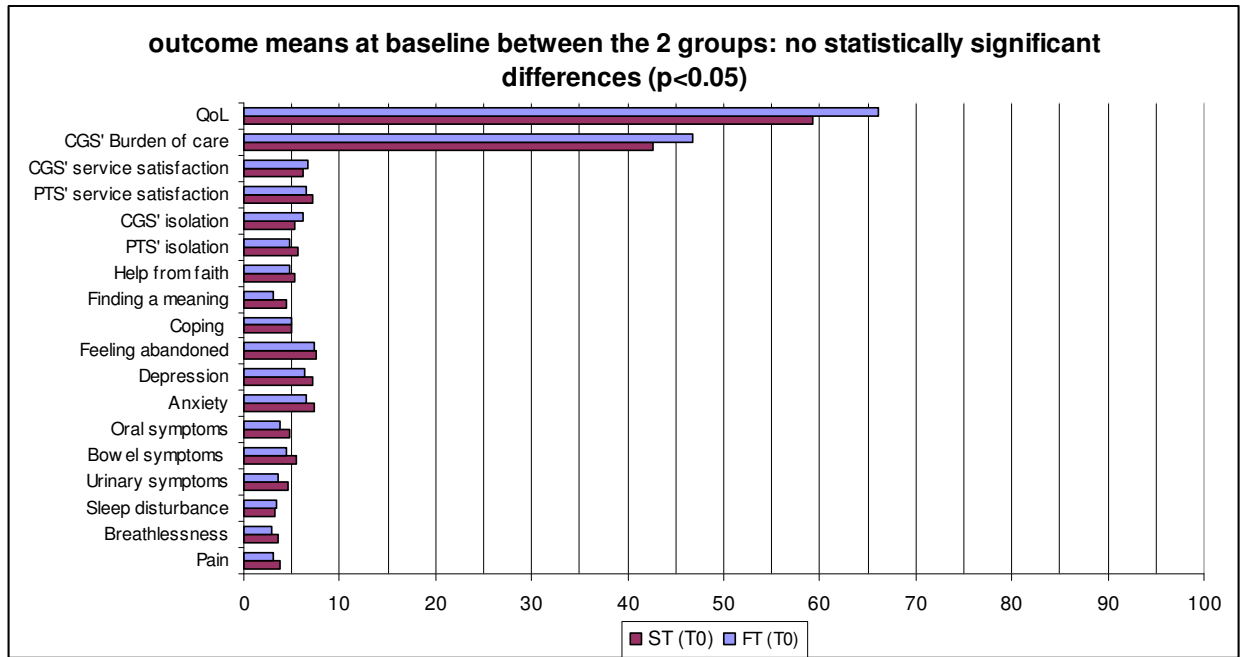


Chart 4.1: the outcome measures mean scores at baseline: comparison between the two groups.

4.5.3.4 Baseline results summary

As result of the recruitment phase of the Ne-Pal explorative randomized controlled study fifty out of the fifty-two referred potential patients were enrolled and randomized in the two groups of the study. Patients were then randomized giving 25 in the two groups. The Fast Track group received the service immediately, the Standard Track group had to wait for 16 weeks before receiving the service. All patients were assessed at baseline, 48 were able to complete the QoL interview, all completed the physical symptoms interview, one or two did not complete the psychosocial, and spiritual tests. The informal carers participated to the baseline assessment, five could not provide data about the burden of care interview because patients did not consent, therefore at baseline only 45 carers answered to the interview.

Main results were a mean of the SEIQoL-DW index of 62.8 %, a prevalence of physical symptoms ranging from the 70% of pain to the 84% of oral discomfort, anxiety and depression 37.5%. No significant differences were found between the two groups for the studied domains. Baseline evaluation will be used as the covariate in the following statistical analysis of covariance. The assumption will be that because no significant differences existed at baseline if differences are found after the intervention it will be justified by the intervention itself.

4.5.4 The results after the intervention (T1)

4.5.4.1 Outcome measures results after the intervention (T1) of the overall sample

In this section the results obtained after 16 weeks are reported. In the next subchapters a general description of what was found in the overall sample and in the two subgroups is shown in terms of mortality, attrition and follow up issues.

Mortality

The sample decreased because four patients died, two in the Fast Track and two in the Standard Track groups. Mortality was 8% over the four months and equal in the two subgroups:

- two ALS/MND who were not tracheostomized
- two with movement disorders (PDs)

No patients with a diagnosis of MS died during the four months of the study.

Attrition

Two patients voluntarily dropped out from the study (4%). Both were in the Fast Track group:

One ALS/MND patient, who was not tracheostomized, and had been keen to participate to the study. He was enrolled and randomized in the intervention group but after three days researchers knew that he was already receiving a palliative home care service provided by the public health system (the only case known of an ALS/MND patient cared for by this service in Turin city at that time). The patient had to decide which service to receive and preferred the public one because he feared that FARO might stop the care provided as this was part of the study. He was therefore discharged from the study. He declared that he did not want to be tracheostomized during the baseline assessment, but the research team knew that after few week he had received a tracheostomy and later was discharged by the public palliative care service because he was not felt that he did not meet the criteria for palliative care.

The second patient who voluntarily left the study was a man with MS who eagerly asked the MS specialists to be involved in the research, participated with enthusiasm at the baseline interview, was randomized and allocated in the intervention group. After few weeks he said to the SPCS professionals that he did not want to see them anymore because he was not a terminally ill patient and was not interested in the service and he was discharged from the study.

Follow up issues

During the study period no major events occurred for the patients who survived. General Practitioners and the district nurses collaborated and families did not have further problems due to their participation at the study.

Two patients were admitted to the local hospitals for acute events:

- A PD patient allocated in the Fast Track group was hospitalized for an acute infection two days after receiving the service . The caring team did not have the

chance to provide the full care because they were only able to see the patient once before the acute event happened when the patient's wife decided to call the ambulance and take him to the hospital where he eventually died after few days.

- A lady with MS, allocated in the Standard Track group, was admitted to hospital with an aspiration pneumonia during the waiting list period. During the hospitalization period a PEG was placed, antibiotics were given and she recovered and was then cared for by the SPCS at home following her discharge from hospital and after the waiting period of 16 weeks had passed.

4.5.4.2 Outcome measures results compared between the two groups after the intervention (T1)

The same battery of tests used at baseline was performed on the participants after 16 weeks. The sample was composed of 44 patients. The 6 missing were the four who died and the two who dropped out - 21 were in the Fast Track group and had already received the service, 23 were in the Standard Track group and were due to receive the SPCS following the T1 examination.

In table 4.12 the results of the overall sample and of the two study groups are presented.

Domain	Overall sample N= number of respondents Mean= mean value SD= Standard Deviation	Fast Track N= number of respondents Mean= mean value SD= Standard Deviation	Standard Track N= number of respondents Mean= mean value SD= Standard Deviation	Statistical comparison between groups Independent t-test
SEIQoL-DW index (0-100)	N=41 mean=66,41 SD=21,10	N=20 mean=78,95 SD=11,76	N=21 mean=54,47 SD=21,25	Significant difference: p=.000*
PAIN (0-10)	N=44 mean=3,86 SD=2,84	N=21 mean=2,19 SD=2,29	N=23 mean=5,39 SD=2,42	Significant difference: p=.000*
BREATHLESSNESS (0-10)	N=44 mean=2,77 SD=2,9	N=21 mean=1,14 SD=1,39	N=23 mean=4,26 SD=3,15	Significant difference: p=.000*
SLEEP DISTURBANCE (0-10)	N=44 mean=2,70 SD=2,44	N=21 mean=1,57 SD=1,93	N=23 mean=3,74 SD=2,43	Significant difference: p=.002*
URINARY SYMPTOMS (0-10)	N=44 mean=2,82 SD= 3,31	N=21 mean=1,57 SD=3,04	N=23 mean=3,96 SD= 3,20	Significant difference: p=.015*
BOWEL SYMPTOMS (0-10)	N=44 mean=3,98 SD=3,61	N=21 mean=2,38 SD=3,32	N=23 mean=5,43 SD=3,28	Significant difference: p=.004*
ORAL SYMPTOMS (0-10)	N=44 mean=4,07 SD=3,46	N=21 mean=2,71 SD=3,16	N=23 mean=5,30 SD=3,31	Significant difference: p=.011*
ANXIETY (HADS-A 0-21)	N=43 mean=7,63 SD=4,1	N=21 mean=7,62 SD=4,18	N=22 mean=7,64 SD=4,12	No significant difference p=.989

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DEPRESSION (HADS-D 0-21)	N=43 mean=7,67 SD=4,3	N=21 mean=7,48 SD=4,27	N=22 mean=7,86 SD=4,48	No significant difference p=.773
FEELING ABANDONED (0-10)	N=43 mean=7,67 SD=2,74	N=20 mean=8,00 SD=2,36	N=23 mean=7,39 SD=3,05	No significant difference p=.474
COPING WITH THE DISEASE (0-10)	N=43 mean=5,60 SD=3,56	N=20 mean=5,10 SD=2,06	N=23 mean=6,04 SD=2,04	No significant difference p=.393
MEANING (0-10)	N=43 mean=3,42 SD=3,86	N=20 mean=3,2 SD=3,63	N=23 mean=3,61 SD=4,12	No significant difference p=.734
HELP FROM FAITH (0-10)	N=43 mean=4,79 SD=3,79	N=20 mean=4,50 SD=3,90	N=23 mean=5,04 SD=3,77	No significant difference : p=.645
SENSE OF SOCIAL ISOLATION (PATIENT 0-10)	N=42 mean=5,36 SD=3,39	N=20 mean=6,25 SD=3,29	N=22 mean=4,55 SD=3,34	No significant difference p=.104
SERVICE SATISFACTION (PATIENT 0-10)	N=42 mean=6,93 SD=2,31	N=20 mean=7,35 SD=2,43	N=22 mean=6,55 SD=2,17	No significant difference p=.265
SENSE OF SOCIAL ISOLATION (CARERS 0-10)	N=43 mean=5,98 SD=3,47	N=21 mean=6,9 SD=3,57	N=22 mean=5,09 SD=3,20	No significant difference p=.087
SERVICE SATISFACTION (CARERS 0-10)	N=43 mean=7,09 SD=2,56	N=21 mean=7,86 SD=2,78	N=22 mean=6,36 SD=2,15	No significant difference p=.055
CAREGIVER BURDEN OF CARE OverallCBI index (0-96)	N=42 mean=41,90 SD=16,38	N=21 mean=41,24 SD=17,83	N=21 mean=42,57 SD=15,22	No significant difference p=.796
Tab 4.12: groups comparison after intervention *Significant difference shown at 0.05 level				

Comparing the two study groups after intervention a significant difference appeared in the QoL and the physical symptoms with an advantage for the Fast Track group. No differences were found for the other domains.

In the following chart 4.2 the mean differences in the two groups after 16 weeks (T1) of the measured outcomes will be displayed graphically.

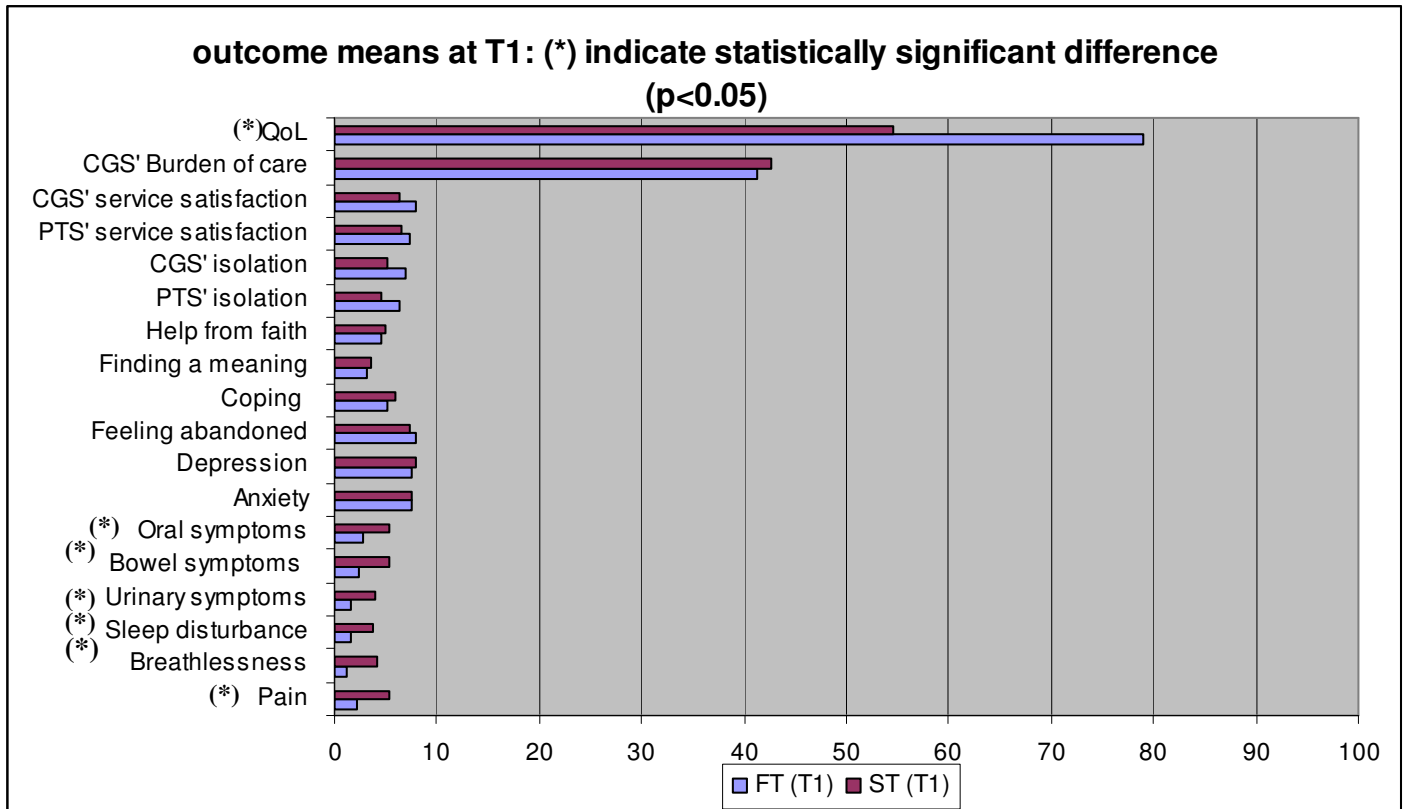


Chart 4.2: the outcome measures mean scores after the intervention (T1): comparison between the two groups.

4.5.4.3 Summary of the results after the intervention (T1)

From the comparison between the groups at T1 it results that:

- no significant differences ($p < 0.05$) were found between the FT group, that had received the SPCS and the ST, that was going to receive it, (but at that moment had just received standard care) for demographics, disease groups, disability, cognitive functions, burden of care for the caregivers, all the psychological, social and spiritual domains included in the evaluated outcomes measures
- patients in the FT group scored significantly higher in the QoL ($p < .001$) compared with the ST.
- a significant difference in all the six assessed physical symptoms was found, always with an advantage for the patients of the FT group versus the ST:
 - pain ($p < .001$), breathlessness ($p < .001$), sleep disturbance ($p = .002$), urinary symptoms ($p = .015$), bowel symptoms ($p = .004$) and oral symptoms ($p = .011$)

4.5.5 Comparison between the groups before and after the intervention

In order to compare the results between the two groups that could be caused by the intervention of the SPCS new variables were created by subtracting the mean results of the test at T0 (baseline) from the ones obtained at T1 (after intervention). These new variables were called “difference variables” and results are shown in Tab 4.13.

Domain	Overall sample N= number of respondents Mean= difference mean T1 – T0 SD= Standard Deviation	Fast Track N= number of respondents Mean= difference mean T1 – T0 SD= Standard Deviation	Standard Track N= number of respondents Mean= difference mean T1 – T0 SD= Standard Deviation	Statistical comparison between FT and ST groups Independent sample t-test
SEIQoL-DW index (0-100)	N=41 mean=3,58 SD=19,01	N=20 mean=12,78 SD=12,82	N=21 mean=-7,41 SD=19,30	$p = .001^*$
PAIN (0-10)	N=44 mean=0,48 SD=3,47	N=21 mean=-0,76 SD=3,20	N=23 mean=1,65 SD=3,36	$p = .019^*$
BREATHLESSNESS (0-10)	N=44 mean=-0,47 SD=3,20	N=21 mean=-1,57 SD=2,92	N=23 mean=0,69 SD=3,12	$p = .017^*$
SLEEP DISTURBANCE (0-10)	N=44 mean=-0,64 SD=3,37	N=21 mean=-1,61 SD=3,23	N=23 mean=0,52 SD=3,23	$p = .034^*$
URINARY SYMPTOMS (0-10)	N=44 mean=-1,32 SD=3,78	N=21 mean=-2,09 SD=2,79	N=23 mean=-0,34 SD=4,40	$p = .128$
BOWEL SYMPTOMS (0-10)	N=44 mean=-0,98 SD=3,81	N=21 mean=-1,90 SD=4,08	N=23 mean=0,17 SD=3,33	$p = .071$

ORAL SYMPTOMS (0-10)	N=44 mean=-0,25 SD=3,24	N=21 mean=-0,80 SD=3,07	N=23 mean=0,43 SD=3,34	p=.207
ANXIETY (HADS-A 0-21)	N=42 mean=0,65 SD=2,97	N=21 mean=0,90 SD=3,49	N=21 mean=0,38 SD=2,41	p=.575
DEPRESSION (HADS-D 0-21)	N=42 mean=0,92 SD=3,44	N=21 mean=1,00 SD=3,14	N=21 mean=0,95 SD=3,80	p=.965
FEELING ABANDONED (0-10)	N=42 mean=0,22 SD=3,81	N=20 mean=0,45 SD=3,69	N=22 mean=-0,22 SD=3,97	p=.572
COPING WITH THE DISEASE (0-10)	N=42 mean=0,58 SD=3,21	N=20 mean=0,3 SD=3,62	N=22 mean=1,22 SD=2,81	p=.358
MEANING (0-10)	N=42 mean=-0,34 SD=3,81	N=20 mean=0,5 SD=3,66	N=22 mean=-0,95 SD=3,90	p=.222
HELP FROM FAITH (0-10)	N=42 mean=-0,29 SD=2,27	N=20 mean=0,0 SD=1,86	N=22 mean=-0,32 SD=2,62	p=.656
SENSE OF SOCIAL ISOLATION (PATIENT 0-10)	N=41 mean=0,09 SD=4,41	N=20 mean=1,35 SD=3,71	N=21 mean=-1,04 SD=4,77	p=.0,82
SERVICE SATISFACTION (PATIENT 0-10)	N=41 mean=0,10 SD=3,09	N=20 mean=0,85 SD=3,78	N=21 mean=-0,52 SD=2,13	p=.158
SENSE OF SOCIAL ISOLATION (CARERS 0-10)	N=41 mean=0,22 SD=4,59	N=20 mean=0,50 SD=4,47	N=21 mean=-0,28 SD=4,77	p=.590
SERVICE SATISFACTION (CARERS 0-10)	N=41 mean=0,68 SD=3,32	N=20 mean=1,25 SD=3,86	N=21 mean=-0,09 SD=2,64	p=.199
CAREGIVER BURDEN OF CARE OverallCBI index (0-96)	N=39 mean=-2,28 SD=9,89	N=20 mean=-4,20 SD=10,01	N=19 mean=0,73 SD=9,37	p=.121
Tab 4.13: groups comparison: differences in the means (after intervention – baseline) for the overall sample and for the two groups.				
*Significant at p<0.05 level				

4.5.5.1 Variations between after and before the intervention in the overall sample

Data shown in the Tab 4.13 represent the new variables obtained from the mathematical difference between the means of each domain at T1 minus the means at T0. These “difference variables” were obtained comparing the means of the analyzed domains before and after the intervention, therefore they provide information about what changed in the four months time between the T0 (baseline) and the T1 assessment. These new data can be interpreted as showing what happened in the sample of participants in the various domains overtime. In this section the variation in the overall

sample is analyzed, in the following sections the two groups of the study will be compared.

Charts 4.3 and 4.4 show respectively the difference between the means in terms of absolute difference (Chart 4.3) and the difference in percentage (Chart 4.4) on the overall sample before and after the intervention. Negative bars reflect the worsening of that domain after the 16 weeks, whereas positive bars indicate that overall that domain improved after the follow up period.

Focusing on the overall sample it results that there have been minimal improvements in QoL (3.58%) and small to moderate improvements in all but one physical symptoms:

- breathlessness (4.7%)
- quality of sleep (6.4%)
- urinary symptoms (13.2%)
- intestinal symptoms (9.8%)
- oral problems (2.5%)

Pain is the only symptom that showed a small worsening in the overall sample (-4.8%).

Looking at the psychological domains it results:

A small worsening in:

- anxiety (-3.1%),
- depression (-4.4%),

A small improvement in:

- feeling abandoned (2.2%)
- coping with the disease (5.8%).

In the spiritual issues the overall sample showed small decrease in the mean results for:

- the meaning of the experience of the disease (3.4%)
- the help received from the faith or religion (2.9%)

About the social explored domains it came out that:

- Patients' sense of isolation improved of 0.9% and patients' satisfaction about the received services improved of 1.0%.
- For the carers the sense of isolation improved of 2.2%, and the service satisfaction improved of 6.8%. The burden of care was reduced of 3%.

Overall only urinary symptoms showed a variation higher than 10% of the scale used to assess the different domains and for this reason can be considered as a clinically moderate change applying the categories declared in the methods section.

We can conclude that no significant changes occurred in the overall sample after the follow up period that coincided with the service provision for the FT group.

Chart 4.3: difference between the means T1-T0 in the overall sample

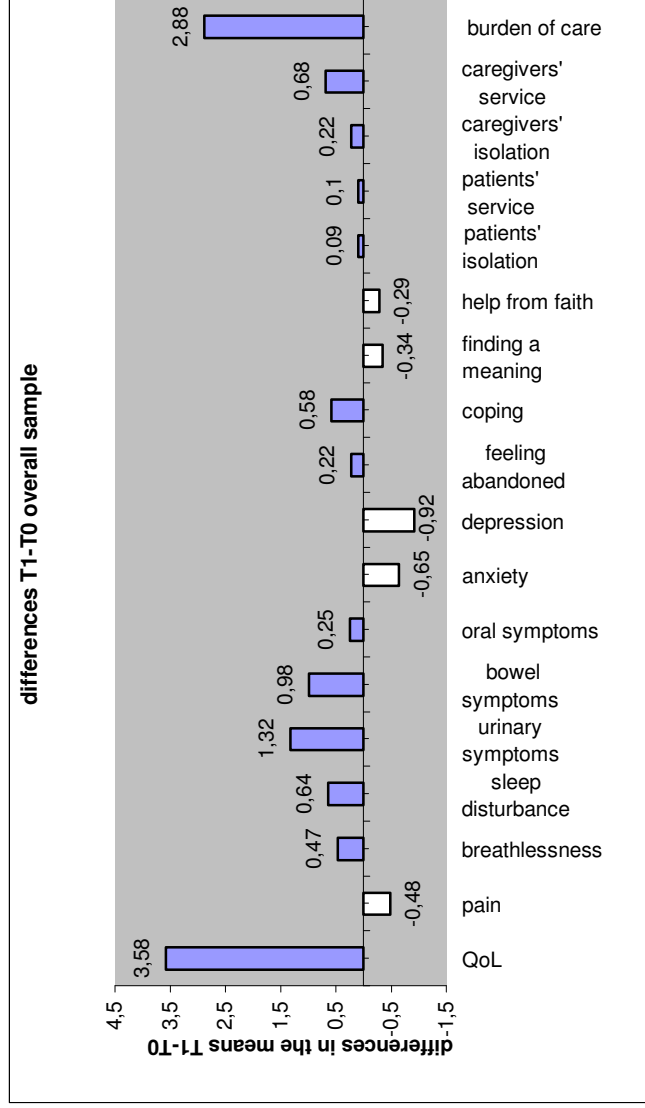
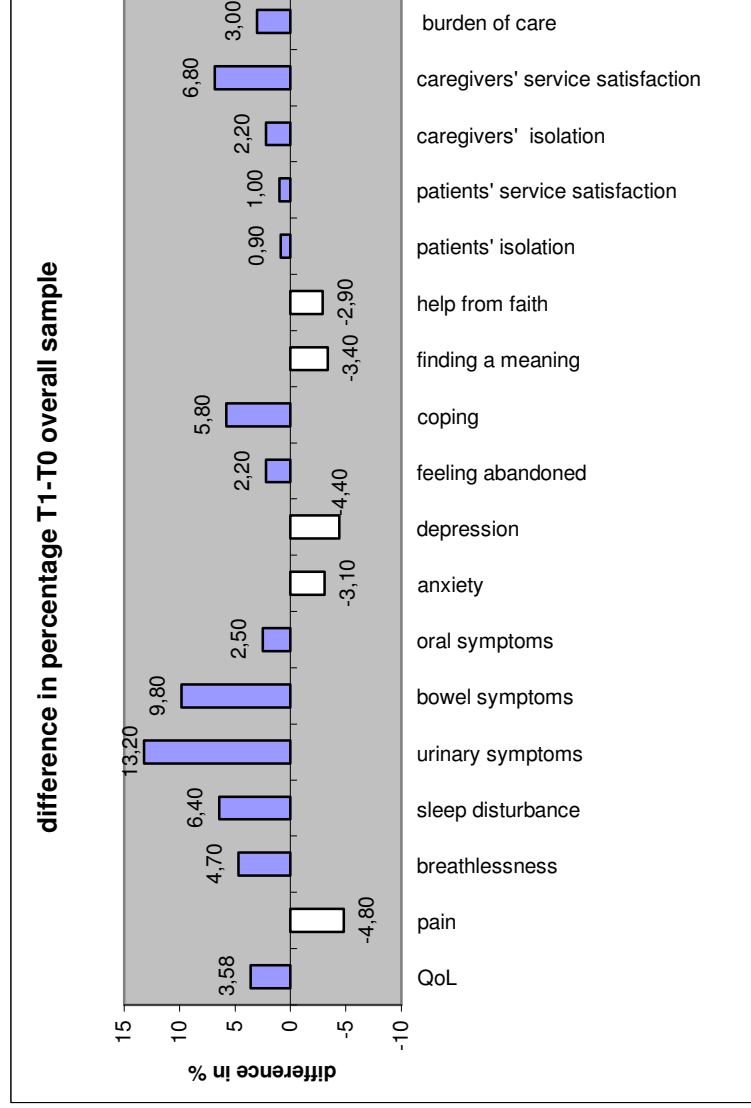


Chart 4.4: % difference between the means T1-T0 in the overall sample



4.5.5.2 Variations between after and before the intervention between the two groups

In Tab 4.13 results obtained by subtracting the means of the various tests at T1 – T0 are shown separately for the two groups of the study (FT and ST). The distance between the means of each domain in the two groups represent what happened in the two groups and can be reported as due to the intervention. In Tab 4.14 the distance in the means of the various domains at T1 – T0 were compared between the study groups and results are displayed in terms of absolute difference, difference in percentage with respect to the scale used to measure that specific item and the statistical significance using an independent t-test to compare the difference of the means.

Domain	Δ in the means T1-T0 FT vs ST (Δ %)	Statistical significance (independent t-test)
SEIQoL-DW index (0-100) Higher scores= better QoL	20,19 (20.2%)	p=.001*
PAIN (0-10) lower score= lower symptom	-2,41 (24.1%)	p=.019*
BREATHLESSNESS (0-10) lower score= lower symptom	-2,26 (22.6%)	p=.017*
SLEEP DISTURBANCE (0-10) lower score= lower symptom	-2,14 (21.4%)	p=.034*
URINARY SYMPTOMS (0-10) lower score= lower symptom	-1,74 (17.4%)	p=.128
BOWEL SYMPTOMS (0-10) lower score= lower symptom	-2,07 (20.7%)	p=.071
ORAL SYMPTOMS (0-10) lower score= lower symptom	-1,24 (12.4%)	p=.207
ANXIETY (HADS-A 0-21) lower score= lower symptom	0,52 (2.5%)	p=.575
DEPRESSION (HADS-D 0-21) lower score= lower symptom	0,04 (0.2%)	p=.965

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FEELING ABANDONED (0-10) Higher scores= lower distress	0,67 (6.7%)	p=.572
COPING WITH THE DISEASE (0-10) Higher scores= better coping	-0,92 (9.2%)	p=.358
MEANING (0-10) Higher scores= higher meaning	1,45 (14.5%)	p=.222
HELP FROM FAITH (0-10) Higher scores= better support	0,31 (3.1%)	p=.656
SENSE OF SOCIAL ISOLATION (PATIENT 0-10) Higher scores= lower sense of isolation	2,39 (23.9%)	p=.082
SERVICE SATISFACTION (PATIENT 0-10) Higher scores= higher satisfaction	1,37 (13.7%)	p=.158
SENSE OF SOCIAL ISOLATION (CARERS 0-10) Higher scores= lower sense of isolation	0,78 (7.8%)	p=.590
SERVICE SATISFACTION (CARERS 0-10) Higher scores= higher satisfaction	1,34 (13.4%)	p=.199
CAREGIVER BURDEN OF CARE CBI index (0-96) Lower score = lower burden of care	-4,93 (5.1%)	p=.121
Tab 4.14: comparison between Δ means T1-T0 between the FT and ST groups *Significant at p=0.05 level		

These results are shown graphically in the chart 4.5. From these data it results that for all the measured outcomes participants in the FT had an advantage (blue bars) if compared with the ST, except for three psychological domains: Anxiety, Depression and Coping with the disease (white bars). The difference for these last three domains was not statistically ($p>0.05$) nor clinically ($\Delta<10\%$) significant.

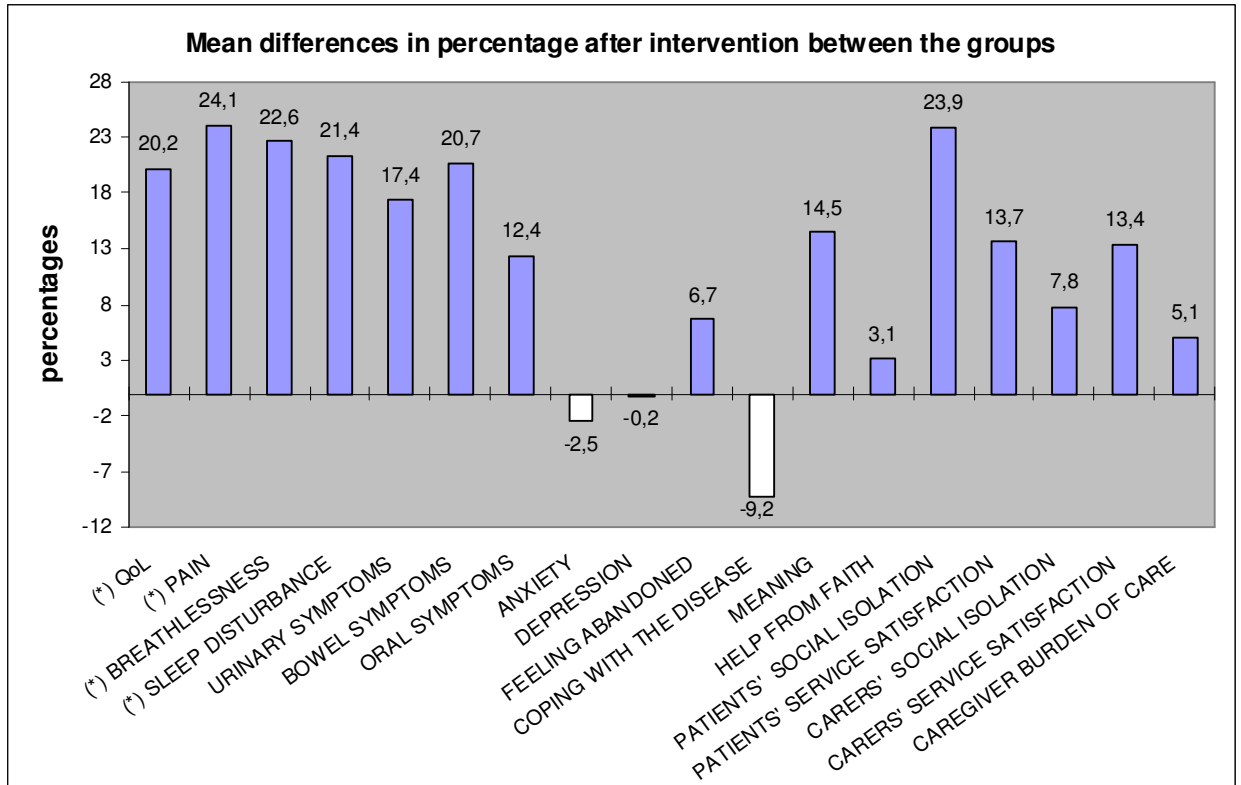


Chart 4.5: Differences in the means T1-T0 (in percentage) between the two groups. Blue bars represent those domains for which the FT showed advantage versus ST, white bars are those domains in which ST showed advantage versus FT. (*) represent the domains for which a statistical significant difference ($p < 0.05$) was found between the groups in the mean comparisons T1-T0.

4.5.5.3 Clinical significance of the comparison between the groups

According to the categories of clinical significance reported in the methods section the results of the Tab 4.14 and chart 4.5 can be grouped in the following categories:

Results could be either favourable to the treated group (FT) if the difference between the means T1-T0 compared to the control group (ST) was better in the former group than in the latter one, or the other way around.

Results could be considered clinically not significant if the difference was lower than 10% of the scale used to measure that specific domain, moderately significant if the difference was between 11 and 19 %, or could be relevantly significant if the difference was 20% or more.

According to this classification the following categories could be found:

1. No clinically significant differences between FT and ST ($\Delta < 10\%$)
2. Moderate clinical advantage in FT versus ST or moderate clinical advantage in ST versus FT ($\Delta 10-19\%$)
3. Relevant clinical advantage in ST versus FT or relevant clinical advantage in ST versus FT ($\Delta \geq 20\%$)

1. No clinically significant differences between FT and ST:

No clinically significant differences ($\Delta < 10\%$) between FT and ST could be detected in the following domains:

- Anxiety
- Depression
- Feeling abandoned
- Coping with the disease
- Help from faith or religion
- Sense of social isolation in carers
- Burden of care for the carers

2. Moderate clinical advantage ($\Delta 10-19\%$) in FT versus ST:

- Moderate clinical advantage was observed in the treated group for the following domains:
 - i. Urinary symptoms and oral symptoms
 - ii. Finding a meaning in the experience of the disease
 - iii. Satisfaction of the service in both patients and carers

3. Moderate clinical advantage ($\Delta 10-19\%$) in ST versus FT

- i. no domains showed moderate improvement in the ST versus the FT

4. Relevant clinical advantage ($\Delta \geq 20\%$) in FT versus ST

Relevant clinical advantage was obtained in the treated group for:

- Quality of life of the patients
- Pain
- Breathlessness
- Quality of sleep
- Intestinal symptoms
- Sense of isolation of the patients

5. Relevant clinical advantage ($\Delta \geq 20\%$) in ST versus FT

In no domains the ST group showed relevant advantages versus the FT one.

4.5.5.4 Statistical significance of the comparison between the groups

4.5.5.4.1 Analysis of Covariance (ANCOVA)

A one-way ANCOVA between-groups analysis was conducted for those variables that showed a statistically significant difference between the two groups after the intervention (T1). The aim of this test was to determine whether adjusting these results for the covariate represented by the baseline assessment (T0) of the same domains the statistical significance was maintained.

The seven variables with these characteristics were: Quality of life, and the physical symptoms (pain control, breathlessness, sleep disorders, urinary symptoms, intestinal symptoms and oral symptoms).

It should be noted that at baseline no statistical significant difference could be found in any of these particular variables.

Quality of life:

One-way ANCOVA between-groups analysis was conducted to compare the effectiveness of the 2 different interventions received by the 2 groups of the NE-PAL study to impact on Quality of Life. The independent variable was the type of intervention: FT (Fast Track= receiving the SPCS), ST (Standard Track= receiving standard care). The dependent variables was the score on the SEIQoL-DW test administered after intervention was completed

Participants' scores on the pre-intervention administration of the SEIQoL-DW test (baseline) were used as the covariate in this analysis. No significant differences were found at baseline in the SEIQoL-DW index between the 2 groups.

After adjusting for pre-intervention scores, there was a significant difference between the two groups on post intervention scores on the QoL ($F_{1,38}=22,57$, $p=.000$, partial eta square = .373).

Symptoms:

One-way ANCOVA between-groups analysis was conducted to compare the effectiveness of the 2 different interventions received by the 2 groups of the NE-PAL study to impact on the following symptoms: Pain, Breathlessness, Sleep disturbances, Urinary problems, Bowel problems and Mouth discomfort. The independent variable was the type of intervention: FT (Fast Track= receiving the SPCS), ST (Standard Track= receiving standard care).

The dependent variables were the scores on the symptoms test administered (NRS 0-10 with high scores meaning high symptom burden) after that the intervention was completed.

Participants' scores on the pre-intervention administration of the symptoms test (baseline) were used as the covariate in this analysis. No significant differences were found at baseline in the symptoms scores (Pain, Breathlessness, Sleep disturbance, Intestinal and Urinary symptoms and Mouth discomfort) between the 2 groups.

Preliminary checks were conducted to ensure that there was no violation of the assumptions of normality, linearity, homogeneity of variance, homogeneity of regression slopes, and reliable measurement of the covariate.

After adjusting for pre-intervention scores, there was a significant difference between the two groups on all considered symptoms, in particular:

Pain control:

post intervention scores on Pain (F1,41)=19,29, p=.000, partial eta square = .320.

Breathlessness:

post intervention scores on Breathlessness (F1,41)=15,78, p=.000, partial eta square = .278.

Sleep disturbances:

post intervention scores on Sleep disturbances (F1,41)=11,47, p=.002, partial eta square = .219.

Urinary symptoms:

post intervention scores on Urinary symptoms (F1,41)=6,08, p=.018, partial eta square = .129.

Bowel symptoms:

post intervention scores on Bowel symptoms (F1,41)=8,26, p=.006, partial eta square = .168.

Mouth discomfort:

post intervention scores on Mouth discomfort (F1,41)=4,38, p=.042, partial eta square = .097.

ANCOVA conclusions

The analysis of covariance shows a significant correlation ($p < 0.05$) between the improvement in QoL and the chance to obtain a better physical symptom control for the patients allocated in the FT group than for those allocated in the ST (control) group.

4.5.5.4.2 Non parametric tests

For the physical symptoms some concern arose about the normal distribution of the results. For this reason a nonparametric test was run on the variable T1 for symptoms to analyze the statistical differences in the two groups.

Mann-Whitney U test for physical symptoms

A Mann-Whitney U performed on symptoms score (pain, breathlessness, sleep disturbances, urinary troubles, bowels troubles and mouth discomfort) at T1 using as independent grouping variable the FT\ST one.

The test (table 4.15) revealed a significant difference in all symptoms in the treated group (FT) compared with the control group (ST)

Symptoms	Study Groups (FT\ST) Medians (md) and groups size (n=)	U= Mann.whitey-U Z= z approximation test p= significance level r= Effect Size
Pain	FT (md=2, n=21) ST (md =5, n=23)	U= 84, z=3,75, p=.000 , r= 0,56
Breathlessness	FT (md=1, n=21) ST (md=4, n=23)	U= 103,5, z=3,32, p=.001 , r=0,50
Sleep disturbances	FT (md=1, n=21) ST (md=4, n=23)	U= 115,5, z=3,01, p=.003 , r=0,45
Urinary symptoms	FT (md=0, n=21) ST (md=5, n=23)	U= 135, z=2,65, p=.008 , r=0,40
Bowels symptoms	FT (md=0, n=21) ST (md=5, n=23)	U= 121,5, z=2,88, p=.004 , r=0,43
Mouth discomfort	FT (md=1, n=21) ST (md=5, n=23)	U= 144,, z=2,32, p=.020 , r=0,35

Table 4.15: Mann-Whittney U at T1 to assess the difference between the two groups for the physical symptoms.

All the symptoms were significantly better for FT ($p < 0.05$ level)

From these tests it appears that in all physical symptoms there have been an improvement in the FT group compared to the control one. In fact, as shown below, there were no statistical differences between the groups at baseline in symptoms scores

Grouping Variable: FT \ ST						
Symptoms baseline assessment	PAIN SCORE T0	BREATH SCORE T0	SLEEP SCORE T0	URIN SCORE T0	BOWEL SCORE T0	MOUTH SCORE T0
Mann-Whitney U	257,000	275,000	309,000	261,500	276,000	247,500
Wilcoxon W	582,000	600,000	634,000	586,500	601,000	572,500
Z	-1,096	-,739	-,069	-1,006	-,715	-1,271
Asymp. Sig. (2-tailed) p =	,273	,460	,945	,315	,475	,204

Table 4.16: SPSS output of Mann-Whittney U test for physical symptoms at baseline (T0). No significant differences between the two groups ($p < 0.05$)

Non parametric test conclusion

The Mann Whitney U non parametric test confirms that in all the six considered physical symptoms there was a statistically significant advantage ($p < 0.05$) for the patients in the treated group compared with the control group after the intervention and that no such differences could be found between the groups at baseline.

4.5.5.4.3 The null hypothesis

The null hypothesis of this study was that no differences could be found in the two groups of the study after the intervention, and therefore that no advantages were showed

in the group that received the SPCS compared to the control group who received the standard care only.

The null hypothesis was accepted for all those domains which did not show any statistical differences after the intervention and that are now listed:

- Caregivers burden of care,
- all the studied psychological domains (anxiety, depression, feeling abandoned, coping with the disease)
- the spiritual domains (finding a meaning in the experience of disease, being helped by faith or religiousness)
- the social aspects (isolation of the carer, service satisfaction for both patients and carers)

For the other domains (QoL, pain, breathlessness, quality of sleep, intestinal symptoms, urinary symptoms and oral symptoms) a statistically significant difference was found between the two groups in all cases with an advantage for the treated groups using an alpha value set at <0.05 .

Being seven the variables that were analyzed using the analysis of covariance (ANCOVA) or the non parametric tests (Mann-Whittney U) a Bonferroni adjustment was used to reduce the probability to fall in a type 1 error, which is to refuse the null hypothesis when in fact it is true. So the alpha value was divided per 7, the number of compared variables, obtaining 0.007. This was the new alpha value to be used to accept or refuse the null hypothesis.

According to this adjustment for the following 5 variables the null hypothesis can be refused:

- Quality of Life: $p=.000$
- Pain: $p=.000$
- Breathlessness: $p=.001$
- Sleep disturbance: $p=.003$
- Intestinal symptoms: $p=.006$

The same hypothesis is to be accepted for the remaining two variables

- Urinary symptoms: $p=.008$
- Oral symptoms: $p=.020$

These results are summarized and reported graphically in chart 4.6

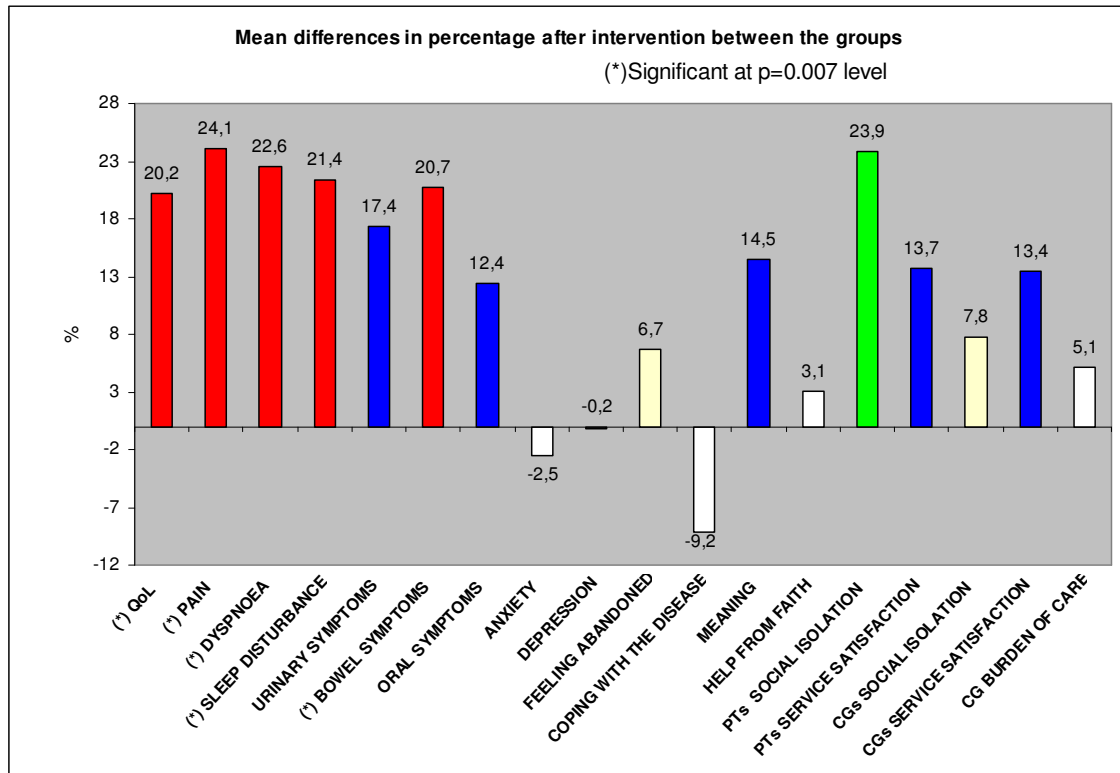


Chart 4.6: comparison between Δ means T1-T0 (in %) between the FT and ST groups.

- Red bars represent domains with both a statistical ($p < 0.007$) and clinically relevant ($\Delta > 20\%$) advantage for FT versus ST
- Green bar represents a clinical relevant advantage of FT vs ST, but no statistically significant difference.
- Blue bars represent clinical moderate advantage ($\Delta 10-19\%$) for FT vs ST, but no statistically significant difference.
- White bars represent domains where no statistical nor clinical significant differences were found between the groups.

(*)Significant at $p=0.007$ level

4.5.5.5 Power of the study

Being an explorative pilot study the sample size was not calculated previously according to the expected number of participants to detect a significant difference between the groups on the main outcomes, but was decided based on the feasibility of the study which was calculated in terms of the affordable increase of the case load for the FARO SPCS. That was defined in 50 patients cared for by the SPCS in one year. When the study was terminated and data analysis conducted, a post hoc calculation of the power of the study was performed based on the main explored outcome, the patients' QoL, measured with the SEIQoL index.

The power of a statistical test is equal to $1 - \beta$, where β is the false negative rate. The power of a statistical test is the probability that the test will reject a false null hypothesis. In general it is defined as large power a study with a power higher than 0,8. According to the mean difference of the SEIQoL index between FT and ST group, the post- hoc power of the study was 0.99 with an Effect size of 1.42 using the Cohen's d, as shown in tab 4.17.

t tests - Means: Difference between two independent means (two groups) SEIQoL OUTCOME		
Analysis:		Post hoc: Compute achieved power
Input:	Tail(s)	Two
	Effect size (Cohen's d)	1.4254497
	α err prob	0.05
	Sample size group 1	20
	Sample size group 2	21
Output:	Noncentrality parameter δ	4.562308
	Critical t	2.022691
	Df	39
	Power (1- β err prob)	0.993566
Tab 4.17: power of the study calculation based upon the QoL index (and estimated sample size at post hoc)		

This result means that the power of this study is very high explaining 98% of the effect for the main outcome (QoL) as due to the intervention, in other words participants who received the service had a significant improvement of the QoL and this allows to reject the null hypothesis.

The same calculation post hoc was performed to calculate the sample size “a posteriori”. The result is that, with that difference observed in the QoL index, a sample of just 14 participants per arm would have been enough to detect the result. In the study 25 patients per arm were enrolled and the analysis was performed on 20 + 21 participants.

The power of the study at post hoc was then calculated on the other statistically significant variables as well as the effect size and the sample size based upon the difference in the means T1-T0 between the two groups. Results are shown in table 4.18.

Symptoms	Effect Size	Estimated patients per arm (estimated sample size)	Power (1- β err prob)
Pain	ES=1.36	16+16=32	0.997
Breathlessness	ES=1.28	17+17=34	0.994
Sleep disturbance	ES=0.99	28+28=56	0.94
Urinary symptoms	ES=0.76	46+46=92	0.797
Intestinal symptoms	ES=0.92	32+32=64	0.912
Oral symptoms	ES=0.80	42+42=84	0.832
Table 4.18: Power of the study calculation based upon the symptoms differences between the groups (and estimated sample size at post hoc)			

These results indicate that for pain, breathlessness, sleep disturbance and intestinal symptoms the power of the study was higher than 90% and the effect size higher than 0,8 that is considered a large effect according to Cohen's classification (Cohen 1988, p.22). for the first two symptoms the post hoc calculation showed that the sample size of this exploratory pilot trial was bigger than what could have been expected “a priori” with the observed mean difference.

4.5.5.6 Summary of the results

To summarize the results of this explorative, pilot, randomized and controlled trial we can say that:

- The study was feasible because no major problems happened during its design, recruitment and follow up.
- Referred patients were keen to participate and only 2/52 (3.8%) refused to enter in the study.
- Mortality was the same in the two groups (8%) during the period of the study.
- Attrition rate was very low (4%). The two patients who dropped out were both in the treated group. All the patients who were randomized in the control group accepted the SPCS after the 16 weeks of wait meaning that they were keen to be cared for by the new service.
- In none of the measured variables the FT group scores were significantly worse (clinically or statistically) than in the ST group.
- In the following domains no clinical nor statistical differences were found between the two groups
 - Anxiety
 - Depression
 - Feeling abandoned
 - Coping with the disease
 - Help from faith or religion
 - Sense of social isolation in carers
 - Caregivers burden of care
- Some domains showed a clinically moderate advantage for the patients in the FT group although no statistical significance was reached:
 - Urinary symptoms
 - Oral symptoms
 - Finding a meaning in the experience of the disease
 - Satisfaction of the received service for the patients
 - Satisfaction of the received service for the carers
- A clinically relevant improvement for the FT was observed for the
 - Sense of social isolation for the patients
- There was a significant clinical and statistical advantage for the patients allocated in the treated group (FT) compared to the ones of the control group (ST) in the following variables:
 - Quality of Life of the patients
 - Pain control
 - Breathlessness
 - Sleep related problems
 - Intestinal symptoms
- The power of the study, calculated at post hoc on the main outcome, patients QoL, was high being $1 - \beta$ of 0,99. the effect size using the Cohen's d was 1,42 and the estimated sample size for this variable was of 14 patients per arm, lower than the analyzed sample size that consisted of 20 + 21 participants. High results at post hoc were confirmed for four physical symptoms: Pain control, Breathlessness, Sleep related problems and Intestinal symptoms, all higher than

80%. These results coincide with the variables listed above for which both statistical and clinical significance difference was found in the FT group versus the ST.

4.6 Nepal discussion

In this chapter of the thesis the discussion of the Ne-Pal explorative pilot randomized and controlled study (RCT) will be presented.

This study represents a phase II RCT of the MRC framework for the design and the evaluation of a new SPCS for patients severely affected by neurodegenerative disorders in Turin city and its metropolitan area (Campbell *et al.* 2000).

According to the author's knowledge this is the first trial aimed at evaluating the impact of a new SPCS on some palliative care outcomes in a sample of patients severely affected by ALS/MND, MS and movement disorders (PD, MSA, PSP). Previously similar research was conducted in the south-east of London by the Supportive and Palliative Care research group of the King's College of London, lead by Professor I. Higginson, for people severely affected by MS (Edmonds *et al.* 2006). The present study tried to follow a similar methodology, but involving participants (and their informal caregivers) with various neurodegenerative conditions all characterized for being incurable, progressive and in the advanced stages.

The study was a randomized, controlled, pilot-explorative trial that, based on the definition provided by Campbell, should "Describe the constant and variable components of a replicable intervention and a feasible protocol for comparing the intervention with an appropriate alternative"(Campbell *et al.* 2000).

The previous phases of the same protocol are described in the chapters of the general methodology and of the qualitative study (Neu-Needs). Having designed the service with the preclinical phase (theory) and having modelled the intervention on the needs of the participants that came out from the qualitative interviews with the potential users and the focus groups with the professionals (phase 1 – modelling), the protocol indicated this phase 2 would be developed as an explorative trial. The explored outcomes were identified among those unmet needs and important concerns that arose from the modelling phase (NeuNeeds study). In particular, for the purpose of this research project, the domains suggested by the participants of the qualitative study that were thought to be addressable by the new SPCS were identified, in particular :

- For the patients:
 - The Quality of life of the patients,
 - Six physical symptoms usually found in palliative care settings: pain, breathlessness, sleep disturbance, urinary symptoms, intestinal symptoms and oral symptoms
 - Four psychological issues: anxiety, depression, feeling abandoned and difficulties in coping with the disease.
 - Two spiritual themes: the meaning of the lived experience of the disease and the help received from faith or religiousness
 - Two social issues: sense of social isolation and the perceived quality of the available services
- For the informal carers:
 - Two social issues: sense of social isolation and the perceived quality of the available services
 - The burden of care

The QoL of the patients and the burden of care for the carers were identified as the main outcomes to be addressed, the others were considered as secondary outcomes. Side outcomes were other aspects such as the feasibility of such a study with very ill patients, the mortality rate, the attrition rate and other potential issues arising during the study.

The sample size was not calculated “a priori” based on the statistical power of the study in view of the explorative nature of the study and a groups comparison without strict rules of sample size was chosen. Sample size was calculated based on the feasibility of the follow up process. This was assumed by considering that FARO foundation SPCS could care for a maximum of 50 patients in one year of time in addition to the routine case load represented by the cancer patients under the care of the service. The framework adopted to design and evaluate the service indicates the setting up of a definitive RCT at the following stage, called phase 3, that has as main outcome to “*Compare a fully defined intervention with an appropriate alternative using a protocol that is theoretically defensible, reproducible, and adequately controlled in a study with appropriate statistical power*” (Campbell *et al.* 2000).

The methodology of the waiting list, also called the Fast Track versus Standard Track, was demonstrated to be feasible and reliable in the previous study conducted by the King’s College group (Higginson, I. J. *et al.* 2006c), and these features were used in the present research project.

4.6.1 Sampling, recruitment, randomization and allocation

The sample size adopted was very similar to the one used in the MS study (52 patients enrolled in that RCT versus 50 in the Ne-Pal RCT).

The enrolment rate was very interesting because out of 52 patients who were referred to the study only 2 denied the consent and therefore 50 were enrolled. This low number of potential participants who refused to participate depends from the fact that they had previously been selected by their professionals (usually neurologists) who had been informed of the inclusion and exclusion criteria of the study. For this reason no “urgent” patients were referred to the trial to avoid the risk that they might have been randomized in the ST group and would have had to wait for care to be provided. This was because being experimental, the service did not have an emergency policy. Thus there was no direct line for patients defined as urgent at the first evaluation leading to a direct care from the SPCS and this is a different approach to that of the King’s College MS study, where they excluded the potential participants if the referring staff and the screening deemed they had very urgent needs or were deteriorating rapidly. In this instance immediate referral to the service was offered, and the patient was withdrawn from the trial (required for 5/69 patients).

In the present study, almost all the referred participants were very keen to participate to the development of an experimental new service potentially useful for them and for other people. For most of them this was the only chance to receive a service tailored on their unmet needs and so they declared to be happy even to wait 4 months, but to have the chance to be cared for by such a service.

The randomization technique adopted with the coupling of two participants per week with similar features (e.g. diagnosis or disability level) allowed contact of potential participants closer to the first assessment and the subsequent randomization and allocation. This procedure was necessary because it was not possible to start the study

with all the sample at the same time. This would have led to a saturation of the service. If participants were to be contacted before the start of the study some would have had a period of several months before being assessed and randomized and this was not acceptable due to the advanced stage of their diseases and the risk of death. It must also be considered that 50% of them would have had a further 16 weeks of wait before receiving the SPCS, for those randomized in the ST group.

The randomization procedure adopted was very simple and did not use specific randomization tools, such as computer based software or random numbers generators. As described in the methods it consisted of the random choice in blind of one of two anonymous and not recognizable folders containing the participants data, done by one of the FARO administrative employees. The first folder chosen was considered in the FT group and the remaining went into the ST one.

Analyzing the results it appears that randomization was successful because the two study groups were homogeneous with an equal distribution of participants in terms of number of participants, gender, age groups, diagnostic groups and disability levels.

4.6.2 Mortality, attrition and missing data

Mortality rate in the 16 weeks of follow up was the same reported for the MS study as 4 patients died during the follow up period: two had been allocated in the FT group and two in the ST, confirming that the input of the service did not increase the mortality rate of the participants. The patients who died during the study were two with ALS/MND and two with PDs.

Attrition was also quite low, just two participants voluntarily left the study before its natural conclusion - both were of the FT group. One patient had to decide whether to choose the FARO experimental service or a public service that was already receiving because the administrators of the latter did not want to collaborate with the study protocol, for this reason he preferred to stay with the public service fearing that the experimental service could be interrupted at the end of the period of follow up. The second patient, despite being severely disabled and totally dependent in the ADL, after 12 weeks of service provision asked to be discharged from the service because he said he was not a terminally ill person and did not want to be cared for by a palliative care unit.

Excluding the four who died and the two who withdrew from the study, 44/50 patients could be assessed at T1, after 16 weeks of service provision for those randomized in the FT or after 16 weeks of wait for those randomized in the ST group. For this reason at T1 21 patients could be reassessed in the FT and 23 in the ST groups.

Not all patients could complete all the tests used to assess the trial outcomes. Some tools, like the SEIQoL-DW required a longer interview and for this reason only 48/50 (96%) patients at baseline could complete this test and only 41/44 (93.2%) at T1. Most patients were so disabled that could not write or keep a pencil in their hands so the researchers did it for them according to their vocal orders or through other way of non verbal communication (for instance the ALS/MND tracheostomized patients answered by electronic communicators or alpha numerical tables). This technique allowed to have 100% of physical symptoms assessment at baseline (50/50) and at T1 (44/44). This means that all patients could correctly utilize the VAS or NRS for the physical symptoms assessment despite being highly disabled. The remaining trial outcomes had lower response rate ranging between 45 – 49 (90-98%) out of 50 eligible at baseline and between 42-43 (95-97%) out of 44 eligible after the 16 weeks representing the duration of the study. The lowest response rate happened at baseline for the caregivers' burden of

care (CBI) (90%) because five patients did not want that their carers (4 paid carers and 1 family carer) to be involved in the research project.

Overall the response rate was satisfactory allowing a good group comparison and few missing data. For these reasons we can conclude that the adopted tools to explore the trial outcomes were acceptable in terms of feasibility and allowed a good compliance in the participants even though they were severely disabled.

4.6.3 Demographics and disability

Analyzing the participants' characteristics it results that there was a moderate prevalence of males (60%) and this can be explained because among the considered diseases it is known that ALS/MND tends to affect more males than females (Bendotti *et al.* 2005) and other studies on the QoL of patients with ALS/MND enrolled 2\3 of males in their sample (Clarke, S. *et al.* 2001), that even though MS affects more frequently women than men, the male gender is a negative prognostic factor and for this reason men are more likely to become severely disabled for MS (Ben-Zacharia and Lublin 2001), and finally some studies report that PDs are more prevalent in males (de Lau and Breteler 2006).

The distribution of the three diagnostic groups (ALS/MND, MS and PDs) resulted homogeneous with respectively 16 patients with ALS/MND (32%), 16 patients with PDs (32%) and 18 with MS (36%). The same proportions were maintained in the two groups of the study after the randomization. This report indicates that even though participants were affected by different neurodegenerative disorders there was a good balance among the diagnostic groups and between the groups of the study.

The results showed that the age groups involved were so distributed: about half of the sample was in the group of 65 years or over, 34% was in the middle age group and the remaining 16% in the group younger than 44. This data is consistent with the epidemiology of the considered diseases, in fact PDs tend to affect the elderly (Wenning *et al.* 2004, Poewe 2006, Litvan *et al.* 1996), in ALS/MND the association with increased age is clear and there is a steep rise in incidence after the age of 50 which continues to increase in age adjusted analysis (Shaw, C. 2006, p 7.), in MS, even though the incidence of the disease is higher in younger people becoming clinically apparent between the age of 20-40 years, high disability level appear in 80% of patients after 30 years from the diagnosis (Goodin *et al.* 2002, Pugliatti *et al.* 2006). The youngest enrolled participants were MS patients whose disease progressed earlier than expected and ALS/MND patients with an early onset of the disease that chose to be tracheostomized. After randomization there was a small increase of participants in the 45-64 group in the FT group versus a higher presence of patients in the under 44 years in the ST. In the group comparison using the Chi-square test for independence however no statistically significant difference appeared between the two groups.

Analyzing the disability degrees of the overall sample at baseline it appears that the sample of patients involved in the present study was affected by a very high disability level. The aim declared in the methods section of enrolling patients severely affected by these neurodegenerative disorders has been achieved. Previous studies or reports about the palliative care involvement for the care of patients with these disorders reported similar or lower disability levels. The prospective RCT analyzing the impact of a SPCS for advanced MS patients reported an EDSS mean level in their sample of 7.7 versus the 8.5 of this study sample (Higginson, I. J. *et al.* 2009). This result shows that in this study the mean disability level was higher and physical functioning of the participants was more impaired.

For the ALS/MND group in a descriptive study about the home care for patients with ALS/MND the mean ALS-FRS score of participants who were receiving the home care was of 13 (Krivickas, Shockley and Mitsumoto 1997), whereas in our study the mean score was 10.2 indicating higher dependency in the ADL of our participants. In other studies aiming at exploring different aspects of the QoL of severely ill patients and the impact on their informal carers, the mean score of ALS-FRS in the considered samples were 22, 22.5, 23.5 and 24.6 respectively (Adelman *et al.* 2004, Clarke, S. *et al.* 2001, Neudert, Wasner and Borasio 2004, Chio *et al.* 2005). Recently in a study evaluating the medical and supportive care among people with ALS/MND who were likely to die in few months for respiratory impairment the reported ALS-FRS-r mean of the patients who died over follow up was 22.3 ± 8.1 and for those who chose tracheostomy 21.3 ± 7.0 (Albert *et al.* 2009). We can conclude that also for ALS/MND patients the mean physical disability level was higher than what is present in the literature in studies set up to evaluate palliative care outcomes such as patients' QoL or caregivers' burden of care.

For the PDs groups it is harder to make comparisons because studies showing the intervention of palliative care services are lacking. Lee's papers discussing the prevalence of pain and the symptom load in idiopathic PD report a mean H&Y stage of 3 among the assessed participants (Lee *et al.* 2007, Lee *et al.* 2006c). Giles published a qualitative study in which 3 patients with a PD diagnosis were interviewed with their family carers and in that case the mean value of the H&Y disability scale was 4 (Giles and Miyasaki 2009). In the Parkinson's disease model of care proposed by Bunting-Perry the advanced stage of the PD disease coincides with 3.5 points of the H&Y scale (Bunting-Perry 2006) and in the paradigm for managing PD Thomas and MacMahon do not report a correlation with the H&Y stages, but identify the criteria to entry into the palliative stage of PD with the inability to tolerate adequate doses of dopaminergic drugs, unsuitability of surgery and advanced co-morbidity leading to a life threatening condition or high disability, conditions that can be considered close to the stage 4 of the H&Y scale (Thomas and MacMahon 2004a, Thomas and MacMahon 2004b). In this study sample the mean H&Y score was of 4.2 - very disabled patients, almost all wheelchair bound or bedridden.

A further confirmation of the high disability of the sample can be detected by the a specific disability scales, the ADL-IADL and the Braden scale analyzing the risk of pressure ulcers due to immobility. ADL mean score was 1.08 out of 6 and IADL 0.98 out of 8 (low values indicate high dependency) and the Braden scale mean value was 15.8 indicating that participants were at risk of developing bed sores.

4.6.4 The trial outcomes

As declared in the methods section the trial outcomes of this pilot-explorative RCT were divided in:

- 2 main outcomes:
 - The patients' quality of life.
 - The burden of care for the caregivers.
- 6 physical symptoms:
 - Pain (considering any painful chronic conditions that could be related or not to the neurodegenerative condition)
 - Breathlessness
 - Sleep disturbance
 - Urinary symptoms

- Intestinal symptoms
- Oral symptoms
- 4 psychological issues:
 - Anxiety
 - Depression
 - Feeling abandoned
 - Coping with the disease
- 2 spiritual domains:
 - Finding a meaning in the experience of the disease
 - Help from religion
- 2 social domains for the patients:
 - Social isolation
 - Satisfaction of the received services
- 2 social domains for the caregivers:
 - Social isolation
 - Satisfaction of the received services

For some of the listed domains it was possible to make comparisons with previously published studies for the various conditions, particularly for the main outcomes, symptoms and some psychological themes . For the others only qualitative studies exist and therefore a numerical comparison was not feasible.

4.6.4.1 The Quality of life

Focusing on the Quality of life of the patients, probably the main outcome of any palliative care intervention, the mean of the SEIQoL-DW at baseline for the overall sample was 62.8 with a Standard Deviation (SD) of 19.45. This data represent the mean of the SEIQoL-DW indexes of 48 out of 50 enrolled patients. Only two participants were not able to complete the SEIQoL-DW interview, even though most of them received help from the researchers to complete the assessment, mostly for difficulties related to the motor impairment which made it difficult for disabled patients to use the disc.. However the information obtained were accurate because it was up to the patients to indicate when the disk position fitted their exact will.

The SEIQoL DW elicited cues represent the areas of individual quality of life chosen by the participants as the most representatives of their personal values. In the sample of this study all participants elicited the “family” as one of the five most important components of their individual QoL. The other more frequently chosen cues were “leisure and entertainment”, including television, sports and personal hobbies (ranging from 62% at baseline and 97% at T1). The health related problems and concerns (about half of the sample at T0 and at T1) were only in third place, even though symptoms were kept separated in this analysis in order to see the comparison between the evolution of the cues and the symptom load and intensity measured with the VAS or NRS that are discussed below. If symptoms are summed to the health domain the percentage rises up to about 80% of participants. The fourth and the fifth most represented cues are occupied at baseline and after the intervention by “friends and relationships” ,“spiritual life” and “ need of independence” all present in about the 50% of the interviews. Less prevalent cues were “financial-economic”, “social themes” and “information-communication” that ranged from 10% to 1/3 of the interviewees. This finding confirms previous researches conducted in ALS patients in Germany (Neudert, Wasner and Borasio 2004) in which “family” was chosen by all patients as determinant of their QoL. Other cues elicited in that study were the friends and social life (54%), “health” (

51%), profession (49%), finances and entertainment (both 22%) and the spiritual life (13%). Further research about the QoL in PD patients living in the UK revealed the following cues elicited by the participants: family (87.8%), health (52.8%), leisure activities (36.6%), marriage (35%), and friends (30.9%)(Lee *et al.* 2006b). Clarke et al demonstrated that in ALS/MND a correlation exist between the type of domains chosen as important determinant of one's own QoL and the physical impairment measured with the ALSFRS scale. In particular when the ALSFRS score is higher than the median, patients tend to elicit cues related to the disease or the health status, whereas when the ALSFRS score is low patients are more likely to chose psychosocial aspects of care (Clarke, S. *et al.* 2001). In our sample the mean disability level is very low and the most frequent cues elicited were in fact the family and entertainment confirming this data.

The SEIQoL index represent the final output of the SEIQoL-DW test. It is a number included in 0-100 range with higher values indicating better QoL. It results from a formula which consider the five elicited areas representing the main aspects of the individual QoL and calculates the sum of the products of the satisfaction of each area (0-100) multiplied for the relative importance of that area (0-1) among the other four elicited.

In the present study the mean SEIQoL-DW index at baseline for the overall sample (N=48) was 62.8 with a SD=19.45. In table 4.19 a comparison with other studies that used this tool in various samples of people in very different conditions is presented

Sample (n)	Mean Index (SD)
ALS/MND (21) (Clarke, S. <i>et al.</i> 2001)	57.8 (22.7)
ALS/MND (21) (Neudert, Wasner and Borasio 2004)	72.3 (not reported)
ALS/MND (80) (Chio <i>et al.</i> 2004a)	73.3 (22.8)
ALS/MND (37) (Lo Coco <i>et al.</i> 2005)	45.56 (not reported)
PD (123) (Lee <i>et al.</i> 2006a)	Not reported
MS (30) (Lintern <i>et al.</i> 2001)	60.99 (16.95)
Palliative Care (62) (Waldron <i>et al.</i> 1999)	60.4 (17.5)
Osteoarthritis (20) (O'Boyle, C. A. <i>et al.</i> 1992)	61.6 (18.8)
Peptic ulcer disease (28) (McGee <i>et al.</i> 1991)	72.6 (10.7)
Healthy adults (42) (McGee <i>et al.</i> 1991)	77.4 (9.5)
Healthy elderly (56) (Browne <i>et al.</i> 1994)	82.1 (12.2)

From this comparison we can conclude that the mean SEIQoL DW index in this study is in line with previously published papers. The SEIQoL-DW of the overall sample (N=41) after 16 weeks (T1) the mean was 66.41 (SD=21,10).

Considering the SEIQoL-DW index in the 2 groups of the study we can see that at baseline the 25 patients who were randomized in the FT group had a mean score of 66.2 (SD= 18.1) and the 23 of the ST who were able to complete the test had a mean score of 59.2 (SD=20,6). This difference was due to chance and there was no statistically significant difference between the 2 groups for this domain (independent samples t-test, p.222). At T1 FT group (N=20) showed an improvement of the QoL measured with the SEIQoL index (mean=78.95; SD=11,76), whereas the control group (N=21) reported a worsening of the same domain (mean=54.47; SD=21,25). The difference between the groups was then statistically significant (p=.000). The analysis of covariance (ANCOVA) using the QoL index at T1 as dependent variable, the belonging to the two

groups as independent variable and the QoL index at baseline as covariate, confirmed this difference between the FT and the ST group (QoL ($F_{1,38}$)=22.57, p =.000, partial eta square = .373).

The difference between the means T0-T1 (baseline – after the intervention) for the SEIQoL- DW index between the FT and the ST groups was of 20,19 points representing an advantage for the treated group of the 20.2% (p =.001). This result with a higher than 20% advantage was considered clinically significant.

No other studies could be found that compared patients with these diagnoses with test-retest methodology using this tool to assess the changes due to an intervention. One study hypothesised that patients with congenital heart diseases experiencing complications leading to a change in health status would not necessarily report a corresponding decrease in the Index. In line with this, the authors found that a deterioration in health status corresponded to an increase in the index score (Moons *et al.* 2004). Another study with patients with metastatic cancer evidenced improvement in the SEIQoL-DW index overtime (Sharpe *et al.* 2005) but both were not comparing the results of the sample with a control group.

The tendency highlighted in this study shows how patients with neurodegenerative disorders in advanced stages tend to improve their individual QoL when are cared for by SPCS, whereas a group that is not receiving the service tend to have a worsening of the same domain. This difference is both statistically and clinically significant. The power of this pilot-explorative RCT, calculated at post-hoc, was very high, being $1-\beta=0.99$. The sample size calculated “a posteriori” using the observed difference of the means for the QoL, resulted of 14 participants per arm. For this reason, even though the study was not aimed at being powerful enough to fulfil the criteria of Phase 3 of the Framework for design and evaluation of complex interventions to improve health (Campbell *et al.* 2000) for the QoL outcome it can be considered a phase 3 definitive randomized controlled study, that in that protocol should “*Compare a fully defined intervention with an appropriate alternative using a protocol that is theoretically defensible, reproducible, and adequately controlled in a study with appropriate statistical power*”.

4.6.4.2 The caregivers burden of care

The caregivers’ burden of care, assessed with the Caregiver Burden Inventory (Novak and Guest 1989) showed a mean of the total index at baseline for the overall sample of eligible caregivers (N=45) of 44.78 (SD=14,9). After the intervention the change in the overall sample was very small: (N=42) mean=41.90 (SD=16,38). Considering what happened in the two groups of the study at baseline the FT carers (N=24) scored a mean of 46.67 (SD=17,5) versus the ST carers (N=21) that scored a mean of 42.62 (SD=11,3). No statistically significant difference between the two groups. After the intervention the FT carers’ mean score (N=21) decreased at 41.24 (SD=17,83) indicating a reduction of the burden of care of 5.1%, whereas the ST carers’(N=21) mean remained stable at 42.57 (SD=15,22).

It was observed a little reduction of the total caregivers’ burden of care in the FT group versus the ST, but this was so small to be considered not clinically nor statistically significant. For this reason the present study did not demonstrate a positive impact of the SPCS on the burden of care for the informal carers involved in the assistance of patients severely affected by neurodegenerative conditions. However the disability of

the patients in both groups was high and may have caused profound caring needs, for which there may be little relief.

4.6.4.3 The physical symptoms

The physical symptoms analyzed in this study were pain, breathlessness, sleep disturbance, urinary, intestinal and oral symptoms. These domains were chosen among the physical problems reported by the sample of patients who participated to the qualitative study and represent typical symptoms faced by traditional SPCS in cancer and non cancer care (Addington-Hall, J., Fakhoury and McCarthy 1998).

Symptoms were measured with simple VAS (0-10 cm) or NRS (0-10 points) tools and both the prevalence (presence of the symptom) and the intensity were reported.

For the overall sample at baseline it was possible to obtain the assessment of the symptoms in the whole sample (N=50) without missing data. The results are shown in the results chapter at the table 4.5. The most prevalent problem were the “oral symptoms” (84%), followed by the intestinal symptoms (80%), breathlessness (72%), pain and urinary symptoms (70%) and sleep disturbance (68%). In the literature review of the symptom prevalence among people affected by advanced and progressive neurological conditions (Saleem, Leigh and Higginson 2007) these problems were among those reported for the considered diagnostic groups (ALS/MND, MS, PD, MSA, PSP). In particular they were included among the 26 common symptoms present in these diseases. Bowel, urinary symptoms and swallowing (which may be seen as an oral symptom) were present in all 5 groups with a prevalence 50% or over. Pain was reported in all but the PSP group, but if spasms and stiffness were added, being painful conditions (Simmons 2005), they would have been present in all the groups with a prevalence higher than 50%. Breathlessness and sleep disturbance were less represented in that review even though they appeared in more than 80% and more than 60% respectively in the NeuNeeds qualitative analysis presented in this thesis.

Considering the symptom intensity at baseline, the highest score was reported for the “bowel symptoms” (mean= 4.96, SD=3.46), the lowest for “breathlessness” (mean=3.24 SD=2.71).

After the randomization and the allocation in the 2 groups of the study no statistically significant difference between the means of the symptoms in the FT and ST group at baseline was found as shown in the table 4.11 of the results section. There was a small advantage in the mean intensity score in the FT group for pain, breathlessness, urinary, bowel and oral symptoms, whereas sleep disturbance was lower in the ST group. These difference were not statistically nor clinically significant.

Conversely at T1 a statistically significant advantage was seen in the FT group for all the symptoms (see table 4.12 in the results chapter). This advantage was maintained after adjusting with the baseline results as shown with the ANCOVA test keeping the alpha value at <0.05. This analysis of covariance allows to compare the means of the symptoms between the groups at T1 adjusting the results for the means of the symptoms at baseline. With this procedure the differences that were present at baseline between the groups, even though not statistically significant, are considered as influencing the final comparison acting as covariates.

Furthermore, because this comparison of means involved 7 variables (the physical symptoms) plus the QoL (the other statistically and clinically significant variable) an adjustment of the alpha value (that had been set at 0.05) was considered to avoid the

risk of a type 1 error (which is to refuse the null hypothesis when in fact it is true). When a Bonferroni adjustment is applied, considering the six symptoms and the QoL as the domains where a statistical significance was seen between the groups after the intervention (in total 7 variables for which a statistical significant improvement were seen in the FT group compared to the ST with a α set at 0.05), keeping the alpha value at $0.05/7 = 0.007$, the statistical advantage is maintained for pain, breathlessness, sleep disturbance and intestinal symptoms (and the QoL described above).

In conclusion we can say that this study showed an improvement in the symptom severity (pain, breathlessness, sleep disturbance and intestinal symptoms) after a 16 weeks SPCS provision for the treated group versus the standard care group.

Clinically, if we consider as a moderate clinical significance an improvement of +10% of the scale used to measure the symptom and as a clinically relevant significance an improvement of +20% of the same scale we can say that this study demonstrated that the SPCS input caused a moderate clinically significant improvement in the treated group for oral and urinary symptoms and a clinically relevant improvement in pain, breathlessness, sleep disturbance and intestinal symptoms. The power of the study, calculated at post hoc was higher than 90% ($1-\beta > 0.90$) for pain, breathlessness and intestinal symptoms. The post hoc calculation of the sample size, using the observed difference in the means between the groups, allowed to say that for pain and breathlessness the sample was bigger than what it would have been expected to be conclusive.

4.6.4.4 The psychological issues

The assessed psychological issues for this study were anxiety, depression, feeling abandoned and coping with the disease.

Anxiety and depression are part of the disorders of the mood category that were described in the qualitative assessment (NeuNeeds). From the content analysis of that study results that in all the interviews to the patients and the caregivers some sentence related to mood disorders appeared. These symptoms are typically measured in clinical research settings and the tool used in this study, the Hospital Anxiety and Depression Scale (HADS) is a very well experimented instrument used in patients with problems similar to this study sample (Marinus *et al.* 2002, Clarke, S. *et al.* 2001, Honarmand and Feinstein 2009).

In this study sample the prevalence of probable or definitive anxiety and depression was of 37.5% for both symptoms. This data were calculating by subtracting from the total results those participants that scored normal values defined as 7 or less in the HADS-A or HADS-D.

Of the total scores obtained from the overall sample at baseline (N=48) thirty patients scored 7 or less for both anxiety and depression (62.5%) which correspond at the normal category. Of the remaining patients nine (18.75%) scored between 8 and 10 for anxiety (borderline category) and the remaining nine (18.75%) scored 11 or more being in the caseness category. For depression, of the eighteen patients who scored more than 7, eight (16.7%) were in the borderline category and 10 (20.8%) in the caseness category.

Anxiety has been reported as affecting 30% of ALS/MND patients in their last month of life whereas depression was at 40% (Ganzini, Johnston and Silveira 2002). In another study the prevalence of depression at baseline for ALS/MND patients was of 19% including minor and major depression (Rabkin *et al.* 2005). In a study for the validation of the HADS for PD patients percentages for possible and probable depression were

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21.5% and 16.9% and 50% showed signs of anxiety (Marinus *et al.* 2002). In a survey with 495 patients severely affected by ALS/MND, MS, PD and Huntington's disease using the HADS to assess these symptoms an overall of 26% on anxiety and 22% of depression was found (Kristjanson, Aoun and Oldham 2006). These results are similar to the ones obtained in the present study.

The mean scores of the HADS-A and HADS-D in the present study are displayed in table 4.20 and are compared with the findings of the same domains obtained in other studies involving patients with advanced neurodegenerative conditions.

Samples (N=)	HADS-A mean (SD)	HADS-D mean (SD)	references
NePAL RCT (48)	6.98 (4.34)	6.75 (4.04)	
ALS/MND, MS, PD and Huntington's disease (495)	7.6 (4.64)	7.00 (4.35)	(Kristjanson, Aoun and Oldham 2006)
ALS/MND (26)	5.8 (3.63)	4.76 (2.8)	(Clarke, S. <i>et al.</i> 2001)
PD (176)	8.74 (4.09)	10.25 (4.38)	(Marinus <i>et al.</i> 2002)
MS (180)	6.69 (4.44)	5.26 (4.05)	(Honarmand and Feinstein 2009)

Table 4.20: mean HADS scores reported in other published studies

The mean values for anxiety and depression in the overall sample at baseline in this study were both in the "normal" category indicating that on average the levels of these symptoms were not significant.

After the randomization in the two groups of the study the mean scores for both anxiety and depression did not show statistically significant differences between the groups (see table 4.11 in the results section). This condition remained after the intervention when the mean score for anxiety in the overall sample raised at 7,63 (SD=4,1) and for depression to 7.67 (SD=4.3), but no difference were found between the groups.

In conclusion it is possible to say that the anxiety and depression assessment in this study, measured with the HADS reported prevalence and intensity of both symptoms similar to other findings shown in studies involving patients with the same diseases. No differences were seen between the groups of this study therefore was no impact on these domains from the SPCS.

Feeling abandoned and coping with the disease are two psychological themes that emerged from the qualitative part of the study.

As reported in results section of the qualitative NeuNeeds assessment 95% of the interviewed patients talked about the personal unpleasant sensation of "feeling abandoned", confused and of fear the future because of their neurological condition.

In the RCT in exam this domain was measured with VAS (0-10cm) or NRS (0-10 points). Lower scores indicated total abandonment whereas higher ones meant absence of this problem. At baseline 49/50 patients completed this item and the mean reported score was 7.45 (SD=2.82). After the intervention (T1) the mean score in the overall

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sample (N=43/44) was 7.67 (SD=2.74) so we can conclude that no difference appeared overtime for the overall sample.

Considering the 2 groups of the study at baseline the results were almost identical:

FT (N=25) mean=7.44 (SD=3.07)

ST (N=25) mean=7.46 (SD=2.6)

After the intervention no significant differences resulted for this test

FT (N=20) mean=8.00 (SD=2.36)

ST (N=23) mean=7.39 (SD=3.05)

The difference between the means in the two groups (6.7%) is lower than 10% of the scale used to assess this domain (no clinical significance) and there is no statistically significant difference between the groups. This difference is positive for the group who has received the service and can reflect the tendency of an improvement of this domain in those who received the service, but the difference is too low to justify such claim.

Similar consideration can be done for the “Coping with the disease” domain. Difficulties related to this issue appeared in 77% of the interviews conducted in the qualitative assessment and confirm what has widely reported in the literature about the difficulty to accept the continuous losses due to the progression of the neurological conditions s described for MS (Edmonds *et al.* 2007b, Wollin, Yates and Kristjanson 2006), the importance of the mastery (Koplas *et al.* 1999) and the prevalence of the mental and psychosocial losses over the difficulties in the ADL’s in determining the QoL in PD patients (Abudi *et al.* 1997). The coping strategies and the relative impact on the QoL of ALS/MND patients (Hughes *et al.* 2005) as well as the shift of personal values observed from conservative values towards self-transcendence values may be related to coping processes of terminally ill patients (Fegg *et al.* 2005).

In the present study at baseline 49/50 patients completed the VAS (0-10cm) or NRS (0-10 points) used to assess this item. The higher score indicated that patients were coping perfectly with the disease and its consequences, lower scores meant difficulties in coping with the problems caused by their conditions.

The mean score for the overall sample at baseline was of 5.02 (SD=3,16). No difference between the groups was found at baseline. After the follow up period the overall mean of the sample (N=43) was 5.60 (SD=3,56) and the results in the two groups indicated that patients who had received the service scored worse than the control group for this domain. The difference was of 0.92 points of the scales corresponding at 9.2% of difference . This difference is not statistically significant (p=.393) and is borderline for the clinical significance that was set at 10%.

The assessment of the psychological domains in this study aimed to evaluate if a SPCS, that includes professionals specifically dedicated to the support care and the accompaniment of severely ill patients, could be helpful. The results indicate that no difference was found between the groups for the psychological domains, even though for coping with the disease it seemed there was a small advantage for patients who were not receiving the SPCS. One possible interpretation of this results (that might be due to chance because its not significant statistically nor clinically) is that patients affected by neurodegenerative disorders, although in far advanced stages, do not see themselves as terminally ill, but usually describe themselves as chronically ill. This vision of their condition can be negatively impacted by the care provided by a SPCS whose professionals are used to deal with dying persons and that tend to talk about death and

dying. This option can worsen the psychological status of patients who are experiencing anxiety and depression symptoms and difficulties in coping with the losses. It is harder to explain why patients did not have improvement in the “abandonment” item when it was clear that they were in charge of a specific service modelled to meet their needs.

One explanation attempted by the researchers was that the negative sensation of feeling abandoned is a profound loss deep-rooted in patients who have a chronic illness and a 16 weeks provision of a service, though with a positive impact on the individual QoL and on symptom control, represented a too short period of time to reverse this feeling. Analyzing the qualitative description of the abandonment it results that participants reported to have been abandoned by their relatives, friends and professional carers. The input of a new service could improve the feeling of being abandoned by the professionals (sensation worsened by the feeling of being abandoned because they are too ill and/or are felt to be incurable, whereas a SPCS should give the message that they will not be abandoned exactly because they are very ill and are incurable). The same SPCS may not necessarily improve the sensation of abandonment from the other family members and friends.

The impact on this domain should probably be assessed in a longer period. Overall the impact on the psychological domains was lower than what could be expected “a priori” and represent a critical point that should be rethought and considered in further research.

4.6.4.5 The spiritual issues

Spirituality is an important component of the total pain, as described by Dame Cicely Saunders (Clark 1999), and is also a determinant part of the QoL of people severely affected by neurodegenerative conditions (Murphy *et al.* 2000, Rabkin, Wagner and Del Bene 2000, Taylor and Herr 2004, Teno 1999).

The two domains that were assessed in this study came out from the qualitative interviews and were the “meaning” of the lived experience represented by the disease and the help received by faith or religiousness.

For the meaning issue what was observed in the qualitative part of the study was that participants seemed divided between those who found a partial or a complete meaning in what was happening to them and those who did not. The fact of having found a meaning could be related to a transcendent signification like a religious experience or because suffering is a normal part of life, or to a social meaning like the sense of justice that can be seen as having received many gifts in life whereas others did not and so falling ill can be a compensation for this.

One of the tasks of a palliative care intervention is to help dying people to obtain a peaceful death and to achieve this result it is very important to help the patients to find some answers to what is happening to them.

The other spiritual issues measured in this trial were the help from faith or religiousness. Even though the FARO SPCS is not a religious charity a number of professionals have religious backgrounds. Three nurses are nuns, one doctor and one nurse have undertaken a masters training in religious support and theology. In the SPCS team there is a Catholic priest that works as spiritual assistant. In the qualitative assessment appeared that faith can be both a positive resource enabling patients to face the losses caused by the neurological disorder and to accept deeply what is happening to them, but can also be an obstacle to the search of the peace when patients live their faith seeking miracles that often do not happen. This can lead to frustration, refusal of their beliefs

and eventually rage. For this reason it was decided to measure if the SPCS, having professionals skilled in spiritual support, could make a difference.

The results, presented in the results session, showed that no statistically significant differences were found between the groups in these two items. This was particularly true for the “help from faith” item, whereas for the “meaning of the lived experience” it was observed an improvement of the 14.5% of the scale used for the assessment in the group who received the service versus the control one. This was reported as a moderate clinical improvement, although the lack of statistical significance could mean that the observed improvement could be due to chance.

4.6.4.6 The social issues

Social problems faced by people severely affected by neurodegenerative conditions affect both patients and their carers (Oliver and Gallagher 1998). The task of caring for terminally ill patients causes social difficulties to the carers independent of the diagnosis (Aoun and Kristjanson 2004). Patients may feel that they are a burden to their carers and this may be related to their depressive status (Chio *et al.* 2005). There are differences among different diagnostic groups in the social needs and in the keenness to accept services focused on palliative care (Kristjanson, Aoun and Oldham 2006, Goy, Carter and Ganzini 2008).

In this study, as part for the specific outcome represented by the caregivers’ burden described above, domains also considered included the sense of social isolation, as this had been commonly described by both the patients and their carers in the qualitative study. The degree of satisfaction or not satisfaction of these services, of both the patients and their carers, was assessed using VAS (0-10cm) or NRS (0-10 points). For the social isolation domain “0” represented feeling totally isolated and “10” do not feeling isolated at all. For the service satisfaction “0” represented absolute absence of satisfaction, whereas “10” a total satisfaction of the received services. On consideration of the social isolation item it appears that at baseline the mean scores for the overall sample were very similar between patients 5.27 (SD=3.23) and carers 5.76 (SD=2.98). After the follow up the scores, for the overall sample, remained quiet similar: 5.36 (SD=3.39) for the patients and 5.98 (SD=3.47) for the carers. Considering the difference between the two groups of the study it results that both the patients and the carers showed an improvement of this domain in the FT group which were not statistically significant, but was clinically relevantly significant for the patients (23.9% of the scale). The carers’ sense of social isolation improved by about 7.8% therefore was considered as not clinically significant.

The trend for social isolation, though not conclusive, appeared to be positive for the participants who received the service compared to the control group. Analyzing the satisfaction degree of the received services, assessed with VAS (0-10cm) or NRS (0-10 points) it seems that both patients and carers showed a moderate improvement of this domain in the group of treatment respect the control one of about 13%. These results are not conclusive because there is not a statistical significance, but they indicate a positive trend in this social issue.

4.6.5 Limitations of the study

There are several limitations in the present study.

- The sample was composed of patients severely affected by neurodegenerative disorders which have different illness trajectories, prognosis and characteristics, even though all are progressive and no cure is available. This was the first published experience of a trial exploring the impact of a SPCS on such heterogeneous group, even though it has been recognized that these patient groups share common palliative care problems (Saleem, Leigh and Higginson 2007).
- Recruitment: patients were referred to the service by specialist neurological services (tertiary clinics) therefore a selection bias could be present due to the exclusion of patients cared for by general neurological departments or too ill to attend the hospital services. A second limitation was due to the design of the study that did not allow to enrol patients with severe cognitive impairment because they had to be able to complete the outcome measures. For this reason patients with severe dementia or psychiatric problems were excluded even though they represent the most disabled and advanced stages of the considered neurodegenerative disorders.
- The explorative nature of the phase 2 study would only allow to elicit a sample size that could be feasible for the new SPCS provided by FARO. For many items the results were not conclusive and therefore the null hypothesis had to be accepted probably because the sample size was not big enough. This does not seem true for the patients' QoL and for pain and breathlessness where the observed difference in the means between the groups show that the sample size was large enough to demonstrate the positive effect of the service provision.
- Only one assessment was done after the intervention. This did not allow a comparison between the groups over time nor allowed a crossover between the groups to evaluate if the same positive effects seen in the treatment group appeared in the control group after a similar period of time of the received service. This is one of the main limitations and is one major area of difference in methodology compared to the other published phase 2 RCT for the MS patients (Higginson, I. J. *et al.* 2009).
- The study was conducted in an area of the North West of Italy and all the social issues can be influenced by the local conditions bound to the regional health service. We can assume that the patients QoL and symptoms were not different from patients with the same pathological conditions living in different places, but the results are to be confirmed by wider and multicentre studies.

4.6.6 Summary

This phase 2 RCT was aimed at comparing two groups of patients severely affected by neurodegenerative conditions and their informal carers: one group received a 16 weeks provision of a new SPCS while the second was receiving the best standard care. From the groups comparison resulted that the SPCS group had significant improvement in QoL and symptom burden versus the control group. There was positive trend (although not significant) for the SPCS group in “social isolation” and “finding a meaning in the experience of the disease” for the patients and in the “service satisfaction” for both

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patients and carers. No improvement could be detected in the SPCS for the psychological domains, on the contrary it seems that “coping with the disease” could be worsened by the SPCS provision.

Overall from this study it appears that the studied population can have a favourable and significant improvement if cared for by a SPCS, above all for the QoL and some symptoms, for which the explorative trial showed a very high statistical power and can be considered conclusive.

Psychosocial and spiritual issues were not conclusive and further research should be undertaken to explore these domains.

Other observations were that the methodology of the waiting list procedure is feasible and reliable and can be used to assess palliative outcomes in palliative care settings for patients with an expected life span of months/years. Low attrition and relatively low missing data confirmed a good impact of the study on the participants that keenly adhered at the proposal of this study and remained in the protocol for the follow up with a very low attrition rate.

Mortality was low, confirming the prognostic difficulties in neurodegenerative disorders, despite the high disability of the enrolled sample. This data could mean that disability alone is not a precise indicator of end of life stages in neurodegenerative conditions representing a confirmation of the difference existing in cancer palliative care.

5. Conclusions and Recommendations

5.1 Conclusions

In this section of the thesis the conclusions of the research project are presented. This chapter consists of a resume of all the previously presented components of the project, a description of the overall effect on the SPCS for neurological patients and some comments of the overall results.

5.1.1 Aim of the study

As stated in the “Idea for the research” chapter this research project was based on a research question composed by two parts:

1. What are the palliative care needs of people severely affected by neurodegenerative conditions?
2. How can a Specialist Palliative Care Service (SPCS) best meet their unmet needs?

The objective of the project was to answer to the research question.

A new SPCS for patients severely affected by ALS/MND, MS and PDs in Turin city and its province was designed, set up, assessed and evaluated. The results of this process were used to answer to the research question.

5.1.2 Methods

The design and the evaluation of the new SPCS for patients severely affected by neurodegenerative disorders was undertaken following the first three phases of the MRC framework for design and evaluation of complex interventions to improve health (Campbell *et al.* 2000).

A literature review aimed at obtaining information about the state of the art of palliative care in neurology was performed. It focused on the characteristics of the considered disorders, specific palliative care needs in this population, previous experiences of palliative care provision or SPCS inputs. Published guidelines, recommendations or official positions from scientific societies, government agencies and research groups were also considered. This step is part of the “theory-preclinical” stage of the MRC framework.

A qualitative study aimed at exploring the palliative care unmet needs of patients severely affected by ALS/MND, MS and PDs and their family carers, living in Turin’s area and cared for by neurologic services operating in two min hospital of the Turin’s province was undertaken. The methodology of this study was based on in-depth interviews with the patients and their carers and focus groups with the professionals involved in their care. This needs assessment study (called Neu-Needs) is part of the “Modelling-phase 1” stage of the MRC framework.

A quantitative study aimed at evaluating the impact of the new SPCS on the PCO of people severely affected by neurodegenerative conditions concluded the research project. It was a pilot-explorative RCT using the waiting list procedure. The latter

allows a comparison of the effects of the intervention on a treated group compared with a control group receiving standard best practise care. After the follow up period (16 weeks) all the enrolled participants can receive the service if desired.

The tools used to assess the trial outcomes were in part validated tests already tested in this population (SEIQoL-DW, HADS, CBI), and other general tools, VAS or NRS, specifically created to assess the impact of the SPCS on the unmet needs reported by the participants of the qualitative needs assessment.

The analysis of the outcomes was conducted with statistical tests to compare the groups (independent samples t-test, non-parametric tests and ANCOVA) and with a clinical judgement based on the clinical significance of the results.

5.1.3 Results

5.1.3.1 The literature

5.1.3.1.1 Needs and problems

The concepts of palliative care unmet needs and the description of the hierarchy of needs by Maslow were the basis of the literature search that looked at the needs and at the problems faced by both the patients severely affected by neurological conditions and their carers. The classic framework of the palliative care needs of the dying, that divides these needs into physical, psychological, social and spiritual, offers a clear definition, but does not provide an straight forward distinction between needs and problems.

Needs and problems are by no means synonymous:

Problems are defined in several dictionaries as - *Perceived gap between the existing state and a desired state, or a deviation from a norm, standard, or status quo. Although most problems turn out to have several solutions (the means to close the gap or correct the deviation), difficulties arise where such means are either not obvious or are not immediately available* -.

Needs are better described as - *A lack of something requisite, desirable, or useful; a physiological or psychological requirement for the well-being of an organism; a condition requiring supply or relief* -.

This difference can be transposed in the present thesis by analysing the number of problems and needs described in the published literature and compared with the findings of the qualitative and the quantitative studies of this research project.

Problems can be seen as the physical impairment caused by the diseases, the loss of independence, the reduction of the life span, the changes in the roles inside the families and in the social environment. These situations are objective and can be detected easily by comparing what the expected functions of healthy people of the same age as those involved in the research. This can clearly show how the gap between the physical and social limitations of the impaired patients and the healthy control group and how this

gap is due to the progressive and relentlessly evolution of the disease, and therefore the “problems”.

Needs are far more subjective. Participants could experience problems but not report them as unmet needs. For instance those participants who accept their disability (which is an objective problem) seem to cope better with the disease and do not see their incapability to be unmet need. On the other hand other situations that are not objective problems, such as the fear of choking to death when there is plenty of evidence that this condition do not happen at the end of life of neurodegenerative conditions and can be easily tackled by simple and well established plans of care, represent a clear unmet need of personal safety and can lead to a rise in the anxiety and in some case to undesired decisions like the wish for a hastened death.

5.1.3.1.2 Palliative care and neurodegenerative disorders

The literature review evidenced an increasing interest in the involvement on palliative care in neurological disorders in recent years.

ALS/MND despite being the rarest condition among the considered, is the neurological disorders with the highest number of papers describing the palliative care involvement. Patients affected by this disease are commonly cared for by SPCS in many western countries. In Italy the hospice care for this population is still underrepresented and most of the care is provided by neurological services.

MS is the only neurodegenerative disorders for which a SPCS prospective evaluation has been attempted (Higginson, I. J. *et al.* 2009). Due to the difficulties in predicting prognosis, people affected by this disorder are rarely considered as terminally ill and their palliative care needs tend to be unrecognized and unmet. Physical symptoms are reported in the literature, but little information is available about the end of life issues and the quality of death of people with end stage MS.

PDs are a group of neurological disorders of which Parkinson’s disease is the most frequent and the best known, but including MSA and PSP, two movement disorders with a poorer prognosis and less response to the available symptomatic therapies. Published studies report the palliative care needs explored qualitatively (including psychosocial issues) and quantitatively (mostly physical symptoms) for Parkinson’s disease only. For the atypical disorders the physiopathology and natural histories only are described.

In the literature a number of articles describe people suffering for neurodegenerative conditions considered as a whole. The main reason to justify the inclusion as a unique group is that these patients are severely affected by neurological relentless progressive conditions for which no cure exist and that are likely to die for a direct cause of their illnesses. Furthermore recent systematic reviews evidenced how the physical needs of patients affected by the disease considered in this study are frequent, many are common among the diseases and are also similar to those experienced by advanced cancer patients or other non malignant conditions (Saleem, Leigh and Higginson 2007, Solano, Gomes and Higginson 2006). Official documents from health organizations suggest improvement of the collaboration among neurology, rehabilitation and palliative care in order to find common pathways for the care of the patients and to exchange expertise (Turner-Stokes *et al.* 2007, Turner-Stokes, Sykes and Silber 2007).

5.1.3.2 The qualitative component of the study

The needs assessment was conducted with 22 in-depth interviews (involving 22 patients and 21 family carers) and three focus groups with 11 professionals directly involved in the care of these patients. The adopted methodology allowed participants to talk freely about the main difficulties that they were experiencing, and how these had been managed. The interaction between patients and their family carers provided information about the lived experience of the family group and helped when patients were impaired in communication and their carer could describe the situation and patients could intervene with non verbal communication. The focus groups included discussions among professionals who were often involved in the care of a specific disease and allowed them to compare their experience with colleagues involved in the care of different conditions. Information about the existing services in the area and suggestions on the organization of the forthcoming SPCS were very helpful.

The main findings of the content analysis of the transcript verbatim of the interviews and the focus groups were:

- High prevalence of physical symptoms:
 - problems with mobility and oral symptoms, including swallowing difficulties, that affected 100% of the patients,
 - pain and respiratory troubles (>80%),
 - intestinal symptoms (>70%),
 - urinary and sleep disorders (>60%)

These results are consistent with those previously published in the literature.

- High prevalence of psycho-social and spiritual unmet needs:
 - anxiety, depression, abandonment and coping issues (70-100%),
 - social isolation (100%),
 - difficulties in finding meanings in the experience of the disease and ambivalence related to faith, religiousness and spiritual support (50-88%).

They were generally dissatisfied with home care services, whereas the hospital based services were described as more efficient and helpful.

Overall all participants (the potential users and the professionals) seemed positive about the forthcoming SPCS, above all for the potential help at home and the possibility of respite admissions into the hospice inpatients facility.

The findings of the needs assessment, summarized above, highlight how both problems and needs were found during the interviews and the focus groups. Within the problems are the objective findings such as the high disability level of the sample of patients (described by the 100% of movement impairment), the lack of available service tailored on the needs of the participants, the absence of effective therapies to arrest or relent the evolution of the diseases. Unmet needs included the necessity of the participants to be considered and treated as human beings, to have the symptoms controlled, to find meaning in their lived experiences, to have their decisions (above all those related to the end of life decisions) respected.

This classification was taken as a lead for the following quantitative assessment of the service. In fact a cohort of measurable domains was chosen from the multitude raised by the participants of the qualitative assessment. The adopted criteria were:

- the importance of the elicited domains was defined by the prevalence of the problems – needs as classified in the content analysis (the most frequently reported conditions were likely to be a reliable outcome measure)
- the needs were considered to be more subjective and potentially reversible than the problems. In particular those unmet needs that usually a SPCS have to face when caring for very ill people (e.g. symptom control, psychosocial issues, spiritual concerns, support to the families etc) are potentially impacted in a positive manner, rather than objective problems like the closeness to death, the physical restrictions, the loss of job or the unrealistic expectations that unfortunately can not be changed and in fact are not considered as palliative care outcomes.

5.1.3.3 The quantitative component of the study

A phase 2 pilot explorative RCT, adopting the waiting list procedure, allowed an assessment of the impact of the new SPCS on the PCO of patients severely affected by neurodegenerative disorders and of their carers.

50 patients and their informal carer were recruited and randomly allocated in the two groups of the study, one receiving the SPCS, the other having to wait 16 weeks before being cared for by the SPCS. At baseline no significant differences were found between the groups. Overall the sample revealed a very high disability level, an equal distribution for the different conditions and for the demographics. The study was feasible, well accepted by the participants, with low attrition, low mortality and little missing data. The groups who had received the service had the same number of deceased patients of the controls, confirming that palliative care does not increase mortality.

Considering the trial outcomes the treated group compared with the control one reported:

- A statistically and clinically significant improvement in the QoL and in the control of pain, breathlessness, sleep disturbance and intestinal symptoms
- A clinically relevant improvement of the social isolation of the patients
- A clinically moderate improvement of
 - the urinary and the oral symptoms
 - one spiritual theme “finding a meaning in the experience of the disease”
 - the satisfaction about the received services for both patients and carers

All the other measured outcomes did not show significant changes between the groups. In particular one of the main outcomes of the trial was the caregivers’ burden of care. This aspect showed a very small improvement for those carers who were supported by the SPCS (5%) that is to be considered too small to be clinically significant and is not statistically significant.

5.1.3.4 The role of the MRC framework

The MRC framework for developing and assessing complex interventions, adopted as a general scheme for both the service development and the research project reported in this thesis, showed some positive aspects and some limits that will now be described.

The most positive aspect of having such a structured programme to follow is that each step can be effectively thought, planned and put in action without incurring into the risk of bypassing some fundamental part.

Chapter 5.1 Conclusions and Recommendations

1. The theory phase precedes the modelling of the service and in the present thesis involves all those processes that led:
 - from the initial awareness of the general problem – the perception that people severely affected by neurodegenerative conditions could receive help from palliative care, but the lack of experience worldwide in this field and the presence of sparse published evidences
 - to the formulation of the research question
 - to the choice of the mixed methods methodology to address the two different parts of the question
 - the literature review concluded this preliminary phase allowed a better understanding of the problem and led to the second step of the framework.

2. The phase 1 (modelling) included:
 - The education process of the personnel of the SPCS, which was stimulated by the evidence from the literature and, later, from the qualitative assessment, of:
 - a. the lack of knowledge of the professionals, expert in cancer care, of the specific clinical features of the neurodegenerative disorders
 - b. the lack of experience of the management of the specific equipment used by these patients (e.g. mechanical ventilators, cough machines, augmentative-alternative communicators)
 - c. the failure of other hospices in starting to care for neurological patients or their decision to stop their admissions in their programmes due to the difficulties that raised during the care and the lack of education in the care for this patient group
 - d. the reports from the potential users (e.g. the participants of the interviews) that stated that they felt totally unsatisfied when they were seen by health professionals (e.g. GP's, emergency services, general wards), who they felt were not prepared for the specific features of their disorders and, sometimes, were scared by the equipment used by the patients and often knew less than the informal caregivers about the correct management of the patient and his or her problems.

 - The needs assessment. By means of the interviews and of the focus groups much useful information was collected and consequently used to model the forthcoming specialist service. The aim was to design it tailored on the needs of the potential users, it was thus essential to ensure that the exploration of the needs of patients and their families living in the area where the SPCS was going to be set up, and collection the professionals' point of view was undertaken. The methodology of the modelling phase was therefore a mix of the analysis of pre-existing experiences from other services, and published in the literature, and new data collected in the qualitative needs assessment of this research.

3. The experimental phase, reflecting the phase 2 of the MRC, represent the final outcome of this research project, but at the same time is just the start of the new service provided by FARO Foundation for people severely affected by neurodegenerative conditions. This is being continued and implemented and that will be better explained in the following paragraph. This phase was explorative by definition and aimed at finding a number of variables potentially modifiable by the input of the new service that are be measured overtime compared to an appropriate control group. In the present piece of research this phase provided the most important news, showing how the service was able to improve some important palliative care outcomes in this population and provide novel data never shown before in the published literature. In this phase a fully quantitative approach was adopted to assess the impact of the new service. A parallel qualitative approach aimed at understanding not only how much the variables changed between the group that received the service and the control group, but also why and how this process would have been very helpful and allowed a broader understanding of the effectiveness of the service. This had not been possible, due to time restraints but future research using this MRC approach would benefit from a qualitative approach in conjunction with the quantitative methodology.

Among the limits of this framework what really makes it difficult the extend use of this tool is its length. It would be very difficult to have to repeat all the procedures if, for instance, FARO Foundation should decide to extend its SPCS to other categories of non cancer patients like those affected by cardiopulmonary advanced conditions, renal failures, or other untreatable disorders. Furthermore the different methodologies adopted in this research and described in this thesis should be conducted in parallel rather than in series. The use of a mixed methods approach with both quantitative and qualitative methods used together, rather than following each others, provide more information and, above all, explain the meaning of the findings in a more straight forward way.

To conclude the iterative process of the MRC framework appears to be a positive pathway to follow to achieve a continuous improvement of the service. The framework is certainly applicable to the palliative care setting, perhaps a little too long to be used in daily clinical settings, whereas it can be an optimal solution for experimental designs.

5.1.3.5 Overall effect

Since the beginnings of this research project the FARO new SPCS for people severely affected by neurodegenerative conditions has cared for 90 patients with a neurological diagnosis. The service is now fully operative for ALS/MND patients who do not have a tracheostomy and is still in a research phase for the other diagnoses and for the ALS/MND tracheostomized patients. The professionals working in the SPCS are satisfied with the new skills learnt during the education process that led to the establishment of the service. The professional network that resulted by the interaction of the new SPCS with the neurologists, rehabilitation specialists, lung specialists and other professionals has aided the development of a comprehensive service for the patients and their families throughout the various aspects of the care pathway. A new inpatients facility is under construction to satisfy the increased number of respite admissions for these people.

Chapter 5.1 Conclusions and Recommendations

In conclusion this project is one of the few evaluations of specialist palliative care for patients with a diagnosis other than cancer. In particular it is the first study that assessed the impact of a SPCS on the palliative outcomes of patients severely affected by different neurodegenerative conditions and in their family carers.

One important result reported in this study is that the involvement of a SPCS caused an improvement of the individual QoL of the patients and a better symptom control above all for pain, breathlessness, quality of sleep and intestinal symptoms. This is the first published study, at the author's knowledge, to show such results in a palliative care setting for neurodegenerative conditions. As there are very few studies on the development of SPCS the study has also shown the effectiveness and importance of SPCS developments generally.

5.2 Recommendations

From the analysis of the results of the overall research project summarized in the Conclusions section the following recommendations can be drawn:

1. Specialist Palliative Care should be offered to patients severely affected by ALS/MND, MS and PDs because of the positive impact on the individual QoL and symptom control.
2. Further studies should be done to evaluate if the positive impact, demonstrated by this study after 16 months of service provision, is maintained over time.
3. Further studies are also needed to reassess the impact of SPCS on the caregivers' burden of care and on the psycho-social and spiritual issues because this study was not conclusive for these aspects, perhaps it was too small to detect significant changes.
4. Comprehensive assessment tools should be constructed and validated to assess the palliative care outcomes for advanced neurological conditions. These instruments should not be disease related, but should consider the common unmet needs of severely impaired neurologic patients independently from their specific diagnosis as it happens for the tools used for cancer patients that are not dependent on the type of cancer, but on the most common problems.
5. Different modalities of service provisions should be experimented for the care of people suffering for long term neurological conditions based on the needs of both the patients and their carers. The positive effect of the input of the SPCS (demonstrated in this study) on the individual QoL and on the symptoms could be exploited not only for the terminal care, but as an "on demand service" to be offered in those occasions of particular sufferance experienced by the patients throughout the long term condition. This idea is similar to the simultaneous care for cancer patients receiving palliative care inputs when chemotherapy is continued and palliative care needs appear.
6. When a palliative care team involved in cancer care decides to extend the service to patients with conditions other than cancer, a dedicated educational project must be set up to enable the professionals to acquire the necessary skills. Improving the knowledge of the specifics of the diseases (trajectories, symptoms, equipment, end of life issues, ethics issues) affect the outcomes of the service.

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7. Appendix 1: abbreviations

Table of abbreviations

ADI	Italian Public Primary Care Home Assistance
AAN	American Academy of Neurologists
A&E	Accident & Emergency Units
ADL	Activities of Daily Living
ALS/MND	Amyotrophic Lateral Sclerosis / Motor Neurone Disease
AMTS	Abbreviated Mental Test Score
ALSFRS-R	Amyotrophic lateral sclerosis functional rating scale revised
ANCOVA	Analysis of Covariance
CBI	Caregiver Burden Inventory
CNS	Central Nervous System
COPD	Chronic Obstructive Pulmonary Disease
CPR	Cardiopulmonary Resuscitation
CRF	Case Record Form
DBS	Deep Brain Stimulation
DN	District Nurse
EoL	End of life
EAPC	European Association for Palliative Care
EC	Ethics Committee
EDSS	Expanded Disability Status Scale
FARO	Foundation for Assistance and Research in Oncology
FDA	Federal Drug Administration
FT – ST	Fast Track – Standard Track
FTD	Fronto-Temporal Dementia
GP	General Practitioner
H&Y	Hoehn & Yahr Score (PDs disability scale)
HADS	Hospital Anxiety and Depression Scale
HIV/AIDS	Human Immunodeficiency Virus / Acquired Immunodeficiency Syndrome
IADL	Instrumental Activities of Daily Living
IGF	Insuline-Like Growth Factor
IV	Invasive Ventilation
MMSE	Mini-Mental State Examination

Chapter 7. Appendix 1: abbreviations

MS	Multiple Sclerosis
MSA	Multiple System Atrophy
NeuNeeds	Neurological Needs (the qualitative study of this thesis)
NePal	Neurology and Palliative care (the quantitative study of this thesis)
NHS	National Health Service
NIV	Non Invasive Ventilation
NRS	Numerical Rating Scales
PCN	Palliative care needs
PCO	Palliative Care Outcomes
PD	Parkinson's Disease
PDs	Parkinson's Disease and related disorders (Parkinson's plus syndromes)
PEG	Percutaneous Endoscopic Gastrostomy
PHT	Physiotherapy
PPMS	Primary Progressive Multiple Sclerosis
PRMS	Progressive/Relapsing Multiple Sclerosis
PSP	Progressive Supranuclear Palsy
QoL	Quality of Life
RCT	Randomized Controlled Study
RRMS	Relapsing Remitting Multiple Sclerosis
SEIQoL-DW	Schedule For The Evaluation Of Individual QoL- Direct Weighting
SLT	Speech and Language Therapy
SIN	Italian Society of Neurologists
SPCS	Specialist Palliative Care Service
SPMS	Secondary Progressive Multiple Sclerosis
VAS	Visual Analogue Scales
WHO	World Health Organization
WMA	World Medical Association

8. Appendix 2: Study documentation

The study documentation of both the qualitative and quantitative studies is provided in the exact form in which it was used in the research (Italian versions) followed by the English translation.

8.1 Participants documents of the qualitative study

8.1.1 Leaflet for the patients and the carers original Italian version:

Allegato 2

Foglietto illustrativo allegato al modulo di richiesta del consenso informato.

Le chiediamo di leggere attentamente questo foglietto illustrativo.

Esso è stato fatto per informarla riguardo ad un progetto di ricerca per il quale sarà richiesta la sua partecipazione. Le verrà richiesto di accettare o meno la partecipazione a tale ricerca, per cui leggendo con attenzione il contenuto del presente avrà la possibilità di conoscerne gli obiettivi, come esse sarà condotta, quali sono i suoi diritti di partecipante e come potrà revocare l'eventuale consenso in qualunque momento lei lo desideri.

Lo studio ha per titolo:

“Quali sono i bisogni di cure palliative specialistiche in pazienti affetti da patologie neurodegenerative in fase avanzata? Uno studio qualitativo.”

Che obiettivi ha questa ricerca?

Si tratta di una ricerca nella quale un gruppo di ricercatori che si occupano di Cure Palliative e di Neurologia cercheranno di capire quali sono i bisogni, i problemi, le preoccupazioni di malati gravemente affetti da patologie come la Sclerosi Laterale Amiotrofica, la Sclerosi Multipla, il morbo di Parkinson, l'Atrofia Multi-sistemica e la Paralisi Sopranucleare Progressiva.

Nella stessa ricerca verranno anche studiati i problemi che colpiscono i famigliari che si prendono cura di questi malati.

Come si svolgerà la ricerca?

Verranno effettuate delle interviste ai malati ed ai loro famigliari da parte di un gruppo di ricercatori.

Le interviste verranno effettuate nel luogo preferito dai partecipanti, presumibilmente al loro domicilio.

Le interviste saranno registrate, poi trascritte e studiate dai ricercatori. Le registrazioni saranno poi cancellate.

I dati che verranno utilizzati saranno completamente anonimi. Solo i ricercatori che avranno condotto le interviste conosceranno l'identità dei partecipanti e non li divulgheranno per nessun motivo.

A chi parteciperà sarà chiesto di esprimere un consenso alla partecipazione. Sarà un modulo scritto dai ricercatori. Esso verrà letto e spiegato ai malati e famigliari interessati. Se verrà dato il consenso alla partecipazione verrà chiesto di firmare il modulo, o comunque di esprimere chiaramente la propria volontà in presenza di un familiare.

Come vengono selezionati i partecipanti?

I neurologi, che hanno in cura gli ammalati e ne conoscono le condizioni cliniche e le caratteristiche famigliari, segnaleranno ai ricercatori queste persone. Poi il gruppo di ricerca contatterà i possibili interessati e ne chiederà il consenso alla partecipazione.

Chi acconsente alla partecipazione dovrà per forza completare le interviste?

Assolutamente no! In qualsiasi momento il consenso potrà essere revocato. L'intervista si bloccherà ed il partecipante potrà acconsentire a che si utilizzi il materiale raccolto fino a quel momento oppure potrà chiedere l'immediata cancellazione delle registrazioni.

A cosa servirà questa ricerca?

- In primo luogo servirà per capire meglio le problematiche specifiche di ammalati e famigliari quando una grave malattia neurologica li colpisce. Siamo interessati a conoscere non solo i problemi fisici, ma anche le difficoltà psicologiche, relazionali, le difficoltà legate alla comunicazione in famiglia e con i medici. Vorremmo conoscere i problemi sociali che si manifestano in queste situazioni: i problemi sul lavoro, le difficoltà economiche, l'isolamento dagli altri, la burocrazia.
Ci interessa anche conoscere come la spiritualità possa giocare un ruolo in queste fasi difficili della malattia.
Tutte queste informazioni saranno raccolte e studiate.
Cercheremo di pubblicare i risultati su riviste specifiche e li presenteremo nei congressi scientifici dedicati alle cure palliative e alla neurologia.
Ci aspettiamo che la conoscenza chiara ed approfondita dei problemi delle persone possa essere da stimolo per migliorare l'assistenza.
- L'obiettivo più prossimo ed interessante sarà però quello che cercheremo di attuare dall'autunno di quest'anno. Infatti verrà istituito un nuovo servizio di cure palliative che la fondazione FARO, che già da anni si occupa di assistere a casa ed in Hospice i malati terminali di cancro, fornirà anche ai malati affetti dalle patologie elencate sopra. Questo servizio sarà modellato il più possibile sui bisogni che saranno evidenziati dall'attuale ricerca. Ecco perché sarà importante la vostra partecipazione. Con i dati raccolti dalle interviste si cercherà di creare un nuovo servizio "cucito" addosso ai bisogni dei malati e delle loro famiglie.

La partecipazione allo studio è dunque graditissima.

Chapter 8. Appendix 2: Study documentation

Non sarà riconosciuta nessuna forma di pagamento ai partecipanti.

L'eventuale rifiuto alla partecipazione sarà ovviamente accettato serenamente dal gruppo di ricercatori.

Chiaramente non ci sarà nessuna disparità di trattamento, in termini di qualità di cure e assistenza, tra chi darà il proprio consenso ad essere intervistato e chi non vorrà partecipare.

Vi porgiamo i più cordiali saluti

Il gruppo di ricerca: Simone Veronese, Gloria Gallo, Chiara Rivoiro

8.1.2 Leaflet for the patients and the carers English version

Attachment 2

Illustrative leaflet appended to the consent form for the participants.

We kindly ask you to read carefully this leaflet.

This document was prepared in order to inform you about a research project in which you have been asked to participate. You will be asked to accept or to decline your participation at the present study, for this reason reading this leaflet will inform you of the aims, how it will be conducted, what are your rights as participant and how you will be able to withdraw your consent at any time.

The title of this study is:

“What are the specialist palliative care needs of patients severely affected by neurodegenerative conditions in advanced stage? A qualitative study.”

What are the aims of this research?

This research project, set up by a group of researchers with an interest in palliative care and neurology, aims at exploring the unmet needs, the worries and the problems faced daily by people severely affected by Amyotrophic Lateral Sclerosis, Multiple Sclerosis, Parkinson’s disease, Multiple System Atrophy and Progressive Supranuclear Palsy. Family carers issues will be also considered in this project.

How will be conducted the research?

A group of researchers will interview you and your family. The interviews will be held in the participant’s preferred location, usually your own home. Interviews will be audio and video recorded, a transcript verbatim of the contents will be performed and analyzed by the researchers. The tapes will be then deleted. Data will be studied and used anonymously. Only the researchers conducting the interviews will know the participants identity and no-one else will have this information. Potential participants are asked to provide their consent to the participation. A specific form will be read and explained to the interested participants. If you consent to be involved you will be asked to sign the form, or to clearly express your consent if you cannot sign it.

How are selected the participants?

Potential participants will be referred to the researchers by the neurologists that take care of them in the hospital services. They are aware of the inclusion criteria for this study. The research group will then contact the potential participants.

If a consent is given, will the participants be bound to complete the interviews?

Not at all! At any time the consent can be withdrawn. The interview will be stopped and then you may decide whether the material collected until that point could be used for the research, or ask the cancellation of the content.

What will be this research used for?

- First of all it will provide important information about the specific problems faced by the patients and by their families with a severe neurologic disorder. Researchers are interested in the physical problems, the psychological and the relational issues, the communication problems within the family and with the professional carers. Further specific points of interest are the social problems that appear in this situations: job related problems, economic difficulties, social isolation and bureaucracy. We also want to know about the role of spirituality in these difficult circumstances. All this information will be collected and studied. We will try to publish the findings in specialized journals and to present in scientific congresses of palliative care and neurology.
- A more ambitious objective is what is going to be set up in Autumn of this year. A new specialist palliative care service provided by the FARO Foundation, an experienced charity that provides a well known similar service for people severely affected by cancer, will be designed and assessed for people severely affected by the disorders listed in this leaflet. This forthcoming new service will be modelled as much as possible on the needs that will emerge from this research project. This is why your participation is so important. Data collected from the interviews will be used to develop a new service tailored on the needs of the patients and of their families.

Your participation is therefore very welcome.

No payment will be provided for the participation. Researchers will accept your refusal if you do not wish to be involved and there will be no discrimination in terms of quality of care and assistance will be applied among who will accept and who will decline the consent to participate at this research.

Yours faithfully,

The research group:

Simone Veronese, Gloria Gallo, Chiara Rivoiro

8.1.3 Leaflet for the patients' General Practitioners Italian version

Allegato 3.

Protocollo di studio sui bisogni:

- di pazienti affetti da patologie neurodegenerative in fase avanzata
- dei loro famigliari (caregivers)

LETTERA INFORMATIVA PER IL MEDICO DI MEDICINA GENERALE.

Egregio Collega,

desidero informarla che è stato proposto al suo paziente, sig.
affetto da..... ed il suo famigliare di riferimento
sig..... Entrambi hanno accettato di partecipare allo studio clinico

“Quali sono i bisogni di cure palliative specialistiche in pazienti affetti da patologie neurodegenerative in fase avanzata? Uno studio qualitativo.”

Tale progetto di ricerca viene condotto da un team di ricercatori formato da medici palliatori e neurologi ed ha l'obiettivo primario di valutare i bisogni di tipo fisico, psicologico, sociale e spirituale degli ammalati affetti da patologie neurodegenerative in fase avanzata (Sclerosi Laterale Amiotrofica, Sclerosi Multipla, morbo di Parkinson, Atrofia Multi-sistemica e la Paralisi Sopranucleare Progressiva).

Anche i famigliari di riferimento (caregivers) saranno coinvolti per conoscere le loro problematiche legate alla gestione della malattia.

I partecipanti saranno intervistati, con interviste semistrutturate ed approfondite, presso il loro domicilio, o comunque dove sarà loro più comodo.

La partecipazione sarà libera, previo consenso informato che sarà comunque revocabile durante le interviste.

I dati saranno trattati con riservatezza. I risultati saranno presentati in forma anonima.

Il progetto nasce dalla collaborazione tra la fondazione F.A.R.O., che da anni si occupa di assistere a casa ed in Hospice i malati terminali di cancro, e le cliniche neurologiche dell'ospedale Molinette di Torino e dell'ospedale S.Luigi di Orbassano.

I risultati dello studio saranno pubblicati su riviste specialistiche, presentati ai congressi dedicati alle cure palliative ed alla neurologia.

Dall'analisi dettagliata di questi bisogni nascerà un nuovo servizio di cure palliative domiciliari, il più possibile modellato sulle reali necessità di queste persone.

Tale servizio sarà fornito dalla fondazione F.A.R.O. a partire dall'autunno 2007, inizialmente in via sperimentale su casi selezionati per valutarne l'efficacia e l'appropriatezza, poi in futuro sarà a disposizione di tutti i pazienti affetti da patologie neurodegenerative le cui caratteristiche siano compatibili con la quelle previste dalla Delibera Regionale sulle Cure Palliative vigente: D.G.R. n. 15-7336 del 14.10.2002.

RingraziandoLa anticipatamente per la sua attenzione rimango a sua disposizione per qualsiasi chiarimento in merito e per qualsiasi informazione che Lei ritenesse necessaria. Cordiali saluti

8.1.4 Leaflet for the patients' General Practitioners English version

Attachment 3

Study Protocol on the needs:

- **Of patients severely affected by neurodegenerative conditions**
- **And their carers (caregivers)**

INFORMATION LETTER FOR THE GENERAL PRACTITIONERS

Dear Colleague,

I want to inform you that it has been proposed to your patient, Mr.... affected by and to his or her family carer Both accepted to participate at the clinical study called:

“What are the specialist palliative care needs of patients severely affected by neurodegenerative conditions in advanced stage? A qualitative study.”

This research project is conducted by a team of researchers formed by palliative care experts and neurologists and its the primary aims is to assess the physical, psychological, social and spiritual unmet needs of people severely affected by neurodegenerative conditions (Amyotrophic Lateral Sclerosis, Multiple Sclerosis, Parkinson's disease, Multiple System Atrophy and Progressive Supranuclear Palsy).

Family carers will be involved to collect their point of view about the problems caused by the diseases. Participants will be interviewed, with in-depth semi structured interviews, in their homes or in other places of their choice.

Participation is free, an informed consent will be obtained beforehand and will be possible to withdraw at any time. Data will be treated confidentially and the results will be presented in anonymous form.

The project was born from the collaboration between FARO Foundation, which has been providing palliative care for cancer patients at home and in hospice for many years, and the neurological clinics of the Molinette Hospital of Turin, and San Luigi Gonzaga Hospital of Orbassano.

Results will be submitted for publication to scientific journals and to specialist congresses.

From the analysis of the unmet needs that will emerge in this study a new specialist palliative care service will be set up, modelled as much as possible on the real problems of this population. This service will be provided by FARO Foundation from Autumn 2007, initially as an experimental phase on selected case to assess its efficacy and appropriateness, later will be available for all the patients with neurodegenerative conditions whose characteristics will be included in the Regional Resolution on Palliative Care in force: D.G.R. n. 15-7336 of the 14.10.2002.

Thanking you in advance I will be available for any enquiry or information that you should need.

The research group:

Simone Veronese, Gloria Gallo, Chiara Rivoiro

8.1.5 Participant's consent form Italian version

Allegato 1

MODELLO DI CONSENSO INFORMATO

Io sottoscritto/a..... attesto di essere stato informato da.....degli obiettivi dello studio:

“Quali sono i bisogni di cure palliative specialistiche in pazienti affetti da patologie neurodegenerative in fase avanzata? Uno studio qualitativo.”

Ho letto il foglio informativo allegato a questo modulo.

Se non ho potuto leggerlo il ricercatore.....lo ha fatto per me.

Ho compreso il contenuto del foglietto allegato.

Se accetterò di partecipare a tale studio lo farò sapendo che il mio consenso potrà essere revocato in qualsiasi momento lo riterrò necessario.

Nel caso in cui decida di negare il mio consenso durante lo svolgimento dello studio stesso potrò decidere se consentire ai ricercatori di utilizzare le informazioni già raccolte o se chiederne l'immediata distruzione.

Sono consapevole dell'utilizzo che verrà fatto dei dati che fornirò ai ricercatori.

Sono stato informato che le registrazioni delle interviste da me rilasciate verranno distrutte dopo essere state trascritte.

Sono anche consapevole che queste trascrizioni saranno conservate dai ricercatori in modo sicuro e che saranno anonime, cioè non ci sarà traccia dei mie dati personali su di esse.

Sono consapevole che la partecipazione o meno allo studio non influirà in alcun modo sulla qualità ed il tipo di cure che riceverò.

In base a tutte queste informazioni da me ricevute e comprese (crociare la risposta precelta):

ACCONSENTO

NON ACCONSENTO

Alla partecipazione a questo studio.

Data e luogo

firma del partecipante

8.1.6 Participant's consent form English version

Attachment 1

CONSENT FORM MODULE

Mr/Mrs states that I have been informed by Dr Simone Veronese about the aims of the study:

“What are the specialist palliative care needs of patients severely affected by neurodegenerative conditions in advanced stage? A qualitative study.”

I have read the leaflet for the participants, in case I could not read it the researcher did it for me.

I understand the content of the leaflet. If I accept to participate to the present study I will do it knowing that my consent can be withdrawn at any time if I decide to do so.

If I will withdraw the consent during the interview I will decide whether consent or not to the researchers to use the data already collected or not.

I am aware of the use that the researchers will do of the data I will provide.

I am aware that the video and audio taped material will be made into a transcript and then deleted.

I am aware that data will be treated anonymously and my personal information will not be given out to anyone else.

I am aware that the decision to participate or not participate will not influence the quality of care and of assistance that I will receive in the future.

According to these conditions I decide to

ACCEPT

NOT ACCEPT

To participate at the present study

Date, Place.

Participant's signature

8.2 Participants documents of the quantitative study

8.2.1 Leaflet for the participants Italian version

FOGLIO INFORMATIVO PER I PAZIENTI

STUDIO ESPLORATIVO PILOTA RANDOMIZZATO, CONTROLLATO SULLA VALUTAZIONE DELL'IMPATTO SUI PALLIATIVE CARE OUTCOMES (PCO) DA PARTE DI UN SERVIZIO SPECIALISTICO DI CURE PALLIATIVE (SPCS) IN PAZIENTI AFFETTI DA PATOLOGIE NEURODEGENERATIVE

Introduzione:

Questo studio è realizzato per verificare l'efficacia di un Servizio di Cure Palliative Specialistiche nell'aiuto a persone affette da malattie neurodegenerative a domicilio ed in Hospice. Si tratta di uno studio randomizzato controllato. Questo significa che alcuni partecipanti riceveranno il servizio, offerto gratuitamente dalla Fondazione F.A.R.O., per un periodo di quattro mesi. Gli altri invece dovranno attendere quattro mesi per poterlo ricevere. Tutti i pazienti riceveranno contemporaneamente i normali servizi che il servizio sanitario mette loro a disposizione (medico di famiglia, servizi ospedalieri e domiciliari come sempre). Il posizionamento nel gruppo che riceverà subito il servizio di cure palliative della F.A.R.O., oppure che si debba attendere quattro mesi è puramente casuale, avviene tramite un sorteggio.

Qual è l'obiettivo dello studio?

L'obiettivo dello studio consiste nel determinare se il servizio di cure palliative domiciliare ed hospice della F.A.R.O. è in grado di migliorare alcune componenti della qualità della vita dei malati affetti da patologie neurodegenerative e nei loro familiari.

Come si eseguirà lo studio?

Nel corso di 8 mesi verranno valutati alcuni ammalati di Sclerosi Laterale Amiotrofica, Sclerosi Multipla, malattia di Parkinson e parkinsonismi. Tutti saranno sottoposti ad una prima visita domiciliare da parte di personale specialistico della F.A.R.O. I pazienti che acconsentiranno alla partecipazione allo studio verranno valutati con dei test specifici per capire quali siano i loro principali problemi. Anche la persona che si prende cura di loro sarà sottoposta ad alcuni test. Dopo questa valutazione i partecipanti saranno divisi nei due gruppi in modo casuale. Un gruppo riceverà subito il servizio della F.A.R.O., l'altro (che chiameremo gruppo di controllo) continuerà a ricevere gli attuali servizi a sua disposizione. Dopo 4 mesi tutti i partecipanti saranno nuovamente sottoposti agli stessi test. Le analisi statistiche ci aiuteranno a capire se e quanto il servizio di cure palliative della F.A.R.O. possa essere stato utile nel migliorare la qualità della vita dei partecipanti. Al termine dei 4 mesi dello studio tutti coloro che hanno partecipato potranno usufruire del servizio della F.A.R.O., se lo vorranno, gratuitamente.

Contiamo di arruolare 50 malati residenti nella città di Torino e nelle aree della provincia dove è attualmente in funzione il servizio domiciliare della F.A.R.O.

Sono tenuto/a a partecipare?

No. Sarà a Lei decidere se prendere parte o meno allo studio. Se decide di parteciparvi, Le daremo questo Foglio informativo da conservare e Le chiederemo di firmare un modulo di consenso. Tuttavia, Lei rimarrà libero/a di ritirarsi in qualsiasi momento, senza dover indicare alcun motivo. Se decide di ritirarsi dallo studio, oppure di non parteciparvi affatto, questo non ridurrà in alcun modo la qualità o lo standard di cura che Lei riceve o che può aspettarsi di ricevere.

Al termine dello studio, Lei potrà scegliere se continuare a ricevere, o meno, il servizio della F.A.R.O., se già lo stava ricevendo, o di iniziare a riceverlo, se si era trovato nel gruppo che doveva attendere i 4 mesi – sempre che non si siano identificati problemi di sicurezza di rilievo e che Lei non abbia ritirato il Suo consenso a partecipare a questo studio.

Cosa mi succederà se partecipo allo studio?

Nel corso della prima visita valuteremo la sua storia di malattia, verrà visitato da un medico della nostra fondazione, le descriveremo l'intento dello studio e le modalità del servizio. Se intenderà partecipare allo studio chiederemo una firma sui moduli del consenso. Se il malato non fosse in grado di firmare, ma riuscisse ad esprimere un chiaro consenso alla partecipazione, allora chiederemo al suo familiare di firmare in sua vece ed in sua presenza.

Se Lei risulta idoneo/a allo studio, La assegneremo a caso al trattamento. Quest'assegnazione casuale, chiamata randomizzazione, significa che Lei potrà ricevere il servizio della F.A.R.O. immediatamente oppure se dovrà attendere 4 mesi. Normalmente per la randomizzazione si utilizza uno speciale programma di computer, in modo che né Lei né il medico saprete in quale gruppo lei finirà.

Se finirà nel gruppo che riceve subito il servizio le sarà assegnato un medico ed un infermiere della fondazione che verranno gratuitamente a visitarla a domicilio e le spiegheranno tutte le possibilità di intervento, finalizzate al miglioramento della qualità della vita, che saranno a sua disposizione. Se finirà nel gruppo di controllo continuerà a ricevere gli attuali servizi a sua disposizione ed alla fine dei 4 mesi sarà nuovamente sottoposto ai test della prima visita. Da quel momento le verrà proposto di ricevere gratuitamente tutti i servizi della nostra fondazione

Se decide di interrompere lo studio, Le chiederemo di completare una valutazione quando decide di sospendere lo studio, oppure il più presto possibile dopo quella data.

Quali sono le alternative di trattamento?

Se decide di non partecipare allo studio, continuerà a ricevere tutti i servizi che ha ricevuto fino ad ora.

Quali sono i rischi e gli effetti indesiderati del trattamento che riceverò durante la partecipazione a questo studio?

Non sono previsti rischi specifici implicabili alla partecipazione allo studio. Ogni intervento, farmacologico e non, che le sarà eventualmente proposto sarà basato sulle più aggiornate linee guida ed indicazioni internazionali e verrà attentamente discusso con lei.

Quali sono i benefici della partecipazione?

Ci auguriamo che il servizio potrà giovare alla Sua qualità di vita, tuttavia non ci è possibile offrire alcuna garanzia a questo riguardo. Le informazioni che ricaveremo dallo studio potrebbero aiutarci a prenderci cura di molti altri malati affetti da patologie neurodegenerative in futuro.

Cosa succede se dovessero emergere nuove informazioni?

Talvolta durante un progetto di ricerca emergono nuove informazioni sui servizi oggetto di studio. In questo caso, il medico della ricerca La informerà e discuterà insieme a Lei se continuare o meno lo studio. Se decide di ritirarsi, il medico prenderà tutte le misure necessarie per il proseguimento delle Sue cure. Se invece decide di continuare lo studio, Le chiederemo di firmare un modulo di consenso aggiornato.

Cosa succede al termine dello studio?

Dopo l'ultima visita (quando Lei avrà completato i 4 mesi di osservazione), potrà scegliere di ricevere, o continuare a ricevere, il servizio della F.A.R.O., se lo desidera. Se non desiderasse ricevere tale servizio le sarà sufficiente comunicarcelo.

La mia partecipazione a questo studio riceverà un trattamento riservato?

Questo studio non è sponsorizzato. Il servizio verrà fornito gratuitamente dalla Fondazione F.A.R.O. di Torino. I dettagli riguardo il Suo trattamento e altre informazioni mediche – ma non i Suoi particolari - saranno resi disponibili solo alle autorità regolamentatrici, per verificare la corretta esecuzione dello studio. Può inoltre darsi che altri enti statali ispezioneranno i Suoi dati, ma manterremo sempre la riservatezza. Tutte le informazioni rimarranno riservate. Potremo pubblicare i risultati di questo studio, ma senza identificarLa in alcun modo. Se Lei acconsente a partecipare allo studio, informeremo il Suo medico. Tuttavia, il Suo nome non verrà divulgato a nessuno al di fuori dell'ospedale/ambulatorio del medico.

Chi posso contattare per avere ulteriori informazioni?

Se Lei ha problemi, preoccupazioni o altre domande circa questo studio, contatti preferibilmente il dottor Simone Veronese presso la Fondazione F.A.R.O. ONLUS telefono 011 888.272

8.2.2 Leaflet for the participants English Version

INFORMATION LEAFLET FOR THE PARTICIPANT

EXPLORATIVE RANDOMIZED AND CONTROLLED STUDY TO EVALUATE THE IMPACT OF A SPECIALIST PALLIATIVE CARE SERVICE (SPCS) ON THE PALLIATIVE CARE OUTCOMES (PCO) FOR PATIENTS WITH NEURODEGENERATIVE CONDITIONS

Introduction:

This study was designed to assess the effects of a new SPCS in the help to people affected by neurodegenerative disorders at home and in the hospice. It is a randomized controlled study, this means that some participants will receive immediately the SPCS provided by FARO Foundation, free of charge immediately whereas others will have to wait for 16 weeks before receiving the service. All the participants will continue to receive the normal services available from the NHS (GP, hospital based services, public home care). The allocation in the group immediately receiving the service, or in the group that has a 16 weeks wait is due to chance and happen for drawing of lots.

What is the aim of this study?

The aim of this study is to determine if the SPCS provided by FARO can improve some components of the Quality of Life in patients affected by advanced neurodegenerative conditions and in their family carers.

How will be conducted the study?

Over a period of 8 months patients with diagnosis of ALS/MND, MS, PDs will be assessed. They will receive a first visit from FARO personnel. Those who will consent to participate will be assessed with specific tools to highlight their main unmet needs. Also the family carer that provides the daily care to the patient will be invited to participate and asked to complete some tests. After this first assessment patients will be randomly allocated in two groups: one group will immediately receive FARO service, and the other group (that we will call control) will continue to receive the standard available services. After 4 months all participants will be reassessed with the same battery of tests. Statistical analysis will help to determine in which aspects the intervention of the SPCS can make the difference to improve the participants' Quality of Life. After 4 months all participants will receive the fullservice from FARO, if they desire it, free of charge. We aim at enrolling about 50 patients and their carers living in Turin city and its metropolitan area.

Must I participate?

No. it is up to you to decide if take part to this study or not. If you decide to be involved you will be given this information leaflet and we will ask you to sign this consent form. However you will remain free to withdraw at any time from the study. If you will leave the study, or you decide not to take part in it this will not affect the standard of quality of care that you are receiving or that you will receive in the future. At the end of the study you will choose if you will want to receive the FARO service for the future.

What happens to me if I participate?

During the first assessment you will be visited by a FARO doctor, the study will be explained to you in details and you will sign the consent form. If you are not be able to

sign your oral consent will be considered valid and a member of your family will sign the form for you.

If you will be considered eligible you will be allocated randomly in the group that receive immediately the FARO service or in the control group that has a 16 weeks wait before receiving the service.

If you will be allocated in the group immediately receiving the service you will be assigned to a mini team composed by a physician expert in palliative care and a nurse. Both will look after you at home or in the hospice. If the input of other professionals of the service will be required they will be organised by your mini team.

If you will be allocated in the control group you will spend 16 weeks receiving the standard services available only. After 4 months you will be reassessed and from that moment you will receive the FARO service.

If you decide to withdraw from the study you will be asked to fill and sign a form.

What are the alternative to the treatment?

If you do not want to receive the service from FARO you can continue to receive the traditional available services.

What are the risks and the possible side effects that I can incur into during this trial?

No risks or specific potential negative factors are foreseen for those who will receive the service.

Any drug or not pharmacological treatment that you will be offered is based upon international published guidelines and you will be informed of any therapeutic option and rationale.

What benefits can I expect from my participation?

We sincerely wish that the service can have positive effects to your own Quality of Life, but this cannot be guaranteed. Information emerging from the study will be of help for the care of other patients in the future.

What if new information emerge during the study development?

Sometimes, during the development of experimental studies, new information may emerge that can alter the activities of the research project. In that case you will be informed by the researchers and you will be able to decide whether withdraw or continue in the study. If you withdraw the research team will provide for you the best therapeutic option available. If you will remain in the study a new up to date consent form will be provided to you.

What happens at the end of the study?

At the end of the assessment that you will receive after 16 weeks you will be asked if you want to receive the service or not. If you will be keen on being cared for by the service this will be continued or initiated free of charge. If you will not be interested in receiving the service you will just have to communicate this to us.

What is the data policy of this study?

This study is not sponsored by any private company. The service is offered by the FARO Foundation that will cover all the costs. Data of the participants will be controlled by the competent authorities to guarantee about the correct development of

the study. No personal sensible data will be spread. The results of the study will be published in specialist journals and in congresses.

Who can I contact for further information?

In case of further doubts or questions please contact Dr Simone Veronese at the FARO Foundation of Turin

8.2.3 Leaflet for the General Practitioners Italian version

LETTERA INFORMATIVA PER IL MEDICO DI MEDICINA GENERALE.

Egregio Collega,

desidero informarla che è stato proposto al suo paziente, sig.....

affetto da ed al suo familiare di riferimento

sig..... di partecipare allo studio clinico

**STUDIO ESPLORATIVO PILOTA RANDOMIZZATO, CONTROLLATO
SULLA VALUTAZIONE DELL'IMPATTO SUI PALLIATIVE CARE
OUTCOMES (PCO) DA PARTE DI UN SERVIZIO SPECIALISTICO DI
CURE PALLIATIVE (SPCS) IN PAZIENTI AFFETTI DA PATOLOGIE**

Tale progetto di ricerca viene condotto da un team di ricercatori della Fondazione F.A.R.O. onlus formato da medici palliatori e neurologi ed ha l'obiettivo primario di verificare l'efficacia di un Servizio di Cure Palliative Specialistiche nell'aiuto a persone affette da malattie neurodegenerative in fase avanzata (Sclerosi Laterale Amiotrofica, Sclerosi Multipla, morbo di Parkinson, e parkinsonismi atipici) a domicilio ed in Hospice.

Si tratta di uno studio randomizzato controllato. Questo significa che alcuni partecipanti riceveranno il servizio, offerto gratuitamente dalla Fondazione F.A.R.O., per un periodo di quattro mesi. Gli altri invece dovranno attendere quattro mesi per poterlo ricevere. Tutti i pazienti riceveranno contemporaneamente i normali servizi che il servizio sanitario mette loro a disposizione (medico di famiglia, servizi ospedalieri e domiciliari come sempre). Il posizionamento nel gruppo che riceverà subito il servizio di cure palliative della F.A.R.O., oppure che si debba attendere quattro mesi è puramente casuale, avviene tramite un sorteggio.

La partecipazione sarà libera, previo consenso informato che sarà comunque revocabile durante lo studio.

I dati saranno trattati con riservatezza. I risultati saranno presentati in forma anonima.

Il progetto nasce dalla collaborazione tra la fondazione F.A.R.O., che da anni si occupa di assistere a casa ed in Hospice i malati terminali di cancro, le cliniche neurologiche ed i servizi ospedalieri dedicati alla cura di questi ammalati dell'ospedale Molinette di Torino e dell'ospedale S.Luigi di Orbassano.

I risultati dello studio saranno pubblicati su riviste specialistiche, presentati ai congressi dedicati alle cure palliative ed alla neurologia.

Questa fase è necessaria per ultimare il percorso intrapreso dalla Fondazione F.A.R.O. finalizzato alla creazione di un nuovo servizio di cure palliative domiciliari ed hospice, il più possibile modellato sulle reali necessità di queste persone.

Tale servizio sarà fornito dalla fondazione F.A.R.O. al termine dell'attuale studio clinico che viene fatto in via sperimentale su casi selezionati per valutarne l'efficacia e l'appropriatezza. In futuro quindi esso sarà a messo a disposizione di tutti i pazienti affetti da patologie neurodegenerative le cui caratteristiche siano compatibili con la quelle previste dalla Delibera Regionale sulle Cure Palliative vigente: D.G.R. n. 15-7336 del 14.10.2002.

RingraziandoLa anticipatamente per la sua attenzione rimango a sua disposizione per qualsiasi chiarimento in merito e per qualsiasi informazione che Lei ritenesse necessaria. Cordiali saluti

DR SIMONE VERONESE

FONDAZIONE F.A.R.O. onlus Torino,

telefono 011.888.272 cellulare 335.588.19.96

8.2.4 Leaflet for the General Practitioners English version

INFORMATIVE LEAFLET FOR THE GENERAL PRACTITIONERS

Dear Colleague,

I want to inform you that your patient, MR/MRS..... affected by His or her family carer MR/MRS Have been proposed to participate to the following trial

<p>EXPLORATIVE RANDOMIZED AND CONTROLLED STUDY TO EVALUATE THE IMPACT OF A SPECIALIST PALLIATIVE CARE SERVICE (SPCS) ON THE PALLIATIVE CARE OUTCOMES (PCO) FOR PATIENTS WITH NEURODEGENERATIVE CONDITIONS</p>
--

This research project is conducted by a team of researchers of the FARO Foundation composed by physicians experts in palliative care and neurologists and aims at assessing the impact of a new SPCS on the help to people severely affected by neurodegenerative conditions (ALS/MND, MS, PDs) at home and in the hospice.

It is a randomized controlled study, this means that some participants will receive immediately the SPCS provided by FARO Foundation, free of charge immediately whereas others will have to wait for 16 weeks before receiving the service. All the participants will continue to receive the normal services available from the NHS (GP, hospital based services, public home care). The allocation in the group immediately receiving the service, or in the group that has a 16 weeks wait is due to chance and happen for drawing of lots.

Participation is free, an informed consent will be asked to the participants and this can be withdrawn at any time if the participants will not want to stay into the study.

Data will be managed safely and will be presented anonymously.

The project is a collaboration between FARO Foundation and the neurologic clinics of the Molinette Hospital in Turin and San Luigi in Orbassano.

The results of the study will be published in scientific journals and presented at international congresses of palliative care and neurology.

This phase is necessary to complete the pathway started by FARO Foundation aimed at creating a new SPCS for people with neurodegenerative disorders modelled as much as possible on the users' needs.

This service will be provided by FARO Foundation from Autumn 2007, initially as an experimental phase on selected case to assess its efficacy and appropriateness, later will be available for all the patients with neurodegenerative conditions whose characteristics will be included in the Regional Resolution on Palliative Care in force: D.G.R. n. 15-7336 of the 14.10.2002.

Thanking you in advance I will be available for any enquiry or information that you should need.

DR SIMONE VERONESE

FONDAZIONE F.A.R.O. onlus Torino, telephone

number 011.888.272 mobile 335.588.19.96

8.2.5 Participant's consent form Italian version

MODULO DI CONSENSO INFORMATO Per i partecipanti

**STUDIO ESPLORATIVO PILOTA RANDOMIZZATO, CONTROLLATO
SULLA VALUTAZIONE DELL'IMPATTO SUI PALLIATIVE CARE
OUTCOMES (PCO) DA PARTE DI UN SERVIZIO SPECIALISTICO DI CURE
PALLIATIVE (SPCS) IN PAZIENTI AFFETTI DA PATOLOGIE
NEURODEGENERATIVE**

Medico sperimentatore: DR VERONESE SIMONE

Io
sottoscritto/a.....
..... nato
a.....il.....indirizzo.....
... telefono.....

DICHIARO

- Di partecipare volontariamente al presente studio avente lo scopo di verificare l'efficacia di un Servizio di Cure Palliative Specialistiche nell'aiuto a persone affette da malattie neurodegenerative a domicilio ed in Hospice.
- Di avere ricevuto dal Dott...VERONESE esaurienti spiegazioni in merito alla richiesta di partecipazione alla ricerca, in particolare sulle finalità e procedure.
- Di avere avuto a disposizione tempo sufficiente per potere leggere attentamente, comprendere ed eventualmente farmi spiegare quanto contenuto nella scheda informativa allegata e da me sottoscritta per presa visione, e che conferma quanto mi è stato spiegato a voce, in particolare che la ricerca sarà condotta nel rispetto dei codici etici internazionali.
- Di aver avuto la possibilità di porre domande e di avere avuto risposte soddisfacenti su tutta la ricerca.
- Di essere stato informato sui disagi ragionevolmente prevedibili.
- Di acconsentire \ non acconsentire che il medico responsabile informi il mio medico di famiglia.
- Di essere consapevole che la partecipazione allo studio è volontaria, con l'assicurazione che il rifiuto a partecipare non influirà nel ricevere il trattamento più idoneo.
- Di essere consapevole che la mia firma a questo Consenso Informato non costituisce una rinuncia ai miei diritti legali o liberagli sperimentatori dalle loro responsabilità legali e professionali.
- Di essere stato assicurato che:
 - Potrò interrompere la mia partecipazione allo studio in qualsiasi momento lo dovessi ritenere opportuno, senza l'obbligo da parte mia di motivarne la decisione.
 - Sarò informato di eventuali variazioni del protocollo che possano influenzare la mia partecipazione.

- E' mio diritto accedere alla documentazione che mi riguarda e alla valutazione espressa dal Comitato Etico (Comitato Etico Fondazione F.A.R.O. onlus) cui potrò rivolgermi se lo riterrò opportuno.
- Una copia del consenso informato e della documentazione di cui ho preso visione rimarrà in mio possesso.
- Per ogni problema o per eventuali ulteriori informazioni potrò rivolgermi al Medico Sperimentatore Dott VERONESE SIMONE
Indirizzo di lavoro FONDAZIONE FARO ONLUS recapito telefonico 011.888.272

Pertanto,
confermo di avere avuto risposte esaurienti a tutti i miei quesiti e, preso atto della situazione illustrata

ACCONSENTO

LIBERAMENTE, SPONTANEAMENTE E IN PIENA COSCIENZA ALLA RICERCA PROPOSTAMI.

Dichiaro inoltre di essere a conoscenza della possibilità di revocare il presente consenso in qualsiasi momento prima dell'avvio dello studio.

Data

.....
Nome e cognome del paziente

.....
firma del paziente

NON ACCONSENTO

LIBERAMENTE, SPONTANEAMENTE E IN PIENA COSCIENZA ALLA RICERCA PROPOSTAMI.

Data

.....
Nome e cognome del paziente

.....
firma del paziente

DICHIARAZIONE DELLA PERSONA CHE ASSISTE (se presente)

Io ho letto e capito tutte le informazioni disponibili sullo studio, sulle procedure che il paziente deve seguire e il mio contributo come persona che assiste.

.....
.....
Nome e cognome della persona che assiste il paziente firma della persona che assiste il paziente data

DICHIARAZIONE DELLO SPERIMENTATORE:

Io sottoscritto Dr VERONESE SIMONE.(nome dello sperimentatore) dichiaro:

- Di avere ricevuto dal Comitato Etico del Centro l'autorizzazione ad effettuare, secondo la normativa attuale, questo studio nell'uomo.
- Di avere ottenuto il presente consenso dopo avere esaurientemente spiegato il Consenso Informato in tutte le sue parti al paziente che parteciperà allo studio e di essermi assicurato che tali spiegazioni siano state comprese dal paziente.
- Di avere consegnato copia del Consenso Informato, firmato e datato dal paziente e copia della lettera per il medico curante.
- Che copia del Consenso Informato, firmato e datato, sarà conservata al Centro nella documentazione relativa a ciascuno persona partecipante allo studio.

SIMONE VERONESE
Nome e cognome dello sperimentatore firma dello sperimentatore

18153
N° iscrizione all'ordine data

8.2.6 Participant's consent form English Version

PARTICIPANTS' CONSENT FORM MODULE

**EXPLORATIVE RANDOMIZED AND CONTROLLED STUDY TO
EVALUATE THE IMPACT OF A SPECIALIST PALLIATIVE CARE
SERVICE (SPCS) ON THE PALLIATIVE CARE OUTCOMES (PCO) FOR
PATIENTS WITH NEURODEGENERATIVE CONDITIONS**

Main researcher: DR VERONESE SIMONE

I undersigned MR/MRS.....born in.....
date.....address.....Telephone
number.....

DECLARE

- To participate voluntarily at the present study aimed at assessing the impact of a new SPCS for people severely affected by neurodegenerative conditions at home or in the hospice.
- To have received by Dr Veronese exhaustive explanations about the request to participate, in particular about the aims and the methods of this study.
- To have had enough time to read carefully the participants' leaflet, understand it and ask about its content. I have been reassured that this research is conducted according to the International Ethics Guidelines on research.
- To have had time to make questions and have received satisfactory answers about all the research project.
- To have been informed about the potential inconvenient that might happen during the study.
- To give my consent / to withhold my consent to inform my GP of my decision to participate at his study.
- To be informed that my participation is voluntarily and that in case of non participation I will not be discriminated in the care and assistance.
- To be conscious that signing this document I am not renouncing to my legal rights of patient and I am not liberating the researchers by their professional and legal responsibility.
- To have be reassured that I can withdraw my consent at any time during the study and that I will not have the obligation to motivate this decision.
- To have been informed that I will be informed of any variation of the study protocol.
- To be aware that I can access to the FARO Fondation EC, and that I can access to my documentation whenever I decide to.
- To have received a copy of the documents that I signed (consent form and participants leaflet)
- To be aware that Dr Veronese is the responsible for this research and that I can contact him through the FARO Foundation at the telephone number 011.888.272

For the previously listed reasons I:

CONSENT

LIBERALLY, SPONTANEOUSLY AND FULLY CONSCIOUSLY TO THIS RESEARCH PROJECT.

My consent is valid now and can be withdrawn at any time before or during the study

DATE.....

SIGNATURE.....

DO NOT CONSENT

LIBERALLY, SPONTANEOUSLY AND FULLY CONSCIOUSLY TO THIS RESEARCH PROJECT.

DATE.....

SIGNATURE.....

DECLARATION OF THE FAMILY CARER (if present)

I read and understood all the available information of this study, its procedures and the role of my contribute

.....

Name and Surname of the family carer

Signature

DECLARATION OF THE MAIN RESEARCHER

I UNDERSIGNED Dr Simone Veronese declare:

- To have obtained by the FARO EC the authorization to undertake, according to the normative law on force, this study on the humans.
- To have obtained the present consent from the participants after having provided accurate information about the study and the consent form.
- To have let a copy of the consent form to the participant with a letter for the GP.
- To keep safe a copy of the documentation and of the consent form of all the participants.

In faith,

Dr Simone Veronese

Turin's Order of Physicians

Registration number:

18153

8.3 Evaluation tools

The tools used to assess the outcome measures in the quantitative study are appended in the next pages. They are reported both in the Italian version, as used in the participants' CRF, and then translated in English.

8.3.1 The SEIQoL-DW Italian version

FOGLIO DI DEFINIZIONE DELLE AREE (CUES)

DESCRIZIONE DELL'AREA

ETICHETTA

1. _____

2. _____

3. _____

4. _____

5. _____

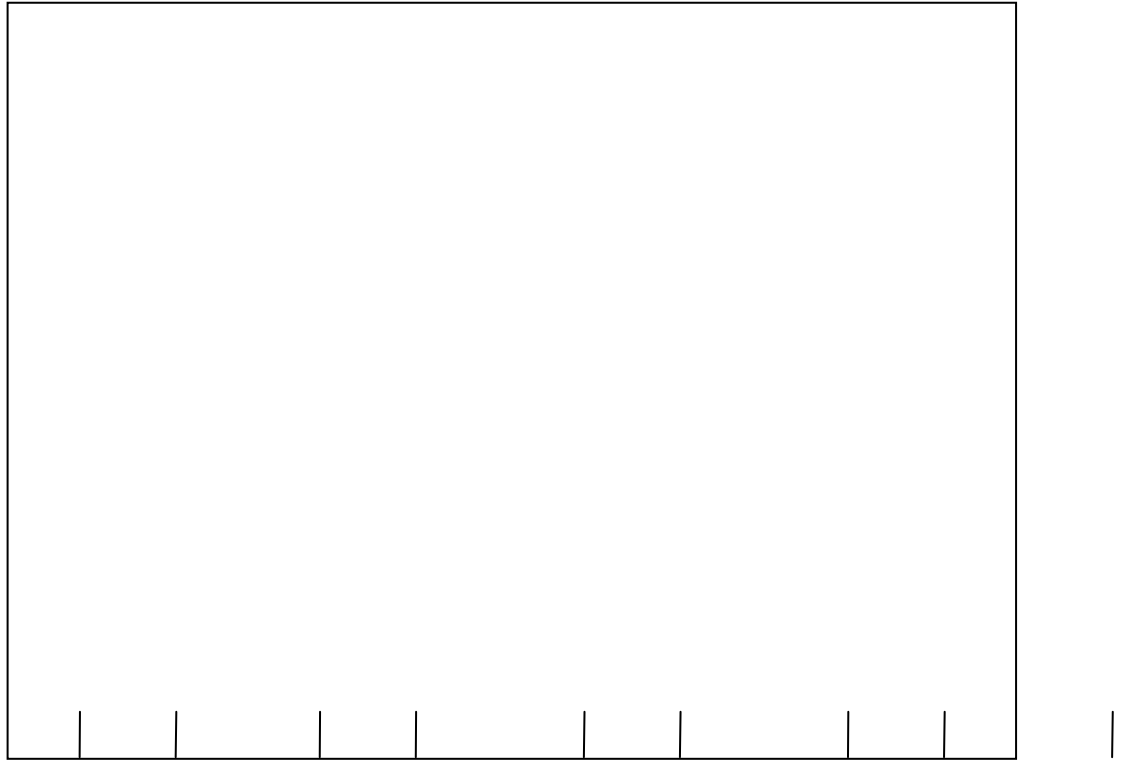
SPUNTARE OGNI ETICHETTA LEGGENDONE LA LISTA AL PARTECIPANTE

FOGLIO DI ESEMPIO DI RILEVAZIONE DEL LIVELLO DI SODDISFAZIONE

Miglior possibile	
Molto bene	
Bene	
Né bene né male	
Male	
Molto male	
Peggior possibile	

FOGLIO DI RILEVAZIONE DEI LIVELLI DI SODDISFAZIONE PER IL PAZIENTE

Meglio possibile



Peggio possibile

FOGLIO DI RILEVAZIONE DATI

1. TEMPO RICHIESTO _____
2. LIVELLO DI COMPrensIONE DEL METODO
 - NON COMPRESO DEL TUTTO
 - SCARSO \ INCERTO
 - BEN COMPRESO
3. AFFATICAMENTO \ ANNOIAMENTO
 - NESSUNO
 - UN PO'
 - MOLTO
4. LIVELLO GENERALE DI VALIDITA' DELLE INFORMAZIONI (alla luce di 2. & 3.)
 - DECISAMENTE NON VALIDE
 - INCERTO
 - VALIDE
5. PESI ASSEGNATI ALLE AREE:
 - AREA 1: PESO (%): _____
 - AREA 2: PESO (%): _____
 - AREA 3: PESO (%): _____
 - AREA 4: PESO (%): _____
 - AREA 5: PESO (%): _____

8.3.2 The SEIQoL-DW English version

CUE DEFINITIONS RECORD FORM

DESCRIPTION OF CUE

CUE LABEL

1. _____

2. _____

3. _____

4. _____

5. _____

(Tick any cues elicited by reading list to person)

SAMPLE CUE LEVELS RECORD FORM

BEST POSSIBLE

VERY GOOD

GOOD

NEITHER GOOD NOR BAD

BAD

VERY BAD

WORST POSSIBLE

The form consists of a large empty rectangular box. On the left side of the box, there are seven vertical tick marks, each aligned with one of the seven quality level labels. At the bottom of the box, there is a horizontal line with several vertical tick marks extending downwards from it, serving as a scale or grid for recording data.

CUE LEVELS RECORD FORM

BEST POSSIBLE

A large empty rectangular box for recording data. The bottom edge of the box is marked with 10 vertical tick marks, indicating a scale or categories. The box is currently empty.

WORST POSSIBLE

INTERVIEW RECORD FORM

1. TIME TAKEN _____

2. UNDERSTANDING OF METHOD

- Not Understood
- Poor/Uncertain Understanding
- Understood

3. FATIGUE/BOREDOM

- None /
- Some /
- A Lot /

4. OVERALL VALIDITY OF INFORMATION (in light of 2 & 3 above)

- Definitely Invalid
- Uncertain
- Valid

5. WEIGHTS ASSIGNED TO CUES

- Cue 1: Weight (%) _____
- Cue 2: Weight (%) _____
- Cue 3: Weight (%) _____
- Cue 4: Weight (%) _____
- Cue 5: Weight (%) _____

8.3.3 The CBI Italian version

caregiver burden inventory

Assistente: _____ Paziente assistito: _____

Data: _____

Mai 0 1 2 3 4 **Sempre**

		scelta
1	La persona di cui mi prendo cura ha bisogno del mio aiuto per molte attività quotidiane	
*		
2	La persona di cui mi prendo cura dipende da me	
*		
3	Devo seguire costantemente la persona di cui mi prendo cura	
*		
4	Devo aiutare la persona di cui mi prendo cura in molte funzioni essenziali	
5	Non ho un minuto di pausa nelle mie attività di cura	
6	Mi sento tagliato fuori dalla vita	
*		
7	Vorrei poter evadere da questa situazione	
8	La mia vita sociale ne ha risentito	
9	Mi sento emotivamente prosciugato dalle cure che fornisco al mio caro	
10	Mi aspettavo che a questo punto della mia vita le cose sarebbero state diverse	
11	Non dormo abbastanza	
12	La mia salute ne ha risentito	
13	Fornire queste cure mi ha debilitato fisicamente	
*		
14	Sono fisicamente esausto	
15	Non vado più d'accordo con i miei familiari come una volta	

Chapter 8. Appendix 2: Study documentation

16	Gli sforzi che faccio per seguire il mio caro non vengono apprezzati dagli altri familiari	
17	Ho avuto problemi con il mio matrimonio	
18	Non lavoro più bene come prima	
*		
19	Mi sento risentito con gli altri familiari che mi potrebbero aiutare ma non lo fanno in una volta	
20	Mi sento imbarazzato per il comportamento della persona di cui mi prendo cura	
21	Mi vergogno della persona di cui mi occupo	
22	Ho motivi di risentimento nei confronti della persona di cui mi occupo	
23	Mi sento a disagio quando degli amici mi vengono a trovare	
24	I miei rapporti con la persona di cui mi prendo cura mi causano rabbia	
Fattore 1:	<input type="checkbox"/>	(Tempo)
Fattore 2:	<input type="checkbox"/>	(Progresso vita)
Fattore 3:	<input type="checkbox"/>	(Fatica fisica)
Fattore 4:	<input type="checkbox"/>	(Fattori sociali)
Fattore 5:	<input type="checkbox"/>	(Fattori emotivi)

Chapter 8. Appendix 2: Study documentation

15	I don't get along with other family members as well as I used to	
16	My caregiving efforts aren't appreciated by others in my family	
17	I've had problems with my marriage	
18	I don't do as good a job at work as I used to	
*		
19	I feel resentful of other relatives who could but do not help	
20	I feel embarrassed over my care-receiver's behaviour	
21	I feel ashamed of my care-receiver	
22	I resent my care-receiver	
23	I feel uncomfortable when I have friends over	
24	I feel angry about my interactions with my care-receiver	
Factor 1:	<input type="checkbox"/>	(Time-dependence burden)
Factor 2:	<input type="checkbox"/>	(Developmental burden)
Factor 3:	<input type="checkbox"/>	(Physical burden)
Factor 4:	<input type="checkbox"/>	(Social burden)
Factor 5:	<input type="checkbox"/>	(Emotional burden)

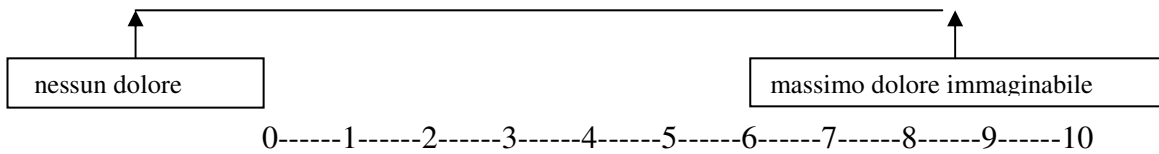
8.3.5 The VAS – NRS used to assess physical symptoms, psychological, social and spiritual issues: the Italian version

SINTOMI FISICI

DOLORE:

Domanda del ricercatore:

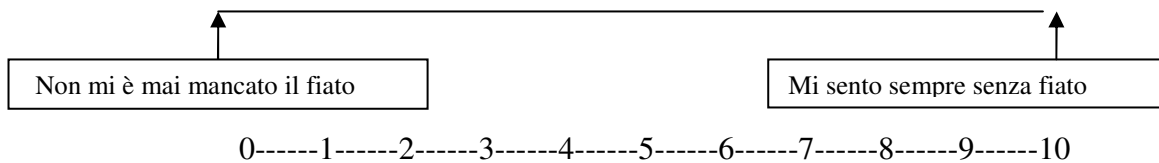
Nell'ultima settimana dovendo definire il livello di dolore fisico da lei provato in un intervallo compreso tra 0= nessun dolore e 10= il massimo dolore immaginabile, come definirebbe il suo dolore?



DISPNEA:

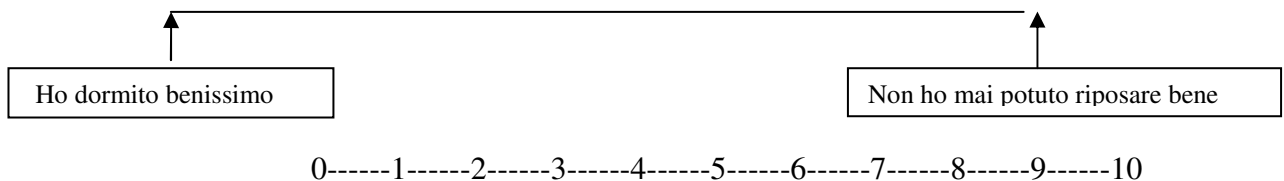
Domanda del ricercatore:

Nell'ultima settimana dovendo descrivere la sua respirazione nell'ultima settimana come la definirebbe in questo intervallo?



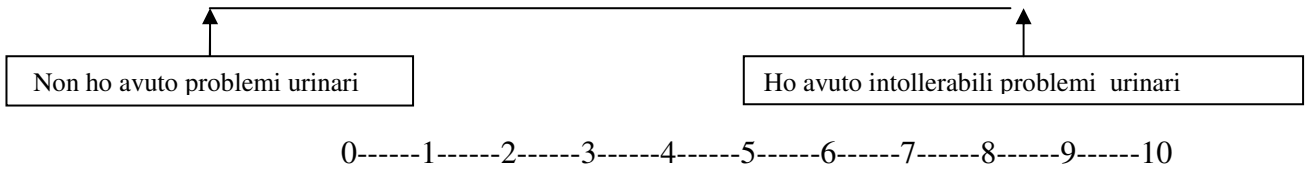
QUALITA' DEL SONNO

Nell'ultima settimana come descriverebbe la qualità del suo sonno?



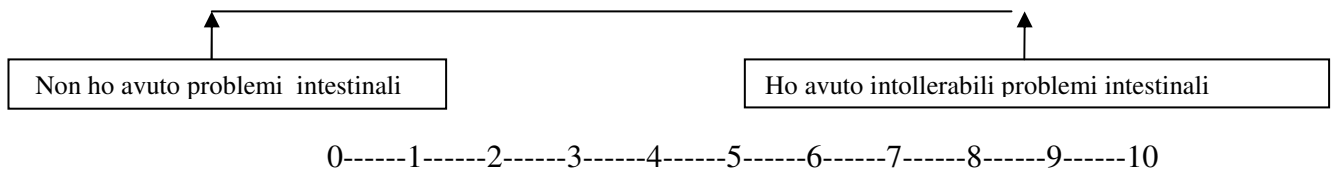
DISTURBI URINARI

Nell'ultima settimana come descriverebbe il controllo delle funzioni urinarie?(suggerimenti: incontinenza?difficoltà ad urinare?)



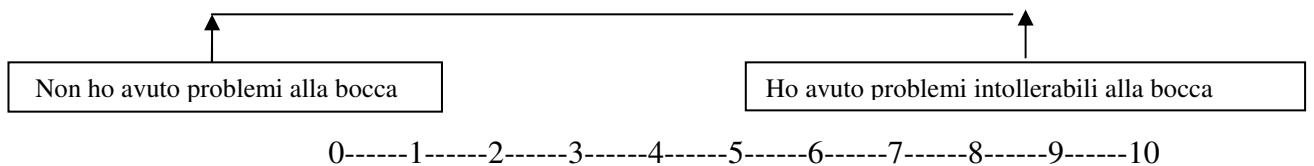
DISTURBI INTESTINALI

Nell'ultima settimana come descriverebbe il controllo delle funzioni intestinali?(suggerimenti: stitichezza? diarrea?)



DISTURBI DEL CAVO ORALE

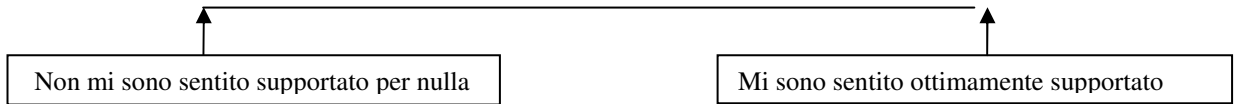
Nell'ultima settimana come descriverebbe lo stato della sua bocca?(suggerimenti: bocca asciutta? Perdita di saliva?)



ASPETTI PSICOLOGICI:

Livello di supporto psicologico:

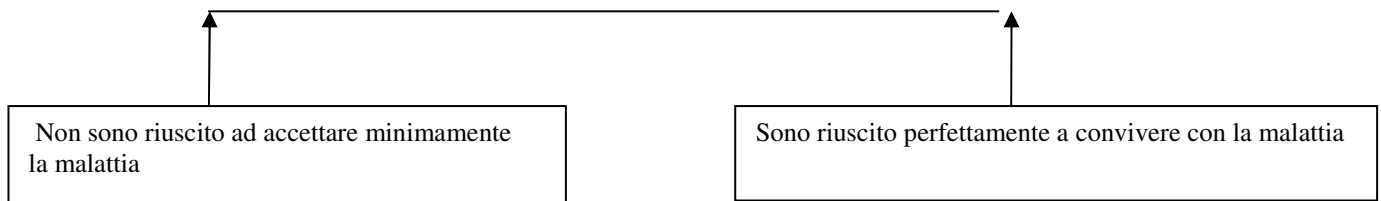
Come definirebbe il livello di supporto psicologico da lei ricevuto nell'ultima settimana?



0-----1-----2-----3-----4-----5-----6-----7-----8-----9-----10

Capacità di affrontare la malattia (livello di "Coping")

Come le sembra di avere affrontato i problemi dovuti alla sua malattia?

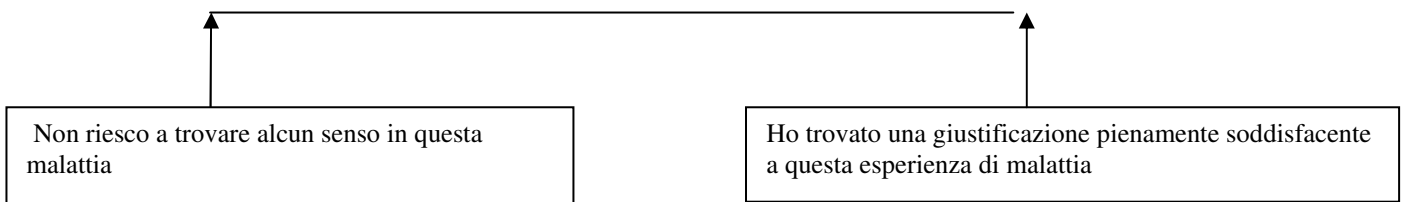


0-----1-----2-----3-----4-----5-----6-----7-----8-----9-----10

ASPETTI SPIRITUALI

Esplorare il livello di "Significato" dell'esperienza di malattia:

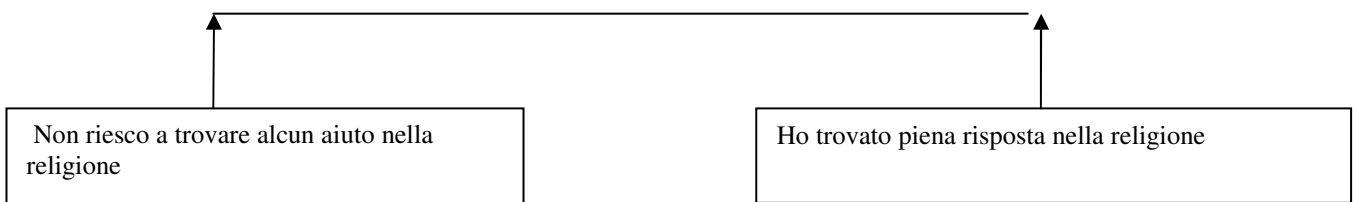
Se le chiedessi di indicare la sua valutazione rispetto al Significato che l'esperienza di malattia che lei sta vivendo ha rispetto alla sua vita, come la definirebbe?



0-----1-----2-----3-----4-----5-----6-----7-----8-----9-----10

Valutare l'importanza della religiosità rispetto al "Significato" della malattia

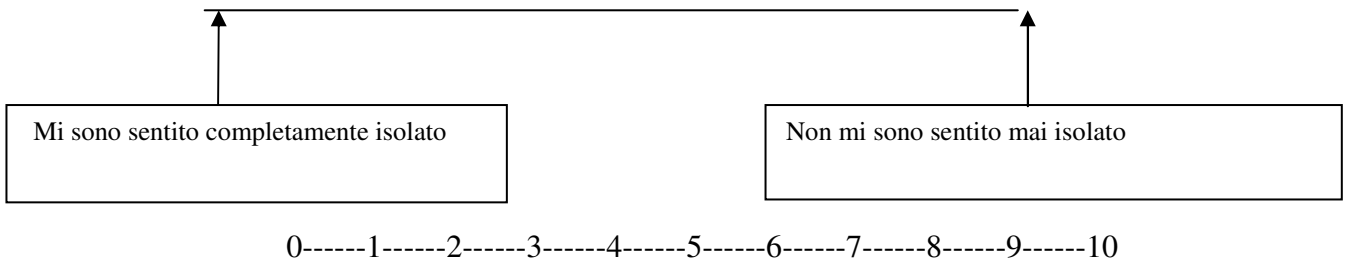
Se le chiedessi di indicare il livello di aiuto che lei riceve dalla religione nell'affrontare questa malattia, come la definirebbe?



0-----1-----2-----3-----4-----5-----6-----7-----8-----9-----10

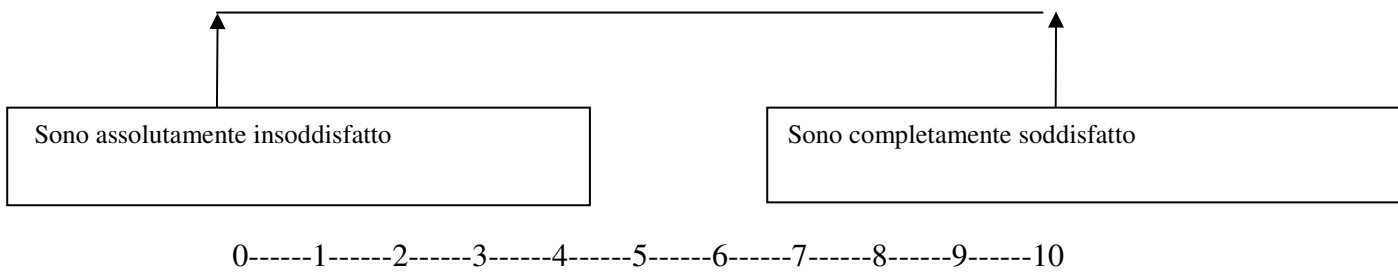
ASPETTI SOCIALI

Valutare il livello di isolamento sociale percepito:
Si è sentito isolato a causa dei problemi dovuti alla sua malattia?



Livello di soddisfazione generale dei servizi ricevuti

Può indicare il suo grado di soddisfazione generale rispetto ai servizi che sta ricevendo?



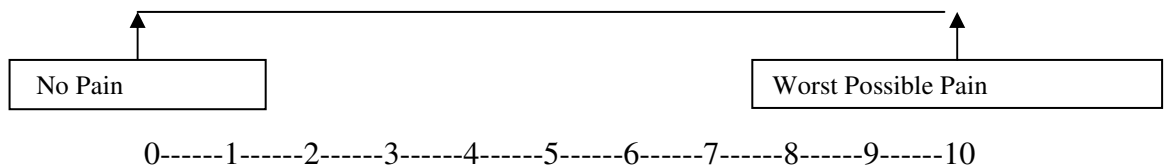
8.3.6 The VAS – NRS used to assess physical symptoms, psychological, social and spiritual issues: the English version

PHYSICAL SYMPTOMS

PAIN

Question:

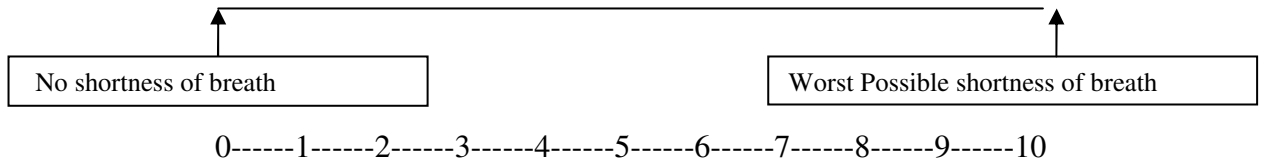
How would you rate the intensity of your pain in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means no pain; an intensity of 10 means pain as bad as you can imagine. Please rate your pain by indicating the number that best describes your pain.



DYSPNOEA

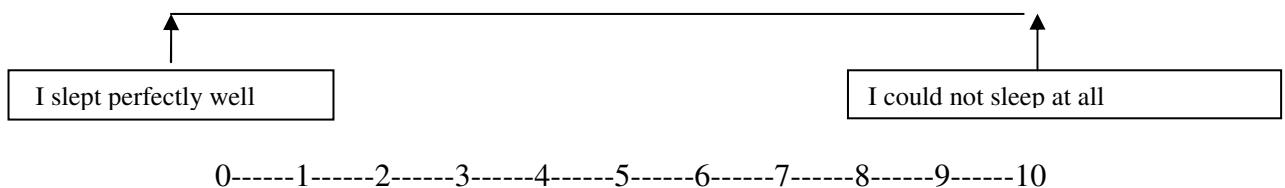
Question:

How would you rate the intensity of your shortness of breath (SOB) in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means no SOB; an intensity of 10 means SOB as bad as you can imagine. Please rate your SOB by indicating the number that best describes your shortness of breath.



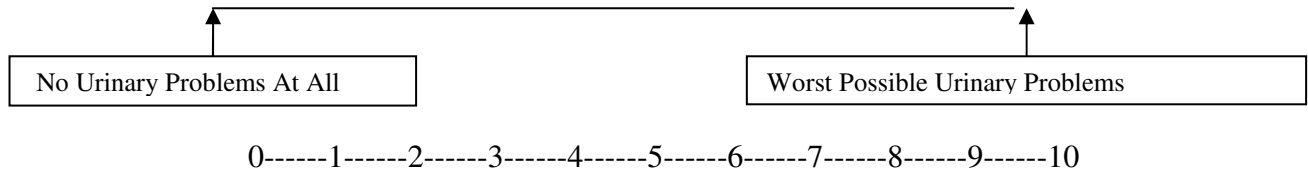
QUALITY OF SLEEP

How would you rate the quality of your sleep (QoS) in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means perfect QoS; an intensity of 10 means absolute impossibility to sleep. Please rate your QoS by indicating the number that best describes your QoS



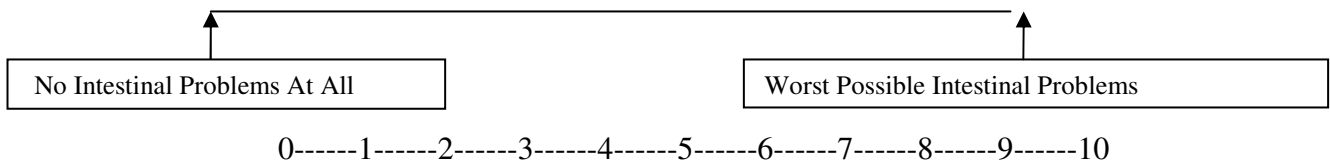
URINARY PROBLEMS

How would you rate your urinary problems in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means did not have any urinary problem; an intensity of 10 means urinary problems as bad as you can imagine. Please rate your urinary problems by indicating the number that best describes your urinary problems



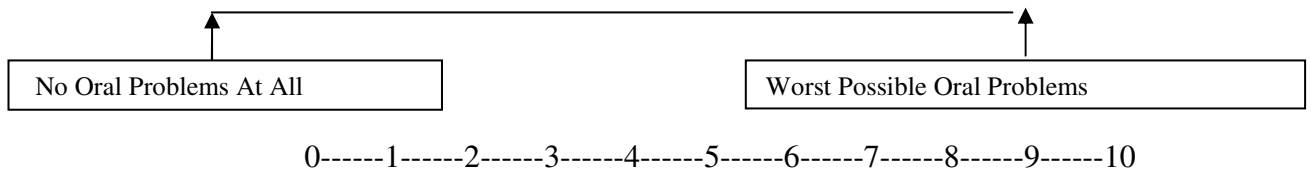
INTESTINAL PROBLEMS

How would you rate your intestinal problems in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means did not have any intestinal problem; an intensity of 10 means intestinal problems as bad as you can imagine. Please rate your intestinal problems by indicating the number that best describes your intestinal problems



ORAL PROBLEMS

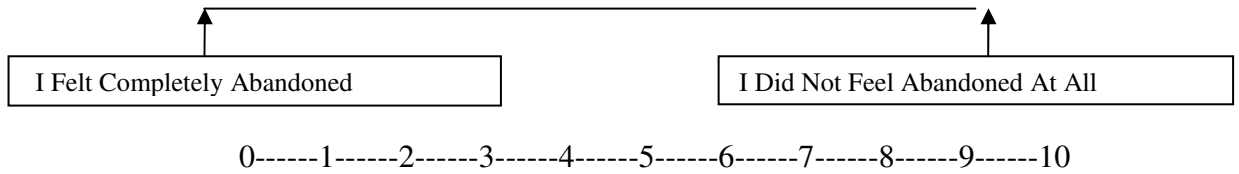
How would you rate your oral problems in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means did not have any problem with your mouth; an intensity of 10 means oral problems as bad as you can imagine. Please rate your oral problems by indicating the number that best describes your oral problems



PSYCHOLOGICAL ASPECTS

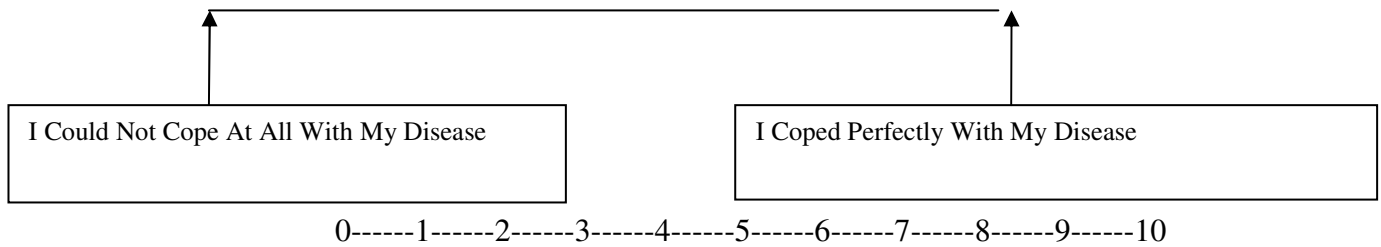
FEELING ABANDONED

How would you rate your feeling of being abandoned in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means you felt completely abandoned; an intensity of 10 means that you did not feel abandoned at all. Please rate your feeling of being abandoned by indicating the number that best describes your condition



COPING WITH THE DISEASE:

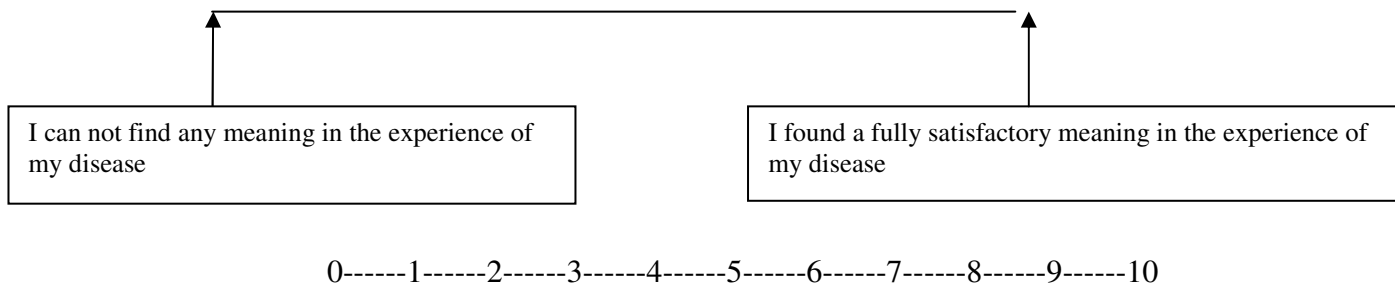
How would you rate your difficulty of coping with your disease in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means you could not cope at all with the problems caused by the disease; an intensity of 10 means that you coped perfectly with your disease. Please rate your difficulty to cope with your disease by indicating the number that best describes your condition



SPIRITUAL ASPECTS

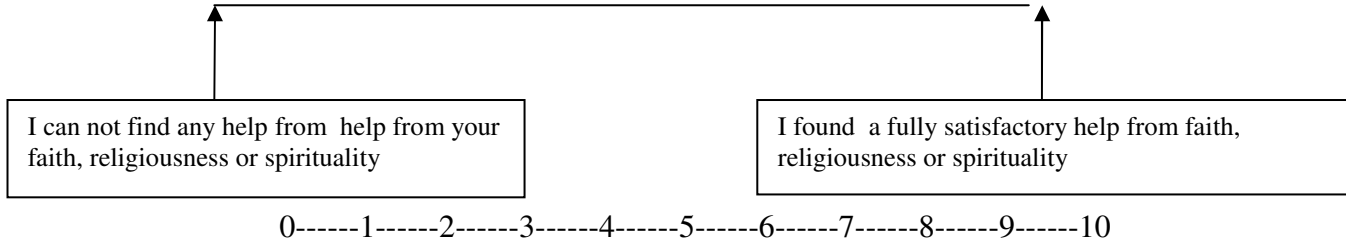
EXPLORING THE MEANING

How would you rate the meaning that you could find in the experience of your disease in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means you did not find any meaning in the experience of your disease; an intensity of 10 means that you found a fully satisfactory meaning in the experience of your disease. Please rate your difficulty find a meaning in the experience of your disease by indicating the number that best describes your condition



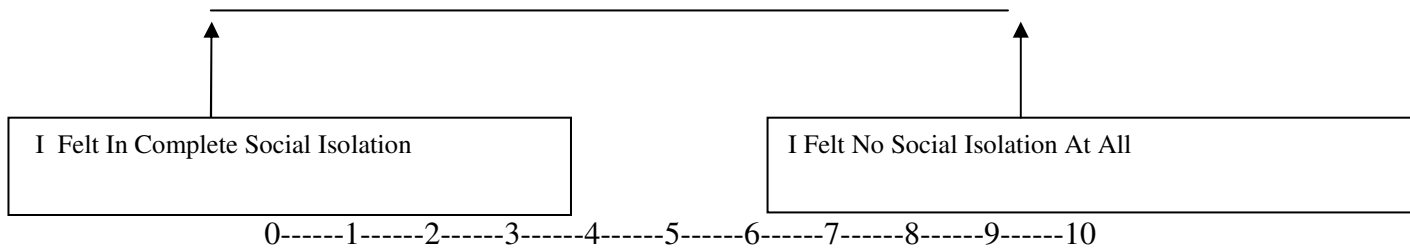
HELP FROM FAITH

How would you rate help that you received from your faith, religiousness or spirituality in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means you did not receive any from your faith, religiousness or spirituality; an intensity of 10 means that you received a fully satisfactory help from your faith, religiousness or spirituality. Please rate your help from your faith, religiousness or spirituality by indicating the number that best describes your condition



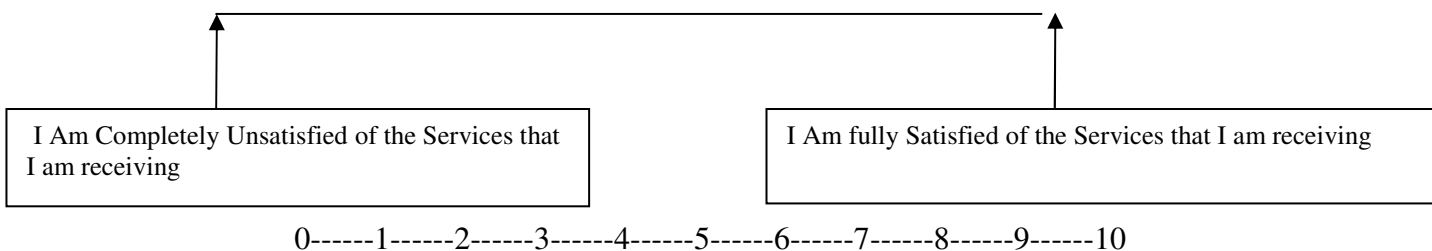
SOCIAL ASPECTS

How would you rate your social relationships in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means you felt totally social isolated; an intensity of 10 means that you felt no social isolation at all. Please rate your social relationships by indicating the number that best describes your condition



SERVICE SATISFACTION


How would you rate your satisfaction about the services that you are receiving in the last week using a scale from 0 (zero) to 10 (ten)? An intensity of 0 means you are completely unsatisfied of the services that you are receiving; an intensity of 10 means that you fully satisfied by the services that you are receiving. Please rate your service satisfaction level by indicating the number that best describes your condition



8.4 Ethics Committees Approvals

8.4.1 San Luigi di Orbassano Hospital Ethics Committee approval

**SERVIZIO SANITARIO NAZIONALE
REGIONE PIEMONTE
Azienda Sanitaria Ospedaliera SAN LUIGI GONZAGA
Regione Gonzole 10 – 10043 ORBASSANO (TO)**



**COMITATO ETICO INTERAZIENDALE
A.S.O. SAN LUIGI GONZAGA DI ORBASSANO**

Prot. n. **8961** Orbassano, - 7 MAG. 2007

Al Direttore Generale
A.S.O. SAN LUIGI GONZAGA
ORBASSANO

PAPEREFAVOREVCL

PRATICA N. 21/INT SEDUTA DEL 23 APRILE 2007

OGGETTO: Studio osservazionale "NeuNeeds"
Promotore: Fondazione FARO – Torino
Sede: S.C.D.U. Neurologia
Sperimentatore: prof. Luca DURELLI

PARERE DIREZIONE SANITARIA: 16 APRILE 2007

ELENCO DOCUMENTAZIONE ESAMINATA:

1. Nota di trasmissione a firma del Responsabile dello studio, sottoscritta da tutti i partecipanti allo stesso
2. Lettera dell'Associazione FARO di Torino
3. Progetto della ricerca
4. Curriculum vitae del/i ricercatore/i, firmato e datato
5. Foglio informativo/Modulo per l'acquisizione del consenso informato per il paziente
6. Foglio informativo/Modulo per l'acquisizione del consenso informato per il familiare
7. Foglio informativo/Modulo per l'acquisizione del consenso informato per partecipanti al focus group
8. Scheda raccolta dati
9. Questionario CRQ-F

Il Comitato Etico dell'A.S.O. San Luigi Gonzaga di Orbassano, costituito con Deliberazione del Direttore Generale n. 81 del 12.02.2007, iscritto nel Registro Regionale dei Comitati Etici con il numero d'ordine 7 nella sezione relativa ai Comitati istituiti ai sensi dell'art. 5 del D.P.G.R. 16.11.2001 della Regione Piemonte e della D.G.R. n.78-4807 del 04.12.2006, nella seduta del 23.04.2007 ha preso in esame la documentazione sopra

Telefono: Centralino 011 9026.1 Segreteria 011 9026.566 E-mail sperimentazioni@sanluigi.piemonte.it 1

**SERVIZIO SANITARIO NAZIONALE
REGIONE PIEMONTE
Azienda Sanitaria Ospedaliera SAN LUIGI GONZAGA
Regione Gonzole 10 – 10043 ORBASSANO (TO)**



**COMITATO ETICO INTERAZIENDALE
A.S.O. SAN LUIGI GONZAGA DI ORBASSANO**

SEGUE: Parere pratica n. 21/INT

descritta, pervenuta in data 16 aprile 2007 e, nell'intesa che lo studio sia condotto in conformità ai principi etici che traggono la loro origine dalla normativa vigente per l'effettuazione delle sperimentazioni cliniche, per quanto di specifica applicazione, ha espresso

PARERE FAVOREVOLE

alla conduzione dello studio osservazionale in oggetto.

In merito dovrà essere informato:

- dell'inizio della sperimentazione e della sua conclusione
- di ogni successivo emendamento e modifica sostanziale del protocollo approvato.

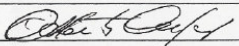

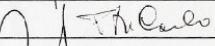

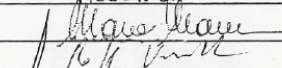
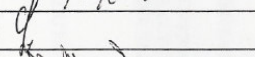
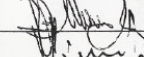
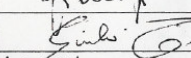



Il responsabile dello studio dovrà, inoltre, far pervenire una relazione annuale sull'andamento dello stesso.

In ogni successiva comunicazione dovrà essere indicato il numero di pratica assegnato a questa sperimentazione.

**IL PRESIDENTE
Prof. Francesco DI CARLO**

Chapter 8. Appendix 2: Study documentation

A.S.O. SAN LUIGI GONZAGA
REGIONE GONZOLE N. 10
10043 ORBASSANO (TO)
COMITATO ETICO INTERAZIENDALE
SEDUTA DEL 23 APRILE 2007

	QUALIFICA		FIRMA
prof. Alberto ANGELI	Clinico di Area Medica - Professore Ordinario di Medicina Interna Università degli Studi di Torino	COMPONENTE	
dott.ssa Elisa Maria BERNO	Biologa Data manager Ospedale Gradenigo - Torino	COMPONENTE	
prof. Francesco DI CARLO	Farmacologo - Professore Ordinario Farmacologia Università degli Studi di Torino	PRESIDENTE	
dott. Giorgio DROETTO	Medico legale - Dirigente medico Dip. Interaz. Medicina Legale ASL 2 e ASL 5	COMPONENTE	
don Paolo FINI	Esperto in Bioetica	VICE PRESIDENTE	ASSENTE
dott. Gabriele GALLONE	Esperto in Biostatistica - Dirigente Medico Direzione Sanitaria di Presidio A.S.O. San Luigi	COMPONENTE	ASSENTE
dott.ssa Mara MANIERO	Rappresentante Professioni Infermieristiche	COMPONENTE	
dott. Umberto ROSSO	Rappresentante delle Associazioni di Volontariato	COMPONENTE	
dott. Roberto SACCO	Direttore S.C. Farmacia A.S.O. San Luigi	COMPONENTE	
prof. Roberto Mario SCARPA	Clinico di Area Chirurgica - Professore Ordinario di Urologia Università degli Studi di Torino	COMPONENTE	
dott. Bruno F. TIRANTI	Esperto di Psicologia - Responsabile U.O.S. Psicologia Età Evolutiva - A.S.L. 5 Col legno	COMPONENTE	
dott. Giulio TITTA	Medico di Medicina Generale A.S.L. 3 - Torino	COMPONENTE	
dott. Massimo VEGLIO	Direttore Sanitario d'Azienda A.S.O. San Luigi di Orbassano	COMPONENTE	

8.4.2 Molinette Hospital Ethics Committee approval



SERVIZIO SANITARIO NAZIONALE
REGIONE PIEMONTE
AZIENDA SANITARIA OSPEDALIERA
"SAN GIOVANNI BATTISTA DI TORINO"
C.so Bramante, 88/90 - 10126 Torino
Cod. Fiscale 05438190018
COMITATO ETICO INTERAZIENDALE
A.S.O. SAN GIOVANNI BATTISTA -
A.S.O. C.T.O./C.R.F./MARIA ADELAIDE

Prot. N. 0046425

Torino, 22 GIU. 2007

Pratica N. CEI/64

Dr. G. Galanzino
Direttore Generale
Azienda Sanitaria Ospedaliera
"San Giovanni Battista" di Torino
SEDE

OGGETTO: Richiesta parere sperimentazione clinica
pervenuta in data 22.05.2007

Prof. A. Chiò
Sperimentatore Principale
S.C. Neurologia I
SEDE

- DOCUMENTAZIONE:
- Protocollo di studio dal titolo: Quali sono i bisogni di cure palliative specialistiche in pazienti affetti da patologie neurodegenerative in fase avanzata? Uno studio qualitativo
- Sponsor: Azienda Sanitaria Ospedaliera "San Giovanni Battista" di Torino
- Allegato 1: Modulo di consenso informato per i pazienti
- Allegato 2: Foglio informativo per il/la paziente (versione 2)
- Allegato 3: Lettera informativa per il medico di medicina generale
- Allegato 4: Modulo di consenso informato per i famigliari
- Allegato 5: Foglio informativo per il famigliare (caregiver) che assiste il paziente (versione 2)
- Allegato 6: Modulo di consenso informato partecipanti ai focus groups
- Allegato 7: Foglio informativo per il/la partecipante al focus group (versione 2=
- Curriculum Vitae dello Sperimentatore Principale
- Collaboratori: Dott.ssa P. Cavalla, Dott. A. Calvo, Dott. M. Vercellino, Dott. S. Veronese, Dott.ssa G. Gallo, Dott.ssa C. Rivoiro
- Richiesta dello Sperimentatore Principale di utilizzare la copertura assicurativa dell'Azienda Sanitaria Ospedaliera "San Giovanni Battista" di Torino
- Dichiarazione pubblica sul conflitto di interessi

Chapter 8. Appendix 2: Study documentation

Il Comitato Etico Interaziendale A.S.O. San Giovanni Battista – A.S.O. C.T.O./C.R.F./Maria Adelaide, istituito in conformità a quanto previsto dal D.M. 12 maggio 2006 e iscritto nel Registro Regionale con determinazione dirigenziale n. 108 del 14.03.2007; nella seduta del 18.06.2007, esaminata la documentazione prodotta, nell'intesa che lo studio clinico sia condotto in conformità ai principi etici che traggono la loro origine dalla Dichiarazione di Helsinki nella sua ultima versione, e che rispetti le GCP e le disposizioni delle normative vigenti, ritiene di esprimere

PARERE FAVOREVOLE.

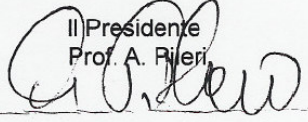
Questa commissione dovrà essere informata dell'inizio della sperimentazione e della sua conclusione od eventuale interruzione nonché di ogni eventuale emendamento al protocollo.

Il responsabile dello studio dovrà, inoltre, far pervenire una relazione annuale sull'andamento dello stesso.

Il Comitato Etico ricorda altresì che deve essere garantito il diritto alla diffusione e/o pubblicazione dei risultati favorevoli o non favorevoli da parte degli sperimentatori che hanno condotto lo studio, nel rispetto delle disposizioni vigenti in tema di riservatezza dei dati sensibili e di tutela brevettuale e che non devono sussistere vincoli di diffusione e pubblicazione dei risultati da parte del Promotore.

In ogni successiva comunicazione dovrà essere indicato il numero di pratica assegnato a questa sperimentazione.

Il Presidente
Prof. A. Pileri



I componenti:

- Prof. A. PILERI in qualità di clinico di area medica (Presente)
- Prof. L. CATTEL in qualità di farmacologo (Assente)
- Dott. O. DAVINI in qualità di direttore sanitario (Presente)
- Dott.ssa S. STECCA in qualità di farmacista (Presente)
- Dott.ssa M. BOERO in qualità di esperto in materie giuridiche (Presente)
- Dott. P. S. BRUNI in qualità di esperto di bioetica (Presente)
- Prof. F. CAMANNI in qualità di clinico di area medica (Presente)
- Dott. G. CARANNANTE in qualità di clinico di area chirurgica (Presente)
- Dott. F. NAPOLETANO in qualità di rappresentante del volontariato (Presente)
- Dott. R. PASSERA in qualità di biostatistico (Presente)
- Sig.ra L. SABA in qualità di rappresentante del settore infermieristico (Presente)
- Dott. F. TALARICO in qualità di medico di medicina generale territoriale (Presente)
- Prof. P. TAPPERO in qualità di medico legale (Presente)
- Dott. A. VENTRE in qualità di psicologo (Assente)

8.4.3 FARO Foundation Ethics Committee approvals

FONDAZIONE F.A.R.O. O.N.L.U.S.
Fondazione Assistenza Ricerca Oncologica Piemonte
Con sede in Torino, Via Cavour, 40 bis
P.IVA 06367710016

Verbale del Comitato Etico del giorno
17 dicembre 2007

Il giorno 17 dicembre 2007, alle ore 17,00 presso la sede legale sita in Torino Via Cavour 40 bis, si è tenuta la riunione del Comitato Etico della Fondazione F.A.R.O. o.n.l.u.s.

Assume la Presidenza la dr.ssa Costanza Calia la quale, con il consenso dei presenti, chiama a fungere da Segretario il dr. Valle Alessandro e constata che:

- sono presenti il dr. Cravetto Giuseppe, il dr. Mirabella Paolo, la dr.ssa Sozzi Marina e il Dr. Zeppegno Giuseppe (hanno giustificato la loro assenza: dr. Peruselli Carlo e dr. Vinciguerra Sergio). Partecipano all'incontro anche il dr. Valle ed il dr. Veronese;
- gli intervenuti si sono dichiarati informati in merito agli argomenti posti all'ordine del giorno;
- il Comitato Etico è pertanto validamente costituito a norma di statuto e quindi atto a deliberare sul seguente

ORDINE DEL GIORNO

- Approvazione progetto pilota destinato all'assistenza dei primi 6 pazienti affetti da patologie neurologiche degenerative in fase avanzata – Proponente: dr. Simone Veronese.

Chapter 8. Appendix 2: Study documentation

- Varie ed eventuali.

Prende la parola il dr. Veronese descrivendo le caratteristiche e finalità del progetto di applicazione delle cure palliative a pazienti affetti da patologie neurologiche degenerative. Traendo spunto da una richiesta telematica del Dr. Peruselli, il dr. Veronese sottolinea il fatto che, essendo lo studio sperimentale, con un risultato in termini di efficacia valutabile solo a posteriori, la Fondazione F.A.R.O. intende farsi carico degli oneri economici dello studio in oggetto, senza gravare sulle risorse del servizio sanitario nazionale.

Il dr. Zeppigno chiede delucidazioni sul fatto che l'ammalato possa fornire consenso/dissenso nell'informare il proprio medico di medicina generale dello studio proposto. Il dr. Veronese risponde affermando che, quantunque si tratti di una evidente criticità, il paziente può riservarsi di informare o meno il proprio medico. In caso negativo, per ovvi motivi, il paziente non verrebbe inserito nello studio.

La cartella clinica utilizzata è quella abitualmente in uso alla Fondazione F.A.R.O.: eventuali necessità di modificare il modello di cartella clinica scaturiranno dallo studio in oggetto.

Su richiesta della dr.ssa Sozzi, il dr. Veronese informa i presenti che tutti gli operatori della Fondazione F.A.R.O. sono stati formati per assistere questa tipologia di ammalati.

L'obiettivo della fase pilota (6 pazienti) oggetto della riunione odierna del Comitato Etico, consiste nella valutazione dei problemi reali delle famiglie assistite e dell'impatto che questa assistenza ha sull'equipe curante F.A.R.O.

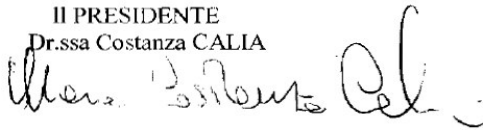
Il dr. Mirabella chiede come verrebbe affrontata un'eventuale discordanza tra il consenso allo studio espresso dall'ammalato e il dissenso della famiglia. Il dr. Veronese afferma che il consenso viene richiesto solo all'ammalato, ma la disponibilità della famiglia è indispensabile per l'attivazione e prosecuzione di un programma di cure palliative domiciliari.

Il dr. Valle ed il dr. Veronese lasciano la sala riunioni per permettere al Comitato Etico di deliberare in merito. Il Comitato Etico approva all'unanimità la fase pilota dello studio.

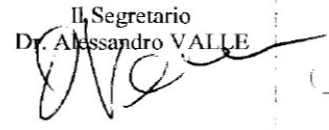
Si dà lettura del verbale che viene condiviso dai membri del Comitato Etico.

La riunione si conclude alle ore 18.45.

Il PRESIDENTE
Dr.ssa Costanza CALIA



Il Segretario
Dr. Alessandro VALLE

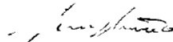


Riunione Comitato Etico del giorno
16 dicembre 2007

MEMBRI

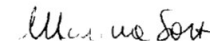
FIRMA

Dr. Cravetto Giuseppe

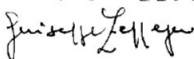


Dr. Mirabella Paolo

Dr.ssa Sozzi Marina



Dr. Zeppigno Giuseppe



Chapter 8. Appendix 2: Study documentation

FONDAZIONE F.A.R.O. O.N.L.U.S.
Fondazione Assistenza Ricerca Oncologica Piemonte
Con sede in Torino, Via Cavour, 40 bis
P.IVA 06367710016

Verbale del Comitato Etico del giorno
3 marzo 2008

Il giorno 3 marzo 2008, alle ore 17.00 presso la sede legale sita in Torino Via Cavour 40 bis, si è tenuta la riunione del Comitato Etico della Fondazione F.A.R.O. o.n.l.u.s.

Assume la Presidenza la dr.ssa Costanza Calia la quale, con il consenso dei presenti, chiama a fungere da Segretario il dr. Valle Alessandro e constata che:

- sono presenti oltre alla di Lei persona il dr. Cravetto Giuseppe, la dr.ssa Marina Sozzi e don Giuseppe Zeppeño. Partecipano all'incontro anche il dr. Valle ed il dr. Veronese;
- gli intervenuti si sono dichiarati informati in merito agli argomenti posti all'ordine del giorno;
- il Comitato Etico è pertanto validamente costituito a norma di statuto e quindi atto a deliberare sul seguente

ORDINE DEL GIORNO

- approvazione protocollo di studio Progetto "NePal";
- varie ed eventuali.

Chapter 8. Appendix 2: Study documentation

Il dr. Veronese prende la parola e aggiorna i presenti sulle prime risultanze scaturite dall'assistenza ai primi sei pazienti neurologici dello studio pilota approvato nel corso dell'ultima riunione del Comitato Etico della Fondazione F.A.R.O. .

Successivamente, il dr. Veronese entra nel dettaglio dello studio randomizzato oggetto dell'incontro odierno, descrivendo le modalità e gli strumenti di misurazione della qualità di vita e di disabilità (fisica e cognitiva) adottati nello studio, ed analizzando i bisogni emersi nella fase preliminare.

I pazienti selezionati verranno randomizzati a ricevere il servizio di assistenza domiciliare della F.A.R.O. subito o dopo 16 settimane (tempo medio per ricevere una consulenza neurologica nel SSN). L'indicazione allo studio scaturisce dall'assenza di evidenze che dimostrino l'efficacia di un servizio di cure palliative domiciliari rispetto ad un servizio di assistenza "tradizionale".

Il dr. Valle precisa che i costi dello studio saranno interamente a carico della Fondazione F.A.R.O. o.n.l.u.s..

La dr.ssa Sozzi chiede se l'assistenza a questi pazienti per un periodo di mesi breve non possa incidere in modo limitato sulla qualità di vita globale delle famiglie assistite. Il dr. Veronese risponde affermando che quanto potrà emergere da questo studio potrebbe in futuro avere una ricaduta anche nella gestione delle fasi più precoci.

Al termine dell'incontro, il Comitato Etico approva all'unanimità l'attivazione dello studio in oggetto.

La riunione si conclude alle ore 18.45.

Il PRESIDENTE
Dr.ssa Costanza CALIA

Il Segretario
Dr. Alessandro VALLE

Riunione del Comitato Etico del giorno
3 marzo 2008

MEMBRI

Zeppegno Don Giuseppe

Mirabella Paolo

Vinciguerra Sergio

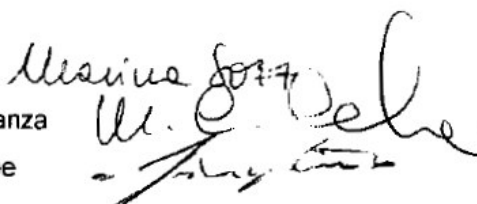
Peruselli Carlo

Sozzi Marina

Calia Maria Costanza

Cravetto Giuseppe

FIRMA



9. Appendix 3: publications

9.1 Research presentations at scientific meetings

- S. Veronese, D.J. Oliver (2007) “*Developing a neurological palliative care service in Turin – a literature review and needs assessment*” Poster. Report 10th Congress of the EAPC Budapest, Hungary, 7-9 June 2007. Published in *Palliative Medicine*, Jun 2007; vol. 21 p.125
- S. Veronese, DJ Oliver (2007) “*Neuneeds: una valutazione qualitativa dei bisogni di cure palliative in pazienti affetti da patologie neurodegenerative e nei loro caregivers*”(Neuneed: a qualitative assessment of palliative care needs for people severely affected by neurodegenerative conditions and their carers) Poster. Published in the Acts of the XIV Congress of Italian Society of Palliative Care (S.I.C.P.), Perugia, 13 - 16 November 2007
- S. Veronese , D.J. Oliver. (2007) “*An international Ph.D. in palliative care: una esperienza formativa e di ricerca possibile per un medico palliativista italiano.*” (An international PhD in palliative care: a concrete research and educational experience for an Italian physician working in palliative care)Poster. Published in the Acts of the XIV Congress of Italian Society of Palliative Care (S.I.C.P.), Perugia, 13 - 16 November 2007
- S.Veronese. (2008) “*Neuneeds: a qualitative assessment for palliative care needs of people severely affected by neurodegenerative conditions*”. Oral Presentation. Report 5th Research Forum of the EAPC, Trondheim, Norway 29-31 May 2008. abstract published in *Palliative Medicine*, June 2008, Volume 22, No. 4, p.435.
- Gallo G. –Veronese S. –Valle A. – Oliver D.J. (2008) “*I bisogni di cure palliative nei pazienti affetti da patologie neurodegenerative: il punto di vista di chi se ne cura.*” (Palliative care needs of people severely affected by neurodegenerative conditions: professional carers’ views) Poster. Acts of the XV Congress of Italian Society of Palliative Care (S.I.C.P.), Giardini Naxos (ME) 3-6/11/2008
- Veronese S., Gallo G., Valle A., Oliver D.J. (2008) “*Quando una equipe di cure palliative si confronta con i malati affetti da sclerosi laterale amiotrofica (sla), sclerosi multipla (sm) malattia di parkinson (pd) e parkinsonismi atipici nasce il bisogno di una formazione specifica: l’esperienza della fondazione faro.*” (Educational needs in a specialist palliative care team facing the needs of people severely affected by amyotrophic lateral sclerosis (ASL/MND), multiple sclerosis (MS), Parkinson’s disease (PD) and related disorders: the F.A.R.O. foundation experience) Poster. Acts of the XV Congress of Italian Society of Palliative Care (S.I.C.P.) Giardini Naxos (ME) 3-6/11/2008. This poster has been awarded with the Ventafridda award as the best poster of the conference.
- Veronese S., Gallo G., Valle A., Oliver D.J. (2008) “*Quando nasce un nuovo servizio di cure palliative per malati neurologici: quali processi seguire? come valutarne gli effetti? l’esperienza della fondazione faro di Torino.*” (A new palliative care service for neurological patients is born: development and evaluation processes. the F.A.R.O. foundation experience). Poster Acts of the XV Congress of Italian Society of Palliative Care (S.I.C.P.) Giardini Naxos (ME) 3-6/11/2008.

Chapter 9. Appendix 3: publications

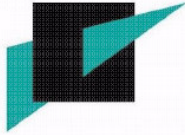
- Veronese S., Gallo G., Valle A., Oliver D.J. (2009) “*The development of a new specialist palliative care service for people severely affected by neurodegenerative conditions in Italy*”
Poster 11th Congress of the European Association for Palliative Care (EAPC) Vienna, Austria, 7–10 May 2009. Published on the Eur. J. Pall Care: Congress Abstracts Issue. P 173
- Veronese S., Gallo G., Valle A., Oliver D.J. (2009) “*Palliative care needs of people severely affected by neurodegenerative conditions: the views of the professional carers.*” Poster 11th Congress of the European Association for Palliative Care (EAPC) Vienna, Austria, 7–10 May 2009. Published on the Eur. J. Pall Care: Congress Abstracts Issue. P 173
- Veronese S., Gallo G., Valle A., Oliver D.J. (2009) “*A qualitative assessment of the physical symptoms of people severely affected by neurodegenerative conditions: the physical symptoms receiving palliative care.*” Poster 11th Congress of the European Association for Palliative Care (EAPC) Vienna, Austria, 7–10 May 2009. Published on the Eur. J. Pall Care: Congress Abstracts Issue. P 173
- Simone Veronese, Gloria Gallo, Chiara Rivoiro, Alessandro Valle, David J. Oliver. (2009) “*Ne-pal: uno studio esplorativo randomizzato e controllato (rct) per valutare l'impatto di un servizio di cure palliative in malati affetti da patologie neurodegenerative. (Ne-pal: a randomized controlled study to evaluate the impact of a specialist palliative care service for patients severely affected by neurodegenerative conditions)*” Poster Acts of the XVI Congress of the Italian Society of Palliative Care (S.I.C.P.) Lecce 27-30/10/2009.
- S. Veronese, G. Gallo, C. Rivoiro, A. Valle. (2009) “*Il servizio di cure palliative F.A.R.O. verso con una nuova sfida: malati oncologici e neurologici in fase avanzata a confronto*” (*F.A.R.O. specialist palliative care service towards a new challenge: advanced oncological and neurological patients and differences in service provision*) Poster Acts of the XVI Congress of the Italian Society of Palliative Care (S.I.C.P.) Lecce 27-30/10/2009.
- S. Veronese, G. Gallo, C. Rivoiro, A. Valle, D.J. Oliver (2010)
“*Specialist palliative care service for people severely affected by neurodegenerative conditions: does this make a difference to palliative care outcomes? results of ne-pal - an explorative randomized controlled trial.*” Oral Presentation. Report 6th Research Forum of the EAPC in Glasgow 9-12/6/2010. Abstract published in Palliative Medicine Vol. 24 Supplement, Number 4 June 2010. P. S24.
- S. Veronese, G. Gallo, C. Rivoiro, A. Valle, D.J. Oliver (2010)
“*Extending palliative care to non cancer patients: challenges and resources*”
Poster. Report 6th Research Forum of the EAPC in Glasgow 9-12/6/2010. Abstract published in Palliative Medicine Vol. 24 Supplement, Number 4 June 2010. P. S183.

9.2 Presented posters

The poster related to this thesis project that were presented at international congresses are appended in the following pages

DEVELOPING A NEUROLOGICAL PALLIATIVE CARE SERVICE IN TURIN – A LITERATURE REVIEW AND NEEDS ASSESSMENT

Simone Veronese - FARO Foundation, Turin, Italy and PhD student
David Oliver - Honorary Senior Lecturer, Kent Institute of Medicine and Health Sciences
University of Kent, UK



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BACKGROUND: There is increasing involvement of palliative care within neurology but there is a little evidence for the efficacy of Specialist Palliative Care Services (SPCS) in

AIMS: Three groups of patients with advanced neurological disease will undergo assessment of their needs: ALS (amyotrophic lateral sclerosis), MS (multiple sclerosis), and PD (Parkinson's disease and related disorders). A new SPCS which aims to meet the needs of neurological patients is being developed and a formal evaluation will be undertaken. This will be undertaken as PhD project at the University of Kent

METHODS: a literature review has been performed showing the following needs and barriers to palliative care provision to these patients

Physical needs

These vary with different diagnoses but common themes are seen

- Weakness
- Speech and communication problems
- Dysphagia
- Pain
- Drooling
- Bowel problems—especially constipation
- Urinary problems

- Poor sleep
- Mobility problems

Psychological themes

- Emotional impact of diagnosis
- High major depression rates
- Anxiety
- Loss and uncertainty about changes in their lives
- Disbelief and devastation
- Sadness and relief
- Anger, Denial and psychosexual problems
- Increased suicidal rate
- Request of hastened death

Social needs

- Tracking down services and information
- Support for personal needs in daily living
- Financial support
- Staying connected and fear of isolation
- Patients with advanced disease are hidden to society
- Prolonged caring for others living with neurodegenerative diseases has substantial costs for carers in terms of loss of social support
- Sense of having to fight for everything, particularly services adaptation and welfare benefits
- Support and palliative care services

Caregivers needs

- information
- access to skilled experts for coordinated action and regular review of their needs in later stage of the disease progression
- bereavement care
- support in caregiving tasks at home, above all when symptoms progress, when assistance are prolonged or patients are ventilated or functional impairment is higher
- Caregivers should be viewed as individuals with their own preferences.
- Respite care
- Emotional support

Spiritual needs

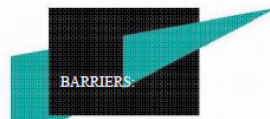
- Worthiness, meaning of life, guilt, hopelessness, sense of being a burden for others.
- Belonging, thankfulness and grace.

Services provided

- Very few palliative care services available for these patients
- Low satisfaction rates in patients and carers about received
- Palliative care service provided more to ALS/MND patients than the other groups
- Services should be tailored and flexible and aimed to find ways to enhance comfort

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- Belief that palliative care should be provided only in the final stages of illnesses
- Difficulty in understanding when patients are terminally ill
- Discomfort in discussing end-of-life issues
- Relief that symptoms are intractable or acceptable
- Ignorance about palliative care
- Neurologists needs of education in palliative care and knowledge about end of life
- Unfamiliarity of the majority of Hospice personnel with these patient's diseases and symptoms
- GP's difficulties to access to resources and to quickly educate themselves in patients management
- Staff training is needed in acute hospital settings to improve knowledge of the diseases

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METHODS:

Needs assessment:

A qualitative study (NEUNEEDS) started in May 2007 in Turin area aimed to assess the needs of patients severely affected by ALS/MND, MS, PD and related disorders, and their carers. Participants are selected by neurologists from 2 major Turin Hospital. Individual semi-structured interviews with patients and carers are performed mostly at patient's home. Focus groups with professional involved are organized in their place of work aimed at understanding their view about patients and carers needs. Data collection will continue until data saturation will be reached. (approximately 20–25 interviews and 3-5 focus groups)

Preliminary results:

To date :
7 patients (3 MS, 2 ALS, 2 PD) and 7 caregivers have been interviewed
1 focus group with 5 neurologists have been performed

Caregivers needs:

- Respite care. Most are exhausted because of the long assistance
- Telephone support by educated professionals in order to be able to manage common problems
- Social help and connection with people happy to help
- Less bureaucracy
- Easy access to aids
- Difficult to find informal paid carers.
- Being alone to manage too many things.

Service satisfaction:

- There are not specific services aimed to satisfy their needs
- Home physiotherapy needs
- Lack of coordination between professionals
- Lack of coordination between patient's associatious volunteers and other services
- Primary domiciliary care not prepared to meet needs other than strictly clinical
- When trouble happens (e.g. infections) patients have to be admitted to hospital where often they are not welcome because of their chronic disease.

Physical needs:

- Lack of symptom assessment and control. In particular uncontrolled pain, constipation, urinary troubles, sleep disturbance
- Disability is a general problem severely affecting Quality of Life
- ALS and MS patients have breathing problems
- High pneumonias and urinary tract infections rates

Psychological needs:

- Feeling isolated
- Fear of losing independence
- Depression
- Anxiety
- Lack of privacy
- Feeling of being an heavy burden
- Feeling unable to communicate their feelings and desires

Social needs:

- Having to fight for everything
- Inadequate social support
- Finance problems
- Difficulty to find paid carers
- Lost jobs
- Friends and relatives tend to vanish when the disease is so long

Spiritual needs:

- Feeling victims of persecution
- Difficult to accept what's happening in life
- Sense of guilt
- Feeling unable to talk to religious assistants
- Need to find different meaning in life

Discussion

These preliminary data confirm that many unmet needs affect quality of life of patients with advanced neurodegenerative conditions and their carers. Data analysis will enable the design of a new specialist palliative care service for patients severely affected by neurodegenerative conditions. This new service will be assessed for its effectiveness and acceptability to patients and their families and professional carers.



Quando una equipe di cure palliative si confronta con i malati affetti da Sclerosi Laterale Amiotrofica (SLA), Sclerosi Multipla (SM) Malattia di Parkinson (PD) e Parkinsonismi atipici nasce il bisogno di una formazione specifica: l'esperienza della Fondazione FARO

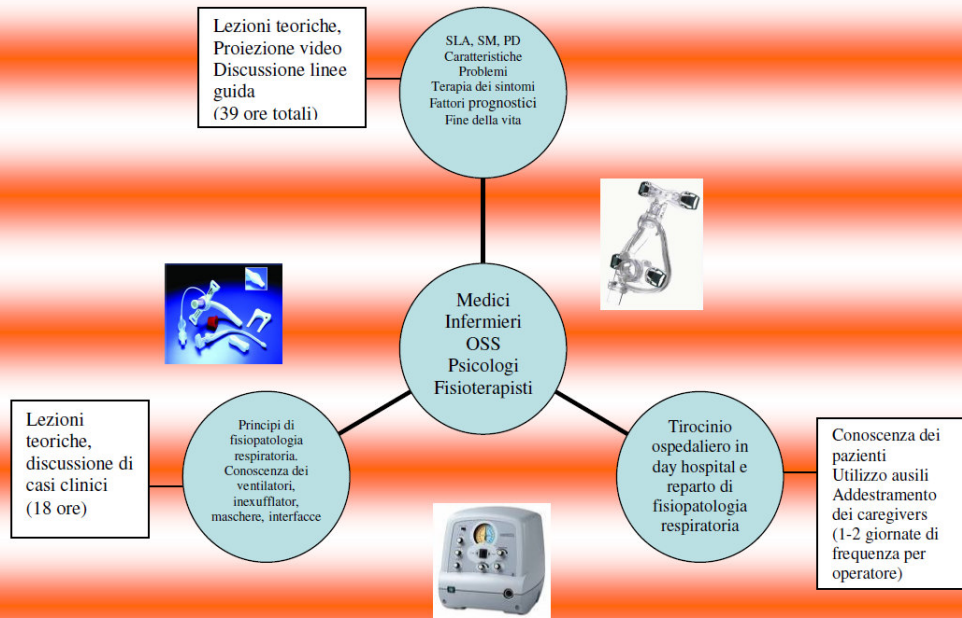
S. Veronese*, G. Gallo*, A. Valle*, DJ Oliver**

* Fondazione F.A.R.O. onlus Torino

** University of Kent at Canterbury (UK)



Dall'analisi della letteratura si evince che la mancanza di conoscenza di patologie come la SLA, SM e PD è spesso causa del fallimento di assistenze in cure palliative, quando non addirittura di interi servizi dedicati. Avendo come obiettivo la creazione di un nuovo servizio di cure palliative domiciliari ed hospice per questi malati, la Fondazione FARO ha organizzato un percorso di formazione dedicato ai suoi operatori.



Il lavoro di formazione di una equipe serve per trasformare il sapere professionale dei suoi componenti, **modellandolo** sui bisogni delle persone di cui ci si dovrà prendere cura



Chapter 9. Appendix 3: publications

Introduzione: I bisogni di cure palliative nei pazienti affetti da patologie neurodegenerative sono molti e non tutti conosciuti. La visione degli stessi da parte dei professionisti che se ne prendono cura non è chiara e non esistono molte pubblicazioni in merito.

Metodi: Nell'ambito di un progetto mirato allo sviluppo di un nuovo servizio di cure palliative specialistiche per tali pazienti è stato condotto uno studio qualitativo che, attraverso 3 focus groups, ha permesso di intervistare 8 neurologi, 1 fisiatra esperto di ventilazione assistita, 1 fisioterapista ed 1 logopedista coinvolti nella cura della Sclerosi Laterale Amiotrofica, Sclerosi Multipla, Malattia di Parkinson e parkinsonismi.



FOCUS GROUPS n=3	Ospedali Molinette e S. Luigi
partecipanti focus groups n=11	esperti in
neurologi n= 8	SLA n=3 (NSLA1-3) SM n=4 (NSM 1-4) MdP n=2 (NMdP 1-2) Pall. Care n=1 (NPC 1)
medico fisiatra n=1	respirazione assistita (NIV \ IV) (MF 1)
fisioterapista n=1	riabilitazione motoria e respiratoria (FKT 1)
logopedista n=1	linguaggio e deglutizione (L1)

Risultati: Tra i risultati emersi dallo studio si rileva la consapevolezza della:

Gravità dei sintomi – (NSM2): *"I bisogni fisici sono molto evidenti, tutti sono trattati per il controllo dei sintomi..."*

(NSM1): *"La fatica è un sintomo molto invalidante e la depressione è sempre presente"*

(NPC1): *"La demenza ha un pessimo impatto sulla famiglia"*

(NSLA1): *"In SLA la sofferenza fisica è molto alta, la scialorrea è un sintomo molto stressante..."*

Bisogni psicosociali – (NSM1): *"Un problema frequente sono i pazienti abbandonati dal proprio sposo"*

(NSM2): *"Problemi relativi al lavoro"*

(NSLA1): *"Nessun servizio di trasporto è previsto per SLA, i benefit sociali sono difficili da ottenere"*

(NMdP1): *"Non ci sono specifici servizi di trasporto per il parkinsoniano, se hanno bisogno di una autoambulanza se la pagano"*

Difficoltà di aiutare a domicilio i propri pazienti – (NMdP2): *"Quando i pazienti non possono più venire in ambulatorio non sappiamo più cosa gli succede". "Alcune volte cerchiamo di trattare le complicanze al telefono, confidando nel MMG, ma non è facile"*

(NSLA1): *"Il problema è che i nostri psicologi non possono uscire dall'ospedale"*

(MF1): *"Spesso i pazienti vengono qui, perché noi siamo qui, raramente vado a visitarli a casa"*

Scarsa conoscenza della terapia del dolore e dell'uso degli oppioidi – (MF1): *"Ne ho sentito parlare, ma non abbiamo esperienza"*

(NMdP1): *"Nella MdP la morfina è scarsamente usata"*

(NSLA1): *"Gli oppioidi sono usati nella SLA anche se per il dolore non sono molto rilevanti".*

"Per il controllo della dispnea non usiamo morfina, perché il sintomo è progressivo, non è segnalato nelle linee guida", così la usiamo solo nella sedazione terminale"
** (vedi linee guida EFNS – Andersen et al. 2005)*

Tutti i partecipanti si sono detti favorevoli allo sviluppo di un servizio di cure palliative domiciliari per migliorare la qualità della vita dei malati e delle famiglie, per aiutare e sostenere nelle scelte terapeutiche e di fine vita, per ridurre le ospedalizzazioni inappropriate. Anche il coinvolgimento dell'Hospice, soprattutto per ricoveri di sollievo e sostegno alla domiciliarità è stato visto come utile.

Conclusioni: Noi ricercatori con i partecipanti auspichiamo lo sviluppo di una rete integrata tra l'ospedale, le cure domiciliari e le cure palliative, con la creazione di percorsi specifici per patologie e condizioni che possano rispondere adeguatamente ai bisogni di cure palliative dei malati.

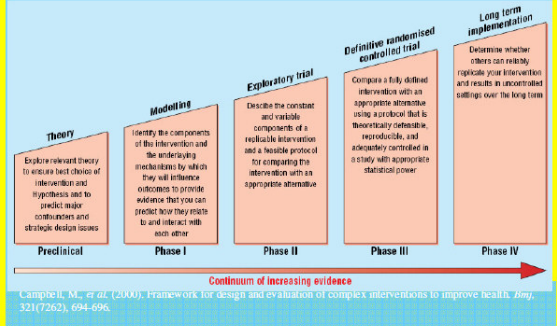


Quando nasce un nuovo servizio di cure palliative per malati neurologici:
 Quali processi seguire? Come valutarne gli effetti?
 L'esperienza della Fondazione FARO di Torino.
 S. Veronese*, G. Gallo*, DJ Oliver**, A. Valle*
 * Fondazione F.A.R.O. onlus Torino
 ** University of Kent at Canterbury (UK)



Fase 1 preclinica (Theory):

- studio del problema teorico con una **revisione della letteratura**
- **l'analisi dei bisogni** attraverso uno studio qualitativo nel quale sono stati intervistati 22 pazienti affetti da Sclerosi Laterale Amiotrofica (SLA), Sclerosi Multipla (SM), Malattia di Parkinson (PD) e parkinsonismi ed i loro famigliari di riferimento e 3 focus groups con neurologi ed altri professionisti coinvolti nelle cure a tali malati.
- Dall'analisi di tali bisogni è emersa la necessità di effettuare un **programma di formazione specifico** per gli operatori di cure palliative che dovranno occuparsi di questi malati. Tale corso è stato completato da tutti gli operatori della Fondazione.



La Fase 2 (Modelling):

- sperimentazione clinica del servizio su un piccolo campione di malati **per valutare l'impatto sull'equipe** di cure palliative:

6 pazienti assistiti per 4 mesi a domicilio ed in hospice tra novembre 2007 e marzo 2008.

- 2 SLA
- 2 SM
- 1 PD
- 1 MSA

La fase 3 (Exploratory trial), ora in atto

- **trial pilota, randomizzato e controllato** nel quale il servizio in esame venga offerto ad un campione di 50 malati (e loro caregivers) con diagnosi di SLA, SM, PD e Parkinsonismi che sono randomizzati in:
- Un gruppo (Fast Track) che riceve il servizio da subito (25 pazienti)
- Un gruppo di controllo (Standard Track) riceverà il servizio stesso, ma solo dopo un periodo di attesa di 16 settimane (25 pazienti)
- Test QoL a pazienti e caregivers
- L'obiettivo è valutare l'impatto del servizio sui Palliative Care Outcomes dei partecipanti. Tale fase di studio è iniziata ad aprile 2008 e si protrarrà fino a febbraio 2009.

The development of a new specialist palliative care service for people severely affected by neurodegenerative conditions in Italy.

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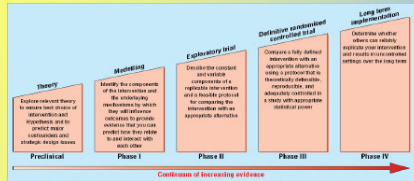
University of Kent

Introduction

The Palliative Care needs of non cancer patients and the effectiveness of palliative care interventions and services have received little research attention. Research and evaluation must play a bigger role in the development of palliative care for non cancer patients than they have in palliative care in general (1). This study shows the development of the specialist palliative care service (SPC) in Turin, Italy, with the new involvement of the care of people with neurodegenerative conditions.

Methods

To address the specific difficulties in defining, developing, documenting, and reproducing complex interventions that are subject to more variation than a drug Irene Higginson reported a novel protocol for the evaluation of a new palliative care service for patients severely affected by Multiple Sclerosis (MS) using the MRC Framework for the Evaluation of Complex Interventions (2)(3).



As a part of a PhD research project a 3 years programme has been developed following the first steps of the MRC framework.

LITERATURE REVIEW

The Ovid Medline, Embase, Cinhalis, Psychinfo and Cochrane library databases were searched (December 2008) looking for relevant papers on this topic. Journals and books were scanned with the aim to find experiences of palliative care services in neurodegenerative disorders, needs assessment and guidelines.

QUALITATIVE ASSESSMENT

22 patients resident in Turin area and severely affected by amyotrophic lateral sclerosis (ALS), multiple sclerosis (MS), Parkinson's disease (PD) and related disorders (PDRD) and their main carers were interviewed and there were 3 focus groups with neurologists and other professionals involved in the care of these patients. Interviews and focus groups were video and audio taped with the aim to catch verbal and non verbal expressions of needs in order to be as accurate as possible in the transcript verbatim. Coding and content analysis of these events were conducted by 2 independent researchers. Inter rater reliability of the content analysis was performed among 3 different researchers.

A PILOT RANDOMIZED CONTROLLED STUDY OF THE NEW SERVICE

A randomised trial of care has been undertaken to evaluate the impact of our new service considering the palliative care outcomes of people severely affected by ALS, MS, PD and PDRD. We conducted randomised controlled trial, of immediate referral to the service (fast-track) versus a 16-week wait (standard best practice). 50 patients, (16 ALS, 18 MS, 12 PD, 2 MSA, 2 PSP) were randomized in two groups: a fast track group (FT) received the service immediately after referral a standard track group (ST) had to wait for a 16 weeks period of time. Both groups were assessed at referrals (T0) and after the 16 weeks (T1). Patients were included if they were considered severely ill by the neurologists and if the clinical prognostic factors published by the Gold Standards Framework for the specific disease group were satisfied (6). Main outcomes were recruitment, attrition and quality of life. Main tools used were SEIQOL-DW (7), HADS (8), CBI (9) and a problem check-list exploring the main needs found out in the previous interviews. Comparison the results of the standardised tests will allow the effectiveness of the new service to be evaluated.

Results

LITERATURE REVIEW revealed several studies focused on needs assessment. Specifically within long-term neurological conditions there is evidence that patients in the final stages neurodegenerative conditions experience very similar symptoms to those observed in cancer patients (4) and experts suggest that the approach to controlling symptoms in progressive non-malignant conditions can be adopted from the strategy for managing cancer-related symptoms. There is strong evidence for the effectiveness of pain relief – in particular in MND, but also in MS and PD. A number of authors have emphasised the need for well coordinated palliative and neurological care, to reduce the call for active euthanasia and physician-assisted suicide, and to reduce the rate of suicides associated with many neurological diseases (5).

QUALITATIVE ASSESSMENT confirmed the high burden of palliative care needs in our sample in terms of physical symptoms, psychosocial issues and spiritual themes. Participants highlighted the lack of specific services available for people severely disabled often bedbound and without the possibility of leaving their homes. The need of a specific educational process for our team members came out from this study, especially for the management of respiratory failure. For this reason specific training was provided for the education of the professionals involved.

SERVICE EVALUATION is in progress. The pilot RCT started on April 2008 and participant recruitment terminated on March 2009. Of 52 evaluated participants only 2 refused consent. 2 out of the 50 enrolled participants dropped out during the study period. 9 patients (6 ALS - all not tracheostomized- and 3 PD) died during the study and could not complete the second assessment. Disability levels of our sample are as shown here:

Scores	N	Range	Minimum	Maximum	Mean
ALSFRS-r (ALS)	16	0-48	0	34	10,19
EDSS (MS)	18	0-10	7	9,5	6,528
H&Y (PD-MSA-PSP)	16	0-5	4	5	4,19

Physiotherapy was provided for 35 of the 50 patients
Results from test analysis will be presented after analysis.

Conclusions

This study is an attempt to provide evidences of efficacy of a SPCS for people severely affected by neurodegenerative disorders in meeting Palliative Care Outcomes. It confirms the high burden of unmet needs for both patients and their carers. Qualitative analysis allowed us a good insight about the multiplicity of problems experience by this population living in Turin area. Although they cannot be generalized they are very similar to previous published studies of needs assessment. One of the most significant need reported was the lack of knowledge of the different diseases by professionals not directly involved in these patients' care, and as a result there was a failure in assistance provided and a lack of trust in the caring teams. For this reason a specific training was set up aimed at improving knowledge about ventilators, cough-assist machines and other respiratory aids for SPCS professionals.

Mortality was lower than what we generally see in cancer patients (9 of 50 participants) . ALS patients are at higher risk of death if not tracheostomized. No MS patients died during the study follow up despite a high disability. Physiotherapy was very helpful for many participants, for 35 of 50 participants (70%) versus the 5% usually required in cancer patients cared for in our service.

The quantitative analysis will not provide definitive statistically significant results about all the items studied, but this pilot study should help to define future studies. This pilot has shown that the feasibility of the methodology of this study. There were only 2 refusals to participate, even though the disability of our sample was very high. This contrasts with the statements often found in literature suggesting that it is difficult to involve severely disabled participants in RCT's.

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Palliative care needs of people severely affected by neurodegenerative conditions:



The point of view of the professional carers

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Aims:

Patients severely affected by neurodegenerative conditions experience many symptoms, as well as spiritual and psycho-social distress. The opinions of the professionals involved in the care of these people were studied to ascertain if they were aware of these issues

Methods:

In the development of a new palliative care service a qualitative assessment of patients needs has been integrated with 3 focus groups, where 8 neurologists, 1 rehabilitation specialist (with expertise in artificial ventilation), 1 physiotherapist and 1 speech and language therapist - all working in department for the care of people with motor neurone disease, multiple sclerosis or Parkinson's disease and parkinsonism - gave their opinions about the main unmet needs of this population. The groups were video and audio taped to enable the researchers to focus on participants' interactions (mean duration about 60 minutes). A verbatim transcript was performed and coding of the different topics was undertaken by 2 independent researchers. A content analysis of the data is presented in this poster

Results:

A content analysis of the groups highlights the participants' awareness of the high burden of physical symptoms of their patients, in particular uncontrolled pain and breathlessness, as well as the psychosocial issues and the spiritual unsolved dilemmas. It appeared that they had limited knowledge of pain therapy and the use of opioids in their clinical practice. All participants supported the development of a new palliative care service, aimed at improving patients quality of life, caregivers support, help and support in end of life choices and issues and to reduce inappropriate hospital admissions and overtreatment. Respite hospice admissions were felt to be of fundamental importance in sustaining care at home, by providing rest and support for family carers.



in PD and related disorders these drugs are scarcely used. There is the problem of chronic constipation the always affect our patients and that can be worsened by opioids (like tramadol). for this reason in my personal experience they're not so frequently used. Patients complain about leg pain gut bound and usually complicated by osteoarthritis, back pain etc. we refer them to the pain clinic where they're treated with local injections of steroids, or prescriptions of acetaminophen, and steroids if necessary. for this reason I must say that my experience with opioids is like zero.

This is a problem in ALS. This disease is unknown to many physicians. Above all those working in emergency dept. It's a quick disease and in short periods of time you happen to make decisions about End of Life. Often resuscitation procedures fail in these patients. some patient want to be tracheostomized, and curiously many of them are happy with their choice. but they're not so many. if a patient goes in an E&A dept for a respiratory insufficiency it's highly possible that he will be tracheostomized. advanced directives in our country aren't recognized to date, so one of our main tasks is to prevent these inappropriate treatments in patients that don't want it.

I don't know what happens when patients are at home: how are they managed, is the carer able to recognize complications? Are GPs able to manage it? It happens that my patients go to A&E of other hospitals for bad dyskinesias, maybe because they swallowed 5 tablets instead of one and their therapy is completely changed when it'd have been enough to wait the drug effect... and consequences are worst!

...respite admission is very important for caregivers. There are some centre that admit patients when they require respiratory management. Patients are very happy because they receive daily PKT, speech therapy and carers are well educated in the use of ventilators and other aids. We can't train them well here in hospitals because we can just do it in day hospital sessions and often aren't so effective. there they spend several days assisting at these procedures. It'd be so important to have places such these here. I think that for ALS respite admissions can be very useful to support carers. maybe they could use 15 days just to sleep! but ALS patients don't want to die in clinics. they want to stay in their places.



I feel impotent. There are structural limitations. We do our best, but.

We saw patients with a tattoo on their throat reporting that they didn't want to be tracheostomized.

we had experience of different professional in the same team providing completely different information about tracheo for instance. We are not prepared for this task. This can be devastating.

...the main question is "Why me?" And above all "Why now?" I worked all my life and now that I could enjoy it... the matter is that we don't know how to answer, there's non one we can refer them to.



Conclusions:

The authors and participants agreed about the importance of networking among hospital services, palliative care and primary medicine. Specific diseases care pathways should include palliative care services in order to assess and take care of the particular palliative care needs of this patient group.

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A qualitative assessment for palliative care needs of people severely affected by neurodegenerative conditions: the physical symptoms



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Introduction

People severely affected by neurodegenerative conditions experience many symptoms and other psychosocial and spiritual problems, similar to those of cancer patients. As a part of a PhD programme in palliative care at the University of Kent a project has been developed to assess people severely affected by Amyotrophic Lateral Sclerosis (ALS), Multiple Sclerosis (MS), Parkinson's Disease (PD), Multisystem Atrophy (MSA) and related disorders and to provide and evaluate, a new specialist palliative care service in the Turin area. In order to assess the palliative care needs of these population living in Turin area a qualitative research project was undertaken.

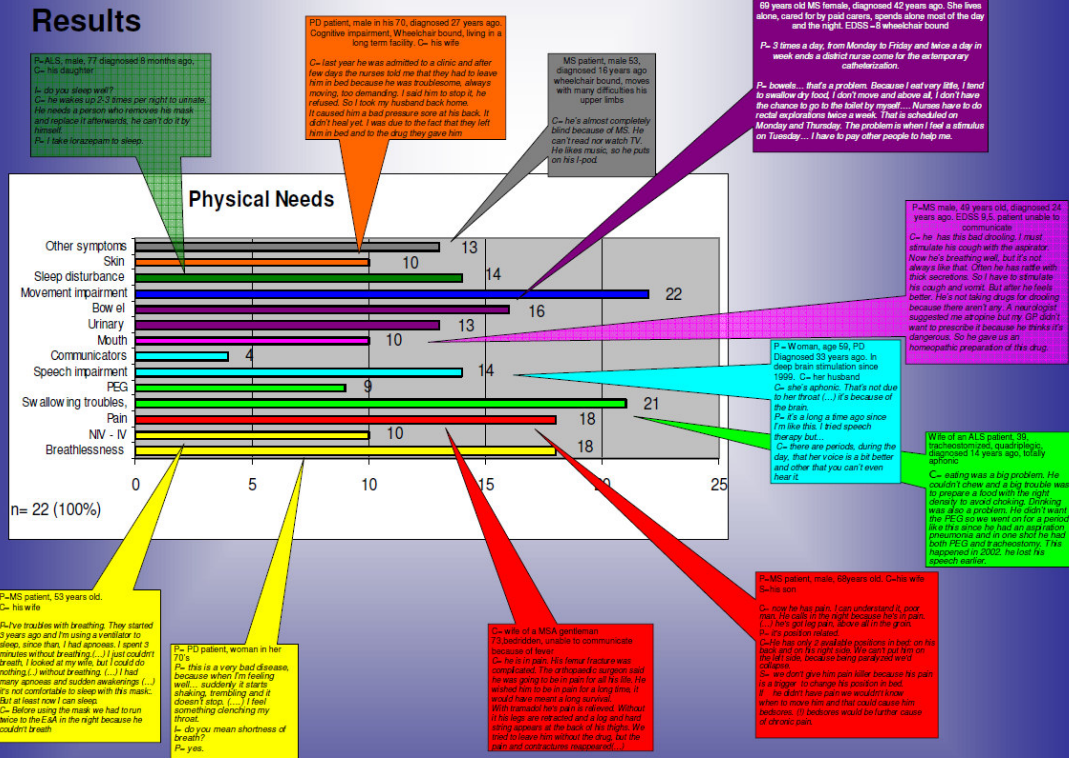
22 patients resident in Turin area and severely affected by amyotrophic lateral sclerosis (ALS), multiple sclerosis (MS), Parkinson's disease (PD) and related disorders (PDRD) and their main carers were interviewed. Interviews were video and audio taped with the aim to catch verbal and non verbal expressions of needs in order to be as accurate as possible in the verbatim transcript. Content analysis of the verbatim transcript has been performed. Coding and content analysis of these events were conducted by 2 independent researchers. Inter-rater reliability of the content analysis was performed among 3 different researchers. Interviews were semi structured and based on the participants' disease stories. The setting was usually the patient's home. Both patient and the main carer were video recorded on the same screen and when patients were very disabled with severe speech impairment the carer talked about the disease. Patients nodded or disagreed with non verbal expression participating as they could.

Methods

Interviews		n=22	
gender	M=18 (82%)	F=4 (18%)	
age	mean=61.5	range (39-80)	
place of interviews	home=20	hospital=1	nursing home=1
paid carer	n=13 (59%)		
diagnosis	ALS n=9	MS n=7	PD n=5 MSA n=1
disability	ALSFPS-R mean=11.3 range= 3-23	EDSS mean=9 range=8-9.5	H&Y mean=4.5 range=4-5 H&Y=5

They were prompted to discuss their physical, psychological, spiritual and social needs and to provide information about the available services. The physical symptoms discussed within the interviews will be discussed in this poster

Results



Conclusions

This initial study confirms the high prevalence of need in this population. Most of these symptoms can be treated with a palliative approach. A specialist palliative care service is being developed to help meet many of these needs, with a specific domiciliary and hospice service for this population in the Turin area.

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Ne-Pal: uno studio esplorativo randomizzato e controllato per valutare l'impatto di un servizio di cure palliative in malati affetti da patologie neurodegenerative

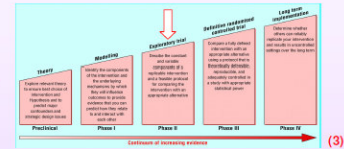
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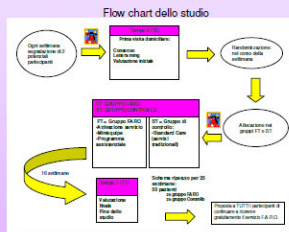
Premessa: Numerose pubblicazioni hanno evidenziato i bisogni di cure palliative nei pazienti affetti da Sclerosi Laterale Amiotrofica (SLA), Sclerosi Multipla (SM), Malattia di Parkinson (PD) e Parkinsonismi atipici (MSA, PSP). Rarissimi sono gli studi che hanno l'impatto che un servizio di cure palliative specialistico (SCPS) può ottenere su questi malati e sul loro famigliari in termini di raggiungimento-soddisfazione dei Palliative Care Outcomes (PCO).

Scopo della ricerca: Nell'ambito del processo di sviluppo di un nuovo SCPS domiciliare ed Hospice la fondazione FARO ha finanziato un progetto di PhD presso la University of Kent con l'obiettivo di valutare l'impatto del nuovo servizio sul PCO in un gruppo di pazienti affetti da queste patologie in fase avanzata di malattia.

Pazienti e metodi: Tra aprile 2008 e marzo 2009, 52 pazienti (ed i loro caregivers) affetti da SLA (n=16), SM (n=20), PD (n=12), MSA (n=2) e PSP (n=2) sono stati arruolati per un RCT utilizzando la Waiting List methodology (1). La randomizzazione prevedeva l'assistenza immediata del SCPS (Fast Track, FT) o un periodo di attesa di 16 settimane prima di poter accedere al SCPS (Standard Track, ST). I partecipanti sono stati segnalati da 6 servizi neurologici ospedalieri di 2 ospedali torinesi. I criteri di inclusione prevedevano l'essere considerati in fase palliativa dal neurologo, alti livelli di disabilità e l'aderenza ai criteri prognostici specifici previsti dal GoldStandardsFramework Inglese (2). Gli strumenti utilizzati per valutare la QoL sono stati il SEIQoL-DW, una serie di NRS per misurarne i sintomi e fattori psico-socio-spirituali, l'HADS ed il Caregiver Burden Inventory (CBI) per i famigliari.



Risultati

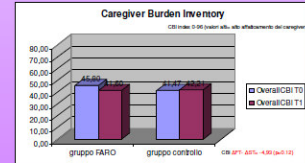
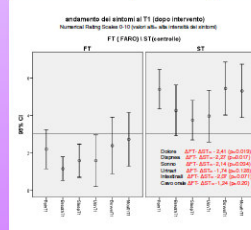
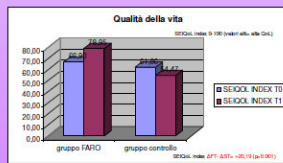


	N	FT	ST	unit
contattati	52			
arruolati	50			
randomizzati	50	25	25	
deceduti	4	2	2	
drop out	2	2	0	
 Sesso				
M:30, 60%	M:16, 64%	M:14, 56%	$\chi^2=0,03$	
F:20, 40%	F:9, 36%	F:11, 44%	$p=,773$	
diagnosi				
AL:16, 32%	AL:7, 28%	AL:9, 36%	$\chi^2=4,72$	
MS:16, 32%	MS:10, 40%	MS:9, 36%	$p=,799$	
PS:18, 36%	PS:9, 36%	PS:8, 32%		
classi di età				
<44:8, 16%	44-64:20, 80%	44-64:20, 80%	$\chi^2=5,242^*$	
45-64:17, 34%	45-64:12, 48%	45-64:15, 60%	$p=,072$	
>65:25, 50%	>65:11, 44%	>65:14, 56%		
disabilità (medici)				
ALSFRS-10:2	ALSFRS-10:7	ALSFRS-9:4	t-test	
EDSS-8:6,2	EDSS-8:6	EDSS-8:4,4	F-test	
H&Y:4,19	H&Y:4,25	H&Y:4,12	EDSS-3:5,83	
			EDSS-5:5,83	
			H&Y:0,55	

Baseline (T0)
 Nessuna differenza clinicamente o statisticamente significativa tra i 2 gruppi nelle variabili misurate:
 •Qualità della vita (SEIQoL-DW index)
 •Caregiver Burden Inventory
 •Sintomi
 •Aspetti psicologici, spirituali e sociali, soddisfazione nei servizi ricevuti

Dopo Intervento (T1)

Confronto tra i 2 gruppi per valutare l'effetto positivo o negativo nel gruppo FARO rispetto al controllo:



Considerando il significato clinico della differenza dei valori dei test -tra il gruppo FARO ed il controllo- con i seguenti parametri (rispetto alla scala di misurazione):
 • $\Delta T1-T0 <10\%$ = basso
 • $\Delta T1-T0 10-20\%$ = moderato
 • $\Delta T1-T0 >20\%$ = importante
 E definendolo:
 • Negativo se il FT mostrava un peggioramento vs ST
 • Positivo se il FT mostrava un miglioramento vs ST

otteniamo→

Parametro	Δ (media T1-T0)	Intervallo	Significatività	Intervallo	Δ (media T1-T0)	Intervallo	Significatività	Intervallo
Qualità della vita	1,74 (3,7%)	0,00-3,48	moderato	positivo	0,00 (0,0%)	0,00-0,00	nessuno	nessuno
Sintomi	0,27 (0,7%)	0,00-0,54	basso	positivo	0,00 (0,0%)	0,00-0,00	nessuno	nessuno
Caregiver Burden Inventory	-0,81 (-1,6%)	-1,62-0,00	moderato	negativo	0,00 (0,0%)	0,00-0,00	nessuno	nessuno
Sintomi (familiari)	0,00 (0,0%)	0,00-0,00	nessuno	nessuno	0,00 (0,0%)	0,00-0,00	nessuno	nessuno
Aspetti psicologici, spirituali e sociali	0,00 (0,0%)	0,00-0,00	nessuno	nessuno	0,00 (0,0%)	0,00-0,00	nessuno	nessuno
Soddisfazione nei servizi ricevuti	0,00 (0,0%)	0,00-0,00	nessuno	nessuno	0,00 (0,0%)	0,00-0,00	nessuno	nessuno

Conclusioni: Questo RCT esplorativo dimostra la fattibilità di trials randomizzati con malati gravemente affetti da patologie neurodegenerative. I partecipanti hanno espresso soddisfazione rispetto al servizio FARO. L'analisi dei test indica che chi ha ricevuto il SCPS FARO ha avuto un bilancio positivo tra aspetti migliorati e quelli peggiorati. In particolare si segnalano un incremento significativo della qualità della vita ed una riduzione di tutti i sintomi studiati. Anche il senso di isolamento sociale sembra decisamente migliorato. Il sollievo ai caregiver è presente, ma la significatività è lieve. Gli aspetti psicologici sono quelli su cui i risultati sono più ambigui. La principale limitazione dello studio è legata al campione ridotto, ma è comunque significativo esplorare con studi di fase 2 (3) ambiti così poco conosciuti per avere indicazioni su come condurre trials più significativi.

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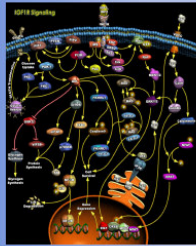
Il servizio di cure palliative FARO verso una nuova sfida: malati oncologici e neurologici in fase avanzata a confronto

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Premessa

Le cure palliative per i malati non oncologici sono una sfida sempre più attuale per i servizi di cure palliative specialistiche (SPCS). I malati affetti da patologie neurodegenerative presentano un carico di sintomi e problemi psicosociali sovrapponibili a quelli dei malati oncologici, ma questo non significa che richiedano servizi identici. Il confronto tra coorti di malati oncologici e neurologici è uno strumento che può consentire di modificare le caratteristiche dei servizi adattandoli alle reali necessità dei pazienti.



Scopo della ricerca

Analizzare le differenze assistenziali tra due campioni di malati assistiti dalla F.A.R.O.: uno formato da pazienti oncologici, l'altro da malati affetti da patologie neurodegenerative.



Pazienti e metodi

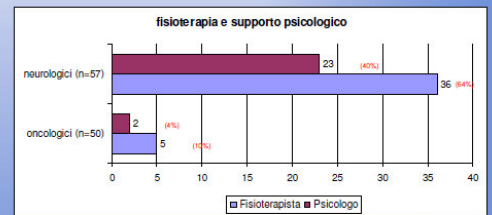
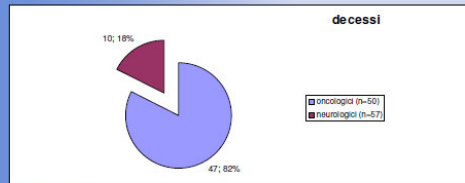
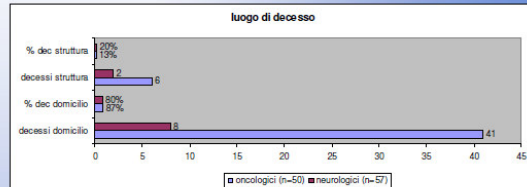
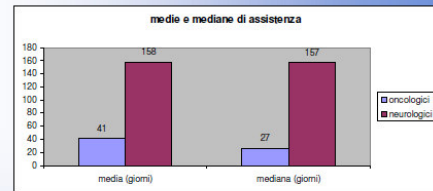
Da settembre 2007 a febbraio 2009 la Fondazione FARO di Torino ha assistito, nell'ambito di un progetto sperimentale, 57 pazienti neurologici affetti dalle seguenti patologie neurodegenerative:

- 13 Malattia di Parkinson (PD)
- 2 Atrofia Multi-sistemica (MSA)
- 2 Paralisi Soprannucleare Progressiva (PSP)
- 20 con Sclerosi Laterale Amiotrofica (SLA)
- 20 con Sclerosi Multipla (SM)

Un campione di 50 malati oncologici, randomizzato tra i 987 assistiti nel 2008, è stato confrontato retrospettivamente con i pazienti neurologici per evidenziare:

- tipologia degli interventi
- sopravvivenza
- durata delle assistenze
- luogo di decesso

Risultati



Discussione

Da questi dati si evince come l'impatto di una coorte di pazienti affetti da patologie neurodegenerative su un SPCS sia stato molto diverso da quello di una consueta popolazione oncologica. Certo è da considerare come la novità rappresentata da questa nuova tipologia di malati possa avere indotto l'attivazione di risorse (come la fisioterapia ed il supporto psicologico) in misura maggiore rispetto a ciò che potrà capitare a regime, ma sono comunque dati molto significativi.

I criteri di presa in carico confermano l'inefficacia nello stabilire una aspettativa di vita coerente con le medie assistenziali usuali in campo oncologico. Ci proponiamo di disegnare delle proposte assistenziali ad hoc rispetto ai reali bisogni riscontrati nei pazienti neurologici partendo da una più approfondita analisi degli interventi praticati e degli outcomes raggiunti.



Extending Palliative Care to Non Cancer Patients: Challenges and Resources

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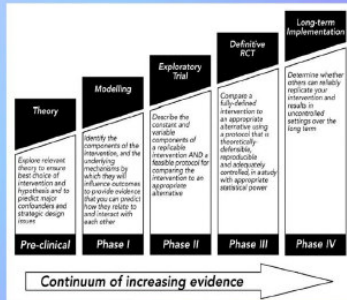
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BACKGROUND

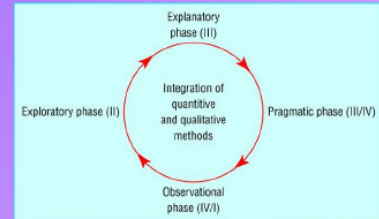
The development of Specialist Palliative Care Services (SPCS) for patients with diagnoses other than cancer has been widely suggested. However these changes presented a challenge for a charity with 20 years of experience in palliative home and hospice care for cancer patients, when it was decided to open its service to people with neurodegenerative conditions. The process of design, modelling, assessment and evaluation of a new service is described.

METHODS



Our design followed the MRC Framework for the Evaluation of Complex(*) Interventions and each phase, from preclinical to phase II is described in terms of methodology and challenges.

(*) Campbell, M. et al. *BMJ* 2006;321:894-898



RESULTS

1. *The charity charter:* the organization had initially cared only for cancer patients, but the evidence of similar unmet needs in neurological patients induced the board to change the charter.



2. *Education of the SPCS team* to the new challenges of caring for this patient group - management of respiratory support, ethics.



3. *Networking:* neurologists, rehabilitation specialists and physiotherapists working in the tertiary clinics caring for Amyotrophic Lateral Sclerosis (Motor neurone disease), Multiple Sclerosis, Parkinson's disease were involved in the service development. Their views of patients needs were explored with focus groups.



4. *Exploration of the needs:* patients and family carers were interviewed in-depth to model the new service on the needs rather than on diagnosis.



5. *Lay patient associations* were informed of the new initiative and their opinions collected.



6. *Prognostic difficulties:* the Gold Standards Framework guidelines were used to help clarify the differing end of life trajectories.



CONCLUSIONS

The new service has grown and 60 patients with neurodegenerative conditions have received hospice care since January 2008, as these challenges were faced and successfully overcome. The next step will be to extend the service to other patients at the end of their lives, independent of their diagnoses, but based on their palliative care needs.



Motor neurone disease
MND patients should be included from diagnosis, as it is a rapidly progressing condition. Indicators of rapid deterioration include:
• Evidence of disturbed sleep related to respiratory muscle weakness in addition to signs of dyspnoea at rest
• Barely intelligible speech
• Difficulty swallowing
• Poor nutritional status
• Needing assistance with ADL's
• Medical complications eg pneumonia, sepsis
• A short interval between onset of symptoms and diagnosis
• A low vital capacity (below 70% of predicted using standard spirometry)

Parinson's Disease
The presence of 2 or more of the criteria in Parkinson disease should trigger inclusion on the Register
• Drug treatment is no longer as effective / an increasingly complex regime of drug treatments
• Reduced independence, need for help with daily living
• Recognition that the condition has become less controlled and less predictable with "off" periods
• Dyskinesias, mobility problems and falls
• Swallowing problems
• Psychiatric signs (depression, anxiety, hallucinations, psychosis)

Multiple Sclerosis
Indications of deterioration and inclusion on register are:
• Significant complex symptoms and medical complications
• Dysphagia (swallowing difficulties) is a key symptom, leading to recurrent aspiration pneumonias and recurrent admissions with sepsis and poor nutritional status
• Communication difficulties e.g. Dysarthria + fatigue
• Cognitive impairment notably the onset of dementia
• Breathlessness may be in the terminal phase