Neurol Sci (2016) 37:1581–1587
DOI 10.1007/s10072-016-2614-x

REVIEW ARTICLE

Need for palliative care for neurological diseases

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Received: 18 April 2016 / Accepted: 17 May 2016 / Published online: 14 June 2016

Abstract The new concept of palliative care supports the idea of palliation as an early approach to patients affected by disabling and life-limiting disease which focuses on the patient's quality of life along the entire course of disease. This model moves beyond the traditional concept of palliation as an approach restricted to the final stage of disease and widens the fields of intervention. There is a growing awareness of the importance of palliative care not only in oncological diseases but also in many other branches of medicine, and it appears particularly evident in the approach to many of the most frequent neurological diseases that are chronic, incurable and autonomy-impairing illnesses. The definition and implementation of palliative goals and procedures in neurology must take into account the specific features of these conditions in terms of the complexity and variability of symptoms, clinical course, disability and prognosis. The realization of an effective palliative approach to neurological diseases requires specific skills and expertise to adapt the concept of palliation to the peculiarities of these diseases; this approach should be realized through the cooperation of different services and the action of a multidisciplinary team in which the neurologist should play a central role to identify and face the patient's needs. In this view, it is paramount for the neurologist to be trained in these issues to promote the integration of palliative care in the care of neurological patients.

Keywords Palliative care · Neurological disease · Specific expertise

Broadening the concept of palliative care to neurological diseases

Palliative care is defined by the World Health Organization as an approach which aims at improving the quality of life of patients and families coping with an incurable disease, through the prevention and relief of suffering by means of identification and treatment of pain and of other physical, social and spiritual problems [1]. The definition provided by the WHO contains an additional key point which radically modifies the traditional concept of palliative care; in fact, it states that *"the palliative approach is applicable during the entire course of illness, in conjunction with other disease-modifying treatments, that are intended to prolong life."* The same principles are adopted by the EAPC (European Association for Palliative Care) stressing the fact that palliative care is the active and total care of the patients whose disease is not responsive to curative treatment.

This new model of palliative care moves beyond the concept of palliation as an approach restricted to the final stage of disease and supports an expert approach to the conditions occurring during the entire course of life-threatening diseases carefully dealing with all symptoms,

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in conjunction with the disease-modifying therapies, including the experimental ones [2]. This model is often referred to as "*simultaneous care*", and it reaffirms the need for an integration of specialised palliative care with the traditional care [3–5]. Early palliative care pursues therefore a threefold aim: improving health condition through the assessment and the treatment of symptoms; strengthening the assistance intended to enhance the patient's and the caregivers' quality of life; searching for the suitability and efficiency of assistance actions [6]. A similar approach has showed satisfying results in the field of oncology [7, 8], where the duration of disease is not so long and heterogeneous as in neurology. In fact, many neurological diseases do not have a resolving treatment and an evolution not influenced by current therapy.

According to this vision, palliative approach aims not only at relieving the patient's suffering in the final stage of the disease, but also at improving his/her quality of life along all the course of illness, hence it is indicated right from the moment of the diagnosis of the incurable disease. This anticipation of palliative care inevitably widens the scope of the issues to be dealt with; the fields of intervention become various, and palliative approach no longer concerns only pain treatment but also deals with all the symptoms which worsen the patient's quality of life during the course of the disease, whether it lasts many years or just a few. When a patient is suffering from disabilities following neurological damage, regarding cognition, motility, feeding, communication and other progressive neurological impairments, interception of the patient's (and his family's) needs acquires a consistent ethical value [9–11].

Within this conceptual framework of palliative care, neurological diseases are distinguished on the basis of different goals and procedures for the implementation and timing of intervention [12]. Both quality and duration of symptoms often require a selective approach of palliative care which are beyond the treatment of pain and feeding problems. Advanced care planning is influenced by the difficulties to predict the time and features of the evolution of the disease in a high percentage of neurological progression. As such, the discussion and communication of the patient's decision has to be planned early in the course of the disease [13]. The same difficulties may be observed in the end of life care because many neurological patients die abruptly due to unexpected complications. A qualifying point is represented by the implementation of a palliative approach which aims to improve the quality of life and well-being for some years over the course of a life-limiting and increasing disability.

People with advanced neurological diseases are currently receiving palliative care from professionals with experience in "primary" palliation, i.e., pain and feeding, but these conditions often are not preeminent in neurological diseases.

Economic reasons support the "primary" palliative care skills based on general medical practice carried out on basic territorial services; on the other hand, limitations of "specialised" palliative care requiring complex interventions may increase the quality of life in long-term assistance and spare repeated and expensive medical actions in both the hospital and in a specific territorial context. Moreover, the composition of specialized teams has to be updated if the concept of palliative care is not to be limited to the end of life and pain treatments occurring in cancer and other conditions (heart, kidney and pulmonary diseases), according to the emerging guidelines previously described [14]. Furthermore, the proposal of practising a palliative approach to complex diseases, such as neurological conditions, and the availability of general practitioner or pain specialists may turn out to be unsatisfactory in terms of the need for long-term care in nervous system diseases. This evidence may support the promotion of a sub-speciality regarding palliative care in neurology to promote the evolution of clinical practice in the advanced and prolonged stages of diseases involving brain, spinal cord, and neuromuscular systems.

General features of palliative care in nervous system diseases

There is an increasing awareness of the importance of palliative care not only in the field of oncology, but also in other branches of medicine, such as congestive heart failure, end-stage renal impairment [15] and, above all, neurological diseases [16], as it has been highlighted for some time in Italy, too. In this regard, it should be recalled that the Italian law has likewise taken account of this evolution, considering that palliative care must also be made accessible to patients with non-oncologic diseases.

In the field of palliative care in neurology, it is paramount to consider the variability of the different conditions in terms of prognosis. On the one hand, there are conditions such as serious brain injury, severe stroke, aggressive neoplasm, characterized by an acute onset for which the survival outlook is measurable in weeks or months. On the other hand, there are progressive diseases entailing a degenerative mechanism, such as irreversible dementia, neuromuscular diseases, Parkinson's disease and movement disorders, and progressive forms of multiple sclerosis whose duration is measurable in years.

In view of this distinction, it is apparent that a "traditional" palliative approach, understood as treatment of the end stage as intended in the past and which seems to persist even today within the medical community, is non-exhaustive for the patients treated by neurologists.

In neurological diseases, the definition and standards of palliative care assistance need specialist skills dealing with the heterogeneity and complexity of nosography, the distinctiveness of clinical course and the impact of disability, the variability and peculiarity of symptoms, the difficulty in identifying the end stage of the disease [17, 18].

Frequently, the prognosis of end stage of neurological diseases does not take advantage of sound predictive indicators and it often does not allow us to determine in advance the last months of life; in many cases, the survival far exceeds the estimated time or, on the contrary, unexpectedly interrupts, as in the case of dementia among other diseases [19].

Some of the general criteria used to identify the end stage of oncological, heart or lung diseases prove inadequate in the field of neurological conditions because they are not necessarily related to the end stages of disease, but describe situations which can occur earlier (e.g., assisted ventilation and enteral nutrition via PEG).

The problem of determining when to turn to a specialized palliative care team may arise. The prognosis of neurological diseases is not always clear and often there is a concern that the request for a specialized palliative care team to support the practitioner may have a depressive and demotivating effect on the patients and their families. In addition, there is a risk of losing the continuity of health professionals, since the integration between the practitioner and specialized palliative care team is not an established reality in some parts of the Italian territory.

Many neurological diseases arise as chronic, progressive and life-limiting conditions and are characterized by different specific neurological symptoms which significantly affect the quality of life of the patients right from the early stages of the disease, such as cognitive impairment, psycho-behavioral disorders, dysphagia, drooling, rigidity and spasticity, postural instability, and autonomic disorders. These aspects unquestionably impose the need for a palliative care approach from the onset of the disease and all along its course [20].

Moreover, it is a known fact that many degenerative diseases of the nervous system set in old age demand particular expertise in the use of neuro- or psychopharmacologic drugs and knowledge about possible interactions between the symptomatic treatments and the developmental features of the disease. For instance, we know how a drug for the treatment of nausea can worsen the problems of reduction in alertness, and how the use of anticholinergics against drooling can impair cognitive performance.

The effects of these diseases on patients' relatives or caregivers, like stress, depression, social withdrawal, and reduced quality of life, should not be underestimated [21–23]. The palliative care approach provides for looking after these aspects as well by involving the relatives and having an accurate and continuous communication and information exchange [24]. The ultimate goal of palliative approach must consist in improving the quality of assistance by reducing hospitalization and emergency admissions [25], hence the achievement of greater appropriateness and efficiency. Palliative care is no longer provided only in hospices and encompasses a comprehensive set of interventions which are practicable in different settings (at home and/or in hospital) [26–28]. The neurologist should activate them on the basis of clinical development, coordinating or integrating a multidisciplinary team in which different professionals converge.

Distinctive features of the palliative approaches to neurological diseases

One of the most challenging aspects of palliative care in neurology is the wide range of symptoms that characterize the long development of the disease and bring to decay the quality of life. The variability and peculiarity of each different disease justify the need for specific expertise in the palliative approach to neurological diseases.

Amyotrophic lateral sclerosis

The first papers that described palliative care in neurology regarded Amyotrophic lateral sclerosis (i.e., Kristianson) [29] and focused on specific aspects such as progressive difficulty in breathing, swallowing, and motility. There is a wide bibliography about this condition, which includes many non-motor symptoms such as depression [25]. A palliative approach that starts at the moment of diagnosis is highly recommended for this disease [30].

The complexity of communication in ALS and in other neurological diseases requires the neurologist to be skillful and sensitive to face different situations along the course of the disease, such as informing the patient about the imminence of life-sustaining invasive procedures or death. Neurological expertise should avoid mistakes in the prognostic evaluation and allow the physician to better inform the patient and his family about the future development, a process that in cohorts of oncological patients has been proved to be very misleading for terminal patients [31–33].

Dementia

Palliative approach in dementia aims at the management of many symptoms which often appear years before death and which determine a progressive cognitive disability and, consequently, a severe burden for the caregiver. The needs most frequently expressed by people with dementia concern both clinical features similar to the terminal phase of other diseases (pain, dyspnoea, agitation, eating problems) [34, 35] and the much frequent comorbidity of these old and fragile patients. However, the palliative approach additionally requires a proper treatment of specific symptoms, such as loss of spatial and temporal orientation, psychiatric and behavioral disorders, and language difficulties. Behavioral disorders have a great impact on dementia; they can be linked to other conditions, such as cognitive impairment, depression, pain, and often represent one of the most difficult aspects of the patient's management. These problems require selective knowledge to carry out a balanced drug therapy, for instance to reduce agitation avoiding excessive sleepiness or a further impairment of ADL, and to manage sleep disorders not resulting in daily somnolence. From this perspective, the treatment must be personalized, because an identical drug approach may induce different clinical effects in patients at the same dementia stage or also in the same patient at different periods of disease evolution. Furthermore, the influence of the co-existence of multiple diseases leading to a multiple drug treatment induces us to promote a single case approach.

The issue of invasive procedure occurs in patients with advanced dementia (PEG, defibrillators, etc.); according to scientific associations, such an approach should be avoided in favor of symptomatic treatments designed to alleviate suffering [36]. Patient comfort should be the primary goal of treatment of advanced dementia, but usually these patients receive treatment which is not consistent with this goal. The prediction of the terminal phase may be extremely complex because a patient can reach the last stage of dementia with severe physical impairment or, alternatively, s/he may even die prematurely due to health problems occurring during the course of dementia (e.g., pneumonia) or because of a concomitant disease. The palliative approach until the late stage of dementia requires relevant steps of clinical management: (a) advanced care planning; (b) adequate prognostic criteria; (c) accuracy in caregiving counseling; (d) evaluation of cultural influences and assistance setting (home or hospice).

Movement disorders

During the late stages of the diseases, movement disorders often show progressive motor and cognitive or behavior impairment, which is further worsened by the reduction of efficacy of symptomatic treatment and frequent appearance of side-effects of long-term therapy [37, 38]. An early palliative approach seems to be largely recommended to face the different aspects of the loss of autonomy [39], such as reduced motor activity and postural impairment in Parkinson's, autonomic disorders in MSA, falls in the PSP, hallucination and fluctuations in dementia with Lewy body, and cognitive impairment in subcortical vascular parkinsonism. These symptoms lead to serious disability. As such, they should be the main target of the palliative approach even if they are not included in the scope defined by the Palliative Performance Scale. These typical manifestations often appear early in the patient's life, long before the common symptoms of terminal stages of other progressive diseases [40] and represent the peculiarity of these diseases. As far as clinical heterogeneity is concerned, the definition of "advanced stage" requires a complex evaluation. The specific scales used to define the severity of these diseases include in themselves general aspects of autonomy which are common to palliative general scales, but at the same time, they identify some specific aspects of these disorders that represent "clinical milestones" whose achievement determines disease progression [41]. In the advanced stages, the broad spectrum of symptoms affects not only motricity but also many other functions and abilities. Advanced Parkinson's disease is characterized by motor complications (hypokinesia/akinesia, dyskinesia, dystonia) often associated with pain, but there are numerous and frequent non-motor symptoms present in both the MP and other parkinsonisms [40, 42]. Such symptoms include autonomic disorders, psychiatric disorders, sleep disturbances, cognitive impairment, bedsore, dysphagia, and respiratory difficulties [43–45]. The complexity of these disorders is also related to pharmacological management, since a treatment targeted to one specific symptom can interfere with other aspects (such as the treatment of dopa-induced psychosis treated with neuroleptics which worsens motor skills, or the treatment of hallucinations of Lewy causing motor block).

Multiple sclerosis

Despite the advances in the treatment of this disease, the development of multiple sclerosis in advanced stages with significant disability can not be slowed down by a treatment based on pathogenic mechanisms. Based on this finding, it is believed that beyond the EDSS 6.5–7, the unmet needs of the patient should be addressed with a symptomatic approach to improve his/her quality of life [46]. This approach corresponds to the requirement of a simultaneous palliative approach and can be carried on until the stage of total dependence and in anticipation of the final outcome. The elements that characterize this condition can be identified in the occurrence of cognitive impairment and multi-organ failure which are frequent complications of the advanced stage of the disease. There are many goals of palliative treatment in the advanced stages of multiple sclerosis. These may include issues such as cognitive impairment, psychiatric and sleep disorders, bladder and sphincter disorders, dysphagia, complications caused by immobility (pressure ulcers, infections), and pain often caused by spasticity, thus requiring a different approach compared to oncological pain [23, 26]. In many cases, the general picture becomes complicated by the presence of frequent infections and moving difficulty related to environmental features. Pharmacological treatment can be effective in treating symptoms such as pain, spasticity, depression, but in some conditions, it may be necessary to carry out a substitutive procedure to face, for example, nutritional problems. Communication and healthcare management have become relevant issues in the advanced and terminal stages of the disease, especially regarding the need for an appropriate and effective communication to avoid isolation, cognitive impairment and affective disorders. Another crucial issue in the palliative approach is also the need to train and protect caregivers from stress related to the complexity of care that these patients require.

Brain tumors

In brain tumors, the palliative approach must deal with problems which are only partially comparable to those of other types of cancer, since the decline of autonomy largely depends on conditions that highly restrict the quality of life, such as seizures, psychiatric disorders, depression, psychosis, and cognitive impairment. These conditions require specific expertise relating to clinical problems and their pharmacological management, with the aim of not only achieving a “relief” from suffering related to the final phase of the disease, but also of optimizing the quality of remaining life. For example, patients often present with agitation and sleep disorders, often associated with steroid therapy, and only a competent and balanced pharmacological management of these symptoms can be effective in avoiding excessive sedation. Patients affected by brain tumors are very fragile not only because of the cerebral disease but also owing to the frequent surgical operations they have undergone which makes them more sensitive to pharmacological treatment and possible side effects. The management of antiepileptic therapy must take into account these peculiarities and fragilities adapting the type and dose of antiepileptic drugs not only to protect the patient from possible critical events but also to reduce side effects, such as excessive sedation, and to maintain their cognitive skills and autonomy. The aim of a palliative approach in these conditions should not only function as a relief from some disturbing symptoms but also as an optimization of the preserved capabilities of the individual so as to achieve a good quality of life. Moreover, the rapid evolution of the disease would also require a frequent re-evaluation of the patient’s conditions which in turn would allow for a continuous updating of the subject’s needs [47].

Cerebrovascular diseases

Considering the clinical heterogeneity of stroke outcomes and the variability of stroke effects related to pre-existing conditions, the palliative approach should be developed individually for each subject also in case of cerebrovascular disease [48].

In March 2015 the AHA/ASA published an extensive document entitled “Palliative and end-of-life care in stroke” which highlights the fundamental features of palliation after stroke [49]. The palliative approach to these conditions must face a wide range of needs and problems affecting patients with stroke and should include many different aspects such as talking with patient and families about preferences, needs, and values to establish the medical decisions and having effective, sensitive discussions about the prognosis, how to deal with physical or mental losses, and, if necessary, of dying. In the most serious condition is important to offer a guidance through choices about life-sustaining treatment options (cardiopulmonary resuscitation, ventilators, feeding tubes, surgery, do-not-resuscitate orders, do-not-intubate orders, and natural feeding). Palliative care should also offer the best treatment options for common post-stroke symptoms, including pain, other physical symptoms, and psychological problems, such as depression and anxiety [48]. This approach, according to some specialists, relates only to more serious condition and situation of compromised autonomy, but in a more extensive vision, all actions taken after an acute cerebrovascular event and designed to understand patient’s needs and desires, to share treatment planning, to alleviate persistent physical and psychological symptoms, to reduce the risk of new damage and functional decay, are a part of the palliative treatment, that includes also the rehabilitation treatment.

Traumatic brain injury

In a traumatic brain injury, the palliative approach can be carried out in two different situations: in conditions of severe autonomy impairment in daily living activities (caused by the presence of motor, sensory and cognitive deficits), and in conditions such as vegetative and minimal conscious state. In conditions of preserved consciousness, patients can present with disability related to serious motor deficits, spasticity, tendons contracture, neurogenic or neuropathic pain syndromes, hemispatial neglect, visual disturbances, and language and speech disorders mainly affecting comprehension. The palliative approach in patients with severe traumatic brain injury is designed to select a life-sustaining treatment, prevent complications and treat pain and vegetative disturbances; it also includes communication and psychological support to the patient's family.

Conclusions

The evolution of palliative care as an early approach which focuses on the patient's quality of life seems to be a shared concept at this point, supported by a lot of evidence; likewise, the need for development and expansion of palliative care beyond oncology is strongly emerging. This is particularly evident within the neurological field, where there has been a growing interest in healthcare management and research regarding palliative care. In neurological diseases, the definition and implementation of palliative care require specific skills and knowledge which are essential to adapt the concept of palliation to the specific features and complexities of these pathologies.

With regard to the professionals in charge of providing palliative care, the EFNS Guidelines, EANS (European Federation of Neurological Societies and European Neurological Society) [17] recommend that the multidisciplinary team of palliative care must be composed of at least three different professionals: a physician, a nurse and a social worker or a psychologist.

In our view, we believe that more professionals should be involved in the palliative approach to neurological diseases, and in relation to the Italian healthcare system, we would propose: a neurologist, a general practitioner, a physiatrist, a psychologist, a social worker, a nurse, a patient care assistant and a palliative care physician.

In our country, we have to face the major problem of the absence of an ad hoc specialist medical training, and the problem of the responsibility of the various professionals composing the team and the coordination of the multidisciplinary team.

Considering the Italian situation, we think that the neurologist is undoubtedly the primary care provider, at least in the first stage of the disease, and that the responsibility for deciding when to involve the above-mentioned different professionals lies with him. The neurologist should be trained in at least the key principles of palliative care, and s/he should be able to directly provide a primary palliative approach, but at present, only few neurologists have received this training.

When the complexity of the disease requires different skills, the next step would be to integrate neurological services with the existing palliative care services for non-neurological diseases (i.e., the existing networks of palliative care, including hospice care), also considering that palliative care services must overcome the idea of "End of Life Care" and also the idea of global and total patient care. These services should provide counseling and, in some cases, temporary patient management aimed at achieving a specific and measurable objective (temporary admission to hospice to alleviate the caregivers' burden, temporary patient management to control specific symptoms, availability of counseling to explain the objectives and practical possibilities of palliation for patients who do not wish life support treatments deemed disproportionate, etc.). From this perspective, it would be useful to identify some "trigger points" [50], which are also markers of clinical deterioration and predictive of a possible "end of life stage" [51].

In conclusion, at this point in history, it is paramount for the neurological community not to delay any further by taking a stance on the prominence of a palliative approach in the field of neurological diseases, which are characterized by so many specific features, as we have sought to demonstrate in this paper.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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