APS Australian Psychological Society

Psychological wellbeing and care for people living with Motor Neurone Disease (plwMND) and their carers

A guide for psychologists

Acknowledgements

We acknowledge the Traditional Custodians of the lands and seas on which we work and live, and pay our respects to their Elders, past, present and emerging, for they hold the dreams of Indigenous Australia.

We acknowledge people with lived and living experience and those who care for them. Their knowledge and experience serve us all and remind us of what is most important in the work we do.

This practice guide was prepared by Mr Paul Cafarella MAPS. The APS would also like to acknowledge and sincerely thank the members of the Expert Reference Group who so kindly contributed their time, knowledge, cultural humility and lived experience to the development of this practice guide.

Ms Vicki Ambrose

Ms Jenne Hartmann

Ms Christy Hunt

Dr Judy Tang MAPS

The Australian Psychological Society (APS) has been able to produce this practice guide thanks to generous funding from FightMND.

Disclaimer and Copyright

Contents

Executive summary

Motor Neurone Disease (MND) is a progressive condition for which there is no cure. Therefore, treatment and care focus on optimising and sustaining quality of life and symptom relief. Numerous psychological sequelae of MND exist, and several studies have demonstrated that psychological factors have a serious and extensive impact on the quality of life of people living with MND (plwMND) and their carers. Psychologists are integral multidisciplinary team (MDT) members, and optimal care requires sufficient knowledge of the disease and the psychological and mental health considerations associated with holistic care of plwMND and their carers. Psychological impairments have important clinical implications for psychologists, including communication, treatment engagement and adherence.

MND diagnostic delay averages around one year, resulting in extended periods of uncertainty, anxiety, frustration, and distress. This delay may negatively impact subsequent service engagement as distrust and dissatisfaction are commonplace. The psychological impact of receiving a terminal diagnosis of MND is immediate and devastating for patients and their loved ones. How the bad news is delivered can impact the initial reaction of the plwMND and influence their attitude toward engaging in symptomatic treatment. Hence, existing protocols for the delivery of bad news should be employed. As part of an MDT, psychologists are sometimes consulted soon after diagnosis. Clinical guidelines recommend psychological care after diagnosis to assist plwMND and their carers to cope with the diagnosis, understand the disease, and accept and cope with a changed life and changes in relationships. These interactions may involve heightened emotions, and the psychologist may assist plwMND in clarifying expectations regarding their ability to manage the progression of their mobility issues and address concerns regarding a loss of independence. Psychologists may optimise coping in the plwMND with tailored communication, facilitating control and assisting the maintenance of identity.

MND characteristics such as muscle weakness, respiratory-related issues, dysphagia, dysarthria, and a range of neuropsychiatric and mental health impairments, as well as poor prognosis, are associated with numerous challenges to the psychological wellbeing of plwMND and their

carers. It is a disease of cumulative, fast-moving, and progressive losses. Each deficit comes with the challenge of adjusting to their new circumstances. Psychologists who are aware of the difficulties associated with these impairments can assist plwMND and their carers with appropriate tools, such as fostering a sense of control and appropriate hope, facilitating independence, using tailored communication, encouraging early engagement in decision-making, being aware of risk factors for mental health issues, and monitoring for psychological distress. As the disease progresses, end-of-life considerations become more prominent. Psychologists who raise or respond to plwMND-initiated advance care planning discussions should do so in the context of patient autonomy, security, control, courage, dignity, and independence.

Psychologists should be aware of a range of factors that contribute to or protect against the development of mental health problems among plwMND and their carers. A range of psychological interventions have been trialled with these populations, with many demonstrating psychological wellbeing benefits. Tailored psychological interventions are preferred by plwMND and their carers, as these methods address their individual needs and values. Psychological interventions should be customised to the personal challenges that plwMND and their carers face at different disease stages. For carers, this includes coping with bereavement. A 'goodness-of-fit' approach to psychological intervention involves the evaluation of the needs and preferences of individual plwMND and carer risks and matching them to tailored treatment.

There are numerous psychological sequelae of Motor Neurone Disease (MND), and several studies have demonstrated that psychological factors have a serious and extensive impact on the quality of life of people living with MND (plwMND) and their carers.[1-3] Optimal care requires that psychologists have sufficient knowledge of the disease and the psychological and mental health considerations associated with holistic care of plwMND and their carers.

What is MND?

The term Motor Neurone Disease refers to a group of rare neurological diseases involving a progressive and ultimately fatal degeneration of motor neurones in the upper and lower extremities.[4] Amyotrophic lateral sclerosis (ALS) is the most common form of MND, and therefore, this term and MND are often used interchangeably. Motor neurones are the nerve cells responsible for muscle movement and control. In the disease process, motor neurones weaken, and paralysis ensues. The progressive loss of motor neurones in the brain and spinal cord commonly leads to muscle weakness in the limbs, difficulties communicating, problems with swallowing and eating, breathlessness, other sensory symptoms, and cognitive impairment.[5]

Prognosis

There is variability in the course of the disease, speed of disability progression and symptoms experienced. However, the disease is relentless in its advancement and is ultimately fatal, with life expectancy averaging 27 months post-diagnosis.[6, 7] Survival also varies, although most commonly, death via respiratory failure occurs in individuals with ALS 3-4 years after onset.[5, 8] Approximately 10% of individuals with ALS have a slower-progressing disease and may survive over 10 years.[9]

MND: Cause, risk factors, prevalence and burden

The cause of MND is unknown. Genetics are thought to be responsible for between 5-15% of cases, as evident in the rates of familial MND.[5] Gender and age are also considered risk factors. Males are at increased risk, with research demonstrating a male-to-female ratio between 1 and 2.9 The age of onset is variable but typically ranges between 50 and 70 years, with peak incidence between 60 and 75 years.[10, 11] While other environmental risk factors have been proposed, causal links with MND have not yet been established.[12]

The Global Burden of Disease (GBD) project estimated the all-age global prevalence of MND to be 4.5 per 100,000 persons.[13] However, the lifetime risk of ALS is approximately 1 in 350, and the prevalence is reduced by the population's limited life expectancy.[14] MND prevalence in Australia is estimated to be 8.7 per 100,000, although these figures are based on statistical modelling rather than epidemiological data. Specific data exists for South Australia, with an estimated prevalence of 6.79 per 100,000 persons. While these numbers are considerably higher than global estimates, the numbers may also result from greater case ascertainment compared to the GBD study.[15] Although MND is not common, the economic burden is massive, with the 2015 MND Australia Deloitte Access Economics Report identifying a $1.13 million total per-person cost, far surpassing the cost of most other chronic diseases.[16]

Types of MND

The group of motor neurone diseases comprises a variety of phenotypes distinguished by the early symptoms experienced by the plwMND and the site of disease onset. MND variants, or the categorisation of subtypes, are determined by evidence of the underlying pathology, onset sites, the areas affected and the rate of disease progression.[17] The most common MND variants are described below.

Amyotrophic lateral sclerosis (ALS) is by far the most common form of MND, comprising 80-90% of cases. Notably, the term MND is used in Australia and the United Kingdom, but ALS is the preferred umbrella term outside these areas. Both upper and lower motor neurones are impacted, usually resulting in an initial wasting and weakening of the limbs and hyperreflexia. Commonly, speech, swallowing and respiratory issues become evident as ALS progresses to later stages. Life expectancy is two to five years post-diagnosis.[18]

Progressive bulbar palsy (PBP) also impacts both upper and lower motor neurones. However, this subtype initially impacts speech and swallowing muscles and deterioration in these domains tends to be rapid. Emotional lability is commonplace. PBP has a life expectancy of 6 months to 3 years from symptom onset.[18, 19]

Progressive muscular atrophy (PMA) is characterised by predominant lower motor neurone involvement with variable upper motor neurone involvement. Presentation often initially involves weakness and wasting in the arms or legs (known as 'flail' arm or leg), reflex absence and muscle twitching. PMA can be difficult to diagnose, and latency to diagnosis tends to be longer than for ALS. It is estimated to account for approximately 10% of plwMND. The involvement of upper neurones may lead to PMA being reclassified as ALS. Life expectancy is usually over 5 years.[18-21]

Primary Lateral Sclerosis (PMS) is a less common (approximately 3%) variant of MND, impacting only the upper motor neurones. It is characterised by earlier onset (usually 40s-50s), leg spasticity, balance problems, bladder hypersensitivity, muscle weakness and cramps. Slurring of speech may also be present. PLS progresses slowly, and life expectancy is significantly longer than other motor neurone diseases. PLS is not considered to be life-limiting, however life expectancy reduces if it develops into ALS.[19, 26]

Psychologists and MND

There is no cure for MND or hope for cessation of progression. Therefore, treatment and care focus on the optimisation and sustainment of quality of life and symptom relief via supportive care.[3, 22, 23]

Numerous studies have demonstrated that psychological factors have a profound impact on the quality of life of plwMND and their carers.[1-3] Multidisciplinary teams (MDTs) have become recommended practice in the care for plwMND and their loved ones since the focus has moved beyond just physical symptom management. The complexity and broad range of needs of plwMND and their carers necessitates the involvement of a variety of different health specialists, with their own specific qualifications and skills.

Psychologists have become integral members of the MDTs and are able to provide important assessment and treatment skills. A multitude of physical and psychological sequelae exist for plwMND and their carers. Standard care necessitates the assessment and management of various individual needs, including psychological and social support.[24] The requirement for this support is evident, appropriate, and necessary right throughout the course of the disease, from diagnosis to the terminal phase, and beyond for remaining loved ones.[24, 25] This evidence-based guide aims to assist psychologists in identifying common psychological and mental health considerations in the management of plwMND and their carers. This knowledge will enable psychologists to adapt practice to meet the needs of plwMND and carers and positively impact their quality of life.

Diagnostic issues and implications for psychological wellbeing

Pre-diagnosis

The time to reach a diagnosis of MND can be lengthy as numerous factors, including the demands on health professionals, local resources and expertise, the requirement for specialised testing, and symptom heterogeneity and overlap with other health conditions, can complicate the process. Diagnostic delay and misdiagnosis are commonplace.[27-29] Diagnostic delay averages around one year, a significant period in the context of a rapidly progressive and fatal disease such as MND.[29]

Awareness of MND among the public and general practitioners remains suboptimal.[30] Australian research identified that the pre-diagnosis period could involve many months of numerous investigations, multiple referrals, misdiagnosis, unsatisfactory interactions with health professionals and sometimes long waiting times for appointments and follow-ups with neurologists before being confronted with the diagnosis. Some plwMND have been dismissed by their doctors when presenting their early symptoms or were referred to non-neurological specialties.[31, 32] Factors contributing to the diagnostic delay led to extended periods of uncertainty, anxiety, frustration, and distress.[33] This period after the onset of symptoms but before diagnosis has been described as "being on a diagnostic roundabout," typified by uncertainty, demoralisation and traumatic experiences. Furthermore, these experiences may negatively impact subsequent service engagement[34] as distrust and dissatisfaction with prolonged investigations are commonplace.[31] Carers have also reported uncertainty, distrust, dissatisfaction, and fear during the pre-diagnosis period.[31]

Diagnosis

Receiving a diagnosis is a pivotal moment for plwMND and their carers.[35] The diagnosis of MND is sometimes provided in an isolated event but is more often communicated incrementally over several appointments.[30, 32, 36] The psychological impact of receiving a terminal diagnosis of MND is immediate[37] and has been described as devastating for patients and their loved ones.[32] It is unsurprising that symptoms of anxiety are highly prevalent in newly diagnosed plwMND and their families.[38, 39] While depression among newly diagnosed plwMND is common, it is even more prevalent among their relatives soon after diagnosis (almost a third).[38, 40]

A neurologist delivers the diagnosis in most cases, and the way the bad news is delivered can impact the initial reaction of the plwMND and influence their attitude toward engaging in symptomatic treatment.[41, 42] Protocols for the delivery of bad news have been developed for other terminal illnesses and have been adapted for MND (e.g., SPIKES protocol[43]; NICE recommendations for information and support at diagnosis[44]; a personalised communication guide developed by van Eenennaam et al., 2020[45]).

In Australia, over a third of plwMND report being dissatisfied with how the diagnosis was communicated.[46] Some important communication elements impacting psychological wellbeing have been identified. PlwMND prefer that an experienced physician deliver the diagnosis. When the doctor breaking the bad news was inexperienced, there was a worsening of the plwMND's mental state.[41, 44] Such news regarding the diagnosis and its implications should be delivered in a clear and empathetic manner and include sufficient information regarding prognosis and treatment options.

Providing reasonable hope when delivering the diagnosis also helps plwMND to cope more effectively. In this sense, hope is maintained via redefining what plwMND and their loved ones hope for. Hope may be redefined on goals rather than a cure,[47] such as maintaining a satisfactory quality rather than quantity of life.[48] Further examples reported by plwMND include the hope of being on the tail part of the survival curve, positive stories of how others have beaten or stretched the odds and recognising that MND statistics are group-based and may not apply to the individual.[45]

PlwMND also value the reassurance of ongoing support, compassion and gentle probes regarding readiness and the amount of prognostic information an individual can handle.[41] The information provided should be based on and sufficient for patient and carer needs without overwhelming them.[49] Neurologists often avoid disclosure of prognostic information[48, 50] due to fear of a negative impact on a patient's mental state, although plwMND prefer honest, compassionate communication.[48, 49] Avoiding topics regarding prognosis has been shown to damage hope in other terminal diseases.[51]

Tailored communication is recommended, as it provides information about a plwMND's personal prognosis and disease progression, is tailored to a patient and carer's emotional and information needs, their illness experiences, coping factors, culture, and facilitates patient and carer control and decision-making.[32, 45, 48, 52] For example, this tailored information enables the plwMND and their carers to make timely and specific plans regarding future care.[45, 48] The negative emotional impact associated with receiving a diagnosis of MND can be mediated, acceptance is fostered, and communication satisfaction is improved with the inclusion of these tailored elements of communication.[48]

The role of psychologists in the post-diagnosis period

The role of psychologists in the diagnostic process has been highlighted in recent times. Ideally, multidisciplinary team involvement should occur immediately after diagnosis,[44] and non-medical healthcare professionals (HCPs), such as psychologists, are sometimes consulted during this period. The psychologist may be one of the first professionals the plwMND has interacted with since the diagnosis of MND.

Research with HCPs, including psychologists and physiotherapists, indicates that these professionals are often 'dealing with the diagnostic aftermath' presented by the newly diagnosed plwMND. Common reactions included anger, frustration and dissatisfaction with the diagnostic process and/or delivery. A further role for psychologists is evident as interactions with newly diagnosed plwMND may assist them to unpack and understand their diagnosis, often through difficult and sometimes distressing conversations.[31, 36]

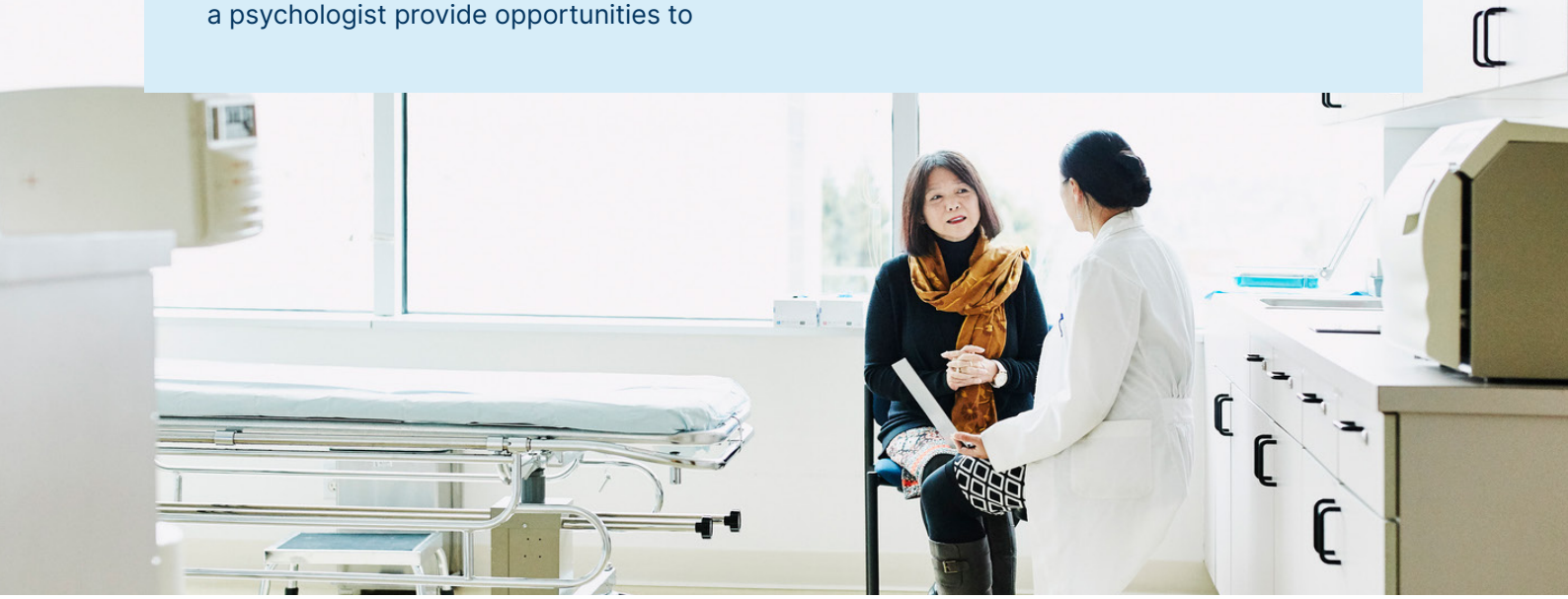
The NICE guidelines recommend psychological care after diagnosis to assist plwMND and their carers to cope with the diagnosis, understand the disease, accept, and cope with a changed life and changes in relationships.[23] Further, psychologists are sometimes involved with early cognitive screening, which may lead to discussions concerning the cognitive changes associated with MND and the reasoning for and implications associated with mental capacity assessments.[44] Another example is the clarification psychologists are sometimes required to provide regarding misconceptions or expectations a plwMND may have about their ability to manage the progression of their mobility issues and address concerns regarding a loss of independence. Consequently, psychologists become involved in the process of breaking further bad news.

Care should be taken to communicate in an inclusive, culturally sensitive manner. These difficult conversations may alert a psychologist to a plwMND's individual needs, such as the requirement for referral to another professional or other sources of support such as an MND Association. These organisations and interactions with other plwMND through these groups may provide meaningful supplementation of information regarding the disease and options for care, as well as compassion, inspiration and hope.[53] Further tips for diagnosis delivery can be found at the MND Connect website[54].

Fostering a perception of control

The perception of a loss of control is consistently reported by plwMND and is a major factor in their psychological wellbeing.[37, 45] Difficult but essential conversations with psychologists help a newly diagnosed plwMND understand their diagnosis, gain a sense of control, and prepare for the future.[32, 36] Debriefing and clarifying the consequences of the diagnosis, including prognosis, can further support acceptance and personal planning for plwMND and their carers. This enables a sense of control as decisions regarding treatment planning and end-of-life considerations can better align with the individual's needs and preferences.[48, 55] Concentrating on the present, maintaining a sense of identity (including culture), and focusing on personal values are additional strategies reported to enhance the perception of control.[37] The role of a psychologist is clear in this context, as interactions with doctors tend to be shorter and may prioritise physical symptoms rather than the psychosocial impact of the terminal diagnosis. Interactions with a psychologist provide opportunities to identify and address these psychosocial needs and preferences of plwMND and their carers.[31, 36, 37]

Psychologists interacting with ATSI plwMND and their families should be aware of guides and resources that assist in communicating the implications of their diagnosis in terms of mobility, independence, and function. Communication and cultural barriers can include a reluctance to acknowledge the news, mistrust based on colonisation, social inequities, fear of a lack of cultural safety, and maintenance of spiritual beliefs regarding the cause of health issues that may not align with Western medicine. A comprehensive list of resources is available at the Australian Indigenous HealthInfoNet[56] website. This site also includes a palliative and end-of-life care portal. Additional resources, training materials and information to help HCPs provide culturally safe and responsive communication and care for ATSI peoples are available at the Gwandalan National Palliative Care Project[57] and Caresearch[58] websites.



Impact of MND and the risk of psychological problems

MND has often been described as a disease of cumulative and progressive losses.[59] The symptoms of MND and its poor prognosis force numerous and regular psychological challenges upon plwMND and their carers. The challenges are in addition to the significant physical, social, and financial problems associated with the disease,[60] and with each deficit comes the challenge of adjusting to their new circumstances while trying to maintain as muchfunctionality, independence, and control as possible.[61]

Muscle weakness

Common presenting symptoms associated with muscle weakness include pain, spasticity, fasciculations, cramps and muscle atrophy.[63] Pain is commonly reported to be frequent and significantly impacts the quality of life of plwMND.[64] Progressive loss of strength and control of muscles affect a plwMND's ability to conduct basic and complex activities of daily living. The lack of mobility and muscle weakness may also lead to pain and spasticity and limit or prohibit core activities such as walking, speaking, eating, and breathing,[65] decreasing autonomy and increasing dependence on family members and others involved in their care.[66] One of the main determinants of quality of life in plwMND is a reduction in valued activity.[3] The inability to participate in meaningful activities contributes to a loss of control and autonomy, which in turn is related to lower psychological wellbeing.[67, 68] These meaningful losses that are beyond the plwMND's ability to control, may lead to an increased external locus of control and a lack of purpose in life. Both of these variables are linked with increased hopelessness in plwMND. Hopelessness is a component of depression and has been associated with a greater degree of suffering and increased suicidal ideation in plwMND.[62, 69, 70]

> Noteworthy for psychologists is that an external locus of control and a lack of meaning in life are more predictive of hopelessness than disease factors,[62] providing some insight into avenues for investigation and management.

Respiratory related issues

A form of respiratory system dysfunction is the usual terminal event for plwMND. As the disease progresses, plwMND will usually experience a complex mix of impairments (e.g., declining airway clearance and bulbar muscle coordination, swallowing and speech impairment, weakened cough, hypermetabolism) and their consequences (e.g., aspiration, communication, need for nutritional support). Dyspnoea (shortness of breath), orthopnoea (shortness of breath when lying down), and hypoventilation become commonplace and progressively worse.[8, 72] ALS-related dyspnoea is one of the most distressing symptoms for plwMND, can be intense and fear-provoking, create a perception of danger, and has a significant

impact on psychological distress, quality of life and emotional wellbeing.[73-76] Dyspnoea and fatigue both impact the causal pathway between anxiety and quality of life in plwMND, and anxiety is the second biggest influence on quality of life.[3, 77] Modelling with a sample of 1022 plwMND demonstrated that dyspnoea has a fundamental impact on both quality of life and depression and that anxiety and fatigue were mediating factors. Dyspnoea, anxiety and fatigue had larger effects on quality of life and depression than disability.[78]

The clinical implications for psychologists from this respiratory research are clear, as quality of life and depression are not straightforward consequences of increasing disability due to MND progression, and mediating factors, such as anxiety, can be identified and treated. Non-invasive ventilation is now standard care for respiratory issues in plwMND and can extend life as well improving quality of life, although adherence plays a significant role in its efficacy.[8, 71, 72] A clear role exists for psychologists to assist plwMND to improve their quality and quantity of life by facilitating adherence.

Dysphagia and its complications

Dysphagia (difficulty swallowing) occurs in most plwMND and has devastating physical and psychological impacts. Dysphagia becomes progressively worse as bulbar and respiratory muscles weaken, resulting in weight loss, nutritional deficiencies, dehydration, choking and aspiration pneumonia, the latter being the leading cause of death for plwMND.[79] Psychological impacts include fear of choking significant enough to severely impact the quality of life of a plwMND, even in the early stages of the illness. Panic and trauma were described in a recent study examining the personal experiences of dysphagia among plwMND.[80] Consequently, there is a relationship between dysphagia and the perception of a loss of control.[68, 81] The environment around mealtimes and food consumption also changes, as eating can be stressful, takes longer and requires more effort, making the experience less pleasurable.[82,83] For some plwMND, the perception of food can change from pleasurable to seeing it as 'fuel' related to survival and 'dangerous.' Recommended modifications, such as changing the texture of foods by pureeing them, may also contribute to this negative change in perception.[80] In essence, the social and relational meaning of food and nutrition may be significantly altered as thoughts and behaviour in these domains become more stressful for plwMND and their carers.[68] Attempts at control by the plwMND may include focusing intently on eating and eliminating social distractions during this time, resulting in eating without company.

In order to reduce social isolation and its inherent emotional risks, psychologists may suggest that carers sharing meals with plwMND allow them to concentrate quietly while in their company.[80]

Psychologists should be aware that the perception of control regarding eating behaviour and related treatment decisions can be influential in the psychological wellbeing of plwMND. Once symptoms of dysphagia are managed, stress levels regarding the issue reduce markedly. PlwMND have reported that focusing on the present could enhance perceptions of dysphagia control, although psychologists are cautioned that plwMND may under-report the severity of their symptoms because they believe they are managing dysphagia effectively and are more concerned with other symptoms of MND.[80]

Inevitably, as dysphagia progresses, alternative modes of feeding will be required, and gastrostomy options, such as percutaneous endoscopic gastrostomy (PEG) or percutaneous radiological gastrostomy (PRG), are recommended by clinical guidelines.[44, 84]

Decision-making regarding gastrostomy is notoriously complex, challenging, and stressful for plwMND, their carers and HCPs.[86] This decision-making is commonly postponed due to a lack of readiness, acceptance issues, anxiety regarding the gastrostomy procedure, and determination to maintain control.[48, 67, 83] Attempts at control and independence by plwMND may be manifested in early acceptance and planning, postponement, and initial refusal.[86, 87] Proactive, early acceptance allows prompt decision-making, while others prefer later decision-making. A key element in the successful maintenance of perception of control is that the plwMND sees gastrostomy as an aid to maintain quality of life and independence rather than a procedure that will compromise these values, especially since the need for it will become inevitable.[86-88]

A 'window of opportunity' where plwMND have a period of time where they are able to exert control over choices regarding gastrostomy decisions may be highlighted by treating psychologists to facilitate the perception of control.[85, 86] This may be manifested in psychologists raising the topic of gastrostomy at regular intervals to facilitate this perception of making decisions in their own time, with choices supported by psychologists.[86]

The involvement of loved ones in decision-making can facilitate better psychological outcomes for plwMND and their carers. The complications associated with feeding, swallowing and decision-making are known sources of worry, burden, and distress for carers,[83, 89] and some studies have reported this may influence the plwMND's choices regarding gastrostomy.[87, 88]

Communication difficulties

Most plwMND will develop difficulty speaking (dysarthria), and it is commonly one of the first presenting signs of MND. The ability to produce clear speech declines as it becomes progressively slurred and slow. Other features interfering with intelligibility include dysphonia (resulting from vocal cord dysfunction), hoarseness, increased jitter, and respiratory muscle weakness.[85, 92, 93] PlwMND have rated speech loss as the worst aspect of the disease.[91, 94] Speech loss affects a plwMND's sense of identity, self-consciousness, participation in daily activities, relationships and engagement with health and social care.

Dysarthria has a consistently negative impact on a plwMND's psychological wellbeing. Consequently, psychologists should be alert to the symptoms and impact of dysarthria and be willing and able to make timely referrals to appropriate services, such as speech pathology services.[90, 91]

Sometimes, temporary measures such as writing or typing are employed when speech becomes unintelligible. However, as the disease progresses, communication becomes more complicated (especially when there is a loss of hand function as well as speech), though it remains essential to the plwMND, their carers and loved ones, and their care team. PlwMND report that communication is strongly linked with personal and social relationships[96] and is integral to their quality of life.[97-99]

Psychologists are likely to benefit from participating in education and training to familiarise themselves with assistive technologies, including augmentative and alternative communication (AAC) devices, and their benefits and limitations, as they have become essential and standard elements of MND care.[95]

Augmentative and alternative communications (AACs) vary in complexity and ease of use. They include communication with no- or low-tech (e.g., gestures, facial expressions, handwriting, topic and alphabet boards, eye-linking systems) or high-tech (e.g., tablets with voice-banking outputs). High-tech AACs require little or no head or limb movement and allow independent communication.[99]

AACs may assist in preserving identity, autonomy, and control in plwMNDs' personal and social lives, as well as concerning their treatment needs and preferences.[98] Communicating in various social contexts and life situations ('communicative participation') is now considered an important treatment outcome,[103] and AACs can be an important support for communicative participation.[101] AACs can be critical in maintaining an emotional connection with loved ones,[104] facilitating social and community participation, reducing depression, and enhancing quality of life.[105] Voice banking or creating a personalised synthetic voice that approximates a plwMND's natural voice, can help preserve identity and social networks and facilitate a sense of control.[106] Furthermore, as the disease progresses, communication regarding a plwMND's physical, emotional, cultural and treatment needs and preferences becomes critical.[96] These communication devices help plwMND and their carers maintain autonomy, control, decision-making ability, and improve quality of life.[98, 105]

Psychologists can assess and monitor the communicative participation of plwMND and their carers and encourage them to obtain timely AAC education, treatment, and equipment.[100, 101] Psychologists should be aware that assistive technology education, access and provision to plwMND and their carers are some of the key roles of MND Associations in Australia.[7] Further information regarding assistive technology is available at the MND Connect[54] website. Psychologists, via discussion and appropriate referral, may facilitate early adoption of AAC assessment since postponement till intelligibility is severely impaired can negatively influence acceptance, uptake, and successful use of the technology.[102] To make plwMND feel comfortable and less frustrated when they are using AAC, it is important that psychologists demonstrate patience and provide sufficient time for the plwMND to express themselves and feel heard (rather than guessing what is trying to be communicated).

Additionally, these technologies allow treatment when plwMND become restricted to the home. Telehealth services, self-reporting, and remote non-invasive ventilation can enhance accessibility, safety, and quality of care.

Psychologists, and other HCPs, can conduct therapy with plwMND and their carers via telehealth, which has a high adoption and adherence rate in this population.[107, 108]

Assistive technologies can empower plwMND and their carers to engage in treatment with an active role for a longer duration as the disease progresses, with clear consequences for psychological wellbeing.[98] Psychotherapy using an AAC device and software has been used successfully with plwMND to reduce psychological distress, better manage symptoms, and enhance dignity, autonomy, and self-esteem.[109] AACs can also enable valued participation in MND clinical trials.[110]

An exaggerated outburst of crying or laughing is often associated with upper motor neurone (pseudobulbar) dysarthria. This behaviour is referred to by several interchangeable terms, including ‘pseudobulbar affect’ (PBA), ‘pathological crying and laughing’ (PCL), ‘emotionality lability’ and ‘involuntary emotional expression disorder’.[23, 85, 111] Prevalence rates of this disorder vary between 25-50% of plwMND and are higher for those with predominantly upper motor neurone dysfunction compared to typical ALS or principally lower motor neurone dysfunction.[111] The PBA reaction is difficult to control, and the intensity of the emotional outburst is disproportionate and often inappropriate to the circumstance, often causing the plwMND to become embarrassed and distressed. PBA is associated with lower quality of life, more negative social interactions and depression. The associated facial movements may also be painful. Consequently, social withdrawal risk increases.[85, 111, 112] Identifying underlying emotions in plwMND may be difficult because of the alteration of emotional expression caused by the disease.

Psychologists working with plwMND should be aware of the prevalence, symptoms, and impact of PBA, as disproportionate or inappropriate laughing or crying may also be associated with mood disorders (e.g., bipolar) or the consumption of illicit drugs and may, therefore, be misinterpreted. Although it can be challenging, psychologists should be able to delineate the differences between PBA and mental disorders, as this exaggerated emotional expression is commonly misdiagnosed as a mood disorder. For example, PBA crying episodes are explosive and brief, without the persistent sadness associated with depression.[112, 113]

Neuropsychiatric impairments

Recently, there has been a movement towards reconceptualising ALS as a multisystem disorder involving a spectrum of cognitive and behavioural changes. The spectrum ranges from plwMND demonstrating a pure motor phenotype to those with ALS–Frontotemporal Dementia (FTD) and accounts for a variety in the severity of cognitive and behavioural changes.[114] Using this approach, approximately 15% meet the diagnostic criteria for ALS-FTD, 35% have a range of less severe and focal neuropsychological and motor impairments, and about half fall within the normal range.[115] Criteria have been developed to assess FTD,[114] categorising plwMND into ALS, ALSci (with cognitive impairment), ALSbi (behavioural impairment), ALScbi (both cognitive and behavioural impairment), and ALS-FTD.

Psychologists receiving referrals for plwMND should be aware of recommended neuropsychological tools, particularly since cognitive screening is a common request.[44]

The Edinburgh Cognitive and Behavioural ALS Screen (ECAS)[116] and the ALS Cognitive Behavioural Screen (ALS-CBS)[117] are the two most commonly used measures that are sensitive and specific to ALS impairments, have been validated and standardised and are recommended for use with plwMND.[115, 118, 119] Common cognitive issues include deficits in verbal fluency, executive functions, social cognition (e.g., abnormal emotional processing, difficulty recognising facial expressions, emotional recognition, and understanding social situations), and language (e.g., word retrieval problems, spoken and pragmatic language) and there is large individual variation in memory problems. In terms of behaviour change, apathy is the most common form, while a loss of sympathy, disinhibition, egocentric behaviour, perseverative and stereotyped behaviours, hyperorality and an increase in eating have been noted.[112, 115, 120]

The cognitive performance of plwMND has been associated with higher levels of depression, with ECAS subtests evaluating social cognitive deficits and inhibitory control demonstrating a relationship.[121, 122] However, research regarding the relationship between anxiety and cognitive function has been inconsistent.[123] The evidence is more consistent regarding an association between behavioural impairments and both depression and hopelessness,[124] although there is a clear overlap between depressive and behavioural symptoms, such as apathy.[123, 125]

Cognitive and behavioural changes may be evident in communication difficulty, passivity, trouble with decision-making, and impaired social interactions. Apart from the negative psychological impacts on the plwMND, these changes are also associated with disease progression, treatment adherence, survival and caregiver psychological wellbeing. The presence of behaviour changes in the plwMND is predictive of lower quality of life, and higher levels of burden and depression in MND carers.[115, 127-129]

Behaviour changes also become challenges for carers and HCPs, such as psychologists, as they have been associated with treatment non-adherence in plwMND manifesting symptoms of FTD.[126]

Treatment adherence is significant as it may extend survival (e.g., non-invasive ventilation), increase quality of life and reduce caregiver burden.[123] Extremely low compliance rates with recommendations for mental health needs have been noted among plwMND with only mild cognitive and behaviour change symptoms.[131]

The consequences of cognitive and behaviour changes further emphasise the importance of psychologists being alert to these changes. Due to the prognostic and therapeutic consequences, early detection and cognitive assessment to assist in the timely management of symptoms is considered a critical aspect of MND care.[114, 118, 123, 130] Cognitive assessments have benefits for plwMND, carers and treating HCPs, as they lead to improvements in clinical care. These may be manifested in the identification and validation of cognitive and behaviour changes, better comprehension of disease impact and progression, informing and directing care (e.g., referrals to specialist services, adapting care plans), and providing a guide to the plwMND's ability to make decisions.[118]

Further information to assist psychologists to link plwMND and their carers to services and support (including the National Insurance Disability Scheme and My Aged Care) is available at the MND Connect[54] website.

A higher prevalence of personal and familial history of psychiatric disorders before MND diagnosis has been demonstrated. There is an association between being diagnosed with MND and being hospitalised for psychiatric reasons (specifically schizophrenia, bipolar disorder, depression, or anxiety) in the preceding year. In this context, the presenting psychiatric symptoms were interpreted as prodromal features of MND.[26] However, there is also a relationship between a diagnosis of depression 5+ years prior to MND diagnosis, further supporting the hypothesis that depression is a risk factor for cerebral neurodegeneration.[26] An association has also been demonstrated

between a history of psychiatric disorders, particularly mood disorders, which may have preceded an MND diagnosis by many years, and the development of cognitive and behavioural disorders in plwMND.[132]

As part of their history-taking, psychologists should familiarise themselves with the plwMND and their relatives' history of psychiatric disorders, as they are risk factors for the development of cognitive and behavioural disorders.

Depression and anxiety among plwMND

Given the impact of the symptoms, the relentless progression, and the poor prognosis, it is unsurprising that mental health issues are commonplace among plwMND and their carers.[133] Depression and anxiety in plwMND are linked with several negative outcomes, including decreased quality of life, reduced length of survival times, and higher risks of suicide and mortality.[64, 134-136]

Research into the prevalence of depression among plwMND has yielded inconsistent results, with prevalence rates ranging from 10-45%.[133, 137] Notably, this epidemiological data has been obtained from a range of self-report scales that vary in sensitivity and specificity for detecting a depressive disorder.[133]

Psychologists should be aware of the strengths and limitations (e.g., somatic overlap) of merely relying on self-report measures of psychological distress.

An additional issue with the use of these questionnaires is that the highly prevalent use of antidepressant medication may have reduced depressive symptoms to a level below the cut-off for 'caseness.' A study in the United Kingdom tracking the trajectory of a cohort of 1120 plwMND over 30 months found that 88% of those using antidepressants reported symptoms below caseness for depression using questionnaires, suggesting that pharmacological treatment may reduce the identification of depression using these tools.[138] Depression questionnaires are often used in clinics and research due to the labour-intensive nature of the gold standard of diagnosis, a clinical interview based on the Diagnostic and Statistical Manual of Mental Disorders (DSM-5-TR).[138]

Psychologists are encouraged to use their assessment skills (e.g., clinical interview) to evaluate the psychological status of the plwMND beyond the questionnaire results. Further information to assist HCPs regarding factors to consider in the provision of psychological care and support can be found in the NICE (2016) international guidelines[44] (Section 1.6 'Psychological and social care support') and on the MND Connect[54] website.

Recent larger studies seem to have confirmed high rates of depression prevalence in plwMND. A systematic review and meta-analysis of the pooled prevalence of depression in plwMND reported by 46 studies yielded an overall summary prevalence of 34%.[133] The study tracking the trajectory of a cohort of 1120 plwMND over 30 months reported a prevalence rate of 23%. However, this figure may be an underestimation as caseness may have been reduced because of antidepressant therapy. This research also found depression to be associated

with being female, younger, level of disability and a poorer quality of life. Depression was also more common shortly before or after diagnosis and as MND progressed through its various stages (known as King's stages of progression).[138, 139] Almost 72% of plwMND with depression as a comorbidity reported onset within three years before their diagnosis of MND, providing further support for the hypothesis that depression may be a prodromal feature of MND.[26, 132, 138]

Psychologists should be regularly investigating for symptoms of depression, particularly early in the disease course.

Some authors have suggested that screening for depression should begin at the initial presentation to a neurologist.[138]

Anxiety in plwMND has not been investigated as widely as depression,[137] and where it has, the reported prevalence rates are also highly variable, ranging from 8-88%.[123, 140, 141] Researchers have suggested that, similar to depression, this may be due to the variety of self-report scales used to detect anxiety.[138] Analogously, perceptions of independence are associated with both depression and anxiety.[142] Also, similar to depression, high prevalence rates for anxiety are evident pre- and post-diagnosis of MND.[26, 143]

For psychologists, the same recommendations for depression apply to anxiety. These include clinical assessment based on the DSM-5-TR, being aware of risk factors and the impact of anxiety, and screening early and regularly in the disease course.[138]

When interacting with ATSI plwMND and their families, psychologists should be aware that many prefer to use the term 'social and emotional wellbeing' (SEWB) rather than mental health or mental illness. SEWB is more holistic and acknowledges social, emotional, spiritual, and cultural aspects of wellbeing, including the impact of policies and past events.[56]

Decision-making

A model has been developed to assist HCPs, such as psychologists, to engage plwMND and their carers in patient-centred decision-making in various domains including gastrostomy. The model emphasises the HCPs role in supporting the perceived control of plwMND by exploring and clarifying values (including cultural), establishing choices and discussing the optimal timing of these choices to ensure that enough resources have been obtained to ensure that these decisions are informed. HCPs, including psychologists, should then support the choices of plwMND, whether that be to adopt early symptom management, defer decision-making to a later stage or refuse treatment. Each of these options should be accepted and presented as valid.[67]

The value of these choices is particularly important in domains where the clinical benefit of the treatment is less clear. For example, compared with non-invasive ventilation (NIV), the benefits derived from gastrostomy in terms of quality and quantity of life are less clear.[144] While such an approach may be challenging for some psychologists, it respects the autonomy and values of the plwMND. This approach increases the perception of control and satisfaction with the decision-making process among plwMND.[86] It has also been suggested that plwMND may value hearing about the experiences of other patients in a manner that demonstrates both the risks and benefits associated with their choices, as this may facilitate more informed decisions. This exposure and specific information regarding the practicalities associated with PEG feeding may contribute to the specific education requested by plwMND and their families regarding interventions.[88]

Patient-centred decision-making can be improved with the input of high-functioning multidisciplinary teams (MDT), as recommended by clinical guidelines.[44, 67] Communication within these teams is paramount to ensure that the plwMND's wishes and needs (e.g., to delay gastrostomy) are respected. In addition, communication, cognitive and behavioural impairments can be significant barriers to engaging plwMND in decision-making. Hence, psychologists should introduce the process early.[145] Clearly, communication, cognitive and behavioural impairments can influence the plwMND's decision-making capacity regarding treatment options, palliative care, and finances. Consequently, timely and appropriate referrals should be made within the MDT to begin sensitive discussions regarding issues such as medical power of attorney.[146]

Psychologists can present these decision-related discussions in the context of preserving the plwMND's preferences and identity.

Gastrostomy feeding tube placement and ventilation options should be regularly discussed with plwMND. However, a synthesis of qualitative research on decision-making in these domains has demonstrated that such choices are not simply functional issues for plwMND, and emotions play a significant role. These interventions may be perceived as a threat to identity, independence and normality, requiring the plwMND to balance quantity versus quality of life values.

HCP interactions may influence decision-making. Therefore, it is critical that these exchanges contribute to the autonomy of the plwMND by promoting, rather than threatening, their perception of agency. Psychologists should maintain appropriate boundaries and high self-awareness as they engage with this complex ethical situation that involves trying to optimise the health outcomes of a plwMND while respecting their right to autonomy. Decision supports should consider emotional factors associated with the plwMND's choices and facilitate autonomy.[147]

End-of-life considerations

The MND Association of Australia has developed guides for counselling regarding treatment options and withdrawal, palliative care, discussing and planning for end-of-life, and exploring and supporting plwMND wishes and preferences (see MND Connect[54]). Unfortunately, evidence suggests that there remains some distance between recommendations and the practice of medical and other health professionals.[148] For example, the role of palliative care is often not discussed until late in the disease trajectory.[149] This lack of best-practice application can have a negative impact on plwMND and their carers. Delaying communication, decision-making and planning, or an HCP ignoring or disregarding a plwMND's wishes, have been described as violations of patients' rights via non-adherence to the ethical principles of autonomy and non-maleficence.[148]

There are numerous challenges for plwMND, carers and psychologists as illness progression becomes more apparent, and HCPs are often concerned that raising the topic of advance care planning (ACP) may diminish hope and create negativity. However, due to the common delay in diagnosis and rapid illness progression, discussing ACP early and at regular intervals is recommended.[149, 150]

Integrating palliative care at an early stage facilitates better acceptance of progressive losses, enhances control, empowers decision-making, enables tailoring of care, and improves psychological and social consequences for patients and carers.[149] While individual variation exists, plwMND and their carers generally prefer to have the chance to discuss what they should expect as MND progresses. ACP can result in greater concordance between patients' wishes and the treating team.[150]

> It is recommended that psychologists raise ACP discussions in the context of patient autonomy, security, control, courage, dignity, and independence.[44, 151] This approach encourages the perception of ACP as a tool facilitating quality of life, rather than inferring imminent death. Further information to assist psychologists in initiating end-of-life discussions and navigating end-of-life care is available at MND Connect[54].

Carers, as well as plwMND, can be reassured by ACP documents[44] with benefits such as a reduction in decisional burden, prevention of regret, improved preparation and enhanced family communication and concordance with patient wishes.[151] Conversely, some plwMND do not wish to discuss ACD till later in the disease course for various reasons, such as a wish to live in the moment.[155] The introduction of a discussion of ACP should match patient readiness.[151]

Assessing and facilitating readiness is a key role for psychologists. Factors such as a positive therapeutic relationship, trust, high-level communication skills and creating an environment of openness and acceptance facilitate readiness towards ACP discussions.[55] For plwMND who are not yet ready to discuss ACP, communication opportunities should still be presented respectfully at regular intervals, and psychologists should offer support for decision-making at significant points of deterioration.[148, 149] Further information to assist HCPs deliver a coordinated multidisciplinary approach to MND symptom management is available at MND Connect[54].

For ATSI plwMND and their families, ACP is referred to as 'advanced care yarning' and entails discussions with a doctor, an Aboriginal Health Worker, or family about future treatment, beliefs, and values.[152] For example, advanced care yarning can help identify the plwMND's preferences as they come close to the end of life, such as returning to Country.[153] Further resources, including templates and videos, to assist HCPs with starting these discussions and facilitating these choices in a culturally safe manner are available at the Palliative Care Australia,[154] Australian Indigenous HealthInfoNet [56] and Gwandalan National Palliative Care Project [57] websites.

Given the symptoms associated with disease progression, it is unsurprising that the wish to hasten death is not uncommon amongst plwMND.[148] Examples may include refusal of invasive and non-invasive treatments and requesting the withdrawal of treatments. More direct methods may include voluntary stopping of eating and drinking, suicide, and voluntary assisted dying (VAD).

PlwMND are at higher risk for suicide compared with people without neurological diseases, and psychologists should be aware that this risk is generally greatest among younger patients and in the first year after diagnosis.[135, 156-158]

Suicidal ideation is not unusual after the shock of an MND diagnosis.[149, 159] Other higher risk times include when the plwMND perceives that the symptoms of the disease have surpassed their ability to tolerate or some may act prior to the onset of paralysis, preventing them from the option.[148, 160, 161] Research has also revealed that carers may also experience suicidal ideation during significant points in the progression of the disease.[162]

Psychologists should be aware of the psychological toll on plwMND and their carers and are strongly encouraged to monitor their mental health and suicidal risk from the time of diagnosis onwards.[158]

VAD has only recently been legalised in most parts of Australia, and initial data from Western Australia indicates that neurological diagnoses (predominantly MND) were the second most common among VAD deaths in 2022-2023.[163] As far as carers are concerned, psychologists should be aware of conflicting evidence on whether VAD increases or decreases bereavement issues. There is anecdotal evidence that some MND carers have experienced disenfranchised grief.[164]

Psychologists treating MND carers should be aware of risk factors for poor bereavement outcomes for those whose family members participated in VAD.

These risk factors include ambivalence, moral disagreement regarding assisted death, and communication difficulty at the end of life. Protective factors included appropriate preparation and planning, a perception of autonomy, less suffering for the patient and supporting the patient's wishes, although these protective factors may also act as risks in some circumstances.[173] There are also unique concerns for bereavement associated with VAD, including potential social stigma and moral conflicts in families.[174]

Psychological support is strongly recommended for family members of those choosing this form of death, both pre- and post-VAD.[165, 166]

End-of-life matters need to be approached in a culturally appropriate and responsive manner.[167] There are customs and sacred practices often conducted by ATSI people prior to, during and after the individual's passing. HCPs should be aware that these practices and choices may vary significantly across different ATSI groups, and communication with the plwMND and their family is essential to ascertain their specific cultural needs and choices. For example, direct reference to terms such as death and dying may cause discomfort in some ATSI people and phrases such as 'passing on' or 'finishing up' may be more culturally appropriate.[168]

Having the choice of location (e.g., on Country, at home, hospital) where they will pass may also be important.[169] Other factors to consider include kinship, communication styles, and 'Sorry Business' (ATSI term for the passing of a person). For example, traditional ATSI practices associated with grief and bereavement may include not using the deceased person's name, avoiding eye contact, and a smoking ceremony. It should also be noted that there is a diversity of perspectives on VAD among ATSI peoples.[170] Psychologists can access a Palliative Care and End-of-Life Care Portal specifically designed to help HCPs provide culturally appropriate care for ATSI people, their families and communities at the Australian Indigenous HealthInfoNet[56] website. Additional training and resources are available at the Gwandalan National Palliative Care Project[57], Caresearch[171], and the Indigenous Program of Experience in the Palliative Approach[172] websites.

Psychologists should be aware of the challenges experienced by diverse populations, including culturally and linguistically diverse (CALD) and lesbian, gay, bisexual, transgender or intersex (LGBTI) groups. For example, barriers to engaging with HCPs in CALD populations include language and literacy, death and dying considered taboo subjects, stigma regarding hospice care, lack of translated and culturally appropriate resources, racism, discrimination, cultural stereotyping, religion, and health beliefs. The Western values of autonomy and self-determination associated with ACP may not align with the values of some CALD individuals. Hence, ACP is less common in CALD populations.

Barriers often experienced by LGBTI Australians include social isolation, discrimination, potentially strained relationships with biological family, and difficulties identifying alternative decision-makers. The Department of Health and Aged Care[175] has summarised the common barriers to palliative care for diverse Australians, including CALD and LGBTI populations. Psychologists should be aware of the concept of intersectionality and be cognisant of the heterogeneity of views and behaviours of individuals within diverse populations. Respectful acknowledgement of different perspectives regarding ACP is important, and consent to begin these discussions is helpful in facilitating trust.[176]

Referral to language support and multilingual resources may also facilitate end-of-life discussions. An extensive range of tips for communicating with CALD individuals has been developed by Diversicare[177]. It is essential that treating psychologists are aware that making heteronormative assumptions may cause distress for LGBTI individuals. For example, ACP should recognise 'family of choice.' Further information on providing appropriate care to CALD and LGBTI populations is available at the ELDAC[176] and Caresearch[58] websites.

Psychological treatment for plwMND

There is a paucity of research into psychological interventions for plwMND, and reviews of these treatments have noted that methodological limitations limit the quality of these studies.

> The existing evidence base is insufficient to recommend specific psychological therapies for plwMND.[178, 179]

Although caution should be used in the interpretation of the results due to methodological deficits, the following provides a brief overview of the key psychological interventions trialled.

MND-tailored Mindfulness-Based Stress Reduction demonstrated reductions in depression and anxiety, and improved quality of life in plwMND. Meditative training involved bringing attention to the present moment and promoting the management of emotions via acceptance of feelings and sensations without judgement.[180, 181]

Acceptance and Commitment Therapy (ACT) has been successfully trialled to improve the psychological wellbeing of people with other neurogenerative diseases. This approach integrates acceptance, mindfulness, motivation, and behaviour change techniques. The aim is to decrease maladaptive attempts to control negative internal experiences (e.g., thoughts, emotions, physical sensations) and to facilitate engagement in activities that enhance one's life. The results of a feasibility and acceptance study were suggestive of small reductions in anxiety and depression and improvements in non-physical quality of life.[60]

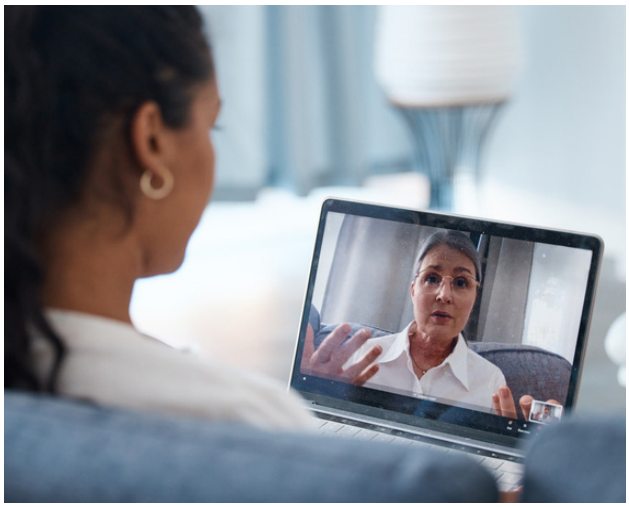
Cognitive-behaviour therapy (CBT) has also been trialled with mixed results. A randomised controlled trial of tailored CBT based on the stress-coping model was delivered to plwMND and their carers. However, the study was halted prematurely due to recruitment problems. Preliminary data was indicative of significantly less decline in mental aspects of quality of life for those in the intervention group.[182] Another study, using a quasi-experimental design, delivered four sessions of CBT coupled with counselling techniques. The results demonstrated a significant decrease in symptoms of depression and anxiety.[183]

A psychodynamic hypnosis intervention has also been trialled with plwMND. An initial uncontrolled pilot intervention included four hypnosis sessions, modified for the plwMND according to their psychological status and physical symptoms. Hypnotic suggestions focused on developing symptom control (e.g., muscle pain, emotional lability), resilience, self-consciousness, and illness acceptance. Domiciliary visits and self-hypnosis training were also integrated over a month. The results demonstrated reductions in depression and anxiety and improvements in quality of life. Participants also reported perceived decreases in physical symptoms such as pain, sleep disorders, emotional lability, and fasciculations.[184] A quasi-experimental design later evaluated the same intervention but also integrated matched controls and evaluation 6 months post-intervention. The results were similar to the pilot, demonstrating a reduction in depression and anxiety symptoms post-intervention.[185]

The results of an uncontrolled feasibility study using a Dignity Therapy (DT) intervention for plwMND and their carers have also been reported. This intervention involved the creation of a document about their life based on a life reflection interview. Salient memories, achievements and events are discussed with the aim of recognising life dignity and meaning and reducing existential distress. No significant differences were observed on quantitative measures of dignity-related distress, hopefulness, and spiritual wellbeing for plwMND or on measures of burden, hopefulness, anxiety, and depression for carers. However, qualitative data indicated that plwMND endorsed the treatment, and carers also found comfort in the DT document.[186, 187]

The impact of expressive disclosure on the psychological wellbeing of plwMND was evaluated with a randomised controlled trial. This intervention involved either writing or speaking about their deepest thoughts and feelings related to their experience with MND for 20 minutes over 3 days, with the hypothesis that expressing rather than withholding stressful events leads to reduced psychological distress. The results demonstrated that those who disclosed thoughts and feelings about MND had higher psychological wellbeing at follow-up. However, ambivalence over emotional expression mediated the effect, and the authors concluded that expressive disclosure may be helpful for plwMND but only for those individuals who have difficulty expressing emotions.[188]

Given the symptoms, burden, and lack of mobility associated with MND, traditional face-to-face psychological interventions may not be suitable for plwMND or their carers. Digital mental health interventions (DMHI) offer a promising alternative. Pinto et al. (2023)[189] describe a DMHI designed for plwMND and their carers which was comprised of an interactive website with self-guided emotion regulation strategies, and tips for managing



distress and improving psychological wellbeing based on CBT, ACT, mindfulness and compassion-focused therapy. Qualitative evaluation indicated that participants found the DMHI useful and acceptable, with benefits noted in developing control and positivity. DMHI can also be helpful in rural and remote areas.

One size does not fit all as far as psychological interventions for plwMND and their carers are concerned. Individual preferences and needs may be a facilitator or barrier to engagement. One study exploring the needs and preferences of plwMND in relation to psychological interventions identified key variables to engagement and success (listed in the box below), and the authors suggested that ACT is well-suited to implementing these factors.[190]

Key factors to engagement and success with psychological interventions included individual tailoring, flexibility, and the facilitation of autonomy. The implications were that psychological intervention should:

1. Assist the person to learn skills to manage negative thoughts and emotions
2. Increase personally meaningful behaviour,
3. Re-direct the focus to what can be controlled
4. Increase attention to experiences in the present.[190]

Challenges to carers' psychological wellbeing

Most of the provision of care for plwMND comes from the community through the role of informal carers, usually spouses or children.[28] Carers also provide physical and emotional support and play an important role in clinical decision-making.[191]

Carer availability and wellbeing are integral to the care of a plwMND. Without the available capacity of a carer, plwMND would not be able to continue to live at home till the end stage of their lives. Consequently, unmarried plwMND are more likely to die in an acute care facility.[192] Further, 80% of MND carers live with the plwMND,[193] and those residing with the plwMND have poorer mental and physical health than carers who did not have primary residence with the plwMND.[194] Importantly, there is a strong bidirectional relationship between the wellbeing of the plwMND and their carers, particularly on psychological variables, further emphasising the importance of addressing the needs of carers.[195-198]

Caring for a plwMND is very demanding. A wide range of tasks may include all activities of daily living and personal care, with an average time of 9.5 hours per day spent in the caring role.[199] The time involved in the provision of care usually increases as the disease progresses.[200] The mental health of MND carers is challenged throughout the course of the disease and beyond. The psychological toll of being the carer of a plwMND is well-established.[201] The disease has been described as unremitting and exhausting, with no hope for recovery.[202] High levels of depression, anxiety, fatigue, strain, social handicap, and increased risk for anticipatory grief (beginning from the time of diagnosis) and prolonged grief disorder (PGD) post-death, as well as lower quality of life, have been regularly demonstrated.[201, 203-207]

> Psychologists should be aware of a range of factors that contribute to or protect against the development of mental health problems among carers of plwMND.[208]

The severity of MND may impact burden, anxiety and depression.[209] Carers providing 22 or more hours per week were 2.65 times more likely to be experiencing psychological distress.193 Positive coping strategies and social support strongly contribute to psychological wellbeing and quality of life.[208, 210] Low self-esteem, worries about end-of-life, existential concerns and changes in identity, roles and relationships are all associated with levels of MND carer psychological distress.[52, 210-213]

A large Australian study found 54% of MND carers to be at moderate risk of developing PGD and almost 10% to be in the high-risk group.[214] Depression increased the risk of PGD by 18 times, while anxiety increased this risk by 8 times. Other significant contributors to PGD included poor family functioning, a recent bereavement (<12 months), being a spouse/ partner of the deceased plwMND, insufficient support during the MND journey, the deceased

being under 60 years old, and a brief period of caring under 1.5 years. Sixty-three per cent reported that they needed bereavement support beyond that provided by family and social networks,[214] highlighting a clear role for psychological intervention.

Anticipatory grief, which can begin from diagnosis, is also highly prevalent in MND carers and increases the risk of developing PGD.[207] Factors associated with anticipatory grief, post-death grief, and prolonged grief in MND carers should be targeted early in the disease process. These include carers' need for substantially more information about MND progression, coping strategies to assist with relationship changes with the plwMND, a decrease in carer depression and anxiety, better planning for the future passing of the plwMND, adjustment and life-rebuilding success post-death of the plwMND. Factors influencing all three types of grief include negative experiences as a carer, loss fixation, insufficient end-of-life and bereavement support, emotional avoidance coping style and a lack of psychological support.[207]

The clinical implications are that early carer education about MND can mitigate grief but should be coupled with psychological support, as increased knowledge may potentially increase the grief response. Psychological interventions for carers should be introduced from the point of diagnosis due to the massive emotional impact associated with role changes and relationships. Psychological interventions with MND carers should aim to reduce emotional avoidance and move towards acceptance and better management of depression and anxiety.[207] Furthermore, psychological support for the carer should not cease at the point of death of the plwMND.

Numerous studies have demonstrated a significant carer burden associated with the role. Carer burden has been defined as the combined physical, emotional, social, and financial impacts linked with the role.[218] Factors associated with higher carer burden include greater behavioural and physical impairment of the plwMND, symptoms of depression in the carer, insufficient information and training regarding MND and care skills, limitations associated with the role, and the impact on relationships.[96, 204, 215] Predictors of burden include higher carer psychological distress (explaining 39% of the variance),[128] hours of care provided and lower carer quality of life.[204] Other carer factors associated with differences in burden include resilience, coping style, problem-solving skills, and use of social supports.[28, 219]

Psychologists can use knowledge of contributing factors to identify vulnerable individuals at risk for high levels of carer burden, and this insight can guide the development and implementation of interventions.[215] Some carer factors may be modifiable, and this is further supported by evidence regarding mediating factors. Psychologists may encourage reappraisal of caring responsibilities, as positive aspects of the role may moderate perceived burden.[204] Protective factors include the appreciation of positive experiences, personal satisfaction, development of a life outside of caring, making time for self-care, finding meaning in the caring role, and a sense of hope.[196, 198, 216, 217] The identification of positive factors associated with being an MND carer may balance, protect and reduce distress among those in this role and should inform psychological interventions.[216]

Psychological interventions for the carers of plwMND

Interventions targeting the psychological wellbeing of MND carers is a relatively new field. Hence, there are limited studies on this topic, and most of the research in this domain has significant methodological weaknesses.

> The current evidence base is insufficient to recommend specific psychological therapies for the carers of plwMND.[220]

The following provides a brief overview of the main psychological interventions trialled. However, given the methodological issues, the results should be interpreted with a degree of caution.

Four studies have implemented mindfulness interventions. One study employed an MND-tailored Mindfulness-Based Stress Reduction treatment, including meditative training to bring focus to the present moment. It demonstrated improvements in psychological wellbeing, including relaxation skills, emotional self-regulation, and acceptance.[180] A similar intervention also using meditation as a core component did not demonstrate any psychological wellbeing benefits on quantitative measures. However, qualitative data indicated that carers valued hearing from others in similar situations, having time explicitly allocated for them rather than focusing on the plwMND, and having professional input.[221] Another study evaluated the effectiveness of a website-based, non-meditative (Langerian) mindfulness intervention. Carers reported decreased burden, depression, and anxiety, with increases in energy and emotional wellbeing.[222] Finally, a randomised controlled trial evaluated a blended psychosocial support program based on Acceptance and Commitment Therapy (ACT), including a mindfulness component. However, qualitative data indicated that MND carers did not find this part of the intervention helpful. Overall, the blended psychosocial support program demonstrated improvements in controlling thoughts about caregiving, but there were no differences in other psychological outcome measures. Qualitative data also indicated higher perceived control over caring, acceptance of thoughts and feelings, more attention to their personal relationships, felt acknowledged and empowered to make choices regarding their needs.[223, 224]

A CBT intervention based on the stress-coping model was halted prematurely due to recruiting problems. Preliminary data demonstrated significant improvements in carer quality of life and reductions in burden.[182]

Chaplains delivered a carer intervention based on self-disclosure and human development to improve wellbeing via an exploration of role-related meaning. No effect was found on a range of quantitative psychological wellbeing measures, although quantitative data revealed benefits associated with role reflection and the opportunity to process emotions.[225]

A mutual support group intervention for family carers found that the experience of caring was profoundly different depending on whether the carer was a son/daughter or a partner of a plwMND. More psychological wellbeing benefits and harmony were evident in the children of the plwMND group. Overall, social comparison with and support from peers assisted participants in coping more effectively, better equipping them for their caring role and reducing isolation. Connecting with these networks facilitated the activation of personal and social resources for wellbeing.[226]

A case-management-based intervention for carers was evaluated in a randomised controlled trial. No effect was found on psychological wellbeing (burden, quality of life).[227]

Youth carers of plwMND tend to be isolated and receive minimal MND-specific care training.[228] A one-day training intervention for youth carers of plwMND was based on the tenets of the individual and family self-management theory and focussed on improving the self-efficacy of these children in their caring role. This study underscored the importance of providing training that targets specific MND care needs, as it demonstrated a significant increase in confidence with several care tasks (communication systems, use of respiratory equipment, using a power chair), translating to the eudaimonic psychological wellbeing aspect of environmental mastery.[229]

The importance of addressing individual needs in carer interventions

MND carers have reported a lack of acknowledgement of their needs and inadequate support. A large national survey of plwMND and their carers indicated that support for carers and family members should be a research priority.[231] Support for carers has been compromised by a lack of integration of services across health and social care, inequities in access to coordinated palliative care, inadequate knowledge base of HCPs and poor monitoring of psychological issues.[232] Generic one-size-fits-all interventions may be short-sighted and unsatisfactory. For example, while many MND carers report that they need help with bereavement from beyond their social networks, they would prefer informal social support from within these networks if they were better equipped. Unfortunately, carers' social networks are often ill-equipped to provide the support is needed. Education within a compassionate communities approach to palliative care allows for the upskilling of carers' social networks for all phases of caregiving, including bereavement. This approach also improves cultural responsiveness and tailored appropriateness of care.[25, 233]

> MND carers have reported a lack of insight, understanding and support from HCPs and their informal social networks,[230] further emphasising the importance of HCPs being cognisant of the psychological issues experienced by this population.

MND carers have derived benefits from interventions that address their individual needs. One study used the Carer Support Needs Assessment Tool (CSNAT) to identify carer needs. Benefits conferred included the acknowledgement of the overwhelming carer journey, validation of the role and empowerment, and reassurance of support.[234] The person-centred care model provided by the Australian MND Advisory service is designed to support the needs of plwMND and their carers. Research has demonstrated improvement in practical and emotional support from this service, as carers reported feeling supported, better equipped to make decisions about their wellbeing, and sufficiently informed.[32] An online intervention involved a 4-month palliative rehabilitation program to support the ability of MND carers

to handle the daily challenges of their role. Videos and virtual groups were conducted based on topics associated with the challenges and needs of MND carers. Carers made ecomaps to explore potential supportive relations during the disease trajectory and post-death of the plwMND. They were also offered customised Mindfulness-based Stress Reduction videos. Qualitative results indicated that carers perceived benefit from the peer support and the videos, felt reduced loneliness and frustration, and were more knowledgeable regarding future challenges.[235] Finally, a psychosocial needs assessment and intervention in low-resource settings was based on Rolland's theory of psychosocial typology for chronic illness. The intervention addressed perceived needs and psychosocial challenges through the different stages of MND progression and integrated ways to handle the challenges. Benefits derived by MND carers included increased confidence in performing the carer role.[236]

Interviews and concept mapping led to the development of a four-cluster map representing different carer needs at each stage: (1) early coping and adjustment; (2) maintenance; (3) transition to end-stage; and (4) coping with change and loss.194 A recent systematic review into MND carer needs across the disease span reclassified these stages into Stage 1) Early coping and adjustment after diagnosis; Stage 2) Across the caregiving course; Stage 3) Transition to the terminal stage; and Stage 4) Coping with change and loss after bereavement. Stage 1 needs include the communication process of the diagnosis, information about the diagnosis, and assistance coping with the diagnosis. Stage 2 needs include information, emotional support, formal support with care at home, needs relating to healthcare services (e.g., access, coordination), financial support, assistance with the dietary requirements of the plwMND, role and social changes. The needs of Stage 3 constitute information provision and access to services, contact with healthcare practitioners, the emotional impact of dying on carers, assistive technology, and ACP. Stage 4 needs include access to bereavement services and support.[237] Another categorised carers' needs into external support and assistance, patient-related factors, and carer psychoemotional factors. As the disease progressed, the frequency of needs based on patient-related factors increased, demonstrating that carers' needs are also dependent on the individual needs of the plwMND. Notably, in relation to the carer's need for external support, an additional source of stress for some carers was associated with uncooperative behaviours of plwMND and resistance to the involvement of external support and services. This was manifested in the plwMND refusing to engage with external services, refusing services and relying only on the carer.[238]

> Psychologists should be aware that, similarly to plwMND, the needs of an MND carer are individual and dynamic, changing as the disease progresses through different stages.

A recent study has integrated MND carer needs and stage-based problems using the Carers' Alert Thermometer (CAT). This tool promoted discussions with MND carers to elicit needs to be addressed and action plans to be developed. The results indicated that the CAT helped carers to clarify needs and work through solutions. Participants in the study suggested that the CAT be used at the time of diagnosis and then at regular intervals as the disease usually progresses quickly and carers' needs change rapidly. The authors also warned against HCPs and other service providers assuming carer needs and encouraged the use of tools such as CAT as they provide a systematic and supportive ongoing review of carer needs before challenges become overwhelming. The tool can provide

important insights and may raise discussions on sensitive topics. In turn, this requires that the treating HCP is educated about factors contributing to carers' psychological wellbeing and appropriate communication skills and obtain the knowledge to signpost carers to relevant information and support.[230]

The variety of individual MND carer needs necessitates the assessment and tailoring of psychological interventions.[220] A 'goodness-of-fit' approach has been proposed where individual carer risks and needs are evaluated and matched to tailored treatment.[25] Psychological interventions for MND carers often focus on hedonic or subjective wellbeing aspects such as burden, anxiety and depression and undervalue the importance of eudaimonic factors (e.g., self-acceptance, environmental mastery, positive relationships, personal growth, purpose in life, and autonomy) in psychological wellbeing.[220]

Treating psychologists should be aware of the factors contributing to the carer's situation when designing carer interventions, such as identifying those at higher risk of poorer psychological wellbeing outcomes[205, 214] and MND stage.[237] Intervention design should consider and incorporate both hedonic ('feeling') and eudaimonic ('function') aspects of psychological wellbeing as both are associated with positive outcomes for MND carers.[220]

References

1. Gentili, D., Deiana, G., Chessa, V., Calabretta, A., Marras, E., Solinas, C., . . . Azara, A. (2023). Quality of Life in Amyotrophic Lateral Sclerosis Patients and Care Burden of Caregivers in Sardinia during COVID-19 Pandemic. *Healthcare (Basel),* 11(11). http://dx.doi.org/10.3390/healthcare11111641

2. Korner, S., Kollewe, K., Abdulla, S., Zapf, A., Dengler, R., & Petri, S. (2015). Interaction of physical function, quality of life and depression in Amyotrophic lateral sclerosis: characterization of a large patient cohort. *BMC Neurol,* 15, 84. http://dx.doi.org/10.1186/s12883-015-0340-2

3. Young, C. A., Ealing, J., McDermott, C., Williams, T., Al-Chalabi, A., Majeed, T., . . . Tennant, A. (2019). The relationships between symptoms, disability, perceived health and quality of life in amyotrophic lateral sclerosis/motor neuron disease. *Amyotroph Lateral Scler Frontotemporal Degener,* 20(5-6), 317-327. http://dx.doi.org/10.1080/21678421.2019.1615951

4. Goutman, S. A. (2017). Diagnosis and Clinical Management of Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders. *Continuum (Minneap Minn), 23(5, Peripheral Nerve and Motor Neuron Disorders),* 1332-1359. http://dx.doi.org/10.1212/CON.0000000000000535

5. van Es, M. A., Hardiman, O., Chio, A., Al-Chalabi, A., Pasterkamp, R. J., Veldink, J. H., & van den Berg, L. H. (2017). *Amyotrophic lateral sclerosis. Lancet,* 390(10107), 2084-2098. http://dx.doi.org/10.1016/S0140-6736(17)31287-4

6. Fight MND. (2018), from https://fightmnd.org.au/

7. MND Australia. (2024).

8. Niedermeyer, S., Murn, M., & Choi, P. J. (2019). Respiratory Failure in Amyotrophic Lateral Sclerosis. Chest, 155(2), 401-408. http://dx.doi.org/10.1016/j.chest.2018.06.035

9. Longinetti, E., & Fang, F. (2019). Epidemiology of amyotrophic lateral sclerosis: an update of recent literature. *Curr Opin Neurol,* 32(5), 771-776. http://dx.doi.org/10.1097/WCO.0000000000000730

10. Marin, B., Fontana, A., Arcuti, S., Copetti, M., Boumediene, F., Couratier, P., . . . Logroscino, G. (2018). Age-specific ALS incidence: a dose-response meta-analysis. *Eur J Epidemiol,* 33(7), 621-634. http://dx.doi.org/10.1007/s10654-018-0392-x

11. Park, J., Kim, J. E., & Song, T. J. (2022). The Global Burden of Motor Neuron Disease: An Analysis of the 2019 Global Burden of Disease Study. *Front Neurol,* 13, 864339. http://dx.doi.org/10.3389/fneur.2022.864339

12. Masrori, P., & Van Damme, P. (2020). Amyotrophic lateral sclerosis: a clinical review. *Eur J Neurol,* 27(10), 1918-1929. http://dx.doi.org/10.1111/ene.14393

13. Global Burden of Disease Motor Neuron Disease Collaborators. (2018). Global, regional, and national burden of motor neuron diseases 1990-2016: a systematic analysis for the Global Burden of Disease Study 2016. *Lancet Neurol,* 17(12), 1083-1097. http://dx.doi.org/10.1016/S1474-4422(18)30404-6

14. Ryan, M., Heverin, M., McLaughlin, R. L., & Hardiman, O. (2019). Lifetime Risk and Heritability of Amyotrophic Lateral Sclerosis. *JAMA Neurol,* 76(11), 1367-1374. http://dx.doi.org/10.1001/jamaneurol.2019.2044

15. Luker, J., Woodman, R., & Schultz, D. (2023). The incidence and prevalence of motor neurone disease in South Australia. *Amyotroph Lateral Scler Frontotemporal Degener, 24(3-4), 195-202*. http://dx.doi.org/10.1080/21678421.2022.2108326

16. Deloitte Access Economics. (2015). Economic Analysis of motor neurone disease in Australia.

17. Dharmadasa, T., Henderson, R. D., Talman, P. S., Macdonell, R. A., Mathers, S., Schultz, D. W., . . . Kiernan, M. C. (2017). Motor neurone disease: progress and challenges. *Med J Aust,* 206(8), 357-362. http://dx.doi.org/10.5694/mja16.01063

18. Yedavalli, V. S., Patil, A., & Shah, P. (2018). Amyotrophic Lateral Sclerosis and its Mimics/Variants: A Comprehensive Review. Journal of Clinical Imaging Science, 8, 53. https://doi.org/10.4103/jcis.JCIS_40_18

19. Turner, M. R., & Talbot, K. (2013). Mimics and chameleons in motor neurone disease. *Pract Neurol,* 13(3), 153-164. http://dx.doi.org/10.1136/practneurol-2013-000557

20. Kim, W. K., Liu, X., Sandner, J., Pasmantier, M., Andrews, J., Rowland, L. P., & Mitsumoto, H. (2009). Study of 962 patients indicates progressive muscular atrophy is a form of ALS. *Neurology,* 73(20), 1686-1692. http://dx.doi.org/10.1212/WNL.0b013e3181c1dea3

21. Riku, Y., Atsuta, N., Yoshida, M., Tatsumi, S., Iwasaki, Y., Mimuro, M., . . . Sobue, G. (2014). Differential motor neuron involvement in progressive muscular atrophy: a comparative study with amyotrophic lateral sclerosis. *BMJ Open,* 4(5), e005213. http://dx.doi.org/10.1136/bmjopen-2014-005213

22. Oh, J., Kim, S. H., & Kim, J. A. (2019). Unmet supportive care needs mediate the relationship between functional status and quality of life in patients with amyotrophic lateral sclerosis. *Palliat Support Care,* 17(6), 650-654. http://dx.doi.org/10.1017/S1478951519000221

23. Oliver, D., Radunovic, A., Allen, A., & McDermott, C. (2017). The development of the UK National Institute of Health and Care Excellence evidence-based clinical guidelines on motor neurone disease. *Amyotroph Lateral Scler Frontotemporal Degener,* 18(5-6), 313-323. http://dx.doi.org/10.1080/21678421.2017.1304558

24. Hogden, A., Foley, G., Henderson, R. D., James, N., & Aoun, S. M. (2017). Amyotrophic lateral sclerosis: improving care with a multidisciplinary approach. *J Multidiscip Healthc,* 10, 205-215. http://dx.doi.org/10.2147/JMDH.S134992

25. Aoun, S. M., Cafarella, P. A., Rumbold, B., Thomas, G., Hogden, A., Jiang, L., . . . Kissane, D. W. (2021). Who cares for the bereaved? A national survey of family caregivers of people with motor neurone disease. *Amyotroph Lateral Scler Frontotemporal Degener,* 22(1-2), 12-22. http://dx.doi.org/10.1080/21678421.2020.1813780

26. Turner, M. R., Goldacre, R., Talbot, K., & Goldacre, M. J. (2016). Psychiatric disorders prior to amyotrophic lateral sclerosis. *Ann Neurol,* 80(6), 935-938. http://dx.doi.org/10.1002/ana.24801

27. Anestis, E., Eccles, F., Fletcher, I., French, M., & Simpson, J. (2020). Giving and receiving a diagnosis of a progressive neurological condition: A scoping review of doctors' and patients' perspectives. *Patient Educ Couns,* 103(9), 1709-1723. http://dx.doi.org/10.1016/j.pec.2020.03.023

28. Galvin, M., Ryan, P., Maguire, S., Heverin, M., Madden, C., Vajda, A., . . . Hardiman, O. (2017). The path to specialist multidisciplinary care in amyotrophic lateral sclerosis: A population- based study of consultations, interventions and costs. *PLoS One,* 12(6), e0179796. http://dx.doi.org/10.1371/journal.pone.0179796

29. Sennfalt, S., Klappe, U., Thams, S., Samuelsson, K., Press, R., Fang, F., & Ingre, C. (2023). The path to diagnosis in ALS: delay, referrals, alternate diagnoses, and clinical progression. *Amyotroph Lateral Scler Frontotemporal Degener,* 24(1-2), 45-53. http://dx.doi.org/10.1080/21678421.2022.2053722

30. Bongioanni, P., Borasio, G. D., Oliver, D. J., Romagnoli, A., Kapitza, K. P., Sidle, K., & Tramonti, F. (2023). Methods for informing people with amyotrophic lateral sclerosis/motor neuron disease of their diagnosis. *Cochrane Database Syst Rev,* 2(2), CD007593. http://dx.doi.org/10.1002/14651858.CD007593.pub2

31. Ozanne, A., & Graneheim, U. H. (2018). Understanding the incomprehensible - patients' and spouses' experiences of comprehensibility before, at and after diagnosis of amyotrophic lateral sclerosis. *Scand J Caring Sci,* 32(2), 663-671. http://dx.doi.org/10.1111/scs.12492

32. Aoun, S. M., O'Brien, M. R., Breen, L. J., & O'Connor, M. (2018). 'The shock of diagnosis': Qualitative accounts from people with Motor Neurone Disease reflecting the need for more person-centred care. *J Neurol Sci,* 387, 80-84. http://dx.doi.org/10.1016/j.jns.2018.01.026

33. O'Brien, M. R., Whitehead, B., Jack, B. A., & Mitchell, J. D. (2011). From symptom onset to a diagnosis of amyotrophic lateral sclerosis/motor neuron disease (ALS/MND): experiences of people with ALS/MND and family carers - a qualitative study. *Amyotroph Lateral Scler,* 12(2), 97-104. http://dx.doi.org/10.3109/17482968.2010.546414

34. King, S., Duke, M., & OConnor, B. (2006). People living with ALS/MND tell the diagnosis story: what happened before they knew *International Symposium on ALS/MND, 7 (Suppl. 1). Amyotrophic Lateral Sclerosis* (pp. 34-36). London: Taylor & Francis.

35. O'Connor, M., Aoun, S. M., & Breen, L. J. (2018). Australian family carer responses when a loved one receives a diagnosis of Motor Neurone Disease-"Our life has changed forever". *Health Soc Care Community,* 26(3), e415-e421. http://dx.doi.org/10.1111/hsc.12541

36. Anestis, E., Eccles, F. J. R., Fletcher, I., Triliva, S., & Simpson, J. (2022). Healthcare professionals' involvement in breaking bad news to newly diagnosed patients with motor neurodegenerative conditions: a qualitative study. *Disabil Rehabil,* 44(25), 7877-7890. http://dx.doi.org/10.1080/09638288.2021.2002436

37. Glennie, N., Harris, F. M., & France, E. F. (2023). Perceptions and experiences of control among people living with motor neurone disease: a systematic review and thematic synthesis. *Disabil Rehabil,* 45(16), 2554-2566. http://dx.doi.org/10.1080/09638288.2022.2104942

38. Larsson, B. J., Nordin, K., & Nygren, I. (2023). Symptoms of anxiety and depression in patients with amyotrophic lateral sclerosis and their relatives during the disease trajectory. *J Neurol Sci,* 455, 122780. http://dx.doi.org/10.1016/j.jns.2023.122780

39. Vignola, A., Guzzo, A., Calvo, A., Moglia, C., Pessia, A., Cavallo, E., . . . Chio, A. (2008). Anxiety undermines quality of life in ALS patients and caregivers. *Eur J Neurol,* *15*(11), 1231-1236. http://dx.doi.org/10.1111/j.1468-1331.2008.02303.x

40. Chen, D., Guo, X., Zheng, Z., Wei, Q., Song, W., Cao, B., . . . Shang, H. (2015). Depression and anxiety in amyotrophic lateral sclerosis: correlations between the distress of patients and caregivers. *Muscle Nerve,* 51(3), 353-357. http://dx.doi.org/10.1002/mus.24325

41. Maksymowicz, S., Libura, M., & Malarkiewicz, P. (2022). Overcoming therapeutic nihilism. Breaking bad news of amyotrophic lateral sclerosis-a patient-centred perspective in rare diseases. *Neurol Sci,* 43(7), 4257-4265. http://dx.doi.org/10.1007/s10072-022-05931-1

42. Silani, V., & Borasio, G. D. (1999). Honesty and hope: announcement of diagnosis in *ALS. Neurology,* 53(8 Suppl 5), S37-39; discussion S40-32.

43. Baile, W. F., Buckman, R., Lenzi, R., Glober, G., Beale, E. A., & Kudelka, A. P. (2000). SPIKES-A six-step protocol for delivering bad news: application to the patient with cancer. *Oncologist,* 5(4), 302-311. http://dx.doi.org/10.1634/theoncologist.5-4-302

44. National Institute for Clinical Excellence. (2016). Motor neurone disease: Assessment and management. *London: NICE Guideline* [NG42] (updated 2019).

45. van Eenennaam, R. M., Kruithof, W. J., van Es, M. A., Kruitwagen-van Reenen, E. T., Westeneng, H. J., Visser-Meily, J. M. A., . . . Beelen, A. (2020). Discussing personalized prognosis in amyotrophic lateral sclerosis: development of a communication guide. *BMC Neurol,* 20(1), 446. http://dx.doi.org/10.1186/s12883-020-02004-8

46. Aoun, S. M., Breen, L. J., Howting, D., Edis, R., Oliver, D., Henderson, R., . . . Birks, C. (2016). Receiving the news of a diagnosis of motor neuron disease: What does it take to make it better? *Amyotroph Lateral Scler Frontotemporal Degener,* 17(3-4), 168-178. http://dx.doi.org/10.3109/21678421.2015.1111907

47. Coulourides Kogan, A., Penido, M., & Enguidanos, S. (2015). Does Disclosure of Terminal Prognosis Mean Losing Hope? Insights from Exploring Patient Perspectives on Their Experience of Palliative Care Consultations. *J Palliat Med, 18(12),* 1019-1025. http://dx.doi.org/10.1089/jpm.2015.0038

48. van Eenennaam, R. M., Koppenol, L. S., Kruithof, W. J., Kruitwagen-van Reenen, E. T., Pieters, S., van Es, M. A., . . . Beelen, A. (2021). Discussing Personalized Prognosis Empowers Patients with Amyotrophic Lateral Sclerosis to Regain Control over Their Future: A Qualitative Study. *Brain Sci,* 11(12). http://dx.doi.org/10.3390/brainsci11121597

49. Genuis, S. K., Luth, W., Bubela, T., & Johnston, W. S. (2023). What do people affected by amyotrophic lateral sclerosis want from health communications? Evidence from the ALS Talk Project. *Muscle Nerve,* 68(3), 286-295. http://dx.doi.org/10.1002/mus.27935

50. Mitsumoto, H. (2018). What if you knew the prognosis of your patients with ALS? *Lancet Neurol,* 17(5), 386-388. http://dx.doi.org/10.1016/S1474-4422(18)30111-X

51. Curtis, J. R., Engelberg, R., Young, J. P., Vig, L. K., Reinke, L. F., Wenrich, M. D., . . . Back, A. L. (2008). An approach to understanding the interaction of hope and desire for explicit prognostic information among individuals with severe chronic obstructive pulmonary disease or advanced cancer. *J Palliat Med*, 11(4), 610-620. http://dx.doi.org/10.1089/jpm.2007.0209

52. Sakellariou, D., Boniface, G., & Brown, P. (2013). Experiences of living with motor neurone disease: a review of qualitative research. *Disabil Rehabil,* 35(21), 1765-1773. http://dx.doi.org/10.3109/09638288.2012.753118

53. Lobban, T. C., & Camm, A. J. (2011). Patient associations as stakeholders: a valuable partner for facilitating access to therapy. *Europace, 13 Suppl 2,* ii21-24. http://dx.doi.org/10.1093/europace/eur086

54. MND Connect. (2024), from https://www.mndaustralia.org.au/mnd-connect

55. Walczak, A., Butow, P. N., Davidson, P. M., Bellemore, F. A., Tattersall, M. H., Clayton, J. M., . . . Epstein, R. M. (2013). Patient perspectives regarding communication about prognosis and end-of-life issues: how can it be optimised? *Patient Educ Couns,* 90(3), 307-314. http://dx.doi.org/10.1016/j.pec.2011.08.009

56. Australian Indigenous HealthInfoNet. (2018). Australian Indigenous HealthInfoNet, from https://healthinfonet.ecu.edu.au/

57. Australian Government Department of Health and Aged Care. (2023). Gwandalan National Palliative Care Project.

58. Caresearch. (2021). Diverse populations, from https://www.caresearch.com.au/Community/Diversity/Diverse-Populations

59. Brownlee, A., & Bruening, L. M. (2012). Methods of Communication at End of Life for the Person With Amyotrophic Lateral Sclerosis. *Topics in Language Disorders,* 32, 168–185.

60. Gould, R. L., Rawlinson, C., Thompson, B., Weeks, K., Gossage-Worrall, R., Cantrill, H., . . . and the, C. C. G. (2023). Acceptance and Commitment Therapy for people living with motor neuron disease: an uncontrolled feasibility study. *Pilot Feasibility Stud*, 9(1), 116. http://dx.doi.org/10.1186/s40814-023-01354-7

61. Oberstadt, M. C. F., Esser, P., Classen, J., & Mehnert, A. (2018). Alleviation of Psychological Distress and the Improvement of Quality of Life in Patients With Amyotrophic Lateral Sclerosis: Adaptation of a Short-Term Psychotherapeutic Intervention. *Front Neurol*, 9, 231. http://dx.doi.org/10.3389/fneur.2018.00231

62. Plahuta, J. M., McCulloch, B. J., Kasarskis, E. J., Ross, M. A., Walter, R. A., & McDonald, E. R. (2002). Amyotrophic lateral sclerosis and hopelessness: psychosocial factors. *Soc Sci Med*, 55(12), 2131-2140. http://dx.doi.org/10.1016/s0277-9536(01)00356-2

63. Hardiman, O., Al-Chalabi, A., Chio, A., Corr, E. M., Logroscino, G., Robberecht, W., . . . van den Berg, L. H. (2017). Amyotrophic lateral sclerosis. *Nat Rev Dis Primers*, 3, 17085. http://dx.doi.org/10.1038/nrdp.2017.85

64. Pizzimenti, A., Aragona, M., Onesti, E., & Inghilleri, M. (2013). Depression, pain and quality of life in patients with amyotrophic lateral sclerosis: a cross-sectional study. *Funct Neurol*, 28(2), 115-119. http://dx.doi.org/10.11138/FNeur/2013.28.2.115

65. Oh, J., & Kim, J. A. (2017). Supportive care needs of patients with amyotrophic lateral sclerosis/motor neuron disease and their caregivers: A scoping review. *J Clin Nurs*, 26(23-24), 4129-4152. http://dx.doi.org/10.1111/jocn.13945

66. Hughes, R. A., Sinha, A., Higginson, I., Down, K., & Leigh, P. N. (2005). Living with motor neurone disease: lives, experiences of services and suggestions for change. *Health Soc Care Community*, 13(1), 64-74. http://dx.doi.org/10.1111/j.1365-2524.2005.00530.x

67. Hogden, A., Greenfield, D., Caga, J., & Cai, X. (2016). Development of patient decision support tools for motor neuron disease using stakeholder consultation: a study protocol. *BMJ Open*, 6(4), e010532. http://dx.doi.org/10.1136/bmjopen-2015-010532

68. Zarotti, N., Coates, E., McGeachan, A., Williams, I., Beever, D., Hackney, G., . . . High, C. S. G. (2019). Health care professionals' views on psychological factors affecting nutritional behaviour in people with motor neuron disease: A thematic analysis. *Br J Health Psychol,* 24(4), 953-969. http://dx.doi.org/10.1111/bjhp.12388

69. Ganzini, L., Johnston, W. S., & Hoffman, W. F. (1999). Correlates of suffering in amyotrophic lateral sclerosis. *Neurology*, 52(7), 1434-1440. http://dx.doi.org/10.1212/wnl.52.7.1434

70. Ganzini, L., Johnston, W. S., McFarland, B. H., Tolle, S. W., & Lee, M. A. (1998). Attitudes of patients with amyotrophic lateral sclerosis and their care givers toward assisted suicide. *N Engl J Med*, 339(14), 967-973. http://dx.doi.org/10.1056/NEJM199810013391406

71. Berlowitz, D. J., Howard, M. E., Fiore, J. F., Jr., Vander Hoorn, S., O'Donoghue, F. J., Westlake, J., . . . Talman, P. (2016). Identifying who will benefit from non-invasive ventilation in amyotrophic lateral sclerosis/motor neurone disease in a clinical cohort. *J Neurol Neurosurg Psychiatry*, 87(3), 280-286. http://dx.doi.org/10.1136/jnnp-2014-310055

72. Berlowitz, D. J., Mathers, S., Hutchinson, K., Hogden, A., Carey, K. A., Graco, M., . . . Sheers, N. L. (2023). The complexity of multidisciplinary respiratory care in amyotrophic lateral sclerosis. *Breathe* (Sheff), 19(3), 220269. http://dx.doi.org/10.1183/20734735.0269-2022

73. Abdulla, S., Vielhaber, S., Kollewe, K., Machts, J., Heinze, H. J., Dengler, R., & Petri, S. (2014). The impact of physical impairment on emotional well-being in ALS. *Amyotroph Lateral Scler Frontotemporal Degener*, 15(5-6), 392-397. http://dx.doi.org/10.3109/21678421.2014.932380

74. Gysels, M. H., & Higginson, I. J. (2011). The lived experience of breathlessness and its implications for care: a qualitative comparison in cancer, COPD, heart failure and MND. *BMC Palliat Care,* 10, 15. http://dx.doi.org/10.1186/1472-684X-10-15

75. Morelot-Panzini, C., Perez, T., Sedkaoui, K., de Bock, E., Aguilaniu, B., Devillier, P., . . . Similowski, T. (2018). The multidimensional nature of dyspnoea in amyotrophic lateral sclerosis patients with chronic respiratory failure: Air hunger, anxiety and fear. *Respir Med*, 145, 1-7. http://dx.doi.org/10.1016/j.rmed.2018.10.010

76. Tripodoro, V. A., & De Vito, E. L. (2008). Management of dyspnea in advanced motor neuron diseases. *Curr Opin Support Palliat Care,* 2(3), 173-179. http://dx.doi.org/10.1097/SPC.0b013e32830c9049

77. Edge, R., Mills, R., Tennant, A., Diggle, P. J., Young, C. A., & group, T. O. s. (2020). Do pain, anxiety and depression influence quality of life for people with amyotrophic lateral sclerosis/motor neuron disease? A national study reconciling previous conflicting literature. *J Neurol*, 267(3), 607-615. http://dx.doi.org/10.1007/s00415-019-09615-3

78. Young, C., Ealing, J., McDermott, C., Williams, T., Al-Chalabi, A., Majeed, T., . . . Tonic Study, G. (2022). Fatigue and anxiety mediate the effect of dyspnea on quality of life in amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Frontotemporal Degener,* 23(5-6), 390-398. http://dx.doi.org/10.1080/21678421.2021.1990343

79. Diver, E. M., & Regan, J. (2022). Use of Pharyngeal High-Resolution Manometry to Evaluate Dysphagia in Adults with Motor Neurone Disease: A Scoping Review. *Dysphagia*, 37(6), 1697-1714. http://dx.doi.org/10.1007/s00455-022-10418-4

80. Lisiecka, D., Kelly, H., & Jackson, J. (2021). How do people with Motor Neurone Disease experience dysphagia? A qualitative investigation of personal experiences. *Disabil Rehabil,* 43(4), 479-488. http://dx.doi.org/10.1080/09638288.2019.1630487

81. Foley, G., Timonen, V., & Hardiman, O. (2014). Understanding psycho-social processes underpinning engagement with services in motor neurone disease: a qualitative study. *Palliat Med*, 28(4), 318-325. http://dx.doi.org/10.1177/0269216313512013

82. Johnson, J., Leigh, P. N., Shaw, C. E., Ellis, C., Burman, R., & Al-Chalabi, A. (2012). Eating-derived pleasure in amyotrophic lateral sclerosis as a predictor of non-oral feeding. *Amyotroph Lateral Scler,* 13(6), 555-559. http://dx.doi.org/10.3109/17482968.2012.704925

83. Stavroulakis, T., Baird, W. O., Baxter, S. K., Walsh, T., Shaw, P. J., & McDermott, C. J. (2014). Factors influencing decision-making in relation to timing of gastrostomy insertion in patients with motor neurone disease. *BMJ Support Palliat Care,* 4(1), 57-63. http://dx.doi.org/10.1136/bmjspcare-2013-000497

84. EFNS Task Force on Diagnosis Management of Amyotrophic Lateral Sclerosis, Andersen, P. M., Abrahams, S., Borasio, G. D., de Carvalho, M., Chio, A., . . . Weber, M. (2012). EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS)--revised report of an EFNS task force. *Eur J Neurol,* 19(3), 360-375. http://dx.doi.org/10.1111/j.1468-1331.2011.03501.x

85. Paynter, C., Mathers, S., Gregory, H., Vogel, A. P., & Cruice, M. (2022). The impact of communication on healthcare involvement for people living with motor neurone disease and their carers: A longitudinal qualitative study. *Int J Lang Commun Disord,* 57(6), 1318-1333. http://dx.doi.org/10.1111/1460-6984.12757

86. van Eenennaam, R. M., Rave, N., Kruithof, W. J., Kruitwagen-van Reenen, E. T., van den Berg, L. H., Visser-Meily, J. A., & Beelen, A. (2023). Control in the absence of choice: A qualitative study on decision-making about gastrostomy in people with amyotrophic lateral sclerosis, caregivers, and healthcare professionals. *PLoS One,* 18(9), e0290508. http://dx.doi.org/10.1371/journal.pone.0290508

87. Greenaway, L. P., Martin, N. H., Lawrence, V., Janssen, A., Al-Chalabi, A., Leigh, P. N., & Goldstein, L. H. (2015). Accepting or declining non-invasive ventilation or gastrostomy in amyotrophic lateral sclerosis: patients' perspectives. *J Neurol,* 262(4), 1002-1013. http://dx.doi.org/10.1007/s00415-015-7665-z

88. Labra, J., Hogden, A., Power, E., James, N., & Flood, V. M. (2020). Gastrostomy uptake in motor neurone disease: a mixed-methods study of patients' decision making. *BMJ Open,* 10(2), e034751. http://dx.doi.org/10.1136/bmjopen-2019-034751

89. Stavroulakis, T., Baird, W. O., Baxter, S. K., Walsh, T., Shaw, P. J., & McDermott, C. J. (2016). The impact of gastrostomy in motor neurone disease: challenges and benefits from a patient and carer perspective. *BMJ Support Palliat Care,* 6(1), 52-59. http://dx.doi.org/10.1136/bmjspcare-2013-000609

90. Charania, S. (2022). *A year in the life: The impact of dysarthria for people with MND.* PhD PhD Thesis, School of Health and Rehabilitation Sciences, The University of Queensland.

91. Leite Neto, L., & Constantini, A. (2017). Dysarthria and quality of life in patients with amyotrophic lateral sclerosis. *Revista CEFAC,* 19(5), 664-673. http://dx.doi.org/10.1590/1982-021620171954017

92. Tomik, B., & Guiloff, R. J. (2010). Dysarthria in amyotrophic lateral sclerosis: A review. *Amyotroph Lateral Scler*, 11(1-2), 4-15. http://dx.doi.org/10.3109/17482960802379004

93. Tomik, J., Tomik, B., Wiatr, M., Skladzien, J., Strek, P., & Szczudlik, A. (2015). The Evaluation of Abnormal Voice Qualities in Patients with Amyotrophic Lateral Sclerosis. *Neurodegener Dis*, 15(4), 225-232. http://dx.doi.org/10.1159/000381956

94. Hecht, M., Hillemacher, T., Grasel, E., Tigges, S., Winterholler, M., Heuss, D., . . . Neundorfer, B. (2002). Subjective experience and coping in ALS. *Amyotroph Lateral Scler Other Motor Neuron Disord,* 3(4), 225-231. http://dx.doi.org/10.1080/146608202760839009

95. Fried-Oken, M., Mooney, A., & Peters, B. (2015). Supporting communication for patients with neurodegenerative disease. NeuroRehabilitation, 37(1), 69-87. http://dx.doi.org/10.3233/NRE-151241

96. Munan, M., Luth, W., Genuis, S. K., Johnston, W. S. W., & MacIntyre, E. (2021). Transitions in Amyotrophic Lateral Sclerosis: Patient and Caregiver Experiences. *Can J Neurol Sci,* 48(4), 496-503. http://dx.doi.org/10.1017/cjn.2020.240

97. Felgoise, S. H., Zaccheo, V., Duff, J., & Simmons, Z. (2016). Verbal communication impacts quality of life in patients with amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Frontotemporal Degener,* 17(3-4), 179-183. http://dx.doi.org/10.3109/21678421.2015.1125499

98. Londral, A. (2022). Assistive Technologies for Communication Empower Patients With ALS to Generate and Self-Report Health Data. *Front Neurol,* 13, 867567. http://dx.doi.org/10.3389/fneur.2022.867567

99. Linse, K., Aust, E., Joos, M., & Hermann, A. (2018). Communication Matters-Pitfalls and Promise of Hightech Communication Devices in Palliative Care of Severely Physically Disabled Patients With Amyotrophic Lateral Sclerosis. *Front Neurol,* 9, 603. http://dx.doi.org/10.3389/fneur.2018.00603

100. McNaughton, D., Giambalvo, F., Kohler, K., Nazareth, G., Caron, J., & Fager, S. (2018). "Augmentative and Alternative Communication (AAC) Will Give You a Voice": Key Practices in AAC Assessment and Intervention as Described by Persons with Amyotrophic Lateral Sclerosis. *Semin Speech Lang,* 39(5), 399-415. http://dx.doi.org/10.1055/s-0038-1669992

101. Peters, B., Wiedrick, J., & Baylor, C. (2023). Effects of Aided Communication on Communicative Participation for People With Amyotrophic Lateral Sclerosis. *Am J Speech Lang Pathol,* 32(4), 1450-1465. http://dx.doi.org/10.1044/2023_AJSLP-22-00346

102. Beukelman, D., Fager, S., & Nordness, A. (2011). Communication Support for People with ALS. *Neurol Res Int, 2011,* 714693. http://dx.doi.org/10.1155/2011/714693

103. Page, A. D., & Yorkston, K. M. (2022). Communicative Participation in Dysarthria: Perspectives for Management. *Brain Sci,* 12(4). http://dx.doi.org/10.3390/brainsci12040420

104. McKelvey, M., Evans, D. L., Kawai, N., & Beukelman, D. (2012). Communication styles of persons with ALS as recounted by surviving partners. *Augment Altern Commun,* 28(4), 232-242. http://dx.doi.org/10.3109/07434618.2012.737023

105. Hwang, C. S., Weng, H. H., Wang, L. F., Tsai, C. H., & Chang, H. T. (2014). An eye-tracking assistive device improves the quality of life for ALS patients and reduces the caregivers' burden. *J Mot Behav,* 46(4), 233-238. http://dx.doi.org/10.1080/00222895.2014.891970

106. Cave, R., & Bloch, S. (2021). Voice banking for people living with motor neurone disease: Views and expectations. *Int J Lang Commun Disord,* 56(1), 116-129. http://dx.doi.org/10.1111/1460-6984.12588

107. Helleman, J., Kruitwagen, E. T., van den Berg, L. H., Visser-Meily, J. M. A., & Beelen, A. (2020). The current use of telehealth in ALS care and the barriers to and facilitators of implementation: a systematic review. *Amyotroph Lateral Scler Frontotemporal Degener,* 21(3-4), 167-182. http://dx.doi.org/10.1080/21678421.2019.1706581

108. Helleman, J., Van Eenennaam, R., Kruitwagen, E. T., Kruithof, W. J., Slappendel, M. J., Van Den Berg, L. H., . . . Beelen, A. (2020). Telehealth as part of specialized ALS care: feasibility and user experiences with "ALS home-monitoring and coaching". *Amyotroph Lateral Scler Frontotemporal Degener,* 21(3-4), 183-192. http://dx.doi.org/10.1080/21678421.2020.1718712

109. Garcia Perez, A. I., & Dapueto, J. J. (2014). Case report of a computer-assisted psychotherapy of a patient with ALS. *Int J Psychiatry Med,* 48(3), 229-233. http://dx.doi.org/10.2190/PM.48.3.g

110. Atassi, N., Yerramilli-Rao, P., Szymonifka, J., Yu, H., Kearney, M., Grasso, D., . . . Cudkowicz, M. E. (2013). Analysis of start-up, retention, and adherence in ALS clinical trials. *Neurology,* 81(15), 1350-1355. http://dx.doi.org/10.1212/WNL.0b013e3182a823e0

111. Finegan, E., Chipika, R. H., Li Hi Shing, S., Hardiman, O., & Bede, P. (2019). Pathological Crying and Laughing in Motor Neuron Disease: Pathobiology, Screening, Intervention. *Front Neurol,* 10, 260. http://dx.doi.org/10.3389/fneur.2019.00260

112. Gondim, F. A. A., Pinto, W., Chieia, M. A. T., Correia, C. D. C., Cunha, F. M. B., Dourado, M. E. T., Jr., . . . Dias-Tosta, E. (2023). Definitions, phenomenology, diagnosis, and management of the disorders of laughter and crying in amyotrophic lateral sclerosis (ALS): Consensus from ALS and Motor Neuron Disease Scientific Department of the Brazilian Academy of Neurology. *Arq Neuropsiquiatr,* 81(8), 764-775. http://dx.doi.org/10.1055/s-0043-1771176

113. Goldin, D. S. (2020). Pseudobulbar Affect: An Overview. *J Psychosoc Nurs Ment Health Serv*, 58(9), 19-24. http://dx.doi.org/10.3928/02793695-20200624-08

114. Strong, M. J., Abrahams, S., Goldstein, L. H., Woolley, S., McLaughlin, P., Snowden, J., . . . Turner, M. R. (2017). Amyotrophic lateral sclerosis - frontotemporal spectrum disorder (ALS-FTSD): Revised diagnostic criteria. *Amyotroph Lateral Scler Frontotemporal Degener,* 18(3-4), 153-174. http://dx.doi.org/10.1080/21678421.2016.1267768

115. Abrahams, S. (2023). Neuropsychological impairment in amyotrophic lateral sclerosis-frontotemporal spectrum disorder. *Nat Rev Neurol,* 19(11), 655-667. http://dx.doi.org/10.1038/s41582-023-00878-z

116. Abrahams, S., Newton, J., Niven, E., Foley, J., & Bak, T. H. (2014). Screening for cognition and behaviour changes in ALS. *Amyotroph Lateral Scler Frontotemporal Degener,* 15(1-2), 9-14. http://dx.doi.org/10.3109/21678421.2013.805784

117. Woolley, S. C., York, M. K., Moore, D. H., Strutt, A. M., Murphy, J., Schulz, P. E., & Katz, J. S. (2010). Detecting frontotemporal dysfunction in ALS: utility of the ALS Cognitive Behavioral Screen (ALS-CBS). *Amyotroph Lateral Scler,* 11(3), 303-311. http://dx.doi.org/10.3109/17482961003727954

118. Gray, D., & Abrahams, S. (2022). International evaluation of current practices in cognitive assessment for motor neurone disease. *British Journal of Neuroscience Nursing,* 18(1), 38-44. http://dx.doi.org/10.12968/bjnn.2022.18.1.38

119. Niven, E., Newton, J., Foley, J., Colville, S., Swingler, R., Chandran, S., . . . Abrahams, S. (2015). Validation of the Edinburgh Cognitive and Behavioural Amyotrophic Lateral Sclerosis Screen (ECAS): A cognitive tool for motor disorders. *Amyotroph Lateral Scler Frontotemporal Degener,* 16(3-4), 172-179. http://dx.doi.org/10.3109/21678421.2015.1030430

120. Lillo, P., Mioshi, E., Zoing, M. C., Kiernan, M. C., & Hodges, J. R. (2011). How common are behavioural changes in amyotrophic lateral sclerosis? *Amyotroph Lateral Scler,* 12(1), 45-51. http://dx.doi.org/10.3109/17482968.2010.520718

121. Carelli, L., Solca, F., Faini, A., Madotto, F., Lafronza, A., Monti, A., . . . Poletti, B. (2018). The Complex Interplay Between Depression/Anxiety and Executive Functioning: Insights From the ECAS in a Large ALS Population. *Front Psychol,* 9, 450. http://dx.doi.org/10.3389/fpsyg.2018.00450

122. Poletti, B., Solca, F., Carelli, L., Madotto, F., Lafronza, A., Faini, A., . . . Silani, V. (2016). The validation of the Italian Edinburgh Cognitive and Behavioural ALS Screen (ECAS). *Amyotroph Lateral Scler Frontotemporal Degener*, 17(7-8), 489-498. http://dx.doi.org/10.1080/21678421.2016.1183679

123. Caga, J., Hsieh, S., Lillo, P., Dudley, K., & Mioshi, E. (2019). The Impact of Cognitive and Behavioral Symptoms on ALS Patients and Their Caregivers. *Front Neurol,* 10, 192. http://dx.doi.org/10.3389/fneur.2019.00192

124. Rabkin, J., Goetz, R., Murphy, J. M., Factor-Litvak, P., Mitsumoto, H., & Group, A. C. S. (2016). Cognitive impairment, behavioral impairment, depression, and wish to die in an ALS cohort. *Neurology,* 87(13), 1320-1328. http://dx.doi.org/10.1212/WNL.0000000000003035

125. Santangelo, G., Siciliano, M., Trojano, L., Femiano, C., Monsurrò, M. R., Tedeschi, G., & Trojsi, F. (2017). Apathy in amyotrophic lateral sclerosis: insights from Dimensional Apathy Scale. *Amyotroph Lateral Scler Frontotemporal Degener*, 18(5-6), 434-442. http://dx.doi.org/10.1080/21678421.2017.1313865

126. Olney, R. K., Murphy, J., Forshew, D., Garwood, E., Miller, B. L., Langmore, S., . . . Lomen-Hoerth, C. (2005). The effects of executive and behavioral dysfunction on the course of ALS. *Neurology,* 65(11), 1774-1777. http://dx.doi.org/10.1212/01.wnl.0000188759.87240.8b

127. Bock, M., Duong, Y. N., Kim, A., Allen, I., Murphy, J., & Lomen-Hoerth, C. (2016). Cognitive-behavioral changes in amyotrophic lateral sclerosis: Screening prevalence and impact on patients and caregivers. *Amyotroph Lateral Scler Frontotemporal Degener,* 17(5-6), 366-373. http://dx.doi.org/10.3109/21678421.2016.1165257

128. Burke, T., Elamin, M., Galvin, M., Hardiman, O., & Pender, N. (2015). Caregiver burden in amyotrophic lateral sclerosis: a cross-sectional investigation of predictors. *J Neurol,* 262(6), 1526-1532. http://dx.doi.org/10.1007/s00415-015-7746-z

129. Cui, B., Cui, L., Gao, J., Liu, M., Li, X., Liu, C., . . . Fang, J. (2015). Cognitive Impairment in Chinese Patients with Sporadic Amyotrophic Lateral Sclerosis. *PLoS One,* 10(9), e0137921. http://dx.doi.org/10.1371/journal.pone.0137921

130. Huynh, W., Ahmed, R., Mahoney, C. J., Nguyen, C., Tu, S., Caga, J., . . . Kiernan, M. C. (2020). The impact of cognitive and behavioral impairment in amyotrophic lateral sclerosis. *Expert Rev Neurother,* 20(3), 281-293. http://dx.doi.org/10.1080/14737175.2020.1727740

131. Fullam, T., Stephens, H. E., Felgoise, S. H., Blessinger, J. K., Walsh, S., & Simmons, Z. (2015). Compliance with recommendations made in a multidisciplinary ALS clinic. *Amyotroph Lateral Scler Frontotemporal Degener,* 17(1-2), 30-37. http://dx.doi.org/10.3109/21678421.2015.1074703

132. McHutchison, C. A., Leighton, D. J., McIntosh, A., Cleary, E., Warner, J., Porteous, M., . . . Abrahams, S. (2020). Relationship between neuropsychiatric disorders and cognitive and behavioural change in MND. *J Neurol Neurosurg Psychiatry,* 91(3), 245-253. http://dx.doi.org/10.1136/jnnp-2019-321737

133. Heidari, M. E., Nadali, J., Parouhan, A., Azarafraz, M., Tabatabai, S. M., Irvani, S. S. N., . . . Gharebaghi, A. (2021). Prevalence of depression among amyotrophic lateral sclerosis (ALS) patients: A systematic review and meta-analysis. *J Affect Disord,* 287, 182-190. http://dx.doi.org/10.1016/j.jad.2021.03.015

134. Bond, L., Bowen, G., Mertens, B., Denson, K., Jordan, K., Vidakovic, B., & Mitchell, C. S. (2020). Associations of Patient Mood, Modulators of Quality of Life, and Pharmaceuticals with Amyotrophic Lateral Sclerosis Survival Duration. *Behav Sci* (Basel), 10(1). http://dx.doi.org/10.3390/bs10010033

135. Fang, F., Valdimarsdottir, U., Furst, C. J., Hultman, C., Fall, K., Sparen, P., & Ye, W. (2008). Suicide among patients with amyotrophic lateral sclerosis. *Brain,* 131(Pt 10), 2729-2733. http://dx.doi.org/10.1093/brain/awn161

136. van Groenestijn, A. C., Kruitwagen-van Reenen, E. T., Visser-Meily, J. M., van den Berg, L. H., & Schroder, C. D. (2016). Associations between psychological factors and health-related quality of life and global quality of life in patients with ALS: a systematic review. *Health Qual Life Outcomes,* 14(1), 107. http://dx.doi.org/10.1186/s12955-016-0507-6

137. Carvalho, T. L., de Almeida, L. M., Lorega, C. M., Barata, M. F., Ferreira, M. L., de Brito-Marques, P. R., & Correia Cda, C. (2016). Depression and anxiety in individuals with amyotrophic lateral sclerosis: a systematic review. *Trends Psychiatry Psychother,* 38(1), 1-5. http://dx.doi.org/10.1590/2237-6089-2015-0030

138. Young, C. A., Ealing, J., McDermott, C. J., Williams, T. L., Al-Chalabi, A., Majeed, T., . . . Tonic Study, G. (2023). Prevalence of depression in amyotrophic lateral sclerosis/motor neuron disease: multi-attribute ascertainment and trajectories over 30 months. *Amyotroph Lateral Scler Frontotemporal Degener,* 24(1-2), 82-90. http://dx.doi.org/10.1080/21678421.2022.2096410

139. Balendra, R., Jones, A., Jivraj, N., Knights, C., Ellis, C. M., Burman, R., . . . Al-Chalabi, A. (2014). Estimating clinical stage of amyotrophic lateral sclerosis from the ALS Functional Rating Scale. *Amyotroph Lateral Scler Frontotemporal Degener,* 15(3-4), 279-284. http://dx.doi.org/10.3109/21678421.2014.897357

140. Cui, F., Zhu, W., Zhou, Z., Ren, Y., Li, Y., Li, M., . . . Huang, X. (2015). Frequency and risk factor analysis of cognitive and anxiety-depressive disorders in patients with amyotrophic lateral sclerosis/motor neuron disease. *Neuropsychiatr Dis Treat,* 11, 2847-2854. http://dx.doi.org/10.2147/NDT.S90520

141. Prado, L. G. R., Bicalho, I. C. S., Vidigal-Lopes, M., Prado, V. G. R., Gomez, R. S., de Souza, L. C., & Teixeira, A. L. (2017). Depression and anxiety in a case series of amyotrophic lateral sclerosis: frequency and association with clinical features. *Einstein (Sao Paulo),* 15(1), 58-60. http://dx.doi.org/10.1590/S1679-45082017AO3870

142. Oh, J., An, J., & Park, K. (2021). Coping in people with amyotrophic lateral sclerosis and motor neuron disease: Systematic review. *J Clin Nurs,* 30(13-14), 1838-1853. http://dx.doi.org/10.1111/jocn.15692

143. Longinetti, E., Mariosa, D., Larsson, H., Ye, W., Ingre, C., Almqvist, C., . . . Fang, F. (2017). Neurodegenerative and psychiatric diseases among families with amyotrophic lateral sclerosis. *Neurology,* 89(6), 578-585. http://dx.doi.org/10.1212/WNL.0000000000004179

144. Castanheira, A., Swash, M., & De Carvalho, M. (2022). Percutaneous gastrostomy in amyotrophic lateral sclerosis: a review. *Amyotroph Lateral Scler Frontotemporal Degener*, 23(3-4), 176-189. http://dx.doi.org/10.1080/21678421.2021.1946089

145. Paynter, C., Cruice, M., Mathers, S., Gregory, H., & Vogel, A. P. (2019). Communication and cognitive impairments and health care decision making in MND: A narrative review. *Journal of Evaluation in Clinical Practice,* 25(6), 1182-1192. http://dx.doi.org/https://doi.org/10.1111/jep.13219

146. Khin Khin, E., Minor, D., Holloway, A., & Pelleg, A. (2015). Decisional Capacity in Amyotrophic Lateral Sclerosis. *J Am Acad Psychiatry Law,* 43(2), 210-217.

147. White, S., O'Cathain, A., Halliday, V., Croot, L., & McDermott, C. J. (2023). Factors influencing decisions people with motor neuron disease make about gastrostomy placement and ventilation: A qualitative evidence synthesis. *Health Expect,* 26(4), 1418-1435. http://dx.doi.org/10.1111/hex.13786

148. Erdmann, A., Spoden, C., Hirschberg, I., & Neitzke, G. (2022). Talking about the end of life: communication patterns in amyotrophic lateral sclerosis - a scoping review. *Palliat Care Soc Pract,* 16, 26323524221083676. http://dx.doi.org/10.1177/26323524221083676

149. Flemming, K., Turner, V., Bolsher, S., Hulme, B., McHugh, E., & Watt, I. (2020). The experiences of, and need for, palliative care for people with motor neurone disease and their informal caregivers: A qualitative systematic review. *Palliat Med*, 34(6), 708-730. http://dx.doi.org/10.1177/0269216320908775

150. Levi, B. H., Simmons, Z., Hanna, C., Brothers, A., Lehman, E., Farace, E., . . . Green, M. J. (2017). Advance care planning for patients with amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Frontotemporal Degener,* 18(5-6), 388-396. http://dx.doi.org/10.1080/21678421.2017.1285317

151. Murray, L., Butow, P. N., White, K., Kiernan, M. C., D'Abrew, N., & Herz, H. (2016). Advance care planning in motor neuron disease: A qualitative study of caregiver perspectives. *Palliat Med*, 30(5), 471-478. http://dx.doi.org/10.1177/0269216315613902

152. Hunter New England Local Health District. (2018). Supportive care into the Dreaming: A palliative care guide for Aboriginal and Torres Strait Islander people through Sorry Business, from https://healthinfonet.ecu.edu.au/key-resources/

153. Aboriginal Health and Medical Research Council of New South Wales. (2021). Journey to dreaming toolkit. Sydney.

154. Palliative Care Australia. (2021). Aboriginal and Torres Strait Islander Peoples Palliative Care Resources, from https://palliativecare.org.au/resource/aboriginal-and-torres-strait-islander-peoples-palliative-care-resources/

155. Gale, C. (2015). Assisting patients with motor neurone disease to make decisions about their care. *Int J Palliat Nurs*, 21(5), 251-255. http://dx.doi.org/10.12968/ijpn.2015.21.5.251

156. Eliasen, A., Dalhoff, K. P., & Horwitz, H. (2018). Neurological diseases and risk of suicide attempt: a case-control study. *J Neurol,* 265(6), 1303-1309. http://dx.doi.org/10.1007/s00415-018-8837-4

157. Erlangsen, A., Stenager, E., Conwell, Y., Andersen, P. K., Hawton, K., Benros, M. E., . . . Stenager, E. (2020). Association Between Neurological Disorders and Death by Suicide in Denmark. *JAMA,* 323(5), 444-454. http://dx.doi.org/10.1001/jama.2019.21834

158. Tsai, C. P., Chang, B. H., & Lee, C. T. (2013). Underlying cause and place of death among patients with amyotrophic lateral sclerosis in Taiwan: a population-based study, 2003-2008. *J Epidemiol,* 23(6), 424-428. http://dx.doi.org/10.2188/jea.je20130045

159. Pavey, A., Allen-Collinson, J., & Pavey, T. (2013). The Lived Experience of Diagnosis Delivery in Motor Neurone Disease: A Sociological-Phenomenological Study. *Sociological Research Online,* 18(2), 36-47. http://dx.doi.org/10.5153/sro.2927

160. Baumrucker, S. J., Stolick, M., Carter, G. T., Mingle, P., Oertli, K. A., & Morris, G. M. (2011). Legal but not always allowed: "physician aid in dying". *Am J Hosp Palliat Care,* 28(6), 449-454. http://dx.doi.org/10.1177/1049909111412540

161. Chamberlain, P. I. (2014). An assisted dying law might save me from a lingering and unpleasant death. *BMJ,* 349, g4784. http://dx.doi.org/10.1136/bmj.g4784

162. Kuhnlein, P., Kubler, A., Raubold, S., Worrell, M., Kurt, A., Gdynia, H. J., . . . Ludolph, A. C. (2008). Palliative care and circumstances of dying in German ALS patients using non-invasive ventilation. *Amyotroph Lateral Scler,* 9(2), 91-98. http://dx.doi.org/10.1080/17482960701830495

163. Voluntary Assisted Dying Board Western Australia. (2023). Voluntary Assisted Dying Board Western Australia Annual Report 2022-23, from https://www.health.wa.gov.au/~/media/Corp/Documents/Health-for/Voluntary-assisted-dying/VAD-Board-Annual-Report-2022-23.pdf

164. Sealey, M., & Aoun, S. M. (2023). Family caregivers bereaved by voluntary-assisted dying with a focus on motor neurone disease: the hidden patients. *Palliat Care Soc Pract, 17,* 26323524231212512. http://dx.doi.org/10.1177/26323524231212512

165. Gamondi, C., Fusi-Schmidhauser, T., Oriani, A., Payne, S., & Preston, N. (2019). Family members' experiences of assisted dying: A systematic literature review with thematic synthesis. *Palliat Med,* 33(8), 1091-1105. http://dx.doi.org/10.1177/0269216319857630

166. Lowers, J., Scardaville, M., Hughes, S., & Preston, N. J. (2020). Comparison of the experience of caregiving at end of life or in hastened death: a narrative synthesis review. *BMC Palliat Care,* 19(1), 154. http://dx.doi.org/10.1186/s12904-020-00660-8

167. Queensland Aboriginal & Torres Strait Islander Health Branch. (2015). Sad news, sorry business: Guidelines for caring for Aboriginal and Torres Strait Islander people through death and dying, from https://healthinfonet.ecu.edu.au/key-resources/resources/21898/?title=Sad+news++sorry+business++guidelines+for+caring+for+Aboriginal+and+Torres+Strait+Islander+people+through+death+and+dying&contentid=21898_1

168. Program of Experience in the Palliative Approach. (2020). Cultural considerations: Providing end of life care for Aboriginal peoples and Torres Strait Islander peoples, from https://healthinfonet.ecu.edu.au/key-resources/resources/29818/?title=Cultural+considerations++providing+end+of+life+care+for+Aboriginal+peoples+and+Torres+Strait+Islander+peoples&contentid=29818_1

169. Gilimbaa. (2020). During sad news and sorry business: Information for family, from https://healthinfonet.ecu.edu.au/key-resources/resources/40671/?title=During+sad+news+and+sorry+business++information+for+family&contentid=40671_1

170. Lewis, S., Willmott, L., White, B. P., La Brooy, C., & Komesaroff, P. (2022). First Nations Perspectives in Law-Making About Voluntary Assisted Dying. *J Law Med,* 29(4), 1168-1181.

171. Caresearch. (2021). Aboriginal and Torres Strait Islander Care - Health Professionals, from https://www.caresearch.com.au/Health-Professionals/Aboriginal-and-Torres-Strait-Islander-Care/Culturally-Safe-and-Responsive-Care

172. Indigenous Program of Experience in the Palliative Approach (IPEPA). (2014). Cultural Considerations: Providing end of life care for Aboriginal peoples and Torres Strait Islander peoples, from https://pepaeducation.com/support-and-education/cultural-considerations-providing-end-of-life-care-for-aboriginal-peoples-and-torres-strait-islander-peoples/

173. Singer, J., Daum, C., Evans, A., Schneider, S., Vugrin, M., & Loggers, E. (2023). An examination and proposed theoretical model of risk and protective factors for bereavement outcomes for family members of individuals who engaged in medical aid in dying: A systematic review. *Palliat Med,* 37(7), 947-958. http://dx.doi.org/10.1177/02692163231172242

174. Andriessen, K., Krysinska, K., Castelli Dransart, D. A., Dargis, L., & Mishara, B. L. (2020). Grief After Euthanasia and Physician-Assisted Suicide. *Crisis,* 41(4), 255-272. http://dx.doi.org/10.1027/0227-5910/a000630

175. Australian Government Department of Health and Aged Care. (2020). Exploratory Analysis of Barriers to Palliative Care – Literature Review.

176. End of Life Directions for Aged Care (ELDAC). (2024). Diverse Population Groups, from https://www.eldac.com.au/Resources/Diverse-Population-Groups

177. Diversicare. (2015). Little book of cultural tips, from https://www.diversicare.com.au/wp-content/uploads/2015/09/Little_Book_of_Cultural_Tips_final_proof_7_May_2015-FINAL-4-27pm.pdf

178. Gould, R. L., Coulson, M. C., Brown, R. G., Goldstein, L. H., Al-Chalabi, A., & Howard, R. J. (2015). Psychotherapy and pharmacotherapy interventions to reduce distress or improve well-being in people with amyotrophic lateral sclerosis: A systematic review. Amyotroph Lateral Scler Frontotemporal Degener, 16(5-6), 293-302. http://dx.doi.org/10.3109/21678421.2015.1062515

179. Zarotti, N., Mayberry, E., Ovaska-Stafford, N., Eccles, F., & Simpson, J. (2021). Psychological interventions for people with motor neuron disease: a scoping review. *Amyotroph Lateral Scler Frontotemporal Degener,* 22(1-2), 1-11. http://dx.doi.org/10.1080/21678421.2020.1788094

180. Marconi, A., Gragnano, G., Lunetta, C., Gatto, R., Fabiani, V., Tagliaferri, A., . . . Pagnini, F. (2016). The experience of meditation for people with amyotrophic lateral sclerosis and their caregivers - a qualitative analysis. *Psychol Health Med,* 21(6), 762-768. http://dx.doi.org/10.1080/13548506.2015.1115110

181. Pagnini, F., Marconi, A., Tagliaferri, A., Manzoni, G. M., Gatto, R., Fabiani, V., . . . Lunetta, C. (2017). Meditation training for people with amyotrophic lateral sclerosis: a randomized clinical trial. *Eur J Neurol,* 24(4), 578-586. http://dx.doi.org/10.1111/ene.13246

182. van Groenestijn, A. C., Schroder, C. D., Visser-Meily, J. M., Reenen, E. T., Veldink, J. H., & van den Berg, L. H. (2015). Cognitive behavioural therapy and quality of life in psychologically distressed patients with amyotrophic lateral sclerosis and their caregivers: Results of a prematurely stopped randomized controlled trial. *Amyotroph Lateral Scler Frontotemporal Degener,* 16(5-6), 309-315. http://dx.doi.org/10.3109/21678421.2015.1038276

183. Diaz, J. L., Sancho, J., Barreto, P., Banuls, P., Renovell, M., & Servera, E. (2016). Effect of a short-term psychological intervention on the anxiety and depression of amyotrophic lateral sclerosis patients. *J Health Psychol,* 21(7), 1426-1435. http://dx.doi.org/10.1177/1359105314554819

184. Palmieri, A., Kleinbub, J. R., Calvo, V., Soraru, G., Grasso, I., Messina, I., & Sambin, M. (2012). Efficacy of hypnosis-based treatment in amyotrophic lateral sclerosis: a pilot study. *Front Psychol,* 3, 465. http://dx.doi.org/10.3389/fpsyg.2012.00465

185. Kleinbub, J. R., Palmieri, A., Broggio, A., Pagnini, F., Benelli, E., Sambin, M., & Soraru, G. (2015). Hypnosis-based psychodynamic treatment in ALS: a longitudinal study on patients and their caregivers. *Front Psychol,* 6, 822. http://dx.doi.org/10.3389/fpsyg.2015.00822

186. Aoun, S. M., Chochinov, H. M., & Kristjanson, L. J. (2015). Dignity therapy for people with motor neuron disease and their family caregivers: a feasibility study. *J Palliat Med,* 18(1), 31-37. http://dx.doi.org/10.1089/jpm.2014.0213

187. Bentley, B., O'Connor, M., Kane, R., & Breen, L. J. (2014). Feasibility, acceptability, and potential effectiveness of dignity therapy for people with motor neurone disease. *PLoS One,* 9(5), e96888. http://dx.doi.org/10.1371/journal.pone.0096888

188. Averill, A. J., Kasarskis, E. J., & Segerstrom, S. C. (2013). Expressive disclosure to improve well-being in patients with amyotrophic lateral sclerosis: a randomised, controlled trial. *Psychol Health,* 28(6), 701-713. http://dx.doi.org/10.1080/08870446.2012.754891

189. Pinto, C., Geraghty, A. W. A., Pagnini, F., Yardley, L., & Dennison, L. (2023). How do people with MND and caregivers experience a digital mental health intervention? A qualitative study. *Front Psychiatry,* 14, 1083196. http://dx.doi.org/10.3389/fpsyt.2023.1083196

190. Weeks, K. R., Gould, R. L., McDermott, C., Lynch, J., Goldstein, L. H., Graham, C. D., . . . Lawrence, V. (2019). Needs and preferences for psychological interventions of people with motor neuron disease. *Amyotroph Lateral Scler Frontotemporal Degener,* 20(7-8), 521-531. http://dx.doi.org/10.1080/21678421.2019.1621344

191. Hogden, A., Greenfield, D., Nugus, P., & Kiernan, M. C. (2013). What are the roles of carers in decision-making for amyotrophic lateral sclerosis multidisciplinary care? *Patient Prefer Adherence,* 7, 171-181. http://dx.doi.org/10.2147/PPA.S40783

192. Goutman, S. A., Nowacek, D. G., Burke, J. F., Kerber, K. A., Skolarus, L. E., & Callaghan, B. C. (2014). Minorities, men, and unmarried amyotrophic lateral sclerosis patients are more likely to die in an acute care facility. *Amyotroph Lateral Scler Frontotemporal Degener,* 15(5-6), 440-443. http://dx.doi.org/10.3109/21678421.2014.924143

193. Mitchell, L. A., Hirdes, J., Poss, J. W., Slegers-Boyd, C., Caldarelli, H., & Martin, L. (2015). Informal caregivers of clients with neurological conditions: profiles, patterns and risk factors for distress from a home care prevalence study. *BMC Health Serv Res,* 15, 350. http://dx.doi.org/10.1186/s12913-015-1010-1

194. Williams, M. T., Donnelly, J. P., Holmlund, T., & Battaglia, M. (2008). ALS: Family caregiver needs and quality of life. *Amyotroph Lateral Scler,* 9(5), 279-286. http://dx.doi.org/10.1080/17482960801934148

195. Boerner, K., & Mock, S. E. (2012). Impact of patient suffering on caregiver well-being: the case of amyotrophic lateral sclerosis patients and their caregivers. *Psychol Health Med,* 17(4), 457-466. http://dx.doi.org/10.1080/13548506.2011.613942

196. Chio, A., Gauthier, A., Calvo, A., Ghiglione, P., & Mutani, R. (2005). Caregiver burden and patients' perception of being a burden in ALS. *Neurology*, 64(10), 1780-1782. http://dx.doi.org/10.1212/01.WNL.0000162034.06268.37

197. Grabler, M. R., Weyen, U., Juckel, G., Tegenthoff, M., & Mavrogiorgou-Juckel, P. (2018). Death Anxiety and Depression in Amyotrophic Lateral Sclerosis Patients and Their Primary Caregivers. *Front Neurol,* 9, 1035. http://dx.doi.org/10.3389/fneur.2018.01035

198. Rabkin, J. G., Wagner, G. J., & Del Bene, M. (2000). Resilience and distress among amyotrophic lateral sclerosis patients and caregivers. *Psychosom Med,* 62(2), 271-279. http://dx.doi.org/10.1097/00006842-200003000-00020

199. Chio, A., Gauthier, A., Vignola, A., Calvo, A., Ghiglione, P., Cavallo, E., . . . Mutani, R. (2006). *Caregiver time use in ALS*. *Neurology,* 67(5), 902-904. http://dx.doi.org/10.1212/01.wnl.0000233840.41688.df

200. Qutub, K., Lacomis, D., Albert, S. M., & Feingold, E. (2014). Life factors affecting depression and burden in amyotrophic lateral sclerosis caregivers. *Amyotroph Lateral Scler Frontotemporal Degener,* 15(3-4), 292-297. http://dx.doi.org/10.3109/21678421.2014.886699

201. Harris, M., Thomas, G., Thomas, M., Cafarella, P., Stocks, A., Greig, J., & McEvoy, R. D. (2018). Supporting wellbeing in motor neurone disease for patients, carers, social networks, and health professionals: A scoping review and synthesis. *Palliat Support Care,* 16(2), 228-237. http://dx.doi.org/10.1017/S1478951517000700

202. Aoun, S. M., Connors, S. L., Priddis, L., Breen, L. J., & Colyer, S. (2012). Motor Neurone Disease family carers' experiences of caring, palliative care and bereavement: an exploratory qualitative study. *Palliat Med,* 26(6), 842-850. http://dx.doi.org/10.1177/0269216311416036

203. Aoun, S. M., Bentley, B., Funk, L., Toye, C., Grande, G., & Stajduhar, K. J. (2013). A 10-year literature review of family caregiving for motor neurone disease: moving from caregiver burden studies to palliative care interventions. *Palliat Med,* 27(5), 437-446. http://dx.doi.org/10.1177/0269216312455729

204. Galvin, M., Corr, B., Madden, C., Mays, I., McQuillan, R., Timonen, V., . . . Hardiman, O. (2016). Caregiving in ALS - a mixed methods approach to the study of Burden. *BMC Palliat Care,* 15(1), 81. http://dx.doi.org/10.1186/s12904-016-0153-0

205. Gluyas, C., Mathers, S., Hennessy Anderson, N., & Ugalde, A. (2017). Factors to consider for motor neurone disease carer intervention research: A narrative literature review. *Palliat Support Care,* 15(5), 600-608. http://dx.doi.org/10.1017/S1478951516000912

206. Pagnini, F. (2013). Psychological wellbeing and quality of life in amyotrophic lateral sclerosis: a review. *Int J Psychol,* 48(3), 194-205. http://dx.doi.org/10.1080/00207594.2012.691977

207. Trucco, A. P., Backhouse, T., Mioshi, E., & Kishita, N. (2024). Factors associated with grief in informal carers of people living with Motor Neuron Disease: A mixed methods systematic review. *Death Stud,* 48(2), 103-117. http://dx.doi.org/10.1080/07481187.2023.2191351

208. Matuz, T., Birbaumer, N., Hautzinger, M., & Kubler, A. (2015). Psychosocial adjustment to ALS: a longitudinal study. *Front Psychol,* 6, 1197. http://dx.doi.org/10.3389/fpsyg.2015.01197

209. Pagnini, F., Rossi, G., Lunetta, C., Banfi, P., Castelnuovo, G., Corbo, M., & Molinari, E. (2010). Burden, depression, and anxiety in caregivers of people with amyotrophic lateral sclerosis. *Psychol Health Med,* 15(6), 685-693. http://dx.doi.org/10.1080/13548506.2010.507773

210. Goldstein, L. H., Atkins, L., Landau, S., Brown, R. G., & Leigh, P. N. (2006). Longitudinal predictors of psychological distress and self-esteem in people with ALS. Neurology, 67(9), 1652-1658. http://dx.doi.org/10.1212/01.wnl.0000242886.91786.47

211. Averill, A. J., Kasarskis, E. J., & Segerstrom, S. C. (2007). Psychological health in patients with amyotrophic lateral sclerosis. *Amyotroph Lateral Scler,* 8(4), 243-254. http://dx.doi.org/10.1080/17482960701374643

212. Bolmsjo, I. (2001). Existential issues in palliative care: interviews of patients with amyotrophic lateral sclerosis. *J Palliat Med,* 4(4), 499-505. http://dx.doi.org/10.1089/109662101753381647

213. Mistry, K., & Simpson, J. (2013). Exploring the transitional process from receiving a diagnosis to living with motor neurone disease. *Psychol Health,* 28(8), 939-953. http://dx.doi.org/10.1080/08870446.2013.770513

214. Aoun, S. M., Kissane, D. W., Cafarella, P. A., Rumbold, B., Hogden, A., Jiang, L., & Bear, N. (2020). Grief, depression, and anxiety in bereaved caregivers of people with motor neurone disease: a population-based national study. *Amyotroph Lateral Scler Frontotemporal Degener,* 21(7-8), 593-605. http://dx.doi.org/10.1080/21678421.2020.1790610

215. de Wit, J., Bakker, L. A., van Groenestijn, A. C., van den Berg, L. H., Schroder, C. D., Visser-Meily, J. M. A., & Beelen, A. (2018). Caregiver burden in amyotrophic lateral sclerosis: A systematic review. *Palliat Med,* 32(1), 231-245. http://dx.doi.org/10.1177/0269216317709965

216. Conroy, E., Kennedy, P., Heverin, M., Hardiman, O., & Galvin, M. (2023). Care, burden and self-described positive aspects of caring in amyotrophic lateral sclerosis: an exploratory, longitudinal, mixed-methods study. *BMJ Open,* 13(1), e064254. http://dx.doi.org/10.1136/bmjopen-2022-064254

217. Roth, D. L., Fredman, L., & Haley, W. E. (2015). Informal caregiving and its impact on health: a reappraisal from population-based studies. *Gerontologist,* 55(2), 309-319. http://dx.doi.org/10.1093/geront/gnu177

218. Zarit, S. H., Todd, P. A., & Zarit, J. M. (1986). Subjective burden of husbands and wives as caregivers: a longitudinal study. *Gerontologist,* 26(3), 260-266. http://dx.doi.org/10.1093/geront/26.3.260

219. Giusiano, S., Peotta, L., Iazzolino, B., Mastro, E., Arcari, M., Palumbo, F., . . . Vasta, R. (2022). Amyotrophic lateral sclerosis caregiver burden and patients' quality of life during COVID-19 pandemic. *Amyotroph Lateral Scler Frontotemporal Degener,* 23(1-2), 146-148. http://dx.doi.org/10.1080/21678421.2021.1912772

220. Cafarella, P., Effing, T., & Chur-Hansen, A. (2022). Interventions targeting psychological well-being for motor neuron disease carers: A systematic review. *Palliat Support Care,* 1-17. http://dx.doi.org/10.1017/S1478951522000311

221. Ugalde, A., Mathers, S., Hennessy Anderson, N., Hudson, P., Orellana, L., & Gluyas, C. (2018). A self-care, problem-solving and mindfulness intervention for informal caregivers of people with motor neurone disease: *A pilot study. Palliat Med,* 32(4), 726-732. http://dx.doi.org/10.1177/0269216317743434

222. Pagnini, F., Phillips, D., Haulman, A., Bankert, M., Simmons, Z., & Langer, E. (2022). An online non-meditative mindfulness intervention for people with ALS and their caregivers: a randomized controlled trial. *Amyotroph Lateral Scler Frontotemporal Degener,* 23(1-2), 116-127. http://dx.doi.org/10.1080/21678421.2021.1928707

223. De Wit, J., Beelen, A., Drossaert, C. H. C., Kolijn, R., Van Den Berg, L. H., SchrOder, C. D., & Visser-Meily, J. M. A. (2020). Blended psychosocial support for partners of patients with ALS and PMA: results of a randomized controlled trial. *Amyotroph Lateral Scler Frontotemporal Degener,* 21(5-6), 344-354. http://dx.doi.org/10.1080/21678421.2020.1757114

224. de Wit, J., Vervoort, S., van Eerden, E., van den Berg, L. H., Visser-Meily, J. M. A., Beelen, A., & Schroder, C. D. (2019). User perspectives on a psychosocial blended support program for partners of patients with amyotrophic lateral sclerosis and progressive muscular atrophy: a qualitative study. *BMC Psychol,* 7(1), 35. http://dx.doi.org/10.1186/s40359-019-0308-x

225. Steinhauser, K. E., Olsen, A., Johnson, K. S., Sanders, L. L., Olsen, M., Ammarell, N., & Grossoehme, D. (2016). The feasibility and acceptability of a chaplain-led intervention for caregivers of seriously ill patients: A Caregiver Outlook pilot study. *Palliat Support Care,* 14(5), 456-467. http://dx.doi.org/10.1017/S1478951515001248

226. Cipolletta, S., Gammino, G. R., Francescon, P., & Palmieri, A. (2018). Mutual support groups for family caregivers of people with amyotrophic lateral sclerosis in Italy: A pilot study. *Health Soc Care Community,* 26(4), 556-563. http://dx.doi.org/10.1111/hsc.12558

227. Creemers, H., Veldink, J. H., Grupstra, H., Nollet, F., Beelen, A., & van den Berg, L. H. (2014). Cluster RCT of case management on patients' quality of life and caregiver strain in ALS. *Neurology,* 82(1), 23-31. http://dx.doi.org/10.1212/01.wnl.0000438227.48470.62

228. Kavanaugh, M. S., Cho, C. C., Howard, M., Fee, D., & Barkhaus, P. E. (2020). US data on children and youth caregivers in amyotrophic lateral sclerosis. *Neurology,* 94(14), e1452-e1459. http://dx.doi.org/10.1212/WNL.0000000000009217

229. Kavanaugh, M. S., Cho, Y., Fee, D., & Barkhaus, P. E. (2020). Skill, confidence and support: conceptual elements of a child/youth caregiver training program in amyotrophic lateral sclerosis - the YCare protocol. *Neurodegener Dis Manag,* 10(4), 231-241. http://dx.doi.org/10.2217/nmt-2020-0004

230. Aoun, S. M., O'Brien, M. R., & Knighting, K. (2024). Using the Carers' Alert Thermometer tool to identify needs and support family caregivers of people with motor neurone disease: moving beyond needs assessments. *Palliat Care Soc Pract,* 18, 26323524241228306. http://dx.doi.org/10.1177/26323524241228306

231. MND Australia. (2018). MND Australia Research Priorities Survey 2018, from https://www.mndaustralia.org.au/getattachment/618cf2f3-6c05-43f9-ad61-1fbe789ecb2e/Advance-June-2018.pdf?lang=en-AU

232. Velaga, V. C., Cook, A., Auret, K., Jenkins, T., Thomas, G., & Aoun, S. M. (2023). Palliative and End-of-Life Care for People Living with Motor Neurone Disease: Ongoing Challenges and Necessity for Shifting Directions. *Brain Sci,* 13(6). http://dx.doi.org/10.3390/brainsci13060920

233. NOUS. (2018). Compassionate communities: An implementation guide for community approaches to end-of-life care, from https://palliativecare.org.au/wp-content/uploads/dlm_uploads/2018/09/An-implementation-guide-for-community.pdf

234. Aoun, S. M., Deas, K., Kristjanson, L. J., & Kissane, D. W. (2017). Identifying and addressing the support needs of family caregivers of people with motor neurone disease using the Carer Support Needs Assessment Tool. *Palliat Support Care,* 15(1), 32-43. http://dx.doi.org/10.1017/S1478951516000341

235. Olesen, L. K., la Cour, K., With, H., Mahoney, A. F., & Handberg, C. (2022). A cross-sectional evaluation of acceptability of an online palliative rehabilitation program for family caregivers of people with amyotrophic lateral sclerosis and cognitive and behavioral impairments. *BMC Health Serv Res*, 22(1), 697. http://dx.doi.org/10.1186/s12913-022-07986-4

236. Thomas, P. T., Warrier, M. G., Arun, S., Bhuvaneshwari, B., Vengalil, S., Nashi, S., . . . Nalini, A. (2023). An individualised psychosocial intervention program for persons with MND/ALS and their families in low resource settings. *Chronic Illn,* 19(2), 458-471. http://dx.doi.org/10.1177/17423953221097076

237. Poppe, C., Kone, I., Iseli, L. M., Schweikert, K., Elger, B. S., & Wangmo, T. (2020). Differentiating needs of informal caregivers of individuals with ALS across the caregiving course: a systematic review. *Amyotroph Lateral Scler Frontotemporal Degener,* 21(7-8), 519-541. http://dx.doi.org/10.1080/21678421.2020.1771735

238. Galvin, M., Carney, S., Corr, B., Mays, I., Pender, N., & Hardiman, O. (2018). Needs of informal caregivers across the caregiving course in amyotrophic lateral sclerosis: a qualitative analysis. *BMJ Open,* 8(1), e018721. http://dx.doi.org/10.1136/bmjopen-2017-018721


APS
Australian
Psychological
Society