New Zealand

Best Practice Recommendations

for the Care of People with Motor Neurone Disease

Whakahara-tau Rangatira mō te Manaaki Mate Tauheke Ioio

mnd
Motor Neurone Disease
New Zealand

Motor Neurone Disease New Zealand

The **MND Association of New Zealand** (MND New Zealand) is a small not-for-profit organisation. Our main purpose is to ensure that people living with MND have the **best quality of life** possible.

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Endorsements

NANZ Neurological Association of New Zealand

Neurological Association of New Zealand (NANZ),

anzspm Australia New Zealand Society of Palliative Medicine

Australian and New Zealand Society of Palliative Medicine (ANZSPM) Aotearoa,

The Thoracic Society of Australia & New Zealand LEADERS IN LUNG HEALTH

Thoracic Society of Australia and New Zealand (TSANZ)

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Acknowledgments

MND New Zealand would like to acknowledge the significant time and effort contributed by the Motor Neurone Disease Clinical Working Group (MNDCWG) in the development of these Best Practice Recommendations. The group includes 18 members from different health sectors around Aotearoa New Zealand, who have an interest in improving care for people with MND and their [1]whānau. The MNDCWG was co-chaired by Claire Reilly and Chris Drennan in 2020, and Claire Reilly and Alan Stanley in 2021.

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The MND Clinical Working Group would like to acknowledge the following contributors who provided expertise and guidance:

- **Graham Cameron**, Pou Tikanga at Bay of Plenty DHB
- **Helen Lappin**, Wheelchair Therapist
- **Ann Smaill**, Speech-language Therapist, TalkLink Trust

Citation

This document and links to all other documents referred to in this document are available on the website at https://mnd.org.nz/for-professionals.

The latest version of this document is available on the MND New Zealand website only. Printed copies may not reflect the most recent updates.

1 The word "whānau" is used throughout this document to refer to immediate family and extended family, and also acknowledges the important role these people frequently play as informal caregivers (paid and unpaid.)

The word "carer" is reserved for reference to trained, formal external caregivers.

Foreword

It is a pleasure and a privilege as the Medical Patron of MND New Zealand to write this foreword for the publication of "New Zealand Best Practice Recommendations for the Care of People with Motor Neurone Disease".

The overriding mission and vision of community-based organisations like MND New Zealand is to provide the very latest information, advice and guidelines on the care and support of people and whānau of Aotearoa New Zealand who are touched by MND. I therefore congratulate MND New Zealand for enthusiastically responding to the request arising out of the hui in Wellington on 18th September 2019 to initiate a project "Improving care, improving lives", aimed at improving the clinical management of Motor Neurone Disease in New Zealand. In fulfilment of this objective, MND New Zealand set up a working group of 18 nationally representative leading clinicians, experts in MND care and treatment.

The working group (MNDCWG) was co-chaired by Drs Claire Reilly, Chris Drennan and Alan Stanley to establish a set of national guidelines and recommendations for the holistic care of people with MND. The MNDCWG have been deliberating and meeting over the last 24 months. In drawing up their recommendations, the MNDCWG have consulted widely with other appropriate clinical experts across New Zealand as well as drawing on best practice clinical guidelines on MND care from the United Kingdom and Canada.

The outcome is this comprehensive forward-looking document especially for the guidance of clinicians which will be periodically reviewed and updated as necessary. This is a first and a milestone for standardising and promoting world class MND holistic clinical care across Aotearoa New Zealand.

The key points that I would like to highlight in these comprehensive and superb recommendations are:

- They represent the first step in an overall framework being developed in response to concerns raised by the MND Community in New Zealand and further resources are being developed
- They have been contextualised for New Zealand and recognise the important roles of Te Tiriti o Waitangi and Te Whare Tapa Whā in the delivery of healthcare services in Aotearoa New Zealand
- They have been written to provide clinical guidance and to standardise care throughout NZ
- They have been peer-reviewed with endorsement from NANZ, TSANZ and ANZSPM Aotearoa and it's hoped that Health NZ will encourage their adoption and implementation

On behalf of all the people and whānau who are touched by MND, I would like to thank MND New Zealand and the Working Group for this superb hallmark comprehensive document which will give hope for standardising the future clinical care of MND people across Aotearoa New Zealand.

Sir Richard Faull KNZM, FRSNZ
Director, Centre for Brain Research, University of Auckland
Medical Patron, MND New Zealand.

Table of Contents

Glossary

The terminology throughout this document is consistent with the NICE Guideline for "Motor neurone disease: assessment and management"(1). It is suggested that the reader refers to the Glossary on page 309 if any clarification or elaboration is required.

https://www.nice.org.uk/guidance/ng42/evidence/full-guideline-pdf-2361774637

Introduction

Why were these recommendations developed?

It is estimated that there are currently around 350 people living with Motor Neurone Disease (MND) in Aotearoa New Zealand. Our aging population will see this number and the associated burden on the New Zealand health system increase. MND is a highly complex disease which requires input from multiple specialties in a coordinated way and integrated across different health and disability sectors. Work carried out by MND New Zealand has confirmed concerns raised by clinicians nationally about variability in clinical practice and the need for national standards to ensure people with MND and their whānau receive best practice care that is timely, cohesive and evidence based.

These recommendations form one part of the "Improving care, improving lives" initiative led by MND New Zealand (Appendix A). This framework has been developed in response to issues that people living with Motor Neurone Disease (*Mate Tauheke Ioio*), and their whānau have raised with MND New Zealand. The aim is to develop a series of resources that will improve the care that people with MND and their whānau receive in the New Zealand health care setting. More information about this initiative can be found here.

Purpose and Scope

The purpose of the Best Practice Recommendations is to provide clinicians, allied health professionals and primary care providers with best practice recommendations for the care and management for people with MND and their whānau in New Zealand. It has been adapted from the 2016 United Kingdom (UK) NICE guideline "Motor neurone disease: assessment and management" and is evidence-based where evidence exists (1). As Aotearoa New Zealand has its own unique cultural heritage and values these recommendations have been contextualised to recognise the important roles of Te Tiriti o Waitangi and Te Whare Tapa Whā in the delivery of healthcare services in New Zealand (Appendix B). They also reflect the variance in population, ethnicity, health systems (including funding and governance), economic constraints and access to care.

The recommendations define a national standard of care for people living with MND for clinical and health policy purposes and are intended to facilitate the development of local MND care pathways throughout New Zealand. They encourage early diagnosis of MND and emphasise the need for integrated health and social services that are person-centred and delivered in a culturally appropriate way.

It is acknowledged that some regions may have limited resources, and regular audit against these standards is encouraged to address these gaps.

Methods

The MND Clinical Working Group (CWG) was formed following an invitation for participants at the MND New Zealand hui in 2019. The group includes 18 members from different health sectors around New Zealand. A lack of knowledge of Tikanga Māori meant external consultation was undertaken. Dr. Melissa Carey, Research Fellow, Māori Health from the University of Auckland, joined the group in September 2021. Raewyn Robinson replaced Heather Brunton as the MND Clinical Nurse Specialist in August 2021.

Development of the Best Practice Recommendations

The CWG first met via teleconference in November 2019 and subsequently met face to face in February 2020. Members met regularly via teleconference throughout 2020 and 2021.

Early in the guideline development process, one other clinician was involved, but they removed themselves from the project because of the time commitments required.

Issues associated with the clinical care of people with MND had already been identified following the hui. A review of existing international guidelines identified that the UK NICE Guideline for "Motor neurone disease: assessment and management" (2016) was the most relevant and up-to-date international guideline available and the group agreed to review this document and contextualise it for the New Zealand setting based on up-to-date research and clinical experience. Each section of the guideline was reviewed by members of the group with relevant experience and then presented to the whole group for any amendments. As many areas lack evidence from clinical trials to direct care in MND, the working group members thought it important that the recommendations be based on group consensus, utilising non-clinical trial literature, evidence in other diseases, or current New Zealand clinical practice. Where necessary, expertise was co-opted from outside the group, such as in the areas of wheelchair seating and assessment, and communication. As a participant from primary health was not available, input on the final draft was requested from General Practitioners (GPs) before publication.

Review process

In November 2020, a working group of Canadian physicians released the "Canadian Best Practice Recommendations for The Management of Amyotrophic Lateral Sclerosis" (2). These were reviewed by the group in early February 2021 to ensure that there were no significant areas of inconsistency.

When the working group was satisfied with the final draft it was emailed to topic experts external to the working group for open-ended feedback. All comments received were considered by the working group and implemented by consensus.

In September 2021, a motion to change the title to "New Zealand Best Practice Recommendations for The Care of People with Motor Neurone Disease" was approved by the group.

During the initial phase of the guideline development, the group attempted to engage a Māori health advisor without success. In the months that followed Graham Cameron, Pou Tikanga at the Bay of Plenty District Health Board provided the group with feedback on the Introduction and Overview. Dr Melissa Carey, health researcher from Auckland University reviewed the final draft.

A complete version of the guideline was prepared and reviewed by all working group members for final approval.

Competing interests

All members of the working group performed their tasks voluntarily and were not paid honoraria for their involvement. The development of the guideline, including out of pocket expenses for travel for face-to-face meetings and preparation of the manuscript for publication, was funded by MND New Zealand.

A conflicts of interest register was completed by members during the planning process. HM had recently commenced working with Cubro but joined the group as an Occupational Therapist (OT) & was not on commission. CR & TF both work for MND New Zealand and CR is a medical doctor living with Motor Neurone Disease. FH, CD and AS were MND New Zealand Council members at some point during the guideline's development. JC is the MND New Zealand Medical Advisor.

Overview of Motor Neurone Disease (MND)

Motor Neurone Disease (MND), the most prevalent form of which is amyotrophic lateral sclerosis (ALS), is a progressive neuro-degenerative disease affecting the motor neurones of the brain and spinal cord (3). Destruction of upper and lower motor neurons results in spasticity, fasciculations, muscle weakness and atrophy. Clinical presentation varies depending upon the site of onset but may include weakness in arms and/or legs with problems related to dexterity and/or mobility (spinal-onset; approximately 2/3), or problems with speech and/or swallowing (bulbar-onset; approximately 1/3). Less commonly, people present with difficulty breathing (less than 3%) (4). Up to 50% of people with MND also demonstrate cognitive or behavioural impairments due to degeneration in the frontal and temporal lobes and 15% meet the criteria for Fronto-temporal Dementia (FTD) (5). Increasingly, ALS and FTD are being recognised as a spectrum of disorders representing the same disease process.

Diagnosis of MND is based on clinical findings, with the coexistence of upper and lower motor neurone signs in the same symptomatic area being characteristic. Clinical diagnosis is supported by deterioration over time, findings on EMG and nerve conduction testing, and exclusion of mimics (4, 6). MND is used synonymously with amyotrophic lateral sclerosis (ALS) in the USA, however, in the UK and Australasia, MND is an overarching term for various diseases of the motor nerves, classified by whether they affect upper or lower motor neurons or both (7). Four main clinical phenotypes have been identified, with site of onset and phenotype being associated with prognosis. The most common form, ALS, accounts for 90% of cases of MND (7). Individuals with ALS show remarkable heterogeneity in both presentation and rate of progression. Ultimately weakness in the respiratory muscles results in respiratory failure and most people die within 3-4 years of symptom onset (6). Slower rates of progression are often seen in 'pure' lower motor neurone or upper motor neurone cases (progressive muscular atrophy (PMA) and primary lateral sclerosis (PLS) (4). Progressive Bulbar Palsy (PBP) is characterised by onset in the bulbar region, often leading to rapidly progressive dysarthria and dysphagia, and overall survival of 6 months-4 years (8). There is considerable overlap between the different variants.

The life-time risk of developing MND is 1 in 300, with the disease being more common after 50 years, but affecting adults of any age (1,9). It is more common in males with most population-based studies finding a male: female ratio of 1.2–1.5 to 1 (6). The cause is not fully understood, but increasingly appears to involve a complex interplay of genetic, epigenetic, internal, and environmental factors resulting in neurodegeneration. Studies suggest that, like carcinogenesis, this pathway requires multiple steps, or 'molecular insults' to cause disease (multistep hypothesis of ALS) (10,11).

The exact prevalence of MND in Aotearoa New Zealand is not known. Based on data from international studies, it is estimated that there are over 350 people living with the disease in New Zealand at any one time (from prospective figures calculated by Chio et al 2013 (12)). Recently published MND mortality data suggest that the prevalence may be higher than in other countries, and rates of MND were significantly lower in Māori compared to those of European descent (13). This is consistent with other studies which suggest a higher incidence of ALS in Caucasian compared to Asian, African, and Hispanic groups (14). Although rates of MND were significantly less in Māori, (raising the possibility that Māori may have protective factors for MND) existing healthcare disparities may mean the lower rate reflects the fact that fewer Māori with MND are accessing health services and therefore fewer are diagnosed.

There is no cure for MND, and management centres on maximising function and quality of life. Interprofessional collaborative practice (ICP) is the cornerstone of care and coordinated interdisciplinary treatment has been shown to improve quality of life and survival (3). Similarly, early intervention with non-invasive ventilation (NIV) for respiratory dysfunction and nutritional support to maintain weight are associated with better outcomes (6). Strategies to reduce diagnostic delays are urgently needed to ensure timely interventions, as well as provision of equipment and aids.

Person-focused care is central to the management of people with MND, and this requires a holistic approach as exemplified by Te Whare Tapa Whā. Ultimately it is the person, in consultation with whānau who decides and consents to treatment and this includes the option of declining interventions.

Assessment, recognition & referral

- The primary care team plays a fundamental role throughout the course of the disease from early recognition and referral to assessment of needs and advocating for and arranging community services.
- If MND is suspected, the person should be referred without delay for neurology review and the possible diagnosis specified in the referral letter.
- If the person needs to be seen urgently (e.g., respiratory compromise, rapid weight loss, rapid loss of function, unable to manage activities of daily living (ADLs) a consultant neurologist should be contacted directly.
- Monitor and assess for symptoms/ signs of respiratory involvement, weight loss and cognitive change – these have implications for decision making and future management.

Presentation

The insidious nature of MND and the wide range of presentations can make diagnosis challenging. The median time from first symptom to diagnosis is 12 months (15). There can be several reasons for delays, including the person either not recognising or ignoring early or intermittent symptoms, lack of familiarity with the condition by clinicians and clinical complexity. It is not uncommon for people to have had assessments by several disciplines, which can also delay diagnosis.

People frequently present with symptoms in one muscle group e.g., foot drop. As the disease progresses, other muscle groups become involved. Progression is variable - not all symptoms happen to everyone, nor do they develop in the same order.

The clinical hallmark is progressive and painless muscular weakness, without sensory disturbance (3).

Common first symptoms

Physical (taha tinana):

- Stumbling, falls or trips, foot drop
- Weakened grip, loss of dexterity
- Speech or swallowing problems e.g., change in speech (quiet or slurred speech), having to 'double swallow'
- Weight loss, muscle twitching (fasciculations), prominent new onset cramps and stiffness

Cognitive (taha hinengaro):

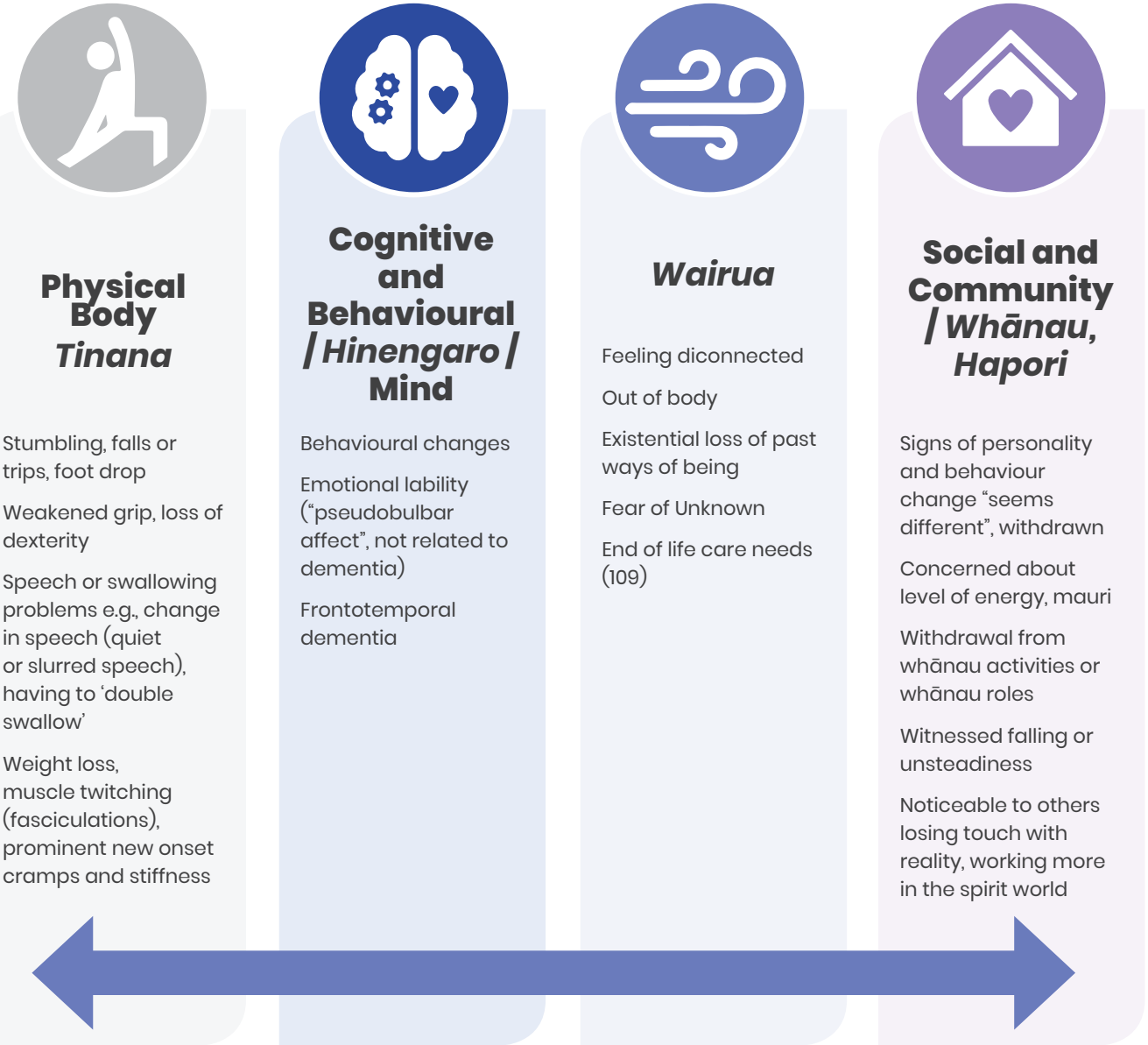
- Behavioural changes
- Emotional lability ("pseudobulbar affect", not related to dementia)
- Frontotemporal dementia

Whānau may report:

- Personality and behaviour change
- Concern about level of mauri
- Withdrawal from whānau activities or whānau roles
- Falling or unsteadiness
- Losing touch with reality, working more in the spirit world
- Dampening of spirit, thinking about end of life
- Loss of life purpose

Less common presentations

- Breathing problems, (in the absence of underlying lung disease) such as shortness of breath lying flat or on exertion, excessive daytime sleepiness, fatigue, early morning headache (Mauri Hā/ Hā ora)
- MND rarely affects bladder and bowel function directly. Upper motor neurone predominant disease can cause bladder overactivity. Constipation is common in advanced disease.



Examination findings

- Muscle wasting and weakness
- Fasciculations (often seen in proximal limb muscles including pectoral, deltoid, and abdomen)
- Mixed upper and lower motor neuron signs (hyperreflexia in wasted weak muscles)
- Many will have normal cognitive function, but some people may experience cognitive and/or behavioural changes
- If cognitive changes are suspected, perform a cognitive screening test (Mini-ACE) (16)

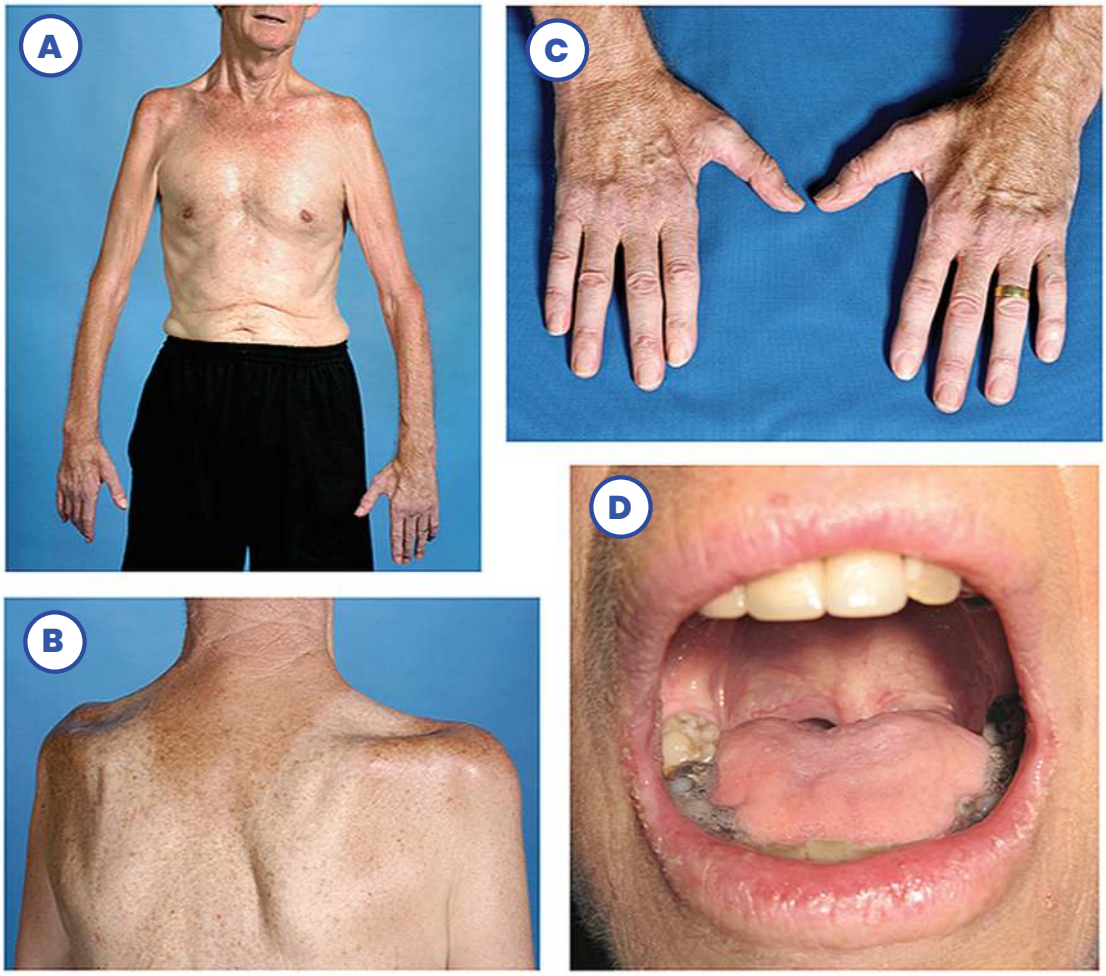


Figure 1: Clinical features of muscles wasting in a patient with ALS (17)

Proximal and symmetrical upper limb wasting (A) results in an inability to lift arms against gravity ("man-in-the-barrel" or flail-arm variant ALS). Note the recessions above and below the scapular spine (B), indicating wasting of supraspinatus and infraspinatus muscles, as well as substantial loss of deltoid muscle. As a consequence, the glenohumeral joint becomes prominent, and prone to subluxation. (C) Disproportionate wasting of the thenar muscles combined with the first dorsal interossei, the so-called "split-hand", is a typical feature in ALS. (D) Substantial wasting of the tongue muscles in bulbar-onset ALS. Note the absence of palatal elevation present on vocalisation. The tongue is often disproportionately affected in comparison to other oropharyngeal musculature in patients with bulbar-onset ALS

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Diagnosis

There is no single test for MND. The most important consideration in the primary care setting is for GPs to be alert to the possibility of MND and refer appropriately. This should take precedence over any investigations when MND is suspected.

The MND UK Association's "Red Flag tool for GPs" (Appendix C), produced in partnership with the Royal College of General Practitioners, is designed to improve timely referrals to neurology and therefore speed up the time to accurate diagnosis. (18).

Management: Key Actions for General Practitioners

1. Prompt referral for diagnosis

It is important that regional protocols and pathways are in place to ensure timely referrals for accurate specialist diagnosis and that all health professionals are aware of these. Use the red flag tool to recognise early signs of MND to refer to neurology in a timely manner (ideally 4-6 weeks).

If MND is suspected

- the person should be referred without delay for neurology review and
- the possible diagnosis specified in the referral letter

If the person needs to be seen urgently (e.g., respiratory compromise, rapid weight loss, rapid loss of function, unable to manage activities of daily living (ADLs) contact the consultant neurologist directly.

The uncertainty that accompanies a possible diagnosis of a neurological disease creates a significant amount of stress for the person involved and their whānau, particularly if there are diagnostic delays. It is important to acknowledge the anxiety and provide appropriate information & support. Where a diagnosis of MND seems likely MND New Zealand will support the person and whānau through this period of diagnostic uncertainty. MND New Zealand can advise of other appropriate agencies that may be available locally e.g., palliative care support, Māori liaison health team.

2. Assess needs

Use a person-centred approach to listen to and ascertain the tinana (physical), mauri and wairua (emotional, and spiritual) and whānau, needs of the person with MND at each appointment; what do they need to uphold their mana/dignity.

Cultural Safety

It is important to incorporate cultural safety principles into the assessment process when communicating with individuals and whānau. This requires a holistic lens with a wide understanding of how different people describe their bodies and what it feels like to be them. A consideration of the power imbalance between the health professional and the person presenting for care is an important factor, particularly in neurological disorders when people are already feeling a loss of power and control over their bodies and the way they interact with the world (19).

3. Arrange Community Services support- a confirmed diagnosis is not necessary.

Changes in mobility, communication, swallowing and breathing can occur quickly, and general practice teams need to be prepared to advocate for rapid attention from providers. Referral for allied health input for speech language therapy, physiotherapy (for mobility/balance/respiratory assessment), and occupational therapy should take place without delay.

4. Refer to appropriate specialist teams as necessary.

In collaboration with consultants in neurology and palliative care, initiate appropriate management and treatment, including anticipatory symptomatic intervention. Include the person with MND on local palliative care registers/lists/ coordination systems, where these exist.

5. Ongoing assessment and care

- Provide support and information throughout the course of the disease.
- Complete necessary forms to support a benefit application.
- Issue repeat prescriptions if the person with MND is prescribed Riluzole by their neurologist: a shared-care protocol should be agreed.
- Assess and monitor any impact the disease is having on the person's ability to drive
- Monitor and assess for symptoms/ signs of:
  - Respiratory involvement – early signs should trigger referral to the specialist respiratory team.
  - Cognitive change – this has implications for decision making and future management.
- Advance care Plan (ACP)- Help the person with MND to talk through management options, including end of life decisions and Advance Directives, including Do Not Attempt Resuscitation (DNACPR), as early as possible (see page 67, "Palliative and End of Life Care").

Communicating the Diagnosis

- Diagnosis must be confirmed by a neurologist, or a physician experienced in the management of MND. Information about the diagnosis, prognosis and management should be given by a neurologist with up-to-date knowledge of the condition (1). Where this is not possible, the physician leading the interdisciplinary team (IDT) should assume this role.
- Tailor discussions to the person's needs, considering their communication ability, cognitive status, health literacy and cultural needs. Stage the provision of information and provide it in both oral and written formats. Provide supporting resources as appropriate.
- Refer person to care coordinator for MND interdisciplinary team and support services including MND New Zealand. A follow up appointment should be arranged within 2-4 weeks with someone from the IDT to answer queries and provide on-going support/management.

Most people with MND initially present to their GP, however referral may also come via other secondary health services e.g., ENT, physiotherapist, speech language therapist, orthopaedics (15). The diagnosis can be difficult and require repeat assessments over several months.

thinksALS- Benefits of Timely Diagnosis

The challenges associated with accurate diagnosis and resulting delays to diagnosis of MND have been well documented (20). The ALS Association's Time to Diagnosis working group published a consensus statement and an associated tool for clinicians on the importance of timely diagnosis. It highlights the need for the earliest possible diagnosis to provide opportunity for timely interventions that can improve outcomes and allow enrollment into clinical trials, whilst also recognising how difficult it can be to give and receive a diagnosis of MND.

New Zealand MND Clinical Network

Many countries have recognised the value of specialist MND centres and the benefits of early referral to such centres. Whilst this is not currently available in New Zealand, there are neurologists and other clinicians who are part of the New Zealand MND Clinical Network with experience treating people with MND and can provide peer support. They can be contacted through MND New Zealand.

Gold Coast Criteria

A consensus meeting held on the Gold Coast in 2019 led to the development of a simplified diagnostic criteria for ALS, abandoning the previous confusing categories of possible, probable, and definite ALS (21).

ALS was defined by the presence of:

- *Progressive motor impairment, documented by history or repeated clinical assessment, preceded by normal motor function.*
- *Upper and lower motor neurone dysfunction in at least one body region (in the same body region if only one body region was involved), or lower motor neuron dysfunction in at least two body regions.*
- *Investigation findings that excluded alternative disease processes.*

Rarely, people present with 'pure' LMN (progressive muscular atrophy (PMA)) or UMN (progressive lateral sclerosis (PLS)) disease, typically associated with slower progression. Progressive Bulbar Palsy (PBP) is characterised by onset in the bulbar region, often leading to rapidly progressive dysarthria and dysphagia.

1. Progressive motor impairment documented by history or repeated clinical assessment, preceded by normal motor function, and
2. Presence of upper1 and lower2 motor neuron dysfunction in at least one body region3, (with upper and lower motor neuron dysfunction noted in the same body region, if only one body region is involved) or lower motor neuron dysfunction in at least two body regions, and
3. Investigations4 excluding other disease processes

Footnotes:

[1]Upper motor neuron dysfunction implies at least one of the following:

1. Increased deep tendon reflexes, including the presence of a reflex in a clinically weak and wasted muscle, or spread to adjacent muscles
2. Presence of pathological reflexes, including Hoffman sign, Babinski sign, crossed adductor reflex, or snout reflex.
3. Increase in velocity-dependent tone (spasticity)
4. Slowed, poorly coordinated voluntary movement, not attributable to weakness of lower motor neuron origin or Parkinsonian features

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[2]Lower motor neuron dysfunction in a given muscle requires either:

Clinical examination evidence of muscle weakness, and muscle wasting

or

Electromyography (EMG) abnormalities that must include:

Both evidence of chronic neurogenic change, defined by large motor unit potentials of increased duration and/or increased amplitude, with polyphasia and motor unit instability regarded as supportive but not obligatory evidence.

and

Evidence of ongoing denervation including:

Fibrillation potentials or positive sharp waves, or fasciculation potentials

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[3]Body regions are defined as bulbar, cervical, thoracic, and lumbosacral. To be classified as an involved region with respect to lower motor neuron involvement, there must be:

1. abnormalities in two limb muscles innervated by different roots and nerves, or
2. one bulbar muscle, or one thoracic muscle either by clinical examination or by EMG.

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[4]The appropriate investigations depend on the clinical presentation, and may include:

3. Nerve conduction studies and needle EMG, MRI or other imaging, fluid studies of blood or CSF, or other modalities as clinically necessary.

Criteria for diagnosis of ALS (21)

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MND and Cognition

Over 50% of people with MND have changes in cognitive function, varying from subtle cognitive impairment to frank fronto-temporal dementia (22). These changes have implications for communication, decision making (including provision of informed consent) and care needs. Ideally, screening for cognitive and behavioral impairment should be performed in people with MND early in their disease.

At diagnosis or if there is a concern about cognition or behaviour

- Explore these areas with the person and their whānau.
- Undertake a formal cognitive assessment, using the Edinburgh Cognitive and Behavioral ALS Screen (ECAS). Where this is not possible, screening should be performed by a clinician with appropriate training using validated tools e.g., ECAS, ALS-CBS (23, 24). Frontal lobe dysfunction may not be apparent on some tests (eg Mini-ACE). If uncertainty remains, then referral to a Neuropsychologist should be considered.
- The MiND-B is a simple tool for the identification and quantification of behavioural symptoms in people with MND (25).
- Documenting cognitive involvement is important. Discussions about advance care plans (ACP) should take place early & people with MND and whānau should be encouraged to ensure a Power of Attorney is appointed. Occasionally a formal assessment of capacity is required, often with the assistance of Mental Health Services. Rarely this results in an application being made to the family court under The Protection of Personal and Property Rights Act 1988 (PPPR Act).

Management of MND

Multidisciplinary Care:

Care by an interdisciplinary team (IDT) with early intervention for respiratory dysfunction with NIV and nutritional support are key in the treatment of MND. People with MND followed through a multidisciplinary clinic have better outcomes than those not followed in a multidisciplinary clinic, including survival, fewer hospital admissions, increased use of adaptive equipment and enhanced quality of life (26-30). Similarly, NIV use is associated with better survival and evidence suggests that the effect is greater than that of Riluzole (31,32). Multiple studies have shown that weight loss is an independent factor for poor survival and whilst there is no definite answer to whether feeding tube insertion affects outcomes, available evidence suggests enteral feeding may be associated with improved quality of life and a modest prolongation of survival in selected patients (33-38).

Disease-modifying therapies

Riluzole, a glutamate agonist, is the only medication currently available for use in New Zealand to extend life in MND. Based on the evidence available, Riluzole is effective throughout the disease course and is generally well-tolerated. It may extend survival by three months although this may be longer in certain groups (39). In people who wish to start Riluzole, this should be commenced soon after diagnosis.

The prescribing decision is taken in collaboration with, or following consultation with, physicians who care for people with MND, neurologists and palliative care physicians. Riluzole requires a special application authority and not all people will meet the criteria for prescription.

Baseline blood tests, including full blood count and liver function tests, should be done before prescribing. Because of the risk of hepatitis and neutropenia, a full blood count and liver function tests should be undertaken every month during the first three months of treatment, every three months during the remainder of the first year, and annually thereafter. If the person develops a dry cough or dyspnoea rapidly worsens, consider arranging a chest x-ray to check for interstitial lung disease and liaise with a Respiratory Physician regarding investigation and management. Riluzole is contraindicated in people with acute porphyria and hepatic disease.

Symptom Management

Pharmaceutical Treatment Options

People with MND should be encouraged to talk about the symptoms they have and treatment options discussed in a timely manner. (See page 44 "Managing symptoms"). Medication dosing in people with MND should be individualised. Practitioners should have a low threshold for seeking advice from specialists familiar with MND. Primary care physicians and specialists should perform intermittent medication reviews and consider discontinuing any non-essential medications.

Rongoā

Māori with MND may want to consider accessing traditional therapies and Rongoā and these referrals should be facilitated via local services. Clinicians need to be aware if people are using Rongoā and the implications this may have for any treatment that they are receiving.

More information can be found in "*Mauri mate – A Māori palliative care framework for hospices*" (40).

Complementary therapies

Many people living with MND use complementary therapies, such as aromatherapy, reflexology, massage, relaxation techniques and acupuncture. There is no controlled trial evidence of the benefits of these therapies in MND, but some people report improvements in wellbeing after having such complementary therapies.

It is important that the therapist has some knowledge of MND and neither the person with MND nor the therapist has false expectations that the complementary therapy will offer a cure. The treatment being undertaken should also be safe and affordable.

Unproven treatments

Costly and unproven treatments are sometimes recommended by well-meaning people.

It is important to discuss the likely benefits of expensive therapy compared with, for example, changes to the home, employment of additional home assistance, or the peace of mind of the person who wishes to leave their whānau well provided for.

- Discuss the potential risks and benefits of unapproved therapies.
- ALS Untangled (www.alsuntangled.com) is an international group of scientists and clinicians who investigate unproven or alternative treatments and conclude with their own recommendations (41).
- "I've Got Nothing to Lose by Trying It" is a useful and thorough guide, which explains how to weigh up claims of cures and interventions (42).

Genetic counselling and testing

Based on international evidence it is recommended that all people with MND be involved in discussions regarding genetic testing, ideally with the support of a genetic counsellor (43-46).

All people with MND should be invited to participate in the University of Auckland MND Genetics study by registering with The New Zealand MND Registry. The Registry connects people living with MND to researchers, enabling New Zealanders to participate in research and helping MND research grow in New Zealand. For more information, email the Registry Curator: MNDRegistry@adhb.govt.nz Phone: 0800 MND REG (0800 663 734).

Prognosis

ALS is a diverse disease, with survival varying greatly from several months to more than 10 years (6). Factors associated with a poorer prognosis if present at diagnosis include (1):

- Older age
- Bulbar presentation (speech and/or swallowing difficulties)
- Weight loss
- Reduced respiratory function
- Shorter time from onset of symptoms to diagnosis
- Evidence of executive dysfunction or dementia
- Lower Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS) or ALSFRS-Revised Score (Appendix D).

Although individual life expectancy varies greatly, most people with ALS are informed that average survival is 3 to 5 years from disease onset. The ENCALS survival model, uses eight predictors (age, El Escorial classification, site of onset, vital capacity, genetic status for C9orf72 expansion, diagnostic delay, cognitive status and functional score) to generate individual survival probabilities based on a composite endpoint, defined as death, tracheostomy, or dependency on non-invasive ventilation for at least 23 hours a day (47). The prediction model is only available to physicians and allows estimation of a more personalised prognosis. It is only suitable for patients with ALS, not for patients with progressive spinal muscular atrophy (PSMA) or primary lateral sclerosis (PLS). The authors have subsequently published a communication guide to use with the tool (48). The ability to predict survival of individual people is still limited but prediction models can be useful in the provision of realistic information for people, facilitate individual care planning and "help patients and their caregivers regain control over the future" (49).

It is important to note that while most people will want to talk about the future, for many, discussing a future event, like an anniversary or birthday, is more important than statistics. This can be much more tangible and practical than a list of probabilities and might result in planning that reflects both hope and realism. VitalTalks is a US-based resource aimed at providing clinicians with verbal tools to assist them with difficult conversations and includes a module on discussing prognosis entitled "Offer Prognostic Information on How to balance hope and realism"(50).

Information and Support

There are no specific best practice guidelines for communicating a diagnosis of MND, but useful strategies have been developed for delivering bad news, such as the SPIKES protocol (51) (Appendix E). People with MND need the diagnosis delivered in a sensitive and culturally responsive manner, with consideration given to their communication ability, cognitive status, and health literacy. This needs to be combined with information about the disease and available support services. Wherever possible prompt the person with MND to bring a support person with them.

The neurologist/physician should have sound knowledge and experience and be prepared to answer questions around the following (1):

- Types and possible causes of MND
- Symptoms of MND
- Options for treatment
- How MND may progress (including cognitive and behavioural changes) and how progression may affect clinical management (see 'Prognosis' above)
- Accessing help in different situations e.g., after hours care, loss of usual carers
- Research opportunities and how to access them (see New Zealand MND Registry)
- Advocacy and Advance Care Planning
- Needs of whānau, and/or carers

Likely questions and concerns of people with MND and whānau, and/or carers

"What is MND?" "Will my children get MND?"	Types and possible causes of MND
"What will happen to me?"	Symptoms of MND & how it may progress
"Is there a treatment?"	Treatment options- Riluzole & IDT Care
"Can it be cured?" "What research is happening?"	Research Opportunities and how to access them (New Zealand MND Registry)
"How will this affect my daily life?" "How long will I live?"	Prognosis (see ENCALS Tool)
"What happens next?"	Health care services-who will be involved, where and when
"What do I do if…"" Who do I call when…?"	Likely needs & care coordinator contact, preventing crises e.g., acute admission to hospital, loss of usual care providers
'How will I die?" "How do I tell my family?" "How can my whānau be included in my care team?"	Local support services available e.g., hospice/palliative care, MND New Zealand
"What about my job?"	Legal rights & social care support e.g., financial aid, carer support, duty of disclosure e.g., driver's license

Good practice tips:

- Establish an appropriate setting, to ensure privacy and avoid interruptions, and allow sufficient time (45-60min) for explanations and questions. An interpreter (not family member) may be needed to support both a person with MND and their family in their own language.
- Tailor discussions to the person's needs, considering their communication ability, cognitive status, and cultural needs. For Māori, whānau play a key role in the health journey and including whānau or a kaitiaki support person in all discussions is particularly important when breaking bad news (52). In the context of neurological disease, it is important to be aware that the brain is tapu and dementia is considered a "spiritual journey ". Consider seeking advice from a local Māori health advisor or cultural support team if appropriate.
- In some Asian cultures, it can be considered cruel to disclose a diagnosis to people with MND, & this can create ethical dilemmas (53).
- Determine how much information the person already knows and wishes to receive, and whether they want whānau members and/or carers involved.
- Stage the provision of information and deliver the diagnosis in a stepwise fashion (many neurologists deliver the diagnosis over two consultations). Avoid information overload- staging the information gives people time to reflect and prepare questions for the next meeting.
- Consider whether some information should be given by other members of the IDT where the content is particularly specialised e.g., NIV, enteral nutrition.
- Provide information in both oral and written formats and, if the person with MND agrees, share the information with whānau (and carers if appropriate). MND New Zealand has information on their website and information packs available.
- Communicate the suspected or confirmed diagnosis with the person's GP without delay.

Ensure the client has:

- A referral to the local care coordinator to arrange appropriate assessments including social services e.g., NASC/ aged care.
- A single point of contact for the specialist MND interdisciplinary team and information about what to do if problems occur out of hours or in an emergency.
- A follow up appointment within 2-4 weeks with someone from the IDT.
- Information about available support services including MND New Zealand.

Interprofessional collaboration

- An interdisciplinary team (IDT) is established in each region to meet and discuss needs of individual people with MND and whānau on a regular basis. The frequency of assessments will be dictated by the person's needs and rate of progression.
- The following should be assessed, managed, and reviewed by the IDT- respiratory care, nutrition, swallowing, cognition, psychosocial issues, communication, mobility, activities of daily living, symptom management and end-of-life care.
- A single point of contact (a care coordinator) within each region is established to ensure an interdisciplinary coordinated approach to care.
- All newly diagnosed people with MND and whānau should be offered referral to the local MND New Zealand Support Advisor.

The complexity of MND necessitates the involvement of multiple different specialists and services. Interdisciplinary care increases use of adaptive equipment, uptake of beneficial

interventions such as NIV and gastrostomy, reduces the need for hospital admission and improves both quality of life and survival (54-60).

Current models of care in New Zealand

- Most clinicians use an interdisciplinary team approach, but the nature of this varies significantly between different regions.
- The recommendations from NICE (including national access to specialist MND clinics with neurologists experienced in MND) would be particularly challenging to implement in New Zealand due to small cohorts of people distributed geographically over large distances and a limited workforce with expert knowledge.

The NICE guideline does acknowledge that the recommended model of care is not used everywhere due to a variety of issues, including, but not limited to, lack of resources and a limited specialised workforce (1).

Instead, it is proposed that:

Nationally:

- A MND 'clinical network' be developed to support clinicians who have limited clinical experience dealing with people with MND.
- 'Local champions' are identified to act as regional experts within their region.

Regionally:

- An IDT is established in each region to meet and discuss needs of individual people on a regular basis. The composition and nature of the IDT will vary between regions depending on local resources and it may meet face-to-face or virtually, and with or without the person with MND present. There must be a clear means of communication between team members to expedite referrals when a person's needs change.
- A single point of contact for the person with MND and whānau (a care coordinator) within each region is established to ensure an interdisciplinary coordinated approach to care. The care coordinator maintains regular contact with the person with MND and whānau and initiates effective and timely response when needs change, liaising with other team members and services. They are responsible for organising regular case conferences and team meetings and ensuring clear communication within the team & with the person with MND and whānau. Their role bridges the gap between home and hospital providing clinical coordination across primary and secondary health services.
- The role of the care coordinator may be filled by appropriately skilled nursing or allied health professionals (AHP) depending on local resources but should be a separately funded role. The care coordinator will be a part of the clinical team and will work closely with all involved services, complementing the services provided by the MND New Zealand Support Team
- All newly diagnosed people with MND and whānau should be offered referral to the local MND New Zealand Support Team Advisor. The MND New Zealand Support Team helps people with MND navigate the healthcare system and ensure needs are met. They offer information, support, and advice, and can initiate contacts to other community services as required.

Interdisciplinary team care:

- The large number of people and organisations involved can be overwhelming and place additional stress on the person with MND and whānau. Ensure the person with MND is involved in planning their care, and understands who is involved, what they do and why.
- The frequency of assessments will depend on individual symptoms and needs but it is important to ensure that there are arrangements in place to trigger an earlier clinical and/or IDT assessment if there is a significant change in symptoms identified by the person, whānau and/or carers (as appropriate), or healthcare professionals.
- Tailor the interdisciplinary team assessment to the person's needs and readiness for information, considering their communication ability, cognitive status, and cultural needs. Discussions may need to involve key whānau/carers for decision making and support.
- Key decisions reached with the person and whānau and/or carers (as appropriate) must be clearly communicated to all healthcare professionals involved in the person's care.
- All healthcare professionals involved in the person's care should be aware that MND symptoms may change quickly, and that people with MND will need repeated ongoing assessments to determine current needs. These should be addressed promptly. Avoid untimely case closure and give priority to ensuring continuity of care.
- It is beneficial to introduce palliative care as part of the IDT at an early stage, particularly where there are current or anticipated significant or complex needs, for example, psychological or social distress, troublesome or rapidly progressing symptoms and complex future care planning needs.
- For guidance on the use of Riluzole for people with MND, see page 20.

The following areas should be assessed, managed, and reviewed by the interdisciplinary team (1):

Te taha tinana: physical health

Nutrition-Weight, diet, fluid intake, feeding and swallowing

Physical function, including muscle problems (weakness, stiffness, and cramps) mobility, balance and activites of daily living

Respiratory function (including sleep quality and fatigue), respiratory symptoms and non-invasive ventilation (NIV)

Saliva problems (siallorhoea),

Speech and communication

Cough effectiveness

Pain and other symptoms eg constipation.

Te taha hinengaro, Te taha wairua: mental & spiritual health

Cognition and behaviour

Psychological support

Information needs

End of life care needs

Wairua

Feeling diconnected

Out of body

Existential loss of past ways of being

Fear of Unknown

End of life care needs (109)

Te taha whānau: extended family/*whānau* health

Information and support for whānau and/or carers (as appropriate)

Connection to place, going home and how to make it happen etc.

The core IDT should consist of healthcare professionals and other professionals with expertise in MND, or access to expert knowledge, and ideally should include the following:

- Neurologist
- GP
- Respiratory physiologist or a healthcare professional who can assess respiratory function
- Specialist nurse
- Dietitian
- Physiotherapist
- Occupational therapist
- Speech-language therapist
- A healthcare professional with expertise in palliative care (MND palliative care expertise may be provided by the neurologist or nurse in the interdisciplinary team, or by a specialist palliative care professional)
- Social worker
- Māori health liaison
- Community pharmacist

The IDT should have prompt access to the following:

- Clinical psychology
- Social care
- Counselling
- Respiratory ventilation services
- Specialist palliative care
- Gastroenterology
- Orthotics
- Seating and wheelchair services
- Alternative and augmentative communication (AAC) services & assistive technology services (TalkLink)
- Community care providers including Needs Assessment and Service Coordination Services (NASC)

Respiratory Management

Te taha tinana, wairua, Breath

- A person with a new diagnosis of MND should have baseline pulmonary function tests (PFTs) and be referred to the local respiratory service whether they have symptoms or not. Expedited referral to a respiratory specialist is recommended if symptoms develop or pulmonary function declines and/or is abnormal at baseline.
- Early referral to a respiratory specialist before respiratory symptoms or signs develop ensures interventions such as NIV are initiated at a time that most benefits the person.
- Respiratory monitoring with clinical assessment and pulmonary function testing should take place every 3 months depending on rate of progression and the person's preference.
- People with MND and whānau/carers should be informed about the possible symptoms and signs of respiratory impairment and the purpose, nature and timing of pulmonary function tests and intervention options.
- Fear, with shortness of breath, managing the existential nature of being breathless or not having enough breath.

Respiratory muscle weakness can cause shortness of breath, fatigue, poor sleep and impact on quality of life. Ventilatory failure can develop at any stage and occasionally is the presenting feature of MND. Early assessment and referral to a respiratory specialist is therefore recommended along with regular monitoring of respiratory symptoms and function. Respiratory monitoring provides guidance on the need for respiratory support and often dictates the timing of other interventions and ultimately survival of a person with MND. This allows for early discussions around both the benefits and implications of interventions for the person with MND and whānau/carers, who can then plan in a more general sense.

Identification and assessment of respiratory impairment:

Subjective assessment

Effects of respiratory muscle weakness are often subtle and non-specific (see below);

- Early signs may include nocturnal hypoventilation resulting in disturbed sleep, nightmares, and headaches, especially on waking.
- Diaphragm weakness can cause orthopnoea. With disease progression the person with MND may experience daytime symptoms including breathlessness, fatigue, sleepiness, weak cough, and voice. Bulbar muscle weakness contributes to weak cough and upper airway obstruction as well as abnormal respiratory patterns.
- Some people with MND have a failure of central or 'brainstem' respiratory drive and are particularly sensitive to even small doses of opiates and benzodiazepines, further reducing ability to clear secretions.

Symptoms and signs of potential respiratory impairment:

Early Symptoms may include (4):

- Breathlessness lying flat (orthopnoea) due to diaphragmatic weakness.
- Fragmented unrefreshing sleep, morning headaches and fatigue due to nocturnal hypoventilation.

Later, as more severe generalised weakness develops, potentially affecting throat and limb muscles, symptoms may include:

- More marked breathlessness e.g., breathlessness on exertion (dressing, speaking, or eating).
- Quiet voice and fewer words per breath.
- Weakened cough and sneeze increasing risk of chest infections.
- Restless sleep, daytime sleepiness, hallucinations, poor concentration and/or memory, nausea, loss of appetite and rapid weight loss may develop as hypoventilation worsens and blood CO2 increases.

Signs
❖ Weak cough & voice
❖ Weak sniff
❖ Shallow breathing
❖ Increased respiratory rate ❖ Abdominal paradox (inward movement of the abdomen during inspiration) ❖ Use of accessory muscles of respiration

Pulmonary function tests

Pulmonary function tests (PFTs) should be performed by a healthcare professional from the interdisciplinary team who has appropriate competencies (1).

1. Resting oxygen saturation measured by pulse oximetry (SpO2) breathing room air.
2. Then one or both of the following:
    a. Sniff nasal inspiratory pressure (SNIP) and/or maximal inspiratory pressure (MIP)
    b. Forced vital capacity (FVC) or vital capacity (VC)

SNIP and MIP are more sensitive than spirometry and oxygen saturation and are the preferred choice for monitoring respiratory muscle weakness. SNIP closely tracks declining respiratory muscle strength and can still be performed by people with facial muscle weakness or bulbar involvement. Portable monitors are available. Be aware that spirometry and oxygen saturation can be normal or show a restricted pattern despite significant respiratory muscle weakness, so it is important to regularly enquire about respiratory symptoms and signs. A drop in supine FVC is more sensitive than sitting FVC.

If a person is unable to perform PFTs (e.g., those with bulbar involvement, cognitive impairment, lack of access) other indicators of respiratory reserve include SpO2, ability to lie flat, neck muscle weakness and single breath count (61). If in doubt, refer to a specialist centre for further assessment.

Arterial or capillary blood gas (ABG/CBG) analysis may be indicated if:

- SpO2 ≤ 94%
- There are symptoms of respiratory failure

ABG/CBG can be used to confirm the presence of daytime hypoventilation, however it is a sign of advanced disease. If the daytime pCO2 is greater than 6 kPa the person requires urgent referral to a respiratory ventilation service (to be seen within one week). It is important to explain the reasons for and implications of the urgent referral to the person and (if the person agrees) to their whānau.

If the ABG/CBG pCO2 is less than or equal to 6 kPa this does not exclude respiratory muscle weakness or nocturnal hypoventilation. If they have symptoms, they should be referred for prompt respiratory specialist assessment.

Overnight oximetry

Respiratory muscle weakness will result in nocturnal hypoventilation before daytime pCO2 is affected. Consider overnight oximetry with ABG, or referral for such, if the person with MND has symptoms of respiratory muscle weakness and other objective criteria for NIV have not been fulfilled or are unavailable. Polysomnography is not required.

Results of respiratory assessment

If respiratory muscle weakness is suspected based on symptoms and clinical assessment discuss with the person and (if appropriate) their whānau/ carers (1):

- Their respiratory impairment and potential treatment options
- Possible referral to a respiratory ventilation service for further assessment based on discussion with the person, and their wishes
- Review end of life planning and goals of care

NIV for treatment of respiratory impairment in people with MND

Timely NIV has been shown to improve a person's quality of life as well as longevity, increasing survival by up to 4 times more than Riluzole in MND (30,31,62,63). It does however come at the cost of increased monitoring, carer burden and potential loss of independence.

Discussions around NIV are complex and involve consideration of multiple factors. People with MND and their whānau /carers need guidance through the decision-making process by members of the IDT familiar with the topic. Some people may decide not to use assisted ventilation or will not tolerate it and need to be reassured that they will be supported regardless.

Potential benefits of NIV	Potential disadvantages of NIV
Significant symptom relief in selected people, with improved sleep, and decreased breathlessness	Can interfere with eating, personal hygiene, & communication, particularly full-face masks if required during the day.
Increased survival	Results in increased carer burden; carers may need to be always available to reposition the mask. This has implications for those living alone and in residential care.
May be used intermittently and time gradually increased e.g., overnight initially	Withdrawal of NIV can complicate end of life care
	Less effective where there is significant bulbar involvement or cognitive impairment.
	Continued risk of aspiration with reduced secretion control

A trial of NIV should be offered if the person's symptoms and signs and the results of the respiratory function tests indicate that the person is likely to benefit from the treatment (1).

Information & Support

Information provided should include:

- Advantages and disadvantages of NIV (see above). Ensure that the person understands that MND will continue to progress even with ventilatory support, but NIV may improve symptoms associated with respiratory impairment and can be life prolonging.
- the need for regular review
- that NIV can be stopped at any time and that the person can ask for help and advice from the respiratory team if they need it
- other options for breathlessness eg medication, psychological techniques
- the possibility of becoming dependent on non-invasive ventilation and that this can affect the end-of-life experience. Reassure that they will be supported by members of the palliative care team at this time.

Ensure whānau and carers have the opportunity to discuss concerns they may have and assess their ability and willingness to assist in providing NIV and any training needs.

Information about the possible use of NIV may be provided at one or more of the following times (1):

- Soon after MND is first diagnosed
- When monitoring pulmonary function
- When pulmonary function deteriorates
- Upon request by the person

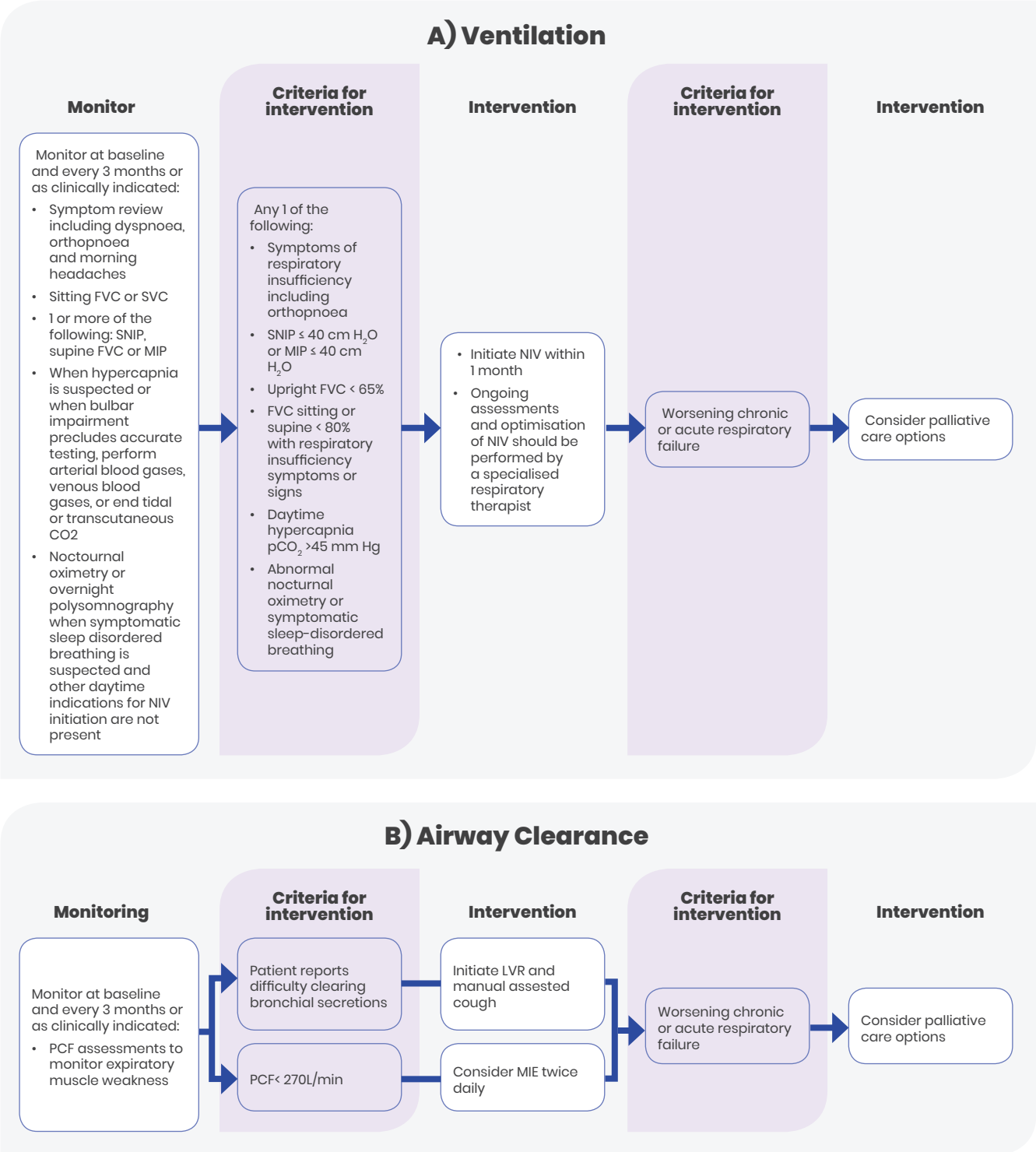


Figure 1: Respiratory decision tree: Summary of recommendations for respiratory management in patients with amyotrophic lateral sclerosis (ALS), including ventilation (A) and airway clearance (B). Note: FVC = forced vital capacity. H2O = water, LVR lung volume recruitment, MIE = mechanical insufflation-exsufflation, MIP = maximal inspiratory pressure, NIV = non-invasive ventilation, PCF = peak cough flow, pCO2 = partial pressure of carbon dioxide, SNIP - sniff nasal inspiratory pressure, SVC = slow vital capacity.

Adapted from Canadian best practice recommendations for the management of amyotrophic lateral sclerosis CMAJ 2020 November 16; E1453-68. Doi: 10.1503/cmaj.191721

Adapted with permission

For those people who have significant cognitive impairment or severe bulbar disease a trial of non-invasive ventilation can be considered if it is thought that they may benefit from an improvement in symptoms related to poor sleep or hypoventilation (1)

People with a diagnosis of frontotemporal dementia

Base decisions about respiratory support for a person with a diagnosis of frontotemporal dementia on considerations specific to their needs and circumstances, such as (1):

- Their capacity to give consent
- Their understanding of the tests and tolerance and/or willingness to undertake them
- Whether they are capable of tolerating NIV
- Whether the person is likely to achieve improvements in sleep-related symptoms and/or behavioural improvements
- The impact on whānau and carers

Discussions about advance care plans (ACP) and EPA should take place early where there is evidence of cognitive decline.

If a trial of NIV is deemed appropriate for the treatment of respiratory insufficiency it should be initiated promptly.

The IDT should consider (1):

- The person's ability to tolerate treatment
- The type of non-invasive ventilator and interface that is most appropriate, based on the individual needs, lifestyle factors and safety
- Regular interface review to address any issues e.g. mask fit, pressure injury
- Need for humidification
- Issues associated with secretion management
- The availability of carers
- How easily the person can get to hospital if necessary
- Risks associated with travelling away from home (especially abroad)
- The risk of ventilator failure and possible consequences. Consider power supply required, including battery back-up, and need for a second machine

When starting NIV, perform initial acclimatisation during the day when the person is awake.

Regular treatment is usually started at night, before and during sleep.

Continue NIV if clinical reviews show symptomatic and/or PFT improvements for a person without severe bulbar and/or cognitive problems. For a person with severe bulbar disease and/or with severe cognitive problems continue NIV if there is an improvement in sleep related symptoms that may be related to respiratory impairment (1). Therapy can be increased gradually depending on symptoms and comfort. Increased survival is associated with average NIV use of >4 hrs/day (64). Be aware that a person's tolerance for, and adherence to, NIV may change overtime and decisions regarding the continuation of NIV may need to be revisited including the option of further trials if desired.

Ongoing management of NIV

Ongoing and regular review of NIV is important to ensure compliance and tolerance, including hours of NIV use, settings, and interface. There is increasing evidence for the benefits of remote monitoring and telehealth for people with MND on NIV which may reduce the need for hospital appointments (65).

People with MND and their whānau should be provided with a comprehensive care plan which covers (1):

- Long-term support provided by the interdisciplinary team including who to contact after hours.
- The frequency of clinical reviews and pulmonary function tests to monitor respiratory impairment.
- Device maintenance and 24-hour emergency support (clinical and technical including contingencies for electric power failure).
- Management of secretions and respiratory physiotherapy assessment, including cough augmentation (if required).
- Training in and support for the use of NIV for the person and their whānau and carers including emergency procedures, equipment failure & how to use the equipment with a wheelchair or other mobility aids if required.
- Services that provide emergency or urgent care including primary health providers should be informed that the person is on NIV. Ambulance services may require notification that NIV is required prior to transfer. Ambulance directives can be included in care plans.

Stopping non-invasive ventilation:

Regular opportunities to discuss the person's wishes in relation to continuing or withdrawing NIV should be offered. At a certain point a person may decide that they want to discontinue assisted ventilation. The process of stopping ventilation should be discussed well in advance with the person with MND and their whānau/family, preferably as part of an ACP, and shared goals of care discussion for hospital admissions (66). Withdrawal of ventilation can be challenging and support from a health professional experienced in withdrawing ventilation is crucial, as deterioration is likely to be rapid. Support from hospice/palliative care and good communication with whānau is also essential in such situations.

Healthcare professionals involved in stopping NIV should have experience and knowledge regarding the management of symptoms of NIV withdrawal and up-to-date knowledge of the law regarding capacity, DNR/ Advance Directive Orders, and Enduring Power of Attorney (EPA).

Invasive ventilation with tracheotomy is controversial & not usually offered in New Zealand.

Management of respiratory impairment without assisted ventilation:

Some people may decide not to use assisted ventilation or will not tolerate it and will require support to manage their respiratory symptoms. People need to be reassured that they will be supported regardless.

- Refer to palliative care for additional strategies and use of medications for dyspnoea.
- ACP/Advanced directive orders - the person's wishes in relation to ventilation for acute events and end-of-life decisions should be clearly documented. This should include instructions on whether oxygen therapy is appropriate or not.

Oxygen therapy needs to be used with great caution due to the possibility of CO2 retention leading to reduced level of consciousness and ultimately, death. This should only be prescribed by a specialist team, with target SpO2 clearly documented.

Airway clearance

An effective cough is essential for the clearance of secretions and prevention of infection. People with MND may have respiratory muscle weakness resulting in reduced chest wall compliance and lung volume, as well as bulbar and glottic muscle dysfunction leading to an impaired cough.

Weak cough can be assessed by measuring peak cough flow (PCF). Ideally this should be performed using an oronasal mask interface rather than a mouthpiece (67). Be aware that it may not be possible for a person with bulbar symptoms/dyspraxia to coordinate a cough.

Cough augmentation techniques can assist with the clearance of mucus from the airways and should be offered to people with MND who cannot cough effectively.

- Recommended breathing exercises and cough augmentation techniques should be initiated when a person with MND reports difficulty clearing airway secretions. These can be taught by a physiotherapist and include lung volume recruitment and manual assisted cough.
- Mechanical insufflation exsufflation (MIE) twice daily should be considered for secretion clearance in people with MND who have reduced peak cough flow (PCF <270 L/min). This can be used more frequently during an acute respiratory infection.
- Use of MIE needs to be individualised. In people with bulbar disease, Lower positive pressures and airflow combined with longer inspiratory times should be considered to minimize laryngeal adduction during insufflation (68).
- Community respiratory support of MIE is required for education, titration, and troubleshooting.

Management of Respiratory Symptoms

Dyspnoea

Shortness of breath is one of the most frightening symptoms of MND and impacts on many other aspects of the disease. Breathlessness can be exacerbated by dysphagia, sialorrhoea, gastroesophageal reflux, weak muscles, general debility, and anxiety. Some people may find neck braces constrictive, exacerbating the problem. Respiratory and palliative care physicians should be involved early in management.

- Optimise non-pharmacological management such as NIV, physical positioning, breathing techniques and use of hand-held fans. These may be used alone or in combination.
- There is good evidence for the use of **morphine** for breathlessness, so this should be used as the first line.

There is no randomised controlled evidence for the use of benzodiazepines in chronic refractory breathlessness (69,70). Intranasal midazolam can be trialled on an individual basis for episodic dyspnoea when associated with significant anxiety. Avoid using for more generalised, pervasive or persistent anxiety as addiction and tolerance to benzodiazepines can occur rapidly. Consider alternative options in patients with a prognosis of greater than 3 months.

Consider prescribing medicines to help ease breathlessness on an 'as needed' basis at home.

Many people have preconceptions and concerns about treatment with opioids. It is important to work with these concerns to reach a therapeutic approach that is acceptable to all people.

Acute shortness of breath is usually triggered by an acute event such as a respiratory tract infection, on top of a background of weakened respiratory muscles. Once reversible causes of worsening respiratory impairment (for example, respiratory tract infections or secretion problems) have been treated, consideration can be given to long-term care, including non-invasive ventilation (NIV) and airway clearance.

Uncontrolled coughing and fear of choking

Choking attacks may occur due to aspiration, impaired respiration, muscle spasm or stridor. Dysphagia and reflux may predispose a person with MND to laryngeal spasm accompanied by stridor. The person may have difficulty breathing or talking, which can be frightening.

Severe, uncontrolled coughing can be distressing for a person with MND & their whānau, and they should be reassured that death caused by choking attacks is very rare. **In the absence of physical obstruction by a food bolus, there is no need to perform back blows or chest thrusts.**

- Consult physiotherapist for airway clearance techniques & SLT for secretion control (including suction equipment)
- Treat any underlying GORD which can exacerbate laryngospasm
- Palliative care can provide advice regarding medications that may assist
- Provide advice to person with MND, & whānau/carers about what to do during a choking attack and when to seek medical advice
    - Assist the person to the most upright position possible
    - Encourage the person to cough and then swallow if they can
    - Use techniques provided by physiotherapist, including cough assist device and/or suction
    - Encourage the person to stay calm and wait for the attack to pass. Focus on slow and steady breathing.
    - If the person has been prescribed medication to help manage choking episodes, use it
    - Open a window to give the feeling of air on the face

Nutritional Management

- Provision of nutritional support is of benefit to people with MND and nutritional status should be documented at diagnosis and monitored regularly by the interdisciplinary team.
- Referral to dietetic services at diagnosis should be considered.
- Referral to SLT for assessment and monitoring of swallowing safety should be made early in the disease process.
- Gastrostomy, including potential benefits and risks should be discussed at an early stage, and as the disease progresses.
- Timing for enteral tube insertion requires consideration of multiple factors including personal preference, nutritional status, respiratory function, and risk of aspiration. Once a decision is made to insert an enteral feeding tube, insertion should be performed without delay.

Weight loss is commonly reported in people with MND with as many as half reporting weight loss >5% at the time of diagnosis and a third with weight loss of more than 10%

(71). Studies have shown that a 5% reduction in usual weight, a 1- point reduction in BMI, or a BMI of less than 24 points, is associated with poorer survival (2). Evidence suggests that people with MND who avoid losing weight during the disease live longer (72,73).

Monitoring of weight and nutritional advice is therefore essential for people with MND and early involvement of a dietitian is recommended. Many will need detailed information about adaptation of their diet and may need food supplements and thickeners prescribed. High-calorie diets can be used to improve nutritional indicators and possibly survival. High-calorie/high-carbohydrate diets may be better than high-calorie/high-fat diets (74,75)

Weight loss and malnutrition can result from:

- Physical difficulties buying, preparing, and eating foods
- Dysphagia
- Fear of choking
- Fatigue associated with prolonged chewing and need for repeated swallowing. This can also result in anxiety and a reluctance to eat with others.
- Loss of muscle mass
- Hypermetabolism
- Cognitive impairment
- Depression
- Loss of appetite
- Constipation
- Respiratory impairment

Effects

- Fatigue
- Reduced physical strength and mobility
- Increased muscle wasting due to muscle catabolism
- Exacerbated respiratory muscle weakness
- Impaired immune function, leading to susceptibility to opportunistic infections
- Susceptibility to pressure injury
- Loss of 'padding' over bony protuberances increasing physical discomfort

Management

i. Assess the person's ability to access food and fluid, taking into account:
   - Reduced oral intake due to limb weakness. Help with food and drink preparation may be required as well as eating and drinking aids and altered utensils to help them take food from the plate to their mouth.
   - Advice and aids for seating, positioning and posture while eating and drinking.
   - Fear of choking and psychological considerations (for example, wanting to eat and drink without assistance in social situations).

ii. Assess the person's nutritional and fluid intake

iii. Enquire about gastrointestinal symptoms, fatigue, appetite, thirst and mood considering need for nutritional supplements

iv. Assess the impact of modified food and fluids on bowel function and palatability

Dysphagia

Difficulty swallowing due to weakness and paralysis of the bulbar muscles will affect two thirds of people with MND during their illness and is associated with increased risk of aspiration.

Weakness of tongue, facial, laryngeal and pharyngeal muscles result in:

- Loss of ability to form lip seal, chew, propel food with the tongue and/or form a bolus.
- Poor or absent swallow reflex.
- Impaired airway protection during swallow.
- Muscle spasm.

This may lead to:

- Sialorrhoea/drooling
- Problems swallowing liquids leading to coughing when drinking (this may be the first sign of dysphagia) and difficulty swallowing medication. A wet, muffled voice may be noticed after eating.
- Aspiration and recurrent chest infections
- Dehydration, malnutrition, and weight loss
- Constipation

Fatigue from difficulty chewing and the need for repeated swallowing can result in lengthy mealtimes and may lead to reluctance to eat out and with others.

Management

Dysphagia requires interdisciplinary and coordinated assessment and care. Deterioration is inevitable, necessitating early referral and regular review.

Swallowing difficulties should be assessed and regularly reviewed by a Speech Language Therapist, who can work with dietitians to provide advice concerning food and fluid consistency, modification of diet and gastrostomy (see below).

The goal of management is to:

1. Maintain nutrition and hydration
2. Reduce choking episodes and risk of aspiration
3. Support quality of life

Airway protection

i. Consult **speech-language therapist (SLT)** to arrange for a clinical swallowing assessment to assess the person's ability to swallow and protect their airway during eating. They may suggest modifying food and drink consistency and palatability (as per the International Dysphagia Diet Standardisation Initiative) and/or suggest prescribing thickeners for fluids. They will perform cranial nerve evaluation and swallow trials of food and fluid. This may also include instrumental evaluation - fibre optic swallow studies (FEES) and Video Fluoroscopy (VFSS).

ii. Consult **physiotherapist and/or occupational therapist (OT)** for advice about head support, seating and positioning. OT can also provide advice in relation to modified eating utensils, cups, plates etc.

iii. Teach safe swallow and airway clearance techniques (**SLT, physiotherapist**). People may also choke on food boluses, so instruct the person with MND and whānau about the correct first aid techniques for choking.

iv. Optimise oral hygiene (see below)

Dependency on others to be fed and for oral care increases respiratory risk. Additional support will be required for people with dysphagia and cognitive change who may need reminders about safe swallowing and require supervision.

Monitoring and review

Consider the ability to manage rehabilitation and compensation for dysphagia and the success of modifications as they change over time.

Gastrostomy

Gastrostomy is recommended for nutritional support in people with MND and significant dysphagia. Gastrostomy should be discussed at an early stage, and as the disease progresses. This gives the person time to make an informed choice.

There is a lack of high-quality research in relation to survival, nutritional outcomes and quality of life associated with gastrostomy. However, available evidence suggests a modest survival advantage as well as increasing/maintaining weight (an independent positive prognostic factor) and improving/maintaining quality of life (76). Early placement of a gastrostomy tube is recommended, even if it isn't used straight away, as there are possible risks with a late gastrostomy (for example, low critical body mass, respiratory complications, risk of dehydration, different methods of insertion, and a higher risk of mortality and procedural complications). Gastrostomy is likely to be most effective at an early stage (5% weight loss). A 2016 study showed that the majority of patients who had lost more than 10% of their premorbid body weight failed to regain weight and even continued to lose weight following gastrostomy (77). Weight loss of more than 10% appears to be associated with significantly shorter survival (78).

Decision-making around enteral nutrition is complex and involves multiple factors being taken into consideration. Gastrostomy should be discussed before there is an urgent need and several conversations are preferable. These discussions should involve the interdisciplinary team and the potential advantages and disadvantages need to be explored with a person with MND and whānau.

In some instances, PEG/ RIG may not be appropriate (e.g., poor prognosis) and some people will not want to have a gastrostomy.

- Consider the person's preferences and issues, such as ability to swallow, weight loss, respiratory function, effort of feeding and drinking, risk of choking and quality of life.
- Some people may need to have a gastrostomy placed ahead of developing significant dysphagia due to respiratory decline, and the risk this poses with delaying tube placement.
- If a person is referred for a gastrostomy, it should take place without unnecessary delay. Liaise with the gastroenterology team regarding planned insertion, as pre-insertion anaesthetic review may be required.

- PEG (percutaneous endoscopic gastrostomy) is the preferred method of gastrostomy when someone has good respiratory function, or PIG/RIG (Per-oral image-guided gastrostomy/Radiologically Inserted Gastrostomy) when there is significant compromise of respiratory function.
- Ensure considerations such as lying flat for the procedure, inability to be understood and secretion management have been discussed prior to admission.
- Be aware that some people will not want to have a gastrostomy. See ACP.

The person with MND & whānau need to be aware that there is an additional level of support needed to live at home with a gastrostomy, such as stomal care and managing feeds

MND Victoria have developed a decision tools guide to aid discussions between people with MND, their health professionals, and family members including gastrostomy https://MNDdecisiontools.com/decision_tools (79). https://mytube.mymnd.org.uk/ is another useful resource for people making decisions about enteral feeding and was developed in the UK by Sheffield MND Care Centre and SITraN, with support from the MND Association.

People with frontotemporal dementia:

It is important to pay particular attention to the nutritional and hydration needs of people with MND who have FTD and who may lack decision-making capacity (1). Before a decision is made on the use of gastrostomy, the neurologist from the interdisciplinary team should assess the following:

- The severity of frontotemporal dementia and cognitive problems.
- The person's capacity to make decisions and give consent.
- Whether the person is likely to accept and tolerate treatment.
- Discuss with the person's enduring power of attorney (EPA) whānau and carers if appropriate and/or refer to any existing ACP.

Managing symptoms

- Pain may occur at any stage of the disease and may have multiple causes. It is important to ask about pain at regular intervals.
- Cramps may cause significant pain and should be managed with non-pharmacological measures and/or anti-spasmodic medication. Fasciculations do not usually require medication.
- Alongside non-pharmacological management consider a trial of anticholinergic medicine as the first-line treatment for sialorrhea. Glycopyrrolate and Hyoscine butyl bromide have less central nervous system penetration and are preferable in people with MND who have cognitive impairment.
- Where available, consider referral to a specialist service for Botulinum toxin, if first-line treatment for sialorrhea is not effective, not tolerated, or contraindicated.

It is important that people and whānau living with MND understand that whilst there is no effective curative treatment for MND, there are effective treatments for many of the symptoms, which can improve quality of life. Medication dosing in people with MND should be individualised. Practitioners should have a low threshold for seeking advice from specialists familiar with MND. Specialist palliative care support can always be sought should symptoms not be adequately managed through standard approaches.

Many symptoms relevant to MND have already been addressed in other chapters. For instance: dysphagia in Nutritional Management and dyspnoea, laryngospasm, and cough in Respiratory Management.

Pain

Pain and discomfort are common, affecting up to 85% of the MND population and can interfere with activities, mood, sleep and relationships, significantly impacting quality of life (80,81). The degeneration of motor neurones does not cause pain directly but the increasing weakness and change in muscle tone can in turn affect mobility and posture contributing to pain. As a result, pain in MND frequently occurs in the limbs, but is also common in the back, neck, shoulders, and buttocks.

People may experience:

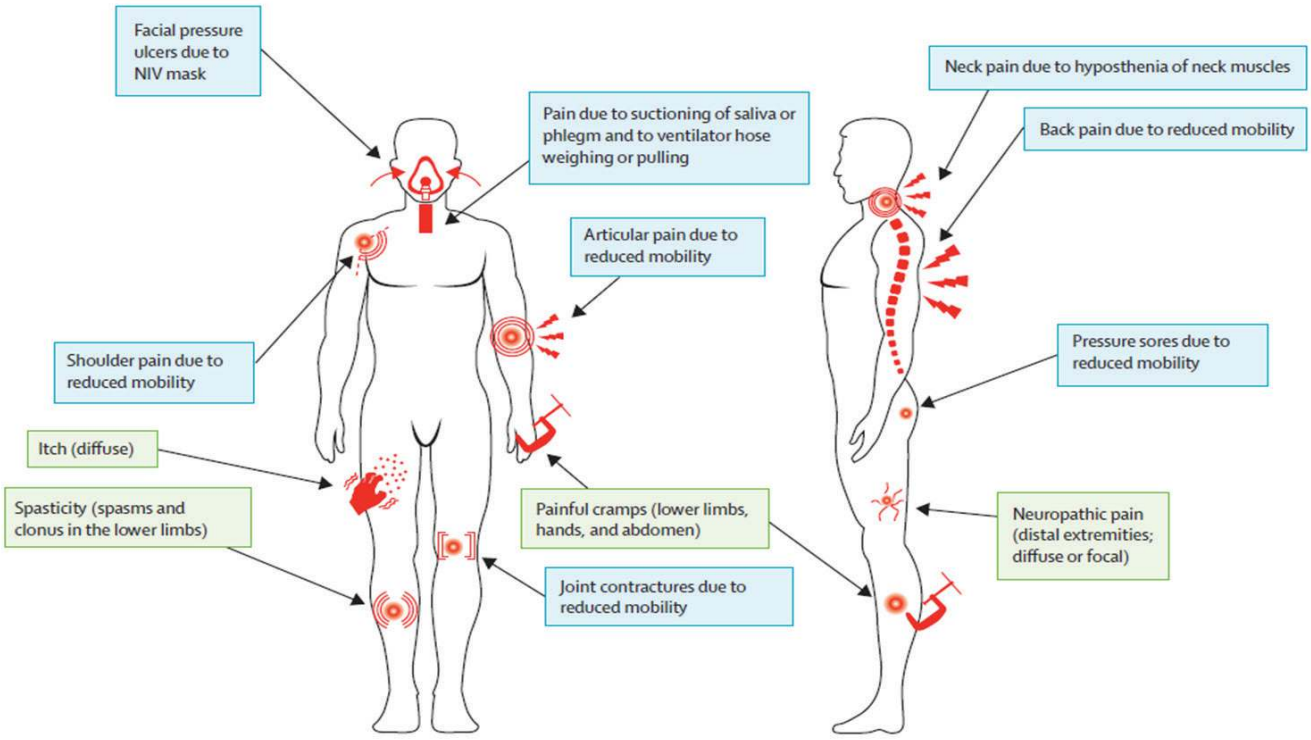
- Cramps
- Spasms
- General aching
- Itching
- Sharp or tender sensations

Causes of pain

Understanding potential underlying causes is essential to management. There may be multiple causes including, but not limited to:

Loss of muscular control necessary to stabilise large joints and maintain a symmetrical spinal posture

- Reduced ability to move resulting in prolonged periods in one position
- Passive injury to joints e.g., shoulder joint damage during assisted transfers
- Muscle cramps, stiffness, and spasticity
- Skin pressure
- Constipation
- Dependent oedema
- Incorrect compensatory equipment in place, such as armchair or mattress not suitable for needs
- Equipment e.g., use of NIV, suction machines



Types of pain in motor neuron disease (77)

Assessment of pain:

Pain can be multidimensional with psychological, emotional, and spiritual components and a holistic approach to management is required. When assessing pain reflect on the Te Whare Tapa Whā model and factors that may impact on a person's experience of pain:

- Te taha tinana: physical health
  Physical factors e.g., certain positions or activities that exacerbate the pain
- Te taha hinengaro: mental health
  Emotional factors such as depression, fear and anxiety
- Te taha wairua: spiritual health
  Spiritual and cultural factors e.g., a perceived loss of mana
- Te taha whānau: extended family/whānau health
- Mana motuhake: autonomy and independence
- Whenua: connection to place and land may assist

Social factors -consider the impact on whānau

It is important to do a complete assessment of the pain including history, type, severity, aggravating/relieving factors. Pain may occur at any stage of the disease. It is important to ask about pain at regular intervals as people may not mention it, choosing instead to focus on more overt muscle symptoms such as weakness (82).

Management of pain

Management of pain requires an interdisciplinary approach, and different treatment options should be discussed. It is important to point out that pain can sometimes be difficult to control and that it may not be possible to get rid of pain completely.

Non-pharmacological treatment

24hr positioning, manual handling & equipment:

Early physiotherapist and OT advice and assessment is essential to identify necessary aids, positioning and transfer techniques, and most appropriate equipment. Good postural care is important for swallowing, respiratory function, reducing fatigue, reducing contracture, skin integrity, general discomfort.

- Education for whānau and carers about optimal positioning and safe manual handling can avoid unnecessary asymmetrical postures and pressure on joints.
- Equipment needs to be reviewed regularly to ensure it is still appropriate. It should promote energy conservation for the individual and their whānau and carers, maximising comfort, safety, and functional ability.
- A physiotherapist can also advise on a suitable exercise programme for people with MND to:
  - Maintain joint range of movement (important for personal cares)
  - Improve mobility by providing suitable aids and advice
  - Prevent contractures
  - Reduce stiffness and discomfort

Other non-pharmacological modalities:

- Superficial heating or cooling, e.g., wheat packs or other means of providing warmth. Sources of heat should be applied safely with appropriate care and wrapping, to avoid risk of burns when people are less able to manage care themselves.
- Gentle massage (mirimiri) can also be utilised for musculoskeletal discomfort.
- Relaxation techniques-guided imagery and distraction techniques may also be useful
- Connecting with whenua and whanau
- Reflection and spirituality including karakia.

These strategies can be very effective in relieving pain and providing comfort as well as empowering the person with MND and whānau to deliver pain relief in a safe manner.

Non-pharmacologic approaches can be used very effectively in conjunction with a pharmacologic approach to pain management.

Pharmacological treatment

- Simple analgesics may be effective, commencing with paracetamol. Adding a NSAID or COX 2 inhibitor may be helpful.
- Neuropathic pain may respond to analgesics such as gabapentin or pregabalin (83). These agents need to be titrated cautiously to prevent adverse effects in frail people.
- Opioids (morphine, oxycodone, fentanyl) may be helpful, but can have unwanted effects and require careful titration as individual requirements vary widely. Due to the complex and individual nature of MND, and risks of respiratory compromise, initiation and titration of opiates should only be done by a physician confident with prescribing opiates in this group of people. Otherwise, consultation with a palliative care physician is recommended. Tramadol is not recommended in older people due to its side effect profile. Prescribing a laxative either regularly or as required as constipation can be very disabling for these people.
- It is important to consider individual needs and preferences when prescribing. There may be problems with swallowing or cognitive issues impacting medication compliance. Where oral dosing is not possible, some slow-release opioid preparations can be given via the gastrostomy tube. Other analgesic agents may also come as liquid preparations to be administered by RIG/PEG. Pharmacists can give advice on formulations and routes of medication in the absence of an oral route. Transdermal Fentanyl patches can be utilised where there is no gastronomy tube but should not be prescribed in opioid naïve individuals. Transdermal opioids have also been shown to have variable efficacy in cachectic people (84). Seek specialist palliative care advice when there are questions around opioid choice and dosing.
- Shoulder pain may be disabling for MND people, due to poor support of the joint or mispositioning. In people with minimal shoulder movement, suprascapular nerve block with long-acting local anaesthetic can be considered (83).

Muscle symptoms

The most common muscle symptoms are fasciculations, (uncontrollable muscle twitching), muscle stiffness, cramps, and weakness.

- **Fasciculations ("twitching")**
  Fasciculations are often among the first symptoms of MND and may be noticed by other people. This is part of the disease process and usually eases over time but may be worse after exertion or during periods of stress. In people who have difficulty sleeping secondary to fasciculations, gabapentin or pregabalin can be considered (2).
- **Cramps**
  These can be bothersome and may be disabling in some people, often earlier in the disease course. Non-pharmacological treatment may include relaxation techniques, massage, hydrotherapy and/or passive exercise programmes in consultation with the physiotherapist. Passive stretching before bedtime can lessen nocturnal cramps. Cramps that are very frequent or disturb sleep may require medication (see below).

Pharmacological treatment

Muscle relaxants, such as baclofen, can be helpful in treating cramp and spasm, as well as painful hypertonia. The response to treatment needs to be monitored, as loss of muscle tone may decrease mobility/ transfers. Coordination with a physiotherapist is helpful to determine the relative role of medication and regular physiotherapy (e.g., passive stretching) in management of cramp and spasm.

- Baclofen can be useful for cramps. Prescribe at night initially. Use smaller doses in frail people. Warn about dry mouth and constipation.
- If baclofen is ineffective, not tolerated, or contraindicated, try gabapentin.
- Consider benzodiazepines (e.g., clonazepam) or mexiletine, as a 3rd line treatment
- NB Mexiletine needs monitoring of ECG before and one month after reaching stable dose due to potential changes to QT interval.
- People seldom tolerate more than small doses of benzodiazepines during the day because of drowsiness and these may be more helpful at night. Observe for respiratory depression associated with sedatives in people with respiratory muscle weakness.

Quinine Sulphate is no longer recommended or available due to concerns regarding side effects (e.g., thrombocytopenia) (83). Some people use tonic water, but 500ml of tonic water contains enough quinine to cause thrombocytopenia in rare susceptible persons.

Mineral and vitamin supplements are unlikely to be beneficial for most people.

There is limited evidence for magnesium outside the setting of pregnancy or magnesium deficiency; however, it may be reasonable to do a limited trial of oral magnesium or lower extremity magnesium baths (Epsom salts) as some people can experience benefit. People should be aware that these can have a laxative effect.

Spasticity, jaw spasm

- Consider baclofen, gabapentin, or benzodiazepines e.g., diazepam, clonazepam, to treat muscle stiffness, spasticity, or increased tone in people with MND.
- If these treatments are not effective, not tolerated, or contraindicated, consider referral to a specialist service for the treatment of severe spasticity.
- SLT can suggest techniques for jaw spasm (e.g., bite box for stretch, education for person and for whānau/ carers) and possibly referral to dental or maxillofacial services.

Cannabinoids

Although cannabinoids in various forms are used by some people with MND, few systematic investigations have been reported. Cannabinoids can be used medicinally as single compounds (THC or CBD) or in various combination ratios

of the two compounds (CBD/THC), or as whole plant extracts. There is limited evidence to suggest that cannabinoids may have a positive effect on spasticity in MND (85). Comparative studies with other oral anti-spasticity medications, such as baclofen, are needed to assess the effectiveness of THC/CBD in relieving MND-related symptoms.

SativexTM is currently the only medicinal cannabis product 'approved' for distribution under the Medicines Act 1981 in New Zealand and is not funded. It is a THC/CBD combination product delivered by nasal spray. In New Zealand, Sativex is indicated as an adjunct treatment for moderate to severe spasticity associated with multiple sclerosis in people who have not responded adequately to other anti-spasticity medication. Cannabidiol only preparations are also commercially available and are listed in the New Zealand Formulary. Similar concerns about the limited evidence apply to these preparations.

Given the above, these products have limited role in the treatment of spasticity in MND at this stage due to lack of evidence, unclear side effects and the cost of current approved medication options.

Muscle weakness and reduced mobility

Early assessment by physiotherapist and occupational therapy allows support required for daily activities to be identified maintaining independence, comfort, safety, and quality of life.

Consider referral to:

- Physiotherapy, for provision of walking aids, orthotics, splints, active and passive exercise, posture management, relaxation techniques & respiratory care. In early disease, regular moderate-intensity exercise is probably beneficial for function and quality of life (86-89). Aerobic, strengthening and range of motion or stretching exercises all appear to be safe and well tolerated in persons with MND, with no evidence to suggest disease or symptom aggravation. An exercise programme may be a resistance, active-assisted or passive programme appropriate to the person's needs, function, abilities, and preferences. Education for whānau and carers about manual handling techniques to minimise injury risk is important.
- Orthotics, for items such as ankle-foot supports, wrist splints and neck support
- Occupational therapy, for 24-hour posture management and equipment for managing activities of daily living. This may include beds, arm neck and trunk support, commodes, riser-recliner chairs, manual and power wheelchairs.

Saliva problems

Many people with MND experience dysphagia and difficulties with saliva control (90). Drooling and feelings of choking may occur, which can cause distress and embarrassment. An interdisciplinary team approach involving the person with MND and whānau will provide the best solutions which will need to adapt as the disease progresses.

Everyone with MND will have different dominant symptoms. Some may have thin, watery saliva leading to drooling, or thick, tenacious saliva or phlegm which is difficult to clear (91). Some people may experience all these difficulties at different times. Additionally, it is important to identify and treat gastroesophageal reflux which is common in MND & may confuse the clinical picture.

Thin, watery saliva:

Excessive saliva (sialorrhoea) is common and people with MND may find they experience pooling of large amounts of thin watery saliva, which can cause drooling. Sialorrhoea is the result of impaired swallowing and poor lip seal, rather than excessive saliva (90).

Thick, tenacious saliva, mucus, and phlegm:

Thick mucus can build up in the mouth and at the back of the throat due to:

- Dehydration
- Evaporation of saliva due to mouth breathing or open mouth posture
- Non-invasive ventilation (NIV) drying out airways

Thickened mucus is difficult to swallow. Additionally, phlegm in the airways may be difficult to cough up due to weakened respiratory muscles and an ineffective cough (91). Stringy mucus can cause airways to become partially blocked, which is very distressing.

Consultation with a speech language therapist is essential.

Assessment

- Assess the volume and viscosity of the saliva- is it watery or tenacious?
- Is it causing drooling or choking?
- Assess the person's respiratory function, swallowing, diet, posture and oral care

Tools to assess the impact of saliva problems, such as the clinical saliva score for MND (CSS-MND) are being developed (92).

Management

Saliva problems can be difficult to control, and different options will need to be explored to find the right solution.

A collaborative team approach involving SLT, OT and physio is important to provide advice:

- SLT for swallowing, diet, and oral care
- OT, physiotherapist for posture, positioning, including head and neck support, and cough management

Strategies and equipment that may help thin, watery saliva (93)

- Encourage a well-supported head position with slightly reclined chair, a collar or chin support
- If the person is able to sleep on their side supported by pillows (postural and respiratory needs should be considered), this will reduce saliva collecting in their throat.
- Careful suctioning if saliva builds up in the mouth (be aware that overuse of suction machines can cause inadvertent salivary gland stimulation)
- Avoid skin irritation- "Promote dab do not wipe" (which can cause inadvertent salivary gland stimulation) and use a barrier cream to protect the surrounding skin
- Adapted clothing which has a waterproof insert will prevent skin irritation and help protect clothes
- Becoming nil by mouth will decrease saliva flow

Pharmacological management:

Anticholinergic medicine is the first-line treatment for sialorrhea-this will reduce the volume and thicken saliva (1). Be aware that anticholinergics can cause confusion in the elderly.

- Tailor dose for each person with MND and monitor closely for side-effects.
- As needed, dosing may be more effective than regular dosing, which may cause tenacious secretions and excessive dryness, and the drying effect can be lost over time.
- If one anticholinergic medication is ineffective, switching to another anticholinergic medication may be considered.
    - Tricyclic antidepressants: nortriptyline or amitriptyline at night, starting at lowest doses.
    - Hyoscine hydrobromide transdermal patch (NB: central nervous side effects)
    - Hyoscine butylbromide, orally or via PEG/RIG. This has fewer central side-effects.
    - Atropine eye drops can be used as a third-line agent for sialorrhoea, but specialist advice should be sought prior to using, as this is an unlicensed indication with potential adverse side effects.

Sialorrhoea medication options in advanced disease towards end of life:

- Glycopyrrolate - can be given via the subcutaneous route.
- Hyoscine butylbromide - can be given by the subcutaneous route.

Both of these medications have less central nervous system penetration and thus are preferable in people with MND who have cognitive impairment.

- Botulinum toxin

If first-line treatment for sialorrhea is not effective, not tolerated, or contraindicated, consider referral to a specialist service for Botulinum toxin, if available (1).

The effect of Botulinum toxin peaks about ten days and wears off over a few months (90). Sialorrhea medication may need to be adjusted accordingly. It is usual to need repeated treatments and changes in dose.

An alternative to Botox is radiotherapy, but the effect is permanent and risks excessive oral dryness. It is rarely used.

Strategies and equipment that may help thick, tenacious saliva, mucus, and phlegm (93)

Provide advice on swallowing, diet, oral care, suctioning, posture & positioning

- Review all current medicines, especially any treatments for sialorrhea.
- Try increasing fluid intake/rehydration. Cold temperatures may stimulate the swallow and jelly, frozen mousse or ice lollies, if safe to swallow, may be helpful.
- Avoidance of mouth breathing, if possible. This may not be an option for people with bulbar symptoms.
- Consider mucolytic agents: papaya enzyme, papaya lozenges; juices and ice cubes – grape, apple, pineapple, and papaya. Pineapple and papaya juices contain proteolytic enzymes, (bromelain and papain), which help break down protein in mucus. The juices can also be part of oral care, applied gently on a sponge. Bromelain and papain enzymes are also available as tablets.
- Sucking on sugarless lozenges can reduce the viscosity of saliva by increasing saliva flow. Ensure this will not cause the person with MND to choke. Avoid lozenges containing menthol as these can have a drying effect. It may be helpful to reduce intake of dairy products where the person isn't reliant on dairy products to maintain their weight.
- Inhaling water vapour or humidification can decrease the viscosity of mucus and help to loosen secretions.
- Teach assisted cough technique and cough assist machine use (CAM). A suction unit can help to remove saliva, mucus, or food particles in the mouth.

Medication for thick, tenacious mucus

Pharmacotherapy with mucolytics, nebulised saline or nebulised ipratropium can be considered (2). There is limited evidence that β-receptor antagonists (e.g., metoprolol or propranolol) can reduce secretions (94).

Dry mouth

- May be related to mouth breathing (e.g., during sleep), oral and pharyngeal thrush, a coated tongue, insufficient fluids and medications (use of anticholinergics).)
- Review medication and consider increasing fluid intake.
- Soft collars or jaw slings (sold as anti-snoring devices) may help to keep the mouth closed.
- Check oral hygiene. Oral lubricants, such as dentamed, oralube, grapeseed oil and biotene moisturising gels (caution with sprays) can help to relieve a dry mouth.

Oral hygiene

Vigilant oral care is important for people with MND and swallowing difficulties. It is important to assess the person's oral care and their ability to maintain these themselves. If they are unable to, provide advice on the following:

- Inspect mouth daily for ulcers or oral thrush. Brush teeth carefully after meals.
- Low froth toothpastes, and toothbrushes with in-built suction or 2 sided bristles can be helpful. Consider forward leaning to avoid toothpaste aspiration.
- Mouth swabbing with half teaspoon of bicarbonate dissolved in a glass of water to help keep mouth clean, or swabs impregnated with the same can be used (be aware of choking on froth).

Dental hygiene appointments may be required for management of gingivitis. Consider referral to specialist dental services in main centres. Patients with a suppressed or overactive gag reflex or active bite reflex, difficulty lying flat, and/or sialorrhoea may require specialist treatment.

Constipation

Sphincter muscles are not normally affected by MND. Changes in bowel function are usually the result of:

- Forced inactivity
- Reduced peristalsis
- Low fluid intake
- Reduced fibre intake
- Weakness of glottic, pelvic floor and abdominal muscles
- Use of analgesics and anticholinergics

Treatment:

- Maintain or increase hydration as required (orally or via PEG/RIG), allowing for seasonal variation in fluid requirements
- Assess and increase fibre intake as required-consider dietitian consultation. Remedies such as bulking agents and fruits with a high sorbitol content (e.g., prunes) are readily available.
- Use of softeners and stimulants may help. Oral laxative options (titrate up cautiously): Docusate sodium + sennoside B,(combined faecal softener and stimulant) Docusate sodium(softener). Bisacodyl(stimulant). Macrogols no longer require a special authority.
- Liquid versions are available that can be given via a feeding tube.
- Use of suppositories, enemas or manual evacuation may be necessary on occasions. E.g., Microlax enemas, glycerine suppositories, bisacodyl suppositories. These interventions may require a district nursing referral to administer.
- Diarrhoea may be due to constipation with overflow.

Bladder problems

The bladder is not usually directly affected by MND; however, some people may experience changes to bladder control, resulting in urinary frequency, urgency and/or incontinence. This is more common in people with predominantly upper motor neurone signs. Increasing fatigue and reduced mobility may also impact ability to maintain continence.

Consider:

- Solifenacin, oxybutynin for urgency & frequency. Be aware that these may cause or exacerbate cognitive impairment in the elderly. Hyoscine butylbromide can also be used and does not cross the blood brain barrier thus it may be safer when there are concerns about cognition or delirium. However, it can exacerbate constipation and dry mouth like all anticholinergic agents.
- Ural sachets can be helpful for bladder spasm.
- A 'uridome' or an indwelling catheter is occasionally appropriate.

Oedema

- Dependent oedema due to restricted activity is common.
- Compression stockings, elevation and light massage may be beneficial.
- Diuretics are rarely appropriate.
- In some areas, referral to the lymphoedema service may be an option.

Skin sensitivity

Good skin and pressure care are vital. Someone with MND may be aware when they need to turn or move but may need help to adjust their position. A need for frequent repositioning at night can have a major impact on sleep for the patient and whānau. Nutrition and continence can also affect skin integrity.

- Early involvement of an OT is recommended. Consider equipment for skin sensitivity relief, such as a bed cradle to relieve the weight of bed clothes, in bed turning aids, lightweight bed clothing and a pressure-relieving mattress and cushions. Equipment does not remove the need for repositioning.
- Refer to a specialist wound care nurse if the issue does not resolve quickly.

Emotional lability (pseudobulbar affect)

Emotional lability is associated with upper motor neuron involvement and presents as disturbed emotional responses, including inappropriate and uncontrollable laughter and crying. In some cases, this can be extreme. It may or may not be associated with more widespread cognitive decline and may also be a manifestation of depression (with or without anxiety). It can be very disabling for people and difficult for those around them, limiting where they go and what they do.

- Treatment is not usually required. The person and whānau should be reassured that it is a symptom of ALS and does not necessarily represent a symptom of depression or impaired cognition. Some people find understanding the symptom helps them to manage the impact.
- Although frequently used, there is insufficient evidence for the use of Amitriptyline or SSRIs but they may be considered if symptoms are distressing to the person, particularly where there are concomitant symptoms such as low mood or altered sleep (2) . Anticholinergics may produce unacceptable side effects including confusion in the elderly.
- Dextromethorphan combined with quinidine (DM/Q; Nuedexta™) may be useful, but access is limited in New Zealand (95).

Equipment and Adaptations

- Most people with MND will require a range of adaptive and mobility equipment to enable them to remain at home for as long as possible and participate in everyday activities. The need for aids and equipment can change rapidly.
- Clinicians involved with assessing needs for equipment should have a good knowledge of the different presentations of MND, as this will assist with decision-making around choices and timing of intervention.
- Equipment and adaptations need to be provided without delay so early referral and on-going review is essential. Anticipating potential problems can prevent delays accessing equipment.

The role of health and social care practitioners, including Physiotherapists and Occupational Therapists, is to compensate for changes in function to enable people to participate in the activities of everyday life; such as providing adaptive equipment, environment modification & exploring alternate ways to complete activities.

Occupational Therapists and Physiotherapists supporting people with MND should:

- Work with health care professionals in different settings to ensure integration of systems (e.g., wheelchairs and assistive technology) and ensure regular reviews
- Coordinate visits with other health care professionals where appropriate
- Facilitate referrals to other services as required

The following should be assessed and advised on (1):

- Activities of daily living, including personal care, dressing, and bathing, housework, shopping, food preparation, eating and drinking, and ability to continue with current work and usual activities.
- Mobility and minimising risk of falls
- Issues arising from upper limb weakness
- Posture and positioning
- Respiratory care and cough management
- Awareness of safety, including safe use of equipment & understanding need for equipment
- The home environment and modifications/adaptations if required
- The need for assistive technology, such as environmental control systems
- Fatigue and conservation of energy

It is important to focus not only on activities essential for physical well-being but consider other aspects of well-being (Te Whare Tapa Whā model). Explore what activities matter to the person with MND and whānau (these may be the same or different) and how you can support them to maintain occupational participation and quality of life.

Management

Occupational Therapists and Physiotherapists need to be familiar with the eligibility criteria and referral process for equipment as well as funding limitations. This is important in terms of "future-proofing" as needs can change rapidly and there may be restrictions on access e.g. a person will only get one main wheelchair seating system, so a good manual wheelchair is important before a powered wheelchair. Current criteria limit use to "in and around home" which can reduce quality of life.

- Equipment and adaptations need to be provided without delay. Early referral is essential. This will require knowledge of the variants of MND and associated prognosis requiring individual variation. Discussing potential problems early can prevent delays identifying needs and accessing equipment.
- Ensure that equipment, adaptations, daily living aids, assistive technology and wheelchairs are provided to meet the changing needs of the person and their whānau and carers (as appropriate) to allow participation in activities of daily living and maximise mobility (1)
- Liaise with other health providers to ensure that all equipment provided can be integrated, for example, integrating augmentative and alternative communication (AAC) aids and devices and environmental control systems with wheelchairs.
- Enable prompt access and assessment for funding for home modifications. It is important to work with the medical team and whānau in relation to what home modifications are available/desirable/acceptable & ensure maximum gain from funding. If the person does not meet criteria for funding, continue to offer information and support in adapting home environments.

Equipment

The Ministry of Health's Equipment and Modification Services provide free or subsidised equipment and modifications to people with disabilities.

Equipment and Modification Services (EMS) cover:

- Equipment
- Housing modifications
- Vehicle purchase and modifications

Currently, District Health Boards are responsible for providing equipment for short-term loan use (i.e., for at least up to 12 weeks). Needs Assessment and Service Coordination services (NASC) may be able to provide additional home support until the equipment and/or modifications can be put in place for a person's long-term use.

NASC works with EMS assessors and providers to determine needs and allocate Ministry-funded disability support services and equipment. The Ministry contracts two providers, Accessible and Enable New Zealand, to administer and provide equipment and modification services. Accessible administers EMS in the Auckland and Northland regions (from Meremere north); Enable New Zealand administers EMS in the rest of the country. The TalkLink Trust has a contract with the Ministry of Health to provide assessments for Communication Assistive Technology, to enable disabled people to communicate more independently and effectively.

Wheelchairs

Refer people to wheelchair services promptly. (1). Wheelchair needs should be determined by an accredited clinical assessor and a manual and/or powered wheelchair that meets the person's needs should be provided without delay.

Mobility vehicles

Currently there is no vehicle funding available unless the person is working. Consider alternate funding e.g., lotteries commission.

Ongoing support

- Regularly review the person's mobility and needs. It is important to continue to monitor their ability to use equipment and to adapt equipment as necessary.

Communication

Mana motuhake, wairua, hinengaro

- Difficulty with communication has a significant psychological impact. Providing access to different modes of communication, including social media, can promote independence, participation, and better quality of life.
- Assessments by Speech Language Therapists and referrals for assistive devices should be made early in the disease course.
- Communication devices may also benefit caregivers and whānau.

Dysarthria is common in MND and 80% people are eventually affected (96). Impairment of speech is due to weakness and/or spasticity of the lips, facial muscles, tongue, larynx and pharynx. Reduced palatal elevation can give the voice a 'nasal' quality and respiratory muscle weakness will impact on speech volume and projection. All the above lead to progressive difficulty with articulation, slurred speech, hoarseness, and/or loss of volume. Eventually anarthria may result. Non-verbal communication may also be affected, with loss of facial expression and body language through physical gestures. Upper limb weakness can impact on access to remote communication, such as handwriting, keyboard and smartphone use (email, internet, social media).

Difficulty with communication has a significant psychological impact, potentially leading to:

- Isolation – communication inadequate or avoided
- Frustration – difficult or impossible to be understood - need time, which may not be available; difficulty using communication aids
- Increased fear and anxiety as unable to discuss these concerns
- Low self-esteem – others make assumptions - deaf, intellectually impaired, or intoxicated
- Loss of control – because misunderstood or opinion ignored or not sought
- Increased sadness – isolation and frustration felt by person with MND and whānau.

Management

The goal of management is to optimise all methods of communication for as long as possible. Knowledge of the variants of MND and associated prognosis is helpful to anticipate potential problems before they become an issue, and plan accordingly (people with bulbar onset have earlier involvement of speech than people with spinal onset).

Early referral to SLT is important:

- For advice on strategies for communication
- To arrange for assessment and provision of communication aids (TalkLink Trust)
- For consideration of voice banking, which should be done early in the disease process as it does require time and energy, and reasonably clear speech for best results

The TalkLink Trust (Assistive Communication Solutions) provides specialised assessment and equipment to people with disabilities throughout New Zealand. There are offices in Auckland, Wellington, and Christchurch, with clinicians regularly traveling to locations around New Zealand, as well as providing support via Telepractice.

Augmentative and Alternative Communication (AAC)

Should.

- Be provided without delay
- Maintain quality of life and maximise participation in daily activities
- Come with adequate support and training for the person with MND, and their person with MND and whānau and/or carers
- Be integrated with other assistive technologies, such as environmental control systems and personal computers or tablets.
- Involve other healthcare professionals, such as OTs, physios, and other support team members as required.

Communication can be aided by various methods:

Low tech

- Writing
- Alphabet board
- Communication books/boards, for conveying a range of communication functions quickly, e.g., to meet needs and feelings
- Perspex eye pointing frame (E-TRAN frame) – letter and phrase based

Mid and high tech

- Telephone and Smart-phone modifications, including hands-free, speech-to-text, stylus to assist text messaging
- Call-bells, including switch adapted call-bells
- Personal alarms
- Voice amplifiers
- Options to support voice and message banking
- Portable speech generating devices to augment and/or replace speech. Includes:
    - Static display devices using recorded speech
    - Dynamic display devices such as iPads, tablets
- Computers to support writing

Computers and speech generating devices support a range of access options

- Touch screen technology
- Speech-to-text
- On-screen keyboard, modified keyboards
- Alternative mouse options such as trackballs, head-mouse
- Eye-gaze
- Switch scanning

The SLT works as part of a team with the OT and Physio who can assist with advice on:

- Seating, positioning, wrist supports, mobile arm supports
- Switches, pointing devices
- Mounting options to access communication devices on wheelchairs, tables, and from bed
- Environmental controls
- Care alerts/call devices

Cognitive impairment can cause problems with communication and the ability to learn and use alternative communication methods.

Useful strategies to aid communication:

- Take time and create a quiet relaxed atmosphere.
- Ascertain individual's own preferred communication strategy and/or equipment used.
- Position – face to face, watch lips, eyes, gestures.
- Clarify the yes/no strategy e.g., eye-blink, head nod/shake, vocalisation, pointing to yes/no cards
- Ask closed questions for "yes" "no" answers; use signals for yes/no.
- Encourage to slow down and over emphasise words.
- Be wary of interruptions or trying to finish sentences.
- Try not to use whānau or carers to translate for the person with MND unless this is what the person wants.

Ongoing support

It is important to have regular, ongoing monitoring of the person's communication needs as MND progresses and review their ability to use AAC equipment. Ensure low tech options are in place as this may be the only functional option at the final stage of the disease progression.

Psychological & Social Support

Te taha hinengaro, whānau

- People with MND and whānau may experience considerable psychological and emotional distress during their journey with MND. The early introduction of palliative care as part of the IDT is beneficial and can assist with difficult conversations and decision-making.
- Regularly discuss psychological & social care needs and provide information about sources of psychosocial and social support. Early referral to the MND New Zealand support team is recommended.
- Caring for someone with MND can be physically and emotionally challenging. Health care providers need to be alert to the physical and emotional well-being of the caregivers.

MND is characterised by a series of losses, associated with grief and bereavement, and the need to continually adapt to change. Feelings of anxiety, depression and isolation are common (97-99). The psychological experience of MND will be influenced by age, gender, stage of life, cultural and educational background, spiritual and religious beliefs, and psychosocial circumstances (100,101). The psychological support provided needs to adapt through the disease course and regular assessment of psychological needs is important. People will access different resources to cope with this distress and the availability of resources will vary. Psychological wellbeing of both the person with MND and whānau impacts hugely on quality of life, and some people may require professional support (1).

Emotional reactions

Research has identified several main themes that impact the person with MND & whānau (102):

1. **Coping with the diagnosis:**
   This may include understanding the diagnosis, reactions to the diagnosis, and support after diagnosis. Before the diagnosis anxiety and low mood may be triggered by worrying symptoms, difficulty in identifying the cause, a protracted period of investigation & the need to be sure before giving the diagnosis.
2. **Understanding the disease:**
   Seeking and processing information and filtering sources of information
3. **Acceptance:**
   Accepting the disease, finding ways to cope, gain control, and have a sense of purpose or meaning
4. **Coping with a changing life:**
   Adjusting to a 'new normal, finding hope, living in the present and looking to the future.
5. **Changes in relationships:**
   Coping with changes in identity and role, intimacy, and the importance of touch.

Fears may include:

- Loss of independence and dignity
- Increasing dependency and becoming a burden
- Choking or 'fighting for breath'
- Inability to move and/or communicate
- Inability to cope and loss of control
- The unknown, death and the process of dying

Denial is a coping mechanism that sometimes operates alongside awareness of the condition and its implications. If denial persists however it can contribute to higher levels of anxiety and depression.

Anger can occur at any stage of the illness. Anger may be displaced onto other members of the whānau.

Sadness may be linked to the recognition that many of life's hopes and expectations will never be realised.

All the above are normal responses to grief. Allowing time and opportunities to acknowledge and discuss these concerns and fears may help alleviate some of these. Carers may experience a similar range of reactions but at different times from the person with MND & these also need addressing.

The importance of sleep

Sleep disruption may be the result of several factors including pain, cramps, spasm, breathing difficulties and choking, and can have a negative impact on quality of life (103,104). Where poor sleep is the result of anxiety and depression treatment can often result in quite rapid improvements in sleep quality. Addressing all issues associated with poor sleep can improve both physical & psychological wellbeing.

Management

Identifying needs

During interdisciplinary team assessments and other appointments, discuss psychological, & social care needs. Topics may include the following: -

- Their understanding of the condition and how it can impact daily life
- coping with diagnosis and prognosis
- concerns around loss & change and fears about dying. This may include spiritual needs or concerns (te taha wairua).
- Their ability to continue with employment and other activities.
- Financial changes which may impact on person with MND and family
- Adjusting to changes in their life and their perception of self
- Changes in the dynamics of the whānau-relationships and roles
- Sexuality and intimacy
- Decision-making and forward planning
- Conversations that count - including EPA and advanced care planning

Provide space for them, their whānau and carers to express thoughts and feelings openly and without judgement.

Providing information

- Respect individuals' desire for information; be aware that information needs may vary during the disease and between individuals.
- Give sufficient information to allow the person with MND to understand what is happening without overwhelming them. Determine what the person with MND has understood and retained of the information provided.
- Give written information and opportunities to return for more information and ask questions.
- Give information that can help them maintain control and make plans.
- Check preferences for involving whānau/carers.
- Be aware that the type, quantity and timing of information and support required may be different for each individual and it may be necessary to revisit some conversations e.g., ACP.
- If relevant, discuss how they might tell whānau, particularly children, about the diagnosis and prognosis.
- Offer the person information about sources of psychosocial and psychological support including the support groups and online forums.

Providing support

- Identify someone to provide ongoing support e.g., MND New Zealand Support Team member, GP, care coordinator
- If needed, refer the person to counselling/psychology/social work services for specialist assessment and support.
- Some people may want spirituality as part of their care plan. It is important that the IDT are aware of the person's spiritual values and how they can accommodate them
- Refer to a social care practitioner/specialist nurse with knowledge of MND or rapidly progressive complex disabilities to discuss the person's needs and preferences for social care.
- Offer information as appropriate to the person with MND assessing their readiness to receive information. Respect the person's wishes about what information they wish to receive.
- Support them to access:
  - Personal care, ensuring there is continuity of care with familiar workers, so that wherever possible, personal care and support is carried out by workers known to the person and their whānau and/or carers (as appropriate). Needs assessment is required to access funded personal and home help.
  - Equipment and practical support
  - Financial support, for example, application for benefits and grants through Work and Income New Zealand and access to carer support through the Ministry of Health. Clients may be eligible for a Community Service Card and a high-user card for medication.
  - Advance care planning
  - Support to engage in work, social activities, and hobbies, such as access to social media and physical access to activities outside their home.
  - Respite care

Te Taha whānau

The psychological and emotional impact on whānau, who may also have roles as carers, is significant and also needs to be addressed. Caring for someone with MND can be physically and emotionally challenging and often leads to feelings of isolation. It is important that whānau/carers have their own time to talk about the impact MND has on them.

Addressing the needs of whānau/carers

Provide advice and information on:

- services available to help them including adequate carer and nursing support and access to specialist care. Discuss what type and frequency of support they would prefer. There may be phases of the illness where this needs to be reviewed, particularly when significant changes happen
- Practical help available: household tasks, personal care, equipment, financial support
- The illness and training in skills to enhance comfort for the person with MND
- Respite care as an inpatient or a home sitting service
- Emotional support directed specifically at the carer including counselling and social work services. Consider referral for additional psychological support. Children and young people may benefit from a referral to a specialist service
- support groups and online forums. whānau / carers may benefit from being in contact with other carers of people with MND.

MND New Zealand is a charitable organisation that supports people living with motor neurone disease, their whānau/carers, and health professionals. A number of information booklets are available from MND New Zealand and there is a library of books. videos and other resources. The website has answers to some of the questions frequently asked by whānau, friends and carers, and has links to more medically detailed sites. National newsletters are published regularly.

The MND New Zealand Support Team provides free, personalised support and advocacy for people with motor neurone disease. There is more information about the service they provide and how to contact the MND New Zealand Support Team on the website.

Anxiety and depression

Appropriate responses to the difficulties faced by people living with MND need to be distinguished from persistent and disabling symptoms of anxiety and depression. Feelings of hopelessness are the most common marker of psychological morbidity in MND & are likely to arise soon after diagnosis. Antidepressants and/or anxiolytics (caution required with benzodiazepines) may be beneficial for some people. They may take several weeks to have an effect. Consider referral for additional support such as those offered through psychology, social work, psychiatry, or spiritual care.

Apathy

This has been reported in some people living with MND. It may be linked to anxiety and depression, or to cognitive change, and may be a barrier to accessing help and support. Apathy can be particularly challenging for caregivers and whānau.

Emotional lability (pseudobulbar affect)

see pg.38

Support as the disease progresses

As MND develops, its impact will be influenced not only by how quickly it progresses, but also the area of the body involved. Adapting to the changes in physical function can be particularly challenging for people with more rapid progression as there is less time available for psychological adjustment.

- As MND progresses, people may have difficulty communicating. The psychosocial effects of communication problems and not being able to share thoughts and feelings are significant. Consider having conversations with an emotional/psychological content from the outset.
- Communication problems can lead to difficulty accessing support and services. Ensure people have a designated contact if possible and different options for contacting support or services
- Ensure people with MND and their whānau understand the likelihood and implications of respiratory problems. This enables everyone to explore the options and likely outcome, thus avoiding uninformed last-minute decisions, e.g., prolongation of unacceptable quality of life

Impact on professionals

MND creates many challenges for professionals and can arouse strong emotions. These can include frustration, powerlessness, inadequacy, and sadness. It highlights attitudes to issues related to disability, quality of life and measures taken to prolong life. Good interdisciplinary teamwork is necessary to provide support and opportunities to discuss concerns and responses to difficult situations. Support, in the form of regular clinical supervision, is recommended.

Planning for end of life

- It is beneficial to introduce palliative care as part of the IDT at an early stage. This allows for supportive care that adapts to a person's needs from diagnosis through to end-of-life care.
- Members of the IDT should be prepared to discuss and/or facilitate discussion about end-of-life concerns with the person with MND and whānau at any point in the course of the disease.
- Conversations about advance care planning help establish care preferences before the disease is advanced and communication is impaired. Ongoing discussions about ACP and goals of care should be part of routine MND follow-up.

The role of palliative care

The prognosis for individuals diagnosed with MND is variable, but overall statistics show that the majority will die within 2-3 years. There is increasing recognition of the importance of palliative care involvement, not just at the end of life, but throughout the course of the illness. It is important to emphasise to the person with MND and whānau that this centres around the provision of optimal supportive care rather than an expectation that end of life is near. The need for co-ordinated interdisciplinary care, that involves difficult conversations and decision-making, lends itself well to a palliative approach with the person's needs and quality of life as the focus.

Discussing end of life issues

People with MND will have questions and concerns about disease progression and dying but may need encouragement to discuss their fears openly both with whānau and with the health care team. Discussing end of life issues can be difficult, but members of the IDT should be prepared to discuss and/or facilitate discussion about end-of-life concerns with the person with MND and whānau at any point in the course of the disease. Palliative care involvement can assist with initiating these discussions. The Health Quality And Safety Commission New Zealand (HQSC) have developed the Serious Illness Conversation Guide Aotearoa to help clinicians without a palliative care background, to have conversations about what is most important to seriously ill people and their whānau if functional abilities change and/or time is limited (105).

When discussing end of life issues:

- Tailor discussions to the person's needs, considering their communication ability, cognitive status, spiritual and cultural needs. and health literacy. Discussions may need to involve key whānau for decision making and support.
- Be sensitive about the timing. Trigger points, such as following diagnosis, intervention with enteral nutrition/NIV or declining respiratory function, can be useful prompts to discuss any concerns about end-of-life care and to review any existing ACPs/Advance Directives, DNACPR orders and the appointment of an EPA for health and welfare. (see below).

- Consider discussing ACP with people at an earlier opportunity if you expect a deterioration in their communication ability, cognitive status, or mental capacity. The appointment of an EPA for health and welfare early is particularly important where there is cognitive decline. This is only enacted if a person loses the capacity to make decisions for themselves.
- Introduce community palliative services (including hospice) early, if people agree. This allows hospice teams to get to know people and whānau and build trust and rapport. They can also be working with/alongside the IDT during the person's journey, avoiding abrupt introductions should clinical issues deteriorate rapidly. Hospice teams can offer much in the early stages of disease in terms of psychosocial care such as counselling, life reviews, and support of children and whānau.
- Be prepared to answer questions about the following:

What could happen at the end of life*	**'How will I die?'***
Symptom control	'Will I be in pain?" "What is happening to my body?" "How will this impact on my mind?"
Preferred place of death, involving specialist palliative care /hospice; providing anticipatory medicines in the home.	'Can I stay at home? Who will look after me?'
Advance care planning including: ❖ What they want/do not want e.g., resuscitation, antibiotics, artificial feeding/ventilation ❖ options for treatment (including telehealth where available) ❖ what should happen in the event of an intercurrent illness ❖ who will represent their decisions, if necessary	'How will my health team know what I want? Do I have to go to the hospital? Can I just go to the local hospital?" "How can I ensure my whānau are involved in discussions around my care if I want them to be ?"
Withdrawal of treatment- What 'is and isn't' euthanasia (see below)	"Do I have to keep using PEG/NIV? "Is withdrawal of feeding or NIV euthanasia?"
Suicide and assisted dying ❖ what is the legal position around assisted dying? ❖ what does capacity to choose this mean?	"Can I choose assisted dying?" "Can others choose this for me?"

People with MND may fear that death will result from choking when in fact this is highly unlikely. In most cases, a person with the condition will die in their sleep due to respiratory failure and/or infection.

Advance care planning

Provide support and advice on:

- ACP/Advance Directives, DNACPR orders, and appointment of an EPA. The Health Quality And Safety Commission (HQSC) has multiple resources available to assist with advance care planning. MND Australia are also in the process of developing ACP & EOL decision-making tools which should be available soon: https://mnddecisiontools.com/decision_tools (79)
- Preferred place of death and how this will be managed. Te Ara Whakapiri (see below) is useful for people with MND and whānau who wish to know about how end of life can be managed at home (106). Rural people with MND may wish to clearly define how far they are willing to travel if complications arise.
- Te ipu aronui https://www.teipuaronui.co.nz/ For Maori End of life care, stories, health professional advice and resources.

- Ensuring ACPs are available when needed, utilising local systems. For example, the Taranaki region has a novel approach whereby they keep these documents in the fridge and all the paramedics know to look there. ACP documents can also be uploaded to GP/secondary care electronic management systems.
- Advanced directives/ACP should be reviewed regularly as choices and preferences around end-of-life care may change over the course of the illness.

It should be noted that no matter how sensitively these topics are broached, some people may never allow conversations about future planning. Continuing to encourage these people to engage can risk alienating them. This does not however preclude involvement of Palliative Care and support of whānau, which can be provided according to a person's goals and wishes.

Respite Care

Respite plays a crucial part in the care of people with MND and whānau. It can relieve carer stress and people's concerns about being burdensome. Respite also helps to facilitate people remaining at home where most people want to be. There is a range of respite options available in each region.

- Provide information and support about respite options to people with MND and whānau. Additional social or nursing care may be required, to enable informal carers to reduce their carer responsibilities and spend time with the person with MND.

Assisted Dying - End of Life Choice Act (2019)

Following a referendum in 2020, the End of Life Choice Act 2019 came into force on 7th November 2021. The Assisted Dying Service allows a person with a terminal illness, who meets the eligibility criteria, to legally request medication to relieve their suffering by ending their life.

The Ministry of Health will be responsible for the Act and has developed a work programme to implement it. This includes developing policy and preparing guidance for health providers. More information, including resources for health professionals can be found here.

People with MND understandably have fears about what will happen as the disease progresses. They may worry about becoming a burden to others or losing independence. These concerns can give rise to questions about assisted dying or euthanasia. Requests for assisted dying should be acknowledged with respect and people should be given the opportunity to explore any concerns should they wish. Efforts should be made to understand, appropriately address, and if desired, remedy, any reversible issues/difficulties that might be underlying such requests, including mental health support if needed. Ongoing optimal supportive care consistent with the goals of palliative care should continue to be provided no matter what choices the person with MND makes.

The Australian and New Zealand Society of Palliative Medicine (ANZSPM) state that (107):

- "Patients have the right to refuse life-sustaining treatments including the provision of medically assisted nutrition and/or hydration. Refusing such treatment does not constitute euthanasia or physician-assisted suicide. "
- "Good medical practice mandates that the ethical principles of beneficence and non-maleficence should be always followed. The benefits and harms of any treatments (including the provision of medically assisted nutrition and/or hydration) should be considered before instituting such treatments. The benefits and harms of continuing treatments previously commenced should be regularly reviewed. Withholding or withdrawing treatments that are not benefiting the patient, is not euthanasia. "
- "Treatment that is appropriately titrated to relieve symptoms and has a secondary and unintended consequence of death, is not euthanasia."
- "Palliative sedation for the management of refractory symptoms (at end of life), is not euthanasia."

Te Ara Whakapiri: Principles and guidance for the last days of life

As the end-of-life approaches, healthcare providers can utilise Te Ara Whakapiri, a guideline developed in New Zealand that outlines "the essential components and considerations required to promote quality care at the end of life for all adults in New Zealand"(106). It is designed to guide the development of individual plans of care that address physical, mental, social, cultural and spiritual issues in the last few days of life and is a useful resource in the care of people with MND. It reflects New Zealand's unique multicultural society having been informed by local research and is endorsed by several key professional health organisations.

The guiding principles and components for the care of adults in Te Ara Whakapiri cover all settings, including the home, residential care facilities, hospitals, and hospices. The document comes in several different versions with terminology adapted to make it easier for whānau to understand.

Te Ara Whakapiri describes three components to care in the last days of life (106):

1. A comprehensive baseline assessment involves identifying the lead practitioner, assessing clinical needs, sensitive and open communication, and clear documentation.
2. Ongoing assessment emphasises the importance of developing individualised care plans.
3. After death care includes verification of death and the need of the whānau for information and privacy.

The Te Whare Tapa Whā model provides a framework for using these components to provide holistic care and increase the total wellbeing of people in their last days of life and that of their whānau. Te Ara Whakapiri is not a care plan but serves as a foundation document for all policies and procedures concerned with care at the end of life and local circumstances, resources and needs have to be taken into consideration. If a person in the last days of life has a level of need that exceeds the resources of the primary palliative care provider, that provider should refer them to specialist palliative care.

Baseline assessment summary

People with MND who are thought to be in the final phase of life, are fragile and can deteriorate quickly compared to other people with terminal illness. Support may need to be mobilised rapidly, particularly if the person wishes to die at home. Ensure planning around end of life incorporates how this might be managed should it occur outside of normal working hours or on whenua which may be another home. This should include access to GP/hospice/nursing support.

Te taha tinana: physical health

Recognition the person is dying

It is important for the health team to identify that the person is dying as early as possible; this can be difficult in neurological disease. Its onset may be signalled by:

- Increased breathlessness
- Dysphagia, weight loss
- Recurring infection, particularly aspiration pneumonia
- Sepsis
- Cognitive difficulties, reduced level of consciousness
- Reduced mobility, pressure sores

The most common cause of death in MND is respiratory failure following upper respiratory tract infection. The terminal phase may be preceded by reduced chest expansion and associated breath sounds, use of accessory muscles, and morning headache from CO2 retention (type 2 respiratory failure).

Early recognition allows for timely, appropriate care and communication, involving the person (where possible) and their whānau. This enables the clinical team to prioritise the provision of comfort and support based on the person's preferences. A lead practitioner (usually the person's primary health care provider) should be appointed to manage care.

- Where appropriate, health practitioners should initiate a conversation with the person to explain the changing nature of their condition.
- Assessment of physical needs:
  - Review current management and initiate prescribing of anticipatory medication for symptom control (see below)
  - Identify physical care needs which may include skin integrity, bowel cares, mouth cares and pressure relief.
- Ensure there is prompt access to the following, if not already provided:
  - Communication method that meets the person's needs, such as an AAC system or alphabet boards.
  - Equipment, if needed, such as syringe drivers, suction machines, riser–recliner chair, hospital bed.
- Advise relevant agencies of the person's deterioration

Te taha hinengaro: mental health

- Encourage person to talk through their preferences for end-of-life care, both with whānau and with the health care team before the need is urgent or before they find it too difficult or tiring to communicate. This should include any advance directives, cultural needs, and the person's wishes in relation to their preferred place of care and death including the method of disposition after death (burial or cremation).
- It is important to maintain communication with the person with MND even in the final stages. Communication aids may be appropriate as well as the use of closed questions which require limited effort (single word answers, eye-blinking, hand squeeze).

Te taha whānau: extended family/whānau health and wider social systems

Discuss the person's changing condition with whānau. When engaging with Māori whānau, ensure assessment occurs to capture and support whānau preferences or needs in respect to Tikanga Māori (40).

Te taha wairua: spiritual health

Spiritual practice, including religion, may become more important to someone as they approach the end of their life. It is an important component of quality of life and is an essential domain of palliative care. Spiritual beliefs and values may impact peoples' and whānau' decisions regarding end-of-life care, therefore it is essential that the medical team are aware of the person's spiritual values and how they can accommodate them.

- Provide the opportunity for the person and whānau to discuss what is important to them at this time and what you can do to help. Everyone's needs are different.
- Various spiritual history taking tools have been developed, which aim to provide clinicians with both the structure and words necessary to uncover the spiritual needs that people commonly have.eg HOPE (108)
- Some people will need more support and may want to talk to you about their spiritual concerns. Encourage the person to explore their worries and fears. Listen without passing judgement or dismissing their concerns. Try to understand and listen to your person's beliefs, without imposing your own. If you don't feel comfortable having these conversations, ask an experienced colleague or a specialist such as a pastor/spiritual leader to be involved.
- Be aware that some people may not want spirituality as part of their care plan.
- Spiritual care workers are usually included as part of palliative care teams. This may include chaplains, spiritual leaders from the community, Māori spiritual leaders, as well as traditional healers (40). When engaged with Māori people and whānau it is important to offer the support of these care coordinators, including local DHB/ Hospice Kaitiaki support.

Symptom control

The five most commonly recognised actual or potential symptoms during the last days of life are:

i. Pain
ii. Agitation/delirium/terminal restlessness
iii. Respiratory tract secretions
iv. Nausea and vomiting
v. Breathlessness

Te Ara Whakapiri has a toolkit with guidelines and algorithms for managing all the above.

i. Pain

- Can be a significant issue in the last few days e.g., muscle spasm, pressure pain.
- Opioid analgesics may be appropriate. Careful titration and individualisation of doses should be considered, to avoid excessive drowsiness and respiratory depression.
- Regular analgesics should be continued and consideration given to alternative routes of administration such as suppositories or parenteral route if swallowing is a problem. Parenteral medication may be given as a continuous subcutaneous infusion using a syringe pump.
- Physiotherapy and/or massage may also be helpful for musculoskeletal pain.
- Ensure whānau and/or carers are able to give medications.

ii. Agitation/delirium/terminal restlessness

- Usually associated with reducing levels of consciousness and can be related to hypercapnoea. This can present as persistent or intermittent agitation which can be very distressing for both the person affected and the people around them. If no reversible cause can be found (consider pain, positioning, urinary problems (retention, UTI) and bowel care), use of antipsychotics and/or benzodiazepines can be helpful.

iii. Respiratory tract secretions

- May develop if the person is unable to clear oropharyngeal secretions. This can usually be managed with repositioning and if necessary antimuscarinics (hyoscine butylbromide and glycopyrrolate). These medications will also reduce saliva production.

iv. Nausea and vomiting

- Consider antiemetics such as metoclopramide, haloperidol or levomepromazine

v. Breathlessness (dyspnoea) and choking

- Shortness of breath and/or respiratory failure may be due to muscle weakness, aspiration, or infection. It can be very frightening, creating anxiety which exacerbates the problem.
- Choking attacks may occur due to aspiration, respiratory muscle weakness, difficulty swallowing and muscle spasm. People with MND may fear dying from choking when in fact this is highly unlikely. It should be stressed that death is usually peaceful. Opioids for dyspnoea, benzodiazepines for anxiety associated with dyspnoea and antimuscarinics for secretions can reduce episodes of choking. People with MND who are fearful of choking and breathlessness will be reassured by knowledge that anticipatory medications are available.
- Time should be spent with the person and whānau explaining how they can manage these choking episodes using correct positioning and cognitive strategies.
- Oxygen therapy needs to be used with great caution in people with MND due to the possibility of CO2 retention leading to reduced loss of consciousness (LOC) and ultimately death. However, when a person is approaching end of life, low flow oxygen may assist with the distress of dyspnoea in combination with medication.
- People may be dependent on non-invasive ventilation (NIV), typically bilevel positive airway pressure ventilation (BPAP or BiPAP); this requires careful consideration if withdrawal is requested. Support from a health professional experienced in withdrawing ventilation is crucial, as deterioration is likely to be rapid following its removal. Support and good communication with whānau is also essential in such situations.

Ongoing assessment of the person's care

An individualised shared care plan for a person in their last days of life should be developed, which addresses the person's physical, mental, social, cultural, and spiritual needs.

This plan should be written in collaboration with the person and their whānau and reviewed regularly. Te Ara Whakapiri is designed to guide care in the last 72hrs of life, thus the plan of care should be reviewed after 3 days should deterioration be more gradual, or a person's condition stabilise or improve.

Care after death

It is essential that the care of people with MND and their whānau continues after death. As per Te Ara Whakapiri certain elements of these policies should be standard

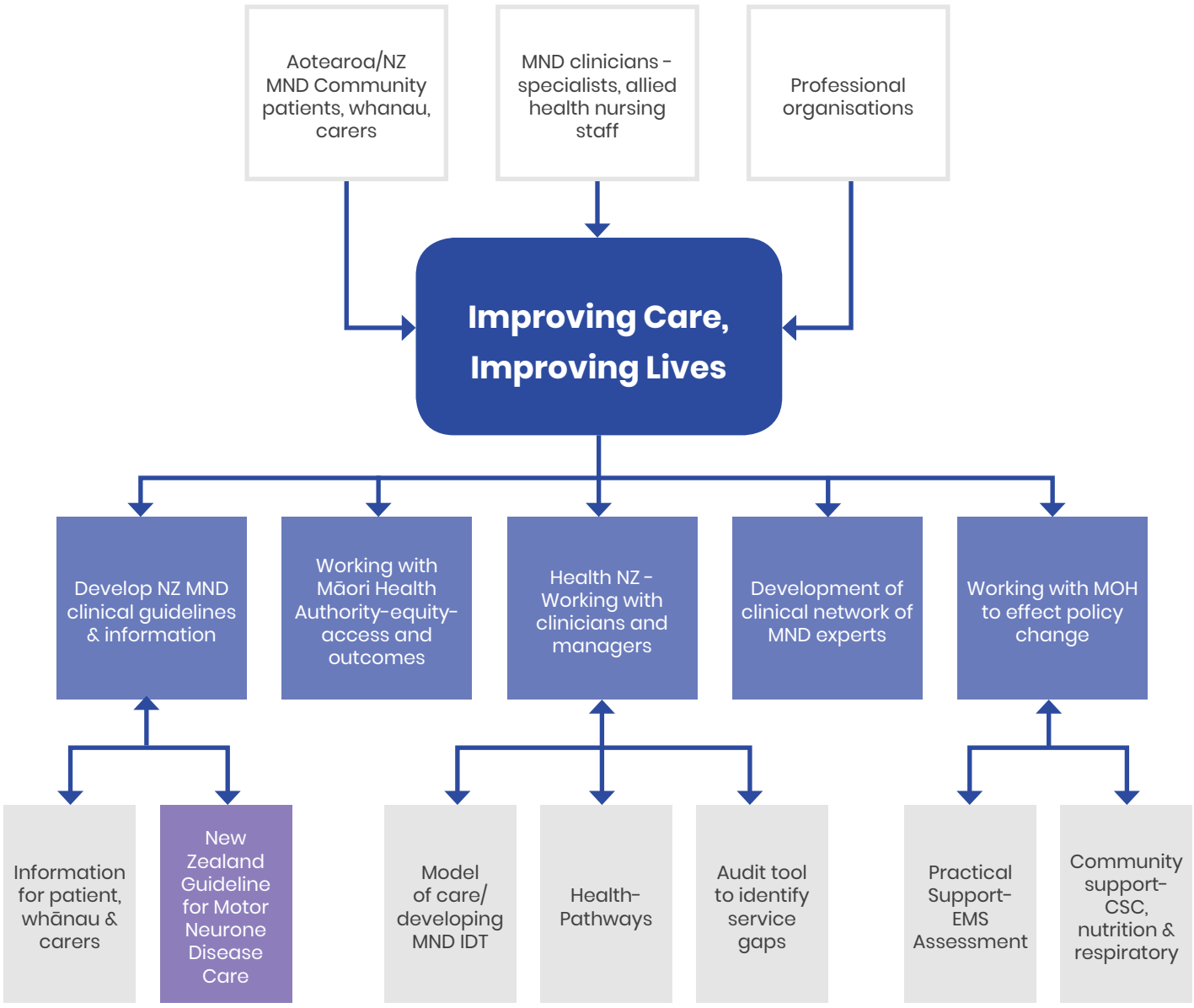
Care after death summary

Te taha tinana: *Physical health*	**Te taha whānau: *Extended family health***
3.1 Verification of death 3.2 Dignity and respect for the person/tūpāpaku	3.3 Provision of information to the family/whānau about what to do next
Te taha hinengaro: *Mental health*	**Te taha wairua: *Spiritual health***
3.4 Assessment of family/whānau bereavement	3.5 Consideration of the spiritual, religious and cultural needs of the family/whānau 3.6 Availability of a private space for the family/whānau

From Te Ara Whakapiri, 2017

Appendix A

Improving care, improving lives framework



Appendix B

Te Tiriti o Waitangi and Te Whare Tapa Whā

Aotearoa New Zealand has an Indigenous population, tangata whenua, and is a multicultural society and health service delivery needs to be provided in a culturally safe way. Special consideration is given to:

- Te Tiriti o Waitangi and acknowledging the principles of protection, partnership, and participation for Māori in relation to the Crown
- Te Whare Tapa Whā, a Māori health framework written by Professor Sir Mason Durie

Te Tiriti o Waitangi

Te Tiriti o Waitangi is the founding document of Aotearoa New Zealand. It is founded on an agreed power sharing partnership between the Crown and whānau, hapū, iwi and kaupapa Māori partners. This is captured in the three Articles of Te Tiriti o Waitangi. The Crown enacts Te Tiriti through five principles:

a. The guarantee of **tino rangatiratanga**, which provides for Māori self-determination and mana motuhake in the design, delivery, and monitoring of primary health care.

b. The principle of **equity**, which requires the Crown to commit to achieving equitable health outcomes for Māori. Existing healthcare disparities may reflect the fact that fewer Māori with MND are accessing health services and therefore fewer are diagnosed.

c. The principle of **active protection**, which requires the Crown to act, to the fullest extent practicable, to achieve equitable health outcomes for Māori. This includes ensuring that it, its agents, and its Treaty partner are well informed on the extent, and nature, of both Māori health outcomes and efforts to achieve Māori health equity.

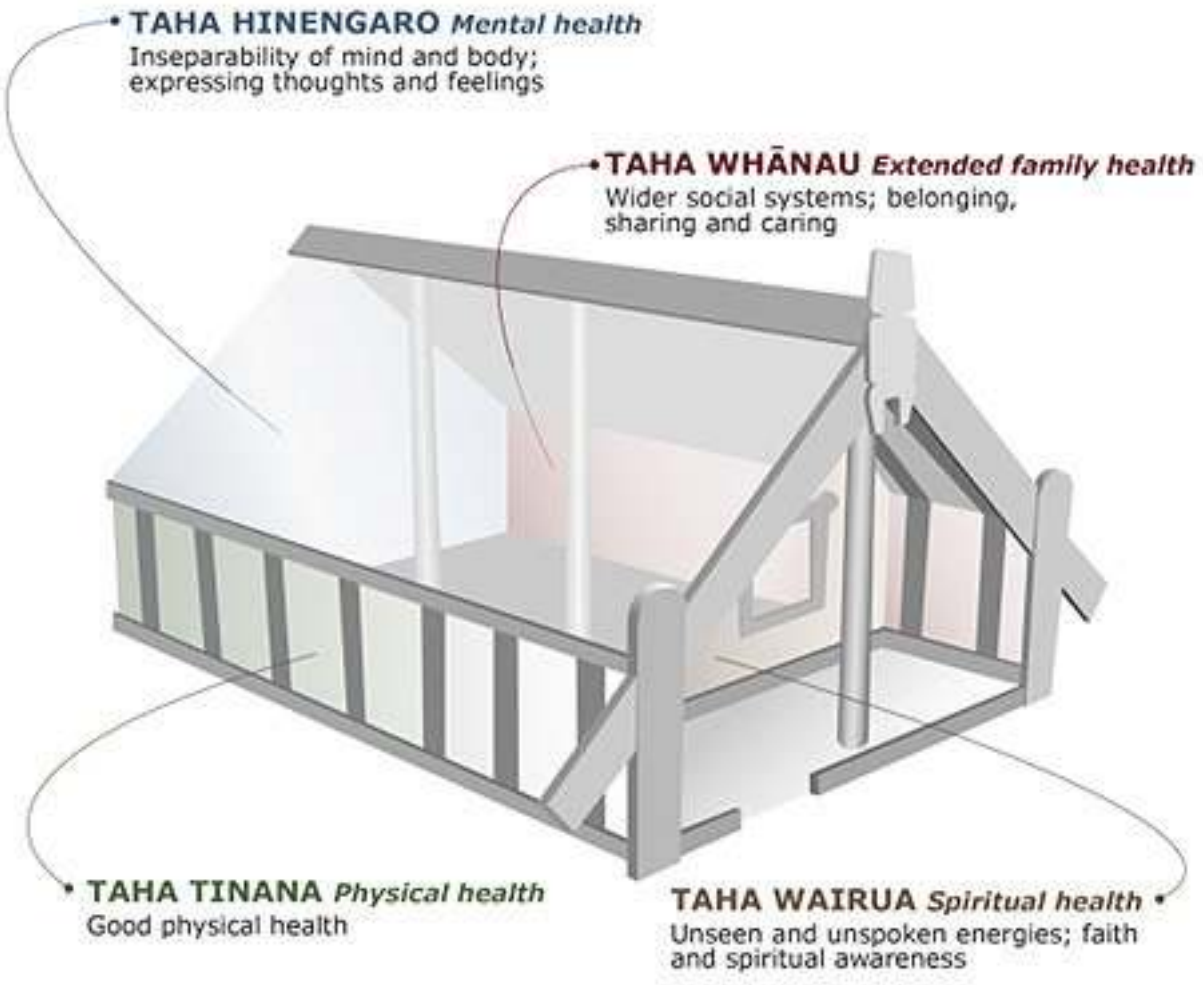
d. The principle of **options**, which requires the Crown to provide for and properly resource kaupapa Māori primary health services. Furthermore, the Crown is obliged to ensure that all [primary health care] services are provided in a culturally appropriate way that recognises and supports the expression of hauora Māori models of care.

e. The principle of **partnership**, which requires the Crown to work in partnership with Māori in the governance, design, delivery, and monitoring of primary health services. Māori must be co-designers, with the Crown, of the [primary health] system for Māori.

Te Whare Tapa Whā

We enact the principles of Te Tiriti o Waitangi using Te Whare Tapa Whā, a model consistent with other frameworks and legislation underpinning quality care, including the Code of Health and Disability Services Consumers' Rights, the Health Practitioners Competence Assurance Act 2003 and competency requirements set out by the Nursing Council of New Zealand and the Medical Council of New Zealand.

Te Whare Tapa Wha is a model of care that is well-suited to the management of people with MND, where patient and whānau centred care which considers all aspects of well-being is important. It is a framework written by Professor Sir Mason Durie that symbolises a holistic view of wellbeing as the four cornerstones of a whare. These walls are taha wairua/spiritual wellbeing, taha hinengaro/mental and emotional wellbeing, taha tinana/physical wellbeing and taha whānau/family and social wellbeing.



Source: Durie M. Whaiora: Māori Health Development. 2nd Ed. 1998. Oxford University Press: Auckland.

Appendix C

Painless, progressive weakness – Could this be MND?

mnda motor neurone disease association

RC GP Royal College of General Practitioners

Painless, progressive weakness – Could this be Motor Neurone Disease?

1. Does the patient have one or more of these symptoms?

Bulbar features

- Dysarthria
  - Slurred or quiet speech often when tired
- Dysphagia
  - Liquids and/or solids
  - Excessive saliva
  - Choking sensation especially when lying flat
- Tongue fasciculations

Limb features

- Focal weakness
- Falls/trips – from foot drop
- Loss of dexterity
- Muscle wasting
- Muscle twitching/ fasciculations
- Cramps
- No sensory features

Respiratory features

- Hard to explain respiratory symptoms
- Shortness of breath on exertion
- Excessive daytime sleepiness
- Fatigue
- Early morning headache
- Orthopnoea

Cognitive features (rare)

- Behavioural change
- Emotional lability (not related to dementia)
- Fronto-temporal dementia

2. Is there progression?

Supporting factors

- Asymmetrical features
- Age – MND can present at any age
- Positive family history of MND or other neurodegenerative disease

Factors NOT supportive of MND diagnosis

- Bladder / bowel involvement
- Prominent sensory symptoms
- Double vision / Ptosis
- Improving symptoms

If yes to 1 and 2 query MND and refer to Neurology

If you think it might be MND please state explicitly in the referral letter.
Common causes of delay are initial referral to ENT or Orthopaedic services.

Additional resources:
MND Association downloads and publications at www.mndassociation.org/gp

Bulbar features

25% of patients present with bulbar symptoms

- Dysarthria
  - Quiet, hoarse or altered speech
  - Slurring of speech often when tired
- Dysphagia – more often liquids first and later solids. Initially can be sensation of catching in throat or choking when drinking quickly.
- Excessive saliva
- Choking sensation when lying flat
- Weak cough – often not noticed by the patient

Painless progressive dysarthria – consider neurological referral rather than ENT.

Limb features

70% of patients present with limb symptoms

- Focal weakness – painless with preserved sensation
- Distal weakness
  - Falls/trips – from foot drop
  - Loss of dexterity eg problems with zips or buttons
- Muscle wasting – hands and shoulders. Typically asymmetrical
- Muscle twitching/fasciculations
- Cramps

Respiratory features

Respiratory problems are often a late feature of MND and an unusual presenting feature. Patients present with features of neuromuscular respiratory failure

- Shortness of breath on exertion
- Excessive daytime sleepiness
- Fatigue
- Early morning headache. Patients often describe a 'muzziness' in the morning, being slow to get going or as if hung over
- Un-refreshing sleep
- Orthopnoea
- Frequent unexplained chest infections
- Weak cough and sniff
- Nocturnal restlessness and/or sweating

Consider MND if investigations for breathlessness do not support a pulmonary or cardiac cause.

Cognitive features

Frank dementia at presentation is rare. Cognitive dysfunction is increasingly recognised, as evidenced by:

- Behavioural change such as apathy or lack of motivation
- Difficulty with complex tasks
- Lack of concentration
- Emotional lability (not related to dementia)

Ask specifically about a family history of these features.

Development group for this resource:
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Appendix D

Revised Amyotrophic Lateral Sclerosis Functional Rating Scale

Table 2: The ALSFRS-R — a functional amyotrophic lateral sclerosis rating scale incorporating assessments of respiratory function[28]

Item	Scoring system	
1. Speech	4	Normal speech process
	3	Detectable speech disturbance
	2	Intelligible with repeating
	1	Speech combined with non-vocal communication
	0	Loss of useful speech
2. Salivation	4	Normal
	3	Slight but definite excess saliva in mouth; may have nighttime drooling
	2	Moderately excessive saliva; may have minimal drooling (during the day)
	1	Marked excess of saliva with some drooling
	0	Marked drooling; requires constant tissue or handkerchief
3. Swallowing	4	Normal eating habits
	3	Early eating problems-occasional choking
	2	Dietary consistency changes
	1	Needs supplement tube feeding
	0	NPO (exclusively parenteral or enteral feeding)
4. Handwriting	4	Normal
	3	Slow or sloppy, all words are legible
	2	Not all words are legible
	1	Able to grip pen, but unable to write
	0	Unable to grip pen
5a. Cutting food and handling utensils*	4	Normal
	3	Somewhat slow and clumsy, but no help needed
	2	Can cut most foods (>50%), although slow and clumsy; some help needed
	1	Food must be cut by someone, but can still feed slowly
	0	Needs to be fed
5b. Cutting food and handling utensils	4	Normal
	3	Clumsy, but able to perform all manipulation independently
	2	Some help needed with closures and fasteners
	1	Provides minimal assistance to caregiver
	0	Unable to perform any aspect of task
6. Dressing and hygiene	4	Normal function
	3	Independent and complete self-care with effort or decreased efficiency
	2	Intermittent assistance or substitute methods
	1	Needs attendant for self-care
	0	Total dependence
7. Turning in bed and adjusting bed clothes	4	Normal function
	3	Somwhat slow and clumsy but no help needed
	2	Can turn alone, or adjust sheets, but with great difficulty
	1	Can initiate, but not turn or adjust sheets alone
	0	Helpless
8. Walking	4	Normal
	3	Early ambulation difficulties
	2	Walks with assistance
	1	Non-ambulatory functional movement
	0	No purposeful leg movement
9. Climbing stairs	4	Normal
	3	Slow
	2	Mild unsteadiness or fatigue
	1	Needs assistance
	0	Cannot do
10. Dyspnea	4	None
	3	Occurs when walking
	2	Occurs with one or more of the following: eating, bathing, dressing
	1	Occurs at rest: considering using mechanical respiratory support
	0	Significant difficulty: considering using mechanical respiratory support
11. Orthopnea	4	None
	3	Some difficulty sleeping at night due to shortness of breath, does not routinely use more than two pillows
	2	Needs extra pillows in order to sleep (more than two)
	1	Can only sleep sitting up
	0	Unable to sleep with mechanical assistance
12. Respiratory insufficiency	4	None
	3	Intermittent use of BiPAP
	2	Continuous use of BiPAP during the night
	1	Continuous use of BiPAP during the day and night
	0	Invasive mechanical ventilation by intubation or tracheotomy

*Patients without G-tube - use 5b if >50% is though G-tube.

**Patients with G-tube - 5b is used if the patient has a G-tube and only if it is the primary method (>50%) of eating.

ADL=activities of daily living;BiPAP=Bilevel Positive Airway Pressure;

G-tube=gastrostomy tube;NPO=nothing by mouth.

Reproduced with permission from Cedarbaum et al., 1999.[28]

Beydoun, Rabeah & Rosenfeld, Jeffrey. (2018). Edaravone in Amyotrophic Lateral Sclerosis Lessons from the Clinical Development Program and the Importance of a Strategic Clinical Trial Design. European Neurological Review. 14. 47-53. 10.17925/USN.2018.14.1.47.

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Appendix E

SPIKES

The SPIKES model was first published in The Oncologist in 2000 as a protocol for delivering bad news to cancer patients.

Since then, it has been adopted more widely and used by clinicians in various circumstances to communicate difficult news to people in a way that is clear, supportive and compassionate.

Set up

- Think about what you want to say in advance.
- You may wish to suggest to the patient that they are accompanied by a friend or relative.
- Choose a time and place which will allow for privacy and quiet, considered discussion. Make sure there is enough seating in the room and turn off or mute any electronic devices so that the patient has your full attention.
- Establish rapport with the patient and maintain eye contact.
- Allow enough time for the patient and/or their relatives to express their emotions and ask questions.

Perception

- Use open-ended questions to determine the patient's understanding of their condition. This will help you tailor the way you deliver the information and where you begin.
- Check that the patient is able and willing to hear what you will say. They may give you an opening to start the discussion, or they may try to avoid hearing what you are saying.

Invitation

- Most people will indicate that they want full information, but some may shun information as a coping mechanism.
- If people do not want to hear details, you can offer to answer any questions in the future or speak to their family or friends.
- Use language appropriate to the patient's level of understanding. It can help to reflect the patient's words and body language.
- Avoid unnecessary jargon and euphemisms, which could impair the patient's comprehension and create a barrier to communication.
- Be sensitive to how the patient is reacting and provide information at an appropriate pace.

Knowledge

- Warning the patient that you have bad news may reduce the shock of disclosure.
- Give the patient and their friend/relative enough time and space to absorb the information and ask questions.
- If there is any doubt about the prognosis, explain this and the options for clarifying uncertainty.
- Give the patient information regarding next steps, such as follow-up appointments. If this is not possible, offer them a realistic timescale of events and reviews. The patient should also be told who will oversee their care and how they can contact them.
- Reassure the patient of ongoing support. This will help them to cope and feel less isolated.
- The patient and relative/friend may differ in how much information they want or require. If you sense a disparity, check that the patient is happy for you to speak to their friend/relative separately.

Empathy

- Don't make assumptions about what the patient might be feeling. Encourage them to express their concerns and respect their wishes about how much information they are prepared to hear.
- Observe and validate the patient's emotions and give them enough time and space to express their reactions.
- Remember that all people are different.

Strategy and summary

- Make sure the patient has understood by asking them to briefly summarise the main points of the conversation. Encourage them to express their concerns.
- Provide reading material for the patient to absorb when they are ready.
- Suggest that the patient note down any questions they'd like to ask you at your next meeting, so you can be sure you are appropriately exploring their understanding.

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