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MND Scotland is indebted to the following specialists for their help and expertise in the preparation of this booklet.

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<th>Contribution</th>
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## Contents

### A Problem Solving Approach for General Practitioners & Allied Health Professionals

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Motor Neurone Disease (MND) is probably one of the most challenging conditions facing the General Practitioner (GP) and the Primary Health Care Team. This is particularly true for the GP who, with an average caseload in Scotland of about 1300 patients, is unlikely to see more than a couple of MND patients in a professional lifetime.

Motor Neurone Disease is an umbrella term which groups together Primary Lateral Sclerosis, Amyotrophic Lateral Sclerosis and Progressive Muscular Atrophy (PLS, ALS and PMA).

Recent genetic discoveries have linked several genes associated with ALS to Frontotemporal Lobar Degeneration so strongly that behavioural and cognitive changes, and associated Frontotemporal Dementia (FTD), are now considered part of the spectrum too. Further, these genetic discoveries suggest the variability of ALS may be because it is several different diseases of different cellular processes, which all look similar, and not a single disease as previously thought.

Recognition of MND
Patients typically present with isolated and unexplained progressive muscular weakness. There is varied presentation and unpredictable, sometimes very rapid, disease progression. The cranial nerves affecting sight and the lower sacral segments of the spinal cord affecting continence are usually spared. Symptoms may include some of the following:

- Functional effects of muscle weakness, e.g. loss of dexterity, falls or trips
- Bulbar features such as speech or swallowing problems, tongue fasciculations, excessive saliva
- Muscle problems, such as weakness, wasting, twitching, cramps and stiffness
- Breathing problems, such as shortness of breath on exertion or respiratory symptoms that are hard to explain
- Effects of reduced respiratory function, such as excessive daytime sleepiness, fatigue, early morning headache or shortness of breath when lying down.

Supportive features include: asymmetrical weakness; progression; family history of MND, dementia or other neurodegenerative disease. See the ‘Think of Motor Neurone Disease’ leaflet on p4.

Note: Sensory lesions are rare.

Memory and intellect frequently remain intact; however frontotemporal lobar degeneration may be present. A significant number of those affected by MND may exhibit psychological changes which can include:

- Behavioural changes
- Cognitive impairment
- Emotional lability (not related to dementia)
- Frontotemporal dementia.

See also the section on Cognitive Change pp16-17.

Motor Neurone Disease is characterised by progressive degeneration of motor neurones in the following areas.

- Anterior horn cells - resulting in Lower Motor Neurone Lesions (LMN)
- Corticospinal tract cells - resulting in Upper Motor Neurone Lesions (UMN)
- Corticobulbar tracts - resulting in UMN lesions in areas controlling the oropharyngeal muscles
- Motor Nuclei in brain stem - resulting in both Upper and Lower Motor Lesions.
If your patient has some of the concerning characteristics below, think of MND.

Not all have to be present. Although more common in those over 60 years it can present at any age.

The disease will often start in one anatomical zone and then spread to affect others. The disease can initially present with a dementia in addition to swallowing difficulties and or limb weakness. Refer to NEUROLOGY URGENTLY.

Delays to diagnosis occur if referred to other services/specialties.

Progressive weakness in a limb which is:
• Asymmetrical in onset e.g. a foot drop or poor grip
• Without pain or loss of feeling.

Progressive weakness in a limb where:
• There is wasting of muscles
• There are fasciculations in muscles
• There are brisk reflexes
• Cramps occur in muscles.

CLINICAL PRESENTATIONS

Amyotrophic Lateral Sclerosis
Limb onset affects 62%, trunk or respiratory onset affects ~4% of all MND diagnoses. Both UMNs and LMNs may be involved. Characterised by: muscle weakness, spasticity, hyperactive reflexes, fasciculations and weight loss. Usually progresses (80% of cases) to include progressive bulbar palsy symptoms including emotional lability. Normal survival range is 2-5 years from onset of first symptoms.

Progressive Bulbar Palsy (PBP) a form of ALS Affects ~22% of all MND diagnoses. UMNs and LMNs may be involved. Typified by dysarthria and dysphagia.

Progressive Speech /Swallowing Disturbance:
• Slurring of speech
• Increasing difficulty with swallowing, saliva often builds with drooling
• Fluids more troubling at first versus solids
• Tongue weakness with fasciculation.

Progressive breathlessness:
• Not explained by a cardiac or respiratory cause
• More difficulty breathing when flat
• Early morning headaches with fatigue
• Increasing daytime sleepiness.

Problems with behaviour and thinking:
• Becoming more passive, problems multi-tasking and or decision making.
• Loss of drive, motivation, empathy, disinhibition
• Increasing problems with language e.g. reduced vocabulary, increasing difficulties with spelling and sentence formation.

Increased risk of MND:
Family history of MND or early onset dementia.

Important Negatives:
• Bladder and bowel dysfunction
• Visual disturbance e.g. double or loss of vision
• Reduced ability to look up or down
• Eyelid drooping (ptosis)
• Pain at onset with tingling/numbness
• Tremor
• Improvement.

LMN damage: nasal speech, regurgitation of fluids via nose, tongue atrophy and fasciculation, pharyngeal weakness. UMN damage: tongue spasticity, explosive dysarthria, emotional lability. Slightly more women than men affected. Usual survival range is 6 months to 3 years from onset of symptoms.

Progressive Muscular Atrophy (PMA) Affects up to 10% of all MND diagnoses. Defined as a disease of purely LMN degeneration, however a significant number of cases develop sub-clinical and, eventually, overt UMN signs. Characterised by: muscle weakness, wasting, weight loss and fasciculation. Affects men more commonly than women. Younger age of onset: Usual survival range is
Relationship of symptoms experienced to lesions

<table>
<thead>
<tr>
<th>Lesion Type</th>
<th>Upper Motor Neurone lesion</th>
<th>Pseudo Bulbar Palsy (other causes - include stroke)</th>
<th>Upper &amp; Lower Motor Neurone lesions</th>
<th>Medulla Lower Motor Neurone lesion</th>
<th>Medulla Lower Motor Neurone lesion</th>
<th>Bulbar Palsy</th>
<th>Corticospinal tract</th>
<th>Anterior Horn cells</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>Tongue - spastic, no fasciculation; Speech - spastic and explosive dysarthria; Dysphagia; Increased reflexes; Emotional lability</td>
<td>Dysarthria; Dysphagia; Wasting of tongue; Jaw jerk reflex increased</td>
<td>Tongue - shrunken, wrinkled, fasciculation; Speech - slurred; Dysphagia; Paralysis of diaphragm</td>
<td>Spastic weakness; Stiffness; Increased reflexes; Extensor plantar responses</td>
<td>Flaccid weakness; Muscle wasting; Muscle fasciculation</td>
<td></td>
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Onset can be insidious
Early symptoms may include: stumbling, foot drop, weakened grip, slurred speech, cramp, muscle wasting and/or tiredness. Note: some patients may present with acute respiratory problems.

Incidence (Scotland):
Currently 4 per 100,000 per annum across the population as a whole, but as high as 17 per 100,000 in the over seventies.

Prevalence (Scotland):
8+ per 100,000

Male/female ratio: 3 : 2 for ALS,
1 : 1.1 for bulbar onset

Average Age of Onset:
Most common in the middle years. Familial cases tend, on average, to the younger end of middle aged; sporadic cases tend, on average, to be closer to or beyond retirement age. MND can strike in any decade of life.

Differential Diagnosis:
No diagnostic tests currently exist but neurological investigations normally include EMG, serological tests and investigations that sometimes include, Lumbar puncture, Myelogram, Muscle Biopsy, MRI and/or CT scan to exclude possibility of other conditions.
Aetiology & Treatment

Recent studies demonstrate the changing epidemiology of motor neurone disease in Scotland.


Sporadic MND

About 95% of MND cases occur in people with no known family history of the condition. Current research suggests that the sporadic form of MND may develop as a result of a combination of genetic susceptibility, lifestyle and environmental factors that occur throughout life. Epidemiological research has failed to identify any significant risk factors to date. A growing body of evidence is indicating that many sporadic MND cases may be due to complex polygenic causes that are not yet fully understood.

Familial MND

Autosomal dominant inheritance patterns are seen in around 5% of MND cases. Approximately one-fifth of these have an autosomal dominant mutation in the copper zinc superoxide dismutase 1 (SOD 1) gene on chromosome 21. Several other genes have been implicated in familial inheritance, notably “C9orf72” which is linked with both ALS and FTLD and is also responsible for around a fifth of familial cases. However, the known genes do not account for all of the familial cases. Research is on-going both to identify the inherited causes in the remaining familial cases and the trigger which initiates disease onset.

Age and site of onset can vary between cases within the same family while penetrance in familial MND is quoted as about 80%, suggesting that some cases may be polygenic in origin.

Clinically the sporadic and familial forms of MND are indistinguishable.

Mechanisms of motor neurone degeneration

The pathogenic processes underlying MND are likely to be multifactorial. Current evidence suggests interplay between several mechanisms including:

- Glutamate mediated excitotoxicity
- Inflammation
- Oxidative stress
- Mitochondrial dysfunction
- Neurotrophic factor dysfunction
- Protein aggregation
- Glial cell dysfunction
- RNA dysfunction (particularly ncRNAs).

Treatments Available

Riluzole (Rilutek) is the only drug currently available in the UK to treat MND. It was licensed in 1996 and NICE approval was obtained for the treatment of MND in 2001. Riluzole acts by modulating the pre-synaptic release of glutamate. Since late 2015 a syrup suspension of possible benefit to those who are tube fed has been available under the brand name Teglutik ®. This is available on prescription from the GP if taking liquids is easier.

In 2017, the FDA in America licensed Edaravone (Radicava ®) for the ALS subtype of MND. However, it is not licensed in Europe or the UK. See MND Scotland website for further information about this drug.

Unfortunately this currently leaves the GP with little to offer beyond supportive and palliative care. Later sections in this booklet discuss many drugs that can be offered for the management of symptoms.
Role of the Primary Health Care Team

Symptomatic treatments commonly used in patients with ALS. Jenkins TM et al. The evidence for symptomatic treatments in amyotrophic lateral sclerosis. Curr Opin Neurol 2014;27(5):524-531

<table>
<thead>
<tr>
<th>SYMPTOM</th>
<th>COMMONLY USED TREATMENTS</th>
</tr>
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<tr>
<td>Disease progression</td>
<td>Riluzole</td>
</tr>
<tr>
<td>Respiratory failure</td>
<td>Non-invasive ventilation (NIV), Breath Stacking, Cough Assist and Tracheostomy</td>
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<tr>
<td>Laryngospasm</td>
<td>Reassurance, prn low dose benzodiazepines (e.g. sublingual Lorazepam 1mg)</td>
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<td>Chest infection</td>
<td>Early antibiotics, chest physiotherapy</td>
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<tr>
<td>Hypersalivation</td>
<td>Hyoscine, salivary gland botox, atropine drops, amitriptyline</td>
</tr>
<tr>
<td>Difficulty expectorating secretions</td>
<td>Carbocisteine, breath stacking, cough assist machine</td>
</tr>
<tr>
<td>Dysphagia and poor nutrition</td>
<td>Soft diet, chin-tuck, supplements, gastrostomy</td>
</tr>
<tr>
<td>Communication problems</td>
<td>Speech therapy, lightwriters, apps, voice banking</td>
</tr>
<tr>
<td>Poor mobility</td>
<td>Physiotherapy, mobility aids</td>
</tr>
<tr>
<td>Spasticity</td>
<td>Physiotherapy, exercise, baclofen, tizanidine, dantrolene</td>
</tr>
<tr>
<td>Pain: Musculoskeletal</td>
<td>Physiotherapy, paracetamol, naproxen, codeine</td>
</tr>
<tr>
<td>Pain: Neuropathic</td>
<td>Amitriptyline, gabapentin</td>
</tr>
<tr>
<td>Cramps</td>
<td>Baclofen, gabapentin, (quinine – with caution)</td>
</tr>
<tr>
<td>Cognitive problems</td>
<td>Neuropsychology, occupational therapy</td>
</tr>
<tr>
<td>Depression</td>
<td>Citalopram, venlafaxine, nortriptyline</td>
</tr>
<tr>
<td>Emotional lability</td>
<td>Citalopram</td>
</tr>
<tr>
<td>Fatigue</td>
<td>Occupational therapy</td>
</tr>
<tr>
<td>Head drop</td>
<td>Collars</td>
</tr>
<tr>
<td>Prevention of deep venous thrombosis</td>
<td>Compression stockings, Dalteparin in inpatients</td>
</tr>
<tr>
<td>Care of the dying</td>
<td>Morphine, hyoscine, midazolam</td>
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</table>

The Role of the Primary Health Care Team (PHCT) has an important role in providing services for people with MND.

Role of the team in MND
A large number of health and social care professionals will need at some stage to be involved with each patient. Communication is therefore vital with other health and social care professionals through multi-disciplinary meetings.

The PHCT has a role in:

a) Monitoring and assessment
Use a patient centred approach to ascertain the patient’s physical, social, emotional and spiritual needs each time you see them.

Where a patient with MND is being seen regularly by a neurologist a shared care approach may be agreed between the neurologist and the GP. This is particularly important where the neurologist is based some distance away and access becomes increasingly difficult and infrequent as the disease progresses.

b) Managing Symptoms
See relevant sections in this booklet.

c) Onward referrals
Whilst the PHCT may be able to meet some of the patient’s needs and manage many of their symptoms, it is inevitable that the input of other health and social care professionals will be necessary at some stage in the disease progression. In many cases the GP acts as the gatekeeper to these other services.
<table>
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<tr>
<th>Needs/Symptoms</th>
<th>In addition to management within the PHCT, consider referral to:-</th>
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<tbody>
<tr>
<td>Mobility e.g. walking and balancing</td>
<td>Physiotherapist</td>
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<tr>
<td>Swallowing</td>
<td>Speech and Language Therapist</td>
</tr>
<tr>
<td>Weight loss, lack of appetite</td>
<td>Dietician</td>
</tr>
<tr>
<td>Communication</td>
<td>Speech and Language Therapist</td>
</tr>
<tr>
<td>Coughing</td>
<td>Physiotherapist and Speech and Language Therapist</td>
</tr>
<tr>
<td>Breathlessness at rest or lying flat, sleep disturbances, daytime sleepiness,</td>
<td>Respiratory consultant, Respiratory physiotherapist, Specialist palliative care services</td>
</tr>
<tr>
<td>morning headaches, tired or lethargic or difficult to rouse</td>
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</tr>
<tr>
<td>Fear, anxiety, depression</td>
<td>Psychologist/counsellor/palliative care services</td>
</tr>
<tr>
<td>Uncontrolled pain</td>
<td>Specialist palliative care services</td>
</tr>
<tr>
<td>Drooling and/or thick viscous mucus</td>
<td>Physiotherapist for advice on positioning or District Nurse/ GP for suction unit/cough assist, GP for medication</td>
</tr>
<tr>
<td>Difficulties in activities of daily living e.g. washing, dressing, cooking</td>
<td>Occupational Therapist or Social Worker</td>
</tr>
<tr>
<td>and leisure</td>
<td></td>
</tr>
<tr>
<td>Support for individual and family</td>
<td>Social Work, MND Clinical Specialist team, Local Hospice Service</td>
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<thead>
<tr>
<th>Question</th>
<th>Y/N</th>
<th>If “No” action to take</th>
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</thead>
<tbody>
<tr>
<td>Can you identify the person with MND as having a neurological condition</td>
<td></td>
<td>Activate the appropriate coding so your system identifies the patient readily.</td>
</tr>
<tr>
<td>Is the patient’s care regularly discussed within the PHCT?</td>
<td></td>
<td>Put the patient’s name on the Supportive Care Register within your practice.</td>
</tr>
<tr>
<td>Is there a member of the PHCT who has been nominated as coordinator</td>
<td></td>
<td>Identify a named person who (a) acts as the single point of contact within the PHCT and (b) can liaise with all health and social care professionals involved.</td>
</tr>
<tr>
<td>Are efforts being made to control all unpleasant symptoms currently</td>
<td></td>
<td>Use the sections in this booklet to look at options for management. If symptoms are still uncontrolled, refer onwards.</td>
</tr>
<tr>
<td>experienced by the patient?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Is the patient with MND on the GP practice Palliative Patients Register?</td>
<td></td>
<td>Notify your out-of-hours service provider/ complete ePCS and KIS (Key Information Summary) - ensure access is given to all relevant facts/wishes.</td>
</tr>
<tr>
<td>Has an electronic palliative care summary submitted?</td>
<td></td>
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<tr>
<td>Do you have a record or copy of any DNACPR order/ADRT/preferred place</td>
<td></td>
<td>Ask to keep a copy in your patient records (Any copy of a DNACPR order should be black and white only in order not to be confused with the original which should be with the patient).</td>
</tr>
<tr>
<td>of care request in your practice records?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Are you aware of the name of the main carer for this patient?</td>
<td></td>
<td>Record the name of the main carer and their contact details on practice system.</td>
</tr>
<tr>
<td>Is the carer registered with your practice, is their record cross</td>
<td></td>
<td>Put a flag on the carer’s record so you are aware of potential impact on carer’s health.</td>
</tr>
<tr>
<td>referenced in your system?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Do you know what the patient’s wishes are in relation to end-of-life?</td>
<td></td>
<td>Use active listening skills to clarify these as the opportunity presents.</td>
</tr>
<tr>
<td>Have end-of-life wishes been recorded so other members of the PHCT are</td>
<td></td>
<td>Encourage patients to record their wishes and make other members of their family aware.</td>
</tr>
<tr>
<td>also aware?</td>
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Useful resources: [www.palliativecarescotland.org.uk/content/living_dying_well/](http://www.palliativecarescotland.org.uk/content/living_dying_well/)
Co-ordinating Care

Thinking Ahead

MND, characterised as it is by a series of losses, inevitably raises concerns for how extensive these losses will be, particularly in relation to mobility and communication. Early consideration should be given to the implications this might have in relation to the home environment, personal care and planning for the future. Frequently those affected by MND require home adaptations, at different stages of the disease, including:

- Additional handrails
- Methods of accessing upper floors or
- Relocation of the bedroom to another part of the house on the same level as the bathroom
- Converting the bathroom to a wet-room
- Widening of doors to accommodate wheelchairs or hoists
- Provision of wheelchair access and egress

The possibility of severely impaired or lost verbal communication due to dysarthria or anarthria should also be considered. If the ability to speak and write are both lost due to the disease, communicating information about developing health issues or the person’s wishes in relation to end-of-life issues can become very difficult for someone. It is therefore of great importance to raise end of life topics earlier with some MND patients than might be normal.

Alternative and augmentative communication (AAC) devices, available via Speech and Language Services, can help with communication difficulties. However, their use is often made difficult and reduced by fatigue in the later stages of the disease so it should not be assumed that a technological solution will always be available.

Anticipatory Care Planning

The PHCT is in an ideal position to help the patient talk through and record their preferred options for their future care and their preferences for the end-of-life stage. These might also include the following:

- Advance Decision to Refuse Treatment (ADRT)
- Advance statement
- Do Not Attempt Cardio-Pulmonary Resuscitation (DNACPR)

All of the above should be recorded, if appropriate, within the patient’s notes in accordance with current guidelines, e.g. see www.resus.org.uk and the recently published My Anticipatory Care Plan information published by Health Improvement Scotland and the Scottish Government.

https://ihub.scot/anticipatory-care-planning-toolkit/

Care requires a truly holistic approach to the needs of the patient with MND and should encompass, where possible, the needs of the principal carers and close family members. As the disease progresses the mounting series of losses to the individual impacts heavily on relationships with carers and family. It is important to advise carers early on of their legal entitlement to a “Carer’s Assessment” and to support them in applying for this.

The Multidisciplinary Team

The large number of people and organisations often involved in the care of a person with MND can be overwhelming. This can place additional stress on the person and their family. Care should be taken to ensure that patient and family are enabled to participate in planning their own care. Good communication from all members of the team is essential.

A MND Clinical Specialist serving patients in your area will have been identified to:

- Act as an expert source of advice to support the activities of all health and social care professionals
- Act as an expert resource to patients and carers; involving appropriate health and social
Co-ordinating Care

care professionals in response to changing needs
• Link with other service providers
• Advise on probable disease progression to aid assessment of needs for the carer as well as informing the formulation of advance/anticipatory care plans.

If you need to find out who your local MND Clinical Specialist is please contact MND Scotland on 0141 332 3903.

Anticipating possible changes and needs depends on:
• Building and maintaining trust
• Confidence in a rapid response to requests for help
• Communication with the MND Clinical Specialists and effective delegation of responsibility.

Needs of people with MND and their carers
Appropriate information and support:
• During the often protracted period of uncertainty around the time of diagnosis
• Throughout the course of the disease in response to progressive deterioration and impact on family life

• In sufficient time and at an appropriate level to make sure patient and family can make informed choices
• Recognising the increased levels of responsibility assumed by carer
• In answering questions honestly, non-avoidance of painful issues.

Typical questions may include:
• What will happen to me?
• Are there any treatments available?
• Is there a cure?
• How long will I live?
• What will the impact on my day-to-day life be?
• What will happen next with my healthcare?
• Will my children get MND?
• How do I tell my family and friends?
• How will I die?
MND Scotland is committed to ensuring support and information, tailored to meet the diverse and changing needs of carers of people with MND, is provided over the duration of the illness and beyond.

Many people caring for someone with MND do not recognise themselves as a carer and are unlikely to describe themselves as such. It is therefore fundamental that health and social care professionals identify unpaid carers so that services and support are offered in a timely manner.

Caring for someone with MND can be physically and emotionally challenging and often leads to feelings of isolation.

It may not be possible for you to provide solutions to every problem, but much can be achieved by listening to carers. After all, they are the experts.

The atypical route MND takes requires timely intervention and interpretation of what is needed to ensure carers’ needs should be fully met.

Evidence based research consistently shows the satisfaction carers have in supporting a loved one. It also finds:

- A sense of abandonment after diagnosis
- Professionals being slow to respond, lacking empathy, seeming detached and having little time to spend with the person with MND and their family
- Depression and/or anxiety affecting substantial numbers of carers
- Quality of sleep and changes to normal routines related to the mental and physical health of the carer
- Financial pressures resulting in increased strain.

Carers’ assessments (Adult carer support plan)
Carers’ assessments should anticipate future needs as well as immediate needs. They must be outcome based and reviewed on a regular, proactive basis, and not only when a crisis arises.

Coping strategies
Carers of people with MND need advice support and information to help them make sense of what is happening to enable them to continue in their caring role and help them plan for the future.

The carers and families of those living with MND often experience considerable psychological and emotional distress. Their whole world is turned upside down and many assumptions, hopes, plans and expectations for the future have to be reviewed.

Information needs will vary, e.g. too much information at the time of diagnosis may be overwhelming for some. However, carers’ feelings about the illness are likely to change as the illness progresses. It is important to check individual information needs on a regular basis so appropriate information is provided when required, is understood and any inaccuracies are corrected. Information needs may also be different from those of people with MND.

You can also help carers by:
- Allowing time for them to talk about the impact MND has on them and their families. Separate discussions with the carer and person with MND may be more appropriate, as their needs may differ
- Allowing time for them to express and explore their feelings by asking questions, will let you know how safe it is to talk about these issues. It is very unlikely a carer would discuss these feelings and fears spontaneously
- Reassuring a carer that their intensity of feelings and conflicting emotions are ‘normal’ can have a positive impact
- MND is all about losses, so allow time to grieve for past, present and future losses
- Helping carers set realistic goals in ‘bite sized’ chunks to make them more achievable
- Good quality care and support in a timely
manner can have a positive effect on emotional well-being and can prevent a crisis situation emerging.

Where the principal carer or spouse is not registered with your practice they should be encouraged to inform their own GP of their circumstances due to the greater psychological, emotional and physical demands placed upon them.

**Respite**

Carers of people living with MND need regular planned respite care. A key feature of the disease is the speed of progression, resulting in many carers continually trying to meet unrelenting physical, emotional and psychological demands. While some hospices might not offer respite per se, many may consider admission of an MND patient for a period of assessment.

MND Scotland also offers “Time Out” grants which can be used to fund periods of respite.

**Support Organisations**

**MND Scotland’s Welfare Rights and Benefits Officers** can review benefit entitlements, handle claims and may be able to access grants from ourselves or other organisations to help with additional costs associated with caring for someone affected by MND.

**MND Scotland**
Telephone 0141 332 3903
Office hours 9am - 5pm, Monday to Friday.

**Care Information Scotland**
Telephone 0800 011 3200
www.careinfoscotland.scot

**Carers Scotland**
Telephone 0808 808 7777
www.carersuk.org/scotland

**Carers Trust Scotland**
Telephone 0300 772 7701
https://carers.org/country/carers-trust-scotland

**Citizens Advice Scotland**
Telephone 03454 040 506
www.cas.org.uk

**Disabled Living Foundation**
Telephone 0300 999 0004
www.dlf.org.uk

**Young carers**
An innovative, service for Young Carers from Quarriers, which provides a breadth of information to support young carers.
https://quarriers.org.uk/how-we-help/young-people/
Much can be done to alleviate psychological distress, help people to adjust and make the most of their coping skills. The major challenges are coping with loss and living with change.

**Before confirmation of the diagnosis**
The patient’s anxiety can increase as a result of:
- Onset of worrying symptoms
- Difficulty in identifying cause
- A protracted period of investigation.

**Support after the diagnosis**

**Preparation**
Find out what the patient knows so far. Take time to sympathize and to convey the potential seriousness of diagnosis and prognosis and avoid the use of euphemisms. Allow the patient and family opportunities to ask any questions or raise issues they may have thought of since receiving the diagnosis.

Most MND patients in Scotland are now routinely assessed for cognitive and behavioural changes. This may be conducted by the MND Clinical specialists or by local Neuro-psychologists. Patients may be offered appointments regarding the outcomes of these assessments and follow up and subsequent help offered if there is an increase in anxiety or depression or cognitive impairment noted.

**Amount of information**
- Anxiety often limits the patient’s ability to absorb information, therefore they may have forgotten key facts given at diagnosis
- Check that the patient understands the information given and language used
- Offer opportunities for expression of thoughts, feelings and concerns. These help to identify support and information needs
- Offer opportunities to set own pace and return for more information
- Ensure appropriate support mechanisms are in place in line with the Health Improvement Scotland Neurological Health Services Clinical Standards (1). This may also include support from local psychological services
- Discuss how the patient and partner might tell other family members, particularly children.

**As the disease progresses**
Ensure people with MND and their families understand the likelihood and implications of respiratory problems, enabling everyone to explore the options and likely outcome, thus avoiding uninformed last minute decisions, e.g. prolongation of unacceptable quality of life. Consider having conversations with emotional/psychological content from the outset. Dysarthria compounds the difficulty of expressing thoughts and feelings.

Earliest possible referral to the palliative care team is recommended.

MND is characterised by a series of losses and appropriate interventions. In consequence there may be unaddressed psychological needs for additional support which are best addressed through anticipatory care planning.

**Emotional Reactions**
Fears may include:
- Fear of “choking to death” or “fighting for breath”
- Loss of independence and dignity
- Loss of identity and role
- Increasing dependency and becoming a burden
- Inability to cope and loss of control
- The unknown death and the process of dying.
Allowing time and opportunities to acknowledge and discuss these concerns and fears may help to alleviate some of these.

Denial is a coping mechanism that sometimes operates alongside awareness of the condition and its implications. Sustained denial is less helpful and can contribute to higher levels of anxiety and depression. The strength of denial can often be assessed by using past or future oriented questions.

**Anger** can occur at any stage of the illness and needs to be acknowledged. If anger is existential in nature, spiritual support may be appropriate if the family have a particular faith or belief system.

**Sadness** may be linked to the recognition that many of life’s hopes and expectations will never be realised. Time is required to explore these issues.

**Depression** is not always easy to differentiate from sadness, but treatment is likely to have a positive effect. Consider formal screening for anxiety and depression in patients and carers where persistent low mood or hopelessness is expressed.

**Emotional lability** is associated with upper motor neurone involvement and can be very distressing for patients and carers. Patients affected by emotional lability might laugh or cry unexpectedly or inappropriately.

**Medication for emotional lability:**
- SSRIs e.g. Fluoxetine, Citalopram, Sertraline

**Adjustment** or coming to terms with the disease is made more difficult by the rate of deterioration and the accompanying changes and losses. Negotiate with the family what type and frequency of support would help reduce anxiety and fear. There may be phases of the illness where this needs to be reviewed, particularly in the transition periods when significant changes to abilities or losses occur.

Carers may experience a similar range of reactions but at different times from the patient.

Hospice and MND Scotland support groups can be a valuable source of support.

**Sleep disturbances**
Sleep disturbances can arise from a number of causes such as cramps, spasms, fear, anxiety and respiratory problems. See sections on Dyspnoea (pp23-24), Pain (pp25-26) and Palliative Care (pp27-28.) Antidepressants decrease the REM sleep periods, which are also the periods where patients with respiratory weaknesses are more likely to experience respiratory insufficiency.

**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 multidisciplinary teamwork is necessary to provide support and opportunities, to discuss concerns and responses to difficult situations. Regular team meetings or establishing a communication pathway between services is essential.

**Families and Carers**
Many carers need considerable support to help them cope with their role, in particular there is a great need for respite care as well as emotional and psychological support. See the section entitled Families and Carers (p11-12) regarding the importance of offering support for the carer.

**MND forces changes in roles and relationships. Consider:**
- Balancing and giving time to needs of other family members
Psychological & Emotional Support

- The social support system of the family
- Meeting the varying information needs of all family members
- Counteracting isolation of individuals and promoting awareness of each person’s needs
- Offering early opportunities for short periods of respite to prevent over-dependence on a single carer
- Other concurrent life events, such as moving house, children leaving home, financial difficulties etc, and link the families with supportive agencies if appropriate
- Exploring the way in which illness changes relationships and acknowledge not only the strain of this but also ways in which they stay the same or are strengthened. Consider referral for additional psychological support if appropriate
- Creating opportunities for expressing negative feelings without feeling guilty
- Physical exhaustion from the caring role exacerbated by powerlessness to prevent suffering and further deterioration.

Bereavement

Bereavement Counselling is a controversial area with some claiming that it should not happen until a minimum of six months has passed since the death. However some family members, including children, may benefit from on-going bereavement support to help adjust to new circumstances. If grief is still a major issue six months later consider referring to MND Scotland’s counselling service or local bereavement organisations such as CRUSE. Bereavement can also result in delayed grief, particularly in those who appear relatively unaffected or unemotional around the time of death.

MND Scotland’s counselling service
www.mndscotland.org.uk/services/counselling
Telephone 0141 332 3903

CRUSE
http://www.cruse.org.uk/

Winston’s Wish
Winston’s Wish is the largest bereavement charity in the UK for children.
www.winstonswish.org.uk

Reference
(1) Clinical Standards - Neurological Health Services October, 2009
NHS Quality Improvement Scotland
Cognitive Change

Up to 50% of ALS patients can show some degree of cognitive and/or behaviour change ranging from mild changes to a more full blown dementia. Cognitive assessment is now an integral part of the management of ALS as recommended in Motor neurone disease: assessment and management management, NICE Guideline NG42, 2016.

Frontotemporal Spectrum Disorder occurs in a significant proportion of ALS cases (1)
• About 15% of ALS cases meet the criteria for frontotemporal dementia (ALS-FTD) with marked cognitive and behavioural change.
• A further 35% have milder cognitive impairment including executive and/or language dysfunction and/or behavioural change including apathy and disinhibition (ALS with cognitive/behaviour impairment) (2,3).

Cognitive changes in ALS FTD
• Marked executive dysfunction (difficulties with higher order cognitive abilities, planning, decision making, problem solving, poor attention span)
• Language changes in some cases (marked aphasia mostly expressive, sometimes also receptive)
• Memory functions (not a primary symptom, but can experience difficulties due to attentional-encoding difficulties, “taking in” information rather than the memory retention problems associated with Alzheimer’s Disease)
• Visuospatial functions preserved.

ALS with cognitive/behavioural impairment
Greater numbers of patients are affected by milder and more focal cognitive and/or behavioural changes that are not classified as dementia.

• Cognitive impairment may manifest as early deficits on neuropsychological tests of executive functions, verbal fluency, social cognition and some people have primary language problems mostly expressive.
• Behavioural changes may partially meet criteria for FTD with the most prominent symptom being one of apathy. Studies have shown that patients have difficulty initiating ideas and thoughts (4).

Behaviour changes can also include disinhibition, loss of sympathy/empathy, stereotypical and ritualistic behaviour and a change in eating behaviour (sweet food preference).
• Behavioural impairment can occur in between 30 to 60% of patients with MND (and can occur in patients with normal cognition or may co-exist in those with cognitive impairment.

Symptom presentation and impact
Executive dysfunction may manifest as difficulties in:
• Learning new tasks, including the use of equipment associated with symptomatic treatment for ALS (e.g. gastrostomy and Non-invasive Positive Pressure Ventilation (NIPPV))
• Making decisions
• Planning for the future
• Managing affairs/finances.

Language changes may manifest as reduced verbal output, spelling difficulties, failure to initiate conversations and patients responding in short phrases or single words.

Behavioural changes can be difficult for interaction with carers and increases carer burden (5). Carers may misinterpret symptoms as failure in their relationships. Carers may be unaware of mild impairment as increasing physical disability results in loss of autonomy and greater reliance on others for daily tasks.

Diagnosis of cognitive impairment
Cognitive and/or behavioural change may occur at the onset of the disease. The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) is a sensitive and valid brief assessment designed to identify these changes https://ecas.psy.ed.ac.uk. The ECAS is suitable for non-neuropsychologist health professionals to use but training is recommended and/or supervision by neuropsychological services in its interpretation.
Cognitive Change

Full neuropsychological assessment and advice by a clinical neuropsychologist is warranted where cognitive impairment has been suspected. Paucity of local neuropsychology services may be an issue but this must not prevent assessment and should not add to delays in this vulnerable group of patients.

Note: Ventilatory failure developed during the course of the illness can exaggerate cognitive symptoms and NIPPV has been shown to improve some cognitive symptoms. Both respiratory muscle weakness (and dehydration) as a cause of cognitive change should therefore be excluded.

Management
In order to explain the need for assessment, should changes occur, it is important to acknowledge to patients and carers that cognitive change may be part of the clinical picture of MND.

In all patients with frontotemporal syndrome support may need to be provided in decision making; capacity issues may need to be considered for severe cases.

Education of carers/clinicians regarding symptoms of dysexecutive syndrome and training strategies is required.

Latest research has shown the cognitive and behaviour change may increase in some patients during the later disease stages (6).

Useful Organisations
Alzheimer’s Scotland, despite its name, deals with all kinds of dementia, not only Alzheimer’s disease. They offer a 24 hour telephone service on 0808 808 3000.
www.alzscot.org.

The Frontotemporal Dementia Support Group provides information and support for carers of people affected by frontotemporal dementia.

http://www.raredementiasupport.org/ftd

Articles Cited


Dysarthria

80% of patients eventually experience this

Causes weakness & wasting of:
- Tongue
- Lips
- Facial muscles
- Pharynx and larynx.

Reduced palatal elevation, which can lead to “hyper-nasal voice quality.”
Reduced breath support, which can lead to reduced volume or “breathy” voice quality.

Impact
Progressive difficulty with articulation, slurred speech and/or loss of volume. May lead to anarthria.

Treatment
Early referral to speech and Language Therapist (SLT) who will:
- Carry out an oromotor examination to assess patient’s range of movement in their orofacial muscles (e.g. lips, tongue, jaw and soft palate)
- Advise on communication strategies
- Arrange for reassessment and provision of communication aids.

The Occupational Therapist (OT) / Neuro-Rehabilitation Service / Orthotics can assist with advice on:
- Seating, positioning, wrist and head supports
- Switches and pointers
- Mobile arm supports, tables to access communication aids
- Environmental controls.

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
- Ask closed questions for “yes” and “no” answers; use signals for yes/no
- Encourage to slow down and over emphasise words
- Be wary of interruptions or trying to finish sentences
- Have a pen and paper handy to encourage patient to write down any difficult words, this can help to reduce frustration and misunderstanding
- Use an alphabet chart and encourage patient to point to letters to spell out words
- The listener can systematically point to letters on an alphabet chart or say them and the patient can nod or blink when the correct letter is reached
- Use an E-tran frame.

Have a full list of frequently used phrases and ask the patient to point or nod when the listener points to the desired phrase.

Ensure spectacles and/or hearing aids are in place if required.

Many patients struggle to speak on the telephone and may prefer to email professionals for questions or changes of appointments. Some use texting on a mobile phone as an alternative method of communication. When talking to patients remember to slow down and over exaggerate words, breaking up longer words into syllables if required. Ensure adequate breath intake (as able) prior to talking, and ‘top-up’ breaths at regular intervals (again as able) when speaking.

Dysarthria exacerbates emotional reactions including
- Isolation - communication inadequate or avoided
- Frustration - difficult or impossible to be understood; need time which may not be available
- Increased fear and anxiety - because unable to discuss these fears and anxieties
- Low self esteem - others shout or think the person is intellectually impaired
- Loss of control - misunderstood or opinions ignored or not sought.
- Increased sadness - isolation and frustration felt by patient, carer and family. Allow time to explore and discuss the above issues.
When a patient presents with dysphagia deterioration is inevitable and referral to appropriate professionals should be made without delay.

**Cause**
Weakness and paralysis of the oropharyngeal muscles resulting from affected glossopharyngeal, vagus, accessory and hypoglossal nerves.

**Effect**
The first sign of early dysphagia (swallowing difficulties) is often reported to be with thin liquids or dry solids. This can result in:
- Coughing or choking when eating and drinking
- A sensation of food “sticking”.

Later stages may involve:
- Loss of ability to form lip seal, chew, propel food with the tongue and/or form a bolus (a bolus is the item of food or drink plus saliva)
- Acid reflux
- Weak pharyngeal muscles resulting in uncoordinated swallow
- Poor or absent swallow reflex
- Failure to close airway or cough
- Muscle spasm.

**Resulting in**
- Drooling
- Dehydration and weight loss
- Aspiration and recurrent chest infection
- Social Isolation
- Choking/airway blockage
- Impaired respiratory function.

Swallowing problems, texture modification, fatigue, mood, constipation, physical difficulties in buying, preparing and eating foods can all result in unintentional weight loss and risk of malnutrition, with functional consequences such as:
- Increasing muscle wastage due to muscle catabolism
- Increasing respiratory muscle weakness
- Increasing cardiac muscle weakness
- Decreasing physical strength and mobility
- Impairing immune function, thus increasing susceptibility to opportunistic infections
- Decreasing tissue viability
- Decreasing morale and quality of life
- Increased risk of mortality.

Many people will need detailed information regarding how to adjust their oral intake. They may require prescription of food supplements and/or fluid thickeners.

**Advice includes**
- Food fortification to improve energy content
- Recipe ideas to increase variety
- Practical solutions to ease food preparation along with physical eating and drinking difficulties by working with a speech and language therapist (SLT) or an occupational therapist (OT)
- Recommending prescribable nutritional supplements
- Discussing gastrostomy as an option for topping up oral intake to meet full nutritional and hydration needs and to administer medications.

There is no consensus for the exact timing of Percutaneous Endoscopic Gastrostomy (PEG) or Radiologically Inserted Gastrostomy (RIG), but there is considered to be a “window of opportunity”. For those individuals whose respiratory function is considered too poor to undergo PEG insertion RIG may be considered. A timely and sensitive discussion about PEG/RIG placement needs to be made before forced vital capacity (FVC) becomes less than 50% of baseline and the effort of eating becomes exhausting. Food and fluid intake is then inadequate resulting in accelerated weight loss/dehydration and worsening dysphagia resulting in aspiration/choking.
Earlier placement is recommended while the individual is well enough and their respiratory function is adequate to undergo this procedure. Early PEG/RIG placement can improve or maintain quality of life. The pros and cons of this option should be discussed to enable the individual to make an informed choice.

Some health boards in Scotland have considerable waiting lists for these procedures and this must be considered as a delaying time factor also. A minority of health boards will place a Naso-gastric tube if the patient is deemed too advanced for gastrostomy insertion, but there is no evidence that Naso-gastric tubes are recommended for this patient group.

A combination of oral and enteral feeding is possible.

Not every patient will choose this type of intervention and their decision should be respected (see section on Palliative Care pp27-28).

Treatment
Assess ability to eat and drink. Consult SLT who can who can monitor swallow function and recommend appropriate consistencies of food and/fluids as well as suggesting strategies to help maximise safety and ease of intake, taking into account the individual’s wishes.

Assess nutritional intake and weight consult dietician. Head support and positioning consult physiotherapist (see section on Co-ordinating Care pp9-10).

Saliva Management
In most patients saliva problems are the result of poor lip seal and/or impaired ability to swallow. Assess the type of saliva to determine treatment, (see below).

Thin, runny saliva
- Suction
- Tricyclic antidepressants such as: Amitriptyline
- Atropine tablets or Atropine sulphate suspension
- Hyoscine (butylbromide or hydrobromide) sublingually, S.C./I.M., 24 hour syringe pump, transdermal patches consult pharmacist for details of availability. (Note: May slow GI transit.)
- Glycopyrrolate (Robinul ® tablets) or I.V./I.M. (can also be given S.C. as intermittent injections or via a syringe pump)
- Botox injection
- Attention to head support and general posture consult physiotherapist or OT.

Thick tenacious saliva
Check fluid intake, rehydrate, avoid mouth breathing. Oral hygiene consult district nurse.

Proteases such as Papain or Bromelaine, from health food stores sprinkled on the tongue can help break down thick saliva. Pineapple, papaya, apple or lemon juices or flavoured ice cubes can also help.

- Nebulise: Hypertonic saline
- Mucodyne ® (Carbocisteine)
- Propranolol
- Metoprolol
- Expectorants
- Guaifenesin
- Robitussin®

Constipation
Sphincter muscles are not normally affected by MND. Altered bowel function is usually the consequence of forced inactivity, reduced peristalsis, low fluid intake, reduced fibre intake and/or weakness of pelvic floor/ abdominal muscles and the use of analgesics.

Treatment
Regular aperients:
- Movicol sachet®
- Lactulose
- Docusate
- Sennoside B
- Co danthramer (where prognosis is limited).

Use of suppository, enemas, manual evacuation may be necessary on occasions. Also remedies such as bulking agents, prunes, linseed etc. available over the counter. Note: Diarrhoea may be due to constipation with overflow. A new International Dysphagia Diet Standardisation Initiative has been established. Please see Appendix C for further details.
Respiratory Management

**Respiratory insufficiency** affects most people with MND.

Respiratory insufficiency arises in most patients during the latter stages of their disease due to involvement of the diaphragm and accessory muscles and is the usual cause of death in patients with MND. However, rarely, patients may experience acute breathlessness and respiratory distress, earlier in their disease without any other symptoms being present. For some patients, breathlessness, reduced vocal power or poor sleep quality and daytime fatigue may be the initial symptom with which they present.

Those with bulbar onset may experience respiratory problems sooner than those with limb onset MND and can also present an increased aspiration risk and a poor cough. A proportion of MND patients have additional “central” brain-stem failure of respiratory drive. These patients may be acutely sensitive to even small doses of benzodiazepines and opioids; oxygen may also exacerbate respiratory depression.

Respiratory muscle function is a strong predictor of both quality of life and of survival. Progressive weakness of the muscles involved in respiration leads to hypoventilation with gradual development of hypercapnoea and hypoxia; respiratory failure is the eventual cause of death in most patients with MND.

**Assisted Ventilation**

**Non-invasive ventilation (NIV)**

**Benefits**
- Improved quality of life through improved sleep, less troublesome symptoms during the day
- Increased survival
- Increased appetite.

**Disadvantages**
- Risk of aspiration if control of oropharyngeal secretions is poor
- Skin breakdown can occur
- Inability to tolerate mask or use machine
- May become less effective as MND progresses
- Increasing dependency over time, which may have adverse impact on quality of life
- Usually requires assistance of a carer and can increase the carer burden.

**Invasive ventilation (IV)**
- Elective tracheostomy and full ventilation remains controversial – it is not routinely offered in the UK but is undertaken when appropriate. There must be significant discussion and planning around tracheostomy provision and all that it entails. Specialist care that may not be able to be provided at home and also the perceived burden on the family/carers. Occasionally a patient with MND will be invasively ventilated during a crisis hospital admission
- Occasionally requested by a person with MND
- Significant cost implications as well as significant impact on carers as the disease progresses
- Complications associated with insertion and long term use of tracheostomy tubes (with significant rates of early morbidity and mortality)
- Total dependency as the illness progresses as well as potential to lose the ability to communicate (become "locked-in") necessitates significant planning with patients and their carers.

**Discontinuation of assisted ventilation**
It is important to remember that anyone using ventilation can ask for it to be stopped at any time. Discontinuation of the NIV/IV should be discussed with the patient, carer and professionals during the initial discussions over its use and again as part of anticipatory care planning.

A decision by a patient to discontinue the use of ventilation should be followed, acknowledging how difficult this can be for professionals as well as the carers and families of the patient. Guidelines are available from the Association for Palliative Medicine and it should always be carefully planned, in consultation with the initiating respiratory physician/team, as well as palliative care teams so that symptoms can be managed to avoid distress or discomfort.
Many hospice doctors have experience in managing this process and if there are concerns they can be consulted and will often provide support and information.

Further Reading

Association for Palliative Medicine of Great Britain and Ireland 2015. Withdrawal of Assisted Ventilation at the Request of a Patient with Motor Neurone Disease - Guidance for Professionals.

https://apmonline.org/committee-pages/apm-professional-guidelines/

The GP role

1. Monitor for signs and symptoms
This is important so that timely interventions can be made to optimise ventilation and therefore improve/maintain quality of life for as long as possible.

At each contact with the patient assess their breathing and ask about symptoms (as below).

• Breathlessness on exertion or when lying flat
• Orthopnoea
• Shallow rapid breathing or use of accessory muscles on inspiration
• Recurrent chest infection
• Weak cough/sniff
• Weak voice
• Sleep disturbances
• Unrefreshing sleep
• Daytime sleepiness
• Headaches at any time
• Poor appetite/easily full
• Very tired/irritable/confused/drowsy.

Early referral to a respiratory specialist is vital if one or more of the above occur.

2. Make onward referrals
• A baseline respiratory assessment and discussion of future options should be undertaken as soon as possible after diagnosis. Refer to the specialist respiratory team for a full assessment and discussion of options particularly if oxygen saturations are less than 94% (or 92% if known lung disease). Check whether the neurologist has done this.

Note: Deterioration can happen quickly, so an urgent referral may be needed.
• Refer to a physiotherapist for advice on positioning, breath stacking, cough assist to prevent emergency intubation
• Refer to district nurse/OT for provision of appropriate equipment (e.g. profiling bed)
• Refer to the specialist palliative care team for advice on psychological strategies, medication, anticipatory care planning. If possible the carer should be involved in all discussions as they will share the burden of any interventions or options.

If respiratory function is deteriorating it is important to introduce the subject of feeding via gastrostomy, even if there are no immediate problems with swallowing or weight control.

3. Preparing for end-of-life GP Checklist
• Is someone helping the patient develop an advance care plan?

• Do you need to involve a specialist palliative care service?
• Have you discussed the situation with patient and family so they can prepare for what is likely to happen?
• Have you continued to reassure the patient and family that, contrary to popular belief, death by choking or fighting for breath is not the norm?
• Have you arranged for appropriate Palliative care/End of Life medications for the home to prevent a crisis admission? Have you included appropriate drug delivery equipment e.g. needles and syringes? (See section on Palliative Care pp27-28, and Dyspnoea pp23-24. Refer also to end-of-life management of symptomatic breathlessness following local palliative care guidelines.)
• Is there adequate physical and emotional support for the family if the person wishes to die at home?
• Have you informed the ambulance service of any relevant advance planning, e.g. DNACPR?
• Have you informed your local primary care out-of-hours service?
Dyspnoea, Weak Cough & Choking

A common symptom in later stages but may occur earlier.

Cause of Dyspnoea
Respiratory muscle weakness (intercostal, diaphragm, abdominals and accessory muscles of the neck.)

Review current medication and review current fluid/calorie intake (see section on Dysphagia and Nutrition pp19-20) since swallow may become affected as respiratory muscle weakness develops, poor intake exacerbates symptoms. Consider anticipatory care planning at this time if not already attended to.

Symptoms or Effect
• Fear, anxiety, panic & disturbed sleep
• Fatigue and reduced functional ability
• Daytime somnolence
• Poor appetite
• Vivid Dreams
• Breathless with or without exertion
• Breathless when lying (orthopnoea)
• Hypoxia or hypercapnia and possible confusion
• Inability to expectorate lung secretions
• Increased CO2 levels resulting in headaches, particularly on waking.

Treatment
• Relieve fear and anxiety with calm reassurance and/or medication.
• Careful positioning, i.e. Position upright in bed or use riser-recliner chair. Consider specialist equipment consult occupational therapist.
• Breathing exercises and chest physiotherapy, teach assisted cough technique consult: physiotherapist. If unsuccessful consider assisted breath stacking.
• Consider use of “Insufflator/Exsufflator” (removes lung secretions, helps prevent pneumonia) – consult physiotherapist.

Consider:
• Referral to the palliative care team (recommended)
• Referral to specialist respiratory team to discuss onset of respiratory problems early in the disease, to explore options before respiratory problems become evident. Involve the family/carers where appropriate.

Note: Implications and concerns about end of life decisions, increased dependency and becoming “locked in” (see sections on Psychological Support (pp13-15), Palliative Care (pp27-28) and Respiratory Management (pp21-22)).

Medication
• Use humidifier
• Use nebuliser
• Low dose beta blockers e.g. Metoprolol, Propranolol, to reduce production of lung secretions and thick tenacious saliva, mucus plugs (contra-indicated in asthmatics)
• Carbocisteine - use with caution if patient’s cough is weak
• Benzodiazepines
• Lorazepam
• Midazolam S.C. or buccal
• Diazepam
• Opioids (Liquid Morphine)
• Morphine Sulphate modified release tablets e.g. MST, Zomorph ®
• Morphine S.C.

Weak Cough
Breath stacking, manually assisted cough. Use of cough-assist machine is vital to enable expectoration of phlegm or mucus from throat or lungs, thus preventing Emergency Department admission and emergency intubation.

For management techniques consult physiotherapist.

Many Scottish Health Boards recommend the following for patients dying at home from any illness:
• An opioid injection S.C. e.g. Morphine or Diamorphine
• An anxiolytic e.g. Midazolam S.C. or buccal (if special order preparation available)
• An antisecretory S.C. e.g. Hyoscine butylbromide or Glycopyrrolate.
Some also include low dose Haloperidol or Levomepromazine as an antiemetic or second line sedative.

The decision to prescribe medication for use in the future should always be based on a risk/benefit analysis. Reasons for not providing anticipatory medicines include risk of drug diversion or misuse.
**Laryngeal spasm**
May be experienced due to impaired swallow. Patients should be encouraged to extend neck backwards, take a slow breath in and a quick breath out. Amitriptyline may be used to eliminate tickle. Can be accompanied by stridor. Arises through adduction of laryngeal muscles. Can be precipitated by gastro-oesophogaeal reflux. Sublingual Lorazepam 0.5mg is commonly used for managing this symptom.

**Choking**
Attacks may be due to aspiration, impaired respiration or muscle spasm. It should be stressed that death caused by choking attacks is rare, and that the final stages of MND are usually peaceful and dignified. (See section on Palliative Care pp27-28.)

**Treatment**
- Glycopyrrolate bromide
- Midazolam (Epistatus ®) – use sublingually if patient starts to cough or choke caused by bubbly frothy secretions. Can cause drowsiness – ensure rest
- Lorazepam sublingually
- Midazolam liquid, buccally
- Midazolam S.C.
- Haloperidol
- Diamorphine.

For stridor try Omeprazole.
Pain

Affects 45-64% of MND patients at some time.

Pain – Causes
Pain may be caused by muscle cramps, spasticity, intractable joint pain, skin sensitivity, immobility, constipation and occasionally neuropathic pain. The need for anticipatory prescribing is crucial to enable the patient to maintain control and a variety of medications including analgesia, sedatives, anticholinergic and antiemetics are likely to be required.

Suggested management

Medication for muscle cramps
• Quinine bisulphate
• Diazepam
• Baclofen
• Consider the use of Anticholinergics.

Medication for spasticity/rigidity
• Baclofen
• Dantrolene sodium (liver function test)
• Gabapentin (Neurontin ®) (renal function test)
• Tizanidine (Zanaflex ®) (liver function test)
• Clonazepam
• Diazepam
• Robaxan ®
• Botulinum toxin may be used for spasticity, but with caution.

Note: Dosage of muscle relaxant should be carefully adjusted. The patient may experience increased weakness and decreased mobility.

Physiotherapy for Joint Stiffness:
• Careful positioning to relieve discomfort
• Passive exercise programme
• Prevention of contractures
• Maintenance of joint mobility
• Regular review of posture.

Medication for Joint Stiffness:
• Simple analgesia may be preferred
• Try non-steroidal anti-inflammatory drugs by mouth or suppository
• Ibuprofen
• Ketoprofen (Not included in the Scottish Palliative Care Guidelines)
• Naproxen
• Diclofenac
• Analgesics should be titrated using the WHO ladder. Some patients may need an opioid analgesic
• Fentanyl patches can be useful if the patient needs more than 30mg of oral Morphine in 24 hours and has stable pain
• Intra-articular injections (use of intrathecal Baclofen or anti-inflammatory agents may be used for frozen shoulder-like symptoms.)

Medication for Spasm
• Baclofen
• Tizanidine
• Dantrolene Sodium
• Clonazepam
• Lorazepam

Jaw spasm
Botulinum toxin may be used with caution for spasticity and jaw spasm.

Skin sensitivity
Good skin and pressure care is vital. Consider appropriate equipment for skin sensitivity relief, e.g. bed cradle to relieve the weight of bed clothes. Use lightweight bed covers. Use pressure relieving mattresses and cushions; warm socks for cold feet.

Oedema
May be related to lack of muscle movement and posture or to intercurrent disease which should be treated accordingly.

Treatment
• Attention to posture and seating requires regular assessment by occupational therapist
• Support TED stockings, effleurage, massage and reflexology may be beneficial
• A thiazidediuretic such as Bendrofluazide.

Note: Diuresis may result in need to urinate more frequently, causing problems for care and even apparent incontinence.
Neuropathic pain
- Tricyclics e.g. Amitriptyline
- Gabapentin/Pregabalin
- Carbamazepine – only recommended for trigeminal neuralgia
- Phenol block (severs the nerve and is rarely used).

Severe pain or respiratory distress
- Morphine/Diamorphine
- Oxycodone
- Fentanyl patches where pain is stable
- Tramadol*
- BuTrans Patches.

*Tramadol may interact with other medications and lower the seizure threshold, it is therefore not recommended in the Scottish Palliative Care Guidelines.  
https://www.palliativecareguidelines.scot.nhs.uk/

BuTrans patches are not approved by the Scottish Medicines Consortium so may not be available in some Health Boards.

Note: For drug dosages see BNF or Scottish Palliative Care Guidelines.  Recent guidelines recommend Amitriptyline or Gabapentin as first line.  Pregabalin is approved by some NHS Boards as second line anticonvulsant if Gabapentin is not tolerated.

If the patient has severe pain or respiratory distress, an opioid may be required.  Morphine (oral or S.C.), Oxycodone (oral or S.C.) or Diamorphine (S.C.) can be used in titrated doses.

With careful titration, excessive drowsiness and respiratory depression can be avoided (see section on Palliative Care pp27-28).

- Morphine Sulphate immediate release liquid/tablets e.g. Oramorph®/Sevredol®
- Morphine Sulphate modified release e.g. MST, Zomorph ® when stabilised.

(About a third of patients have short term nausea when starting a regular opioid – treat with Metoclopramide for 5-7 days.  All patients on opioids are likely to need a regular laxative.)

Constipation
Maintain hydration and assess fibre intake.  Prescribe a concurrent laxative (see section on Dysphagia and Nutrition on pp19-20.)

Colic
May consider Buscopan (Hyoscine butylbromide) for colic and abdominal distension, however may slow GI transit.
Palliative Care

The concepts and good practice of palliative care should apply from diagnosis. MND is characterised by a series of losses with the accompanying issues of grief and bereavement.

Discussions about anticipatory care planning including DNACPR orders ensure that the patient’s wishes are respected at each stage of the disease.

Working with Palliative Care Services
Introducing the concept of palliative care and specialist services can be difficult for patients to accept, but establishing links via outpatient clinic, day care, respite (where available) and complementary therapies may be a good way in. Emphasising that good quality supportive care is linked to better patient outcomes may be helpful.

It is important that all people with MND should be placed on the GP supportive and palliative care register and discussed regularly in a practice multidisciplinary meeting. This will ensure that planning for the future care can be undertaken while linking the opinions of the multidisciplinary care team. If not already done, anticipatory care planning and discussion of DNACPR should be initiated (but not necessarily concluded) at this stage.

The Terminal Stage
The commonest cause of death is respiratory failure: usually a gradual process (weeks/months).

Death in the majority of cases is very peaceful, following lengthening periods of sleepiness, gradually resulting in a coma and death. Very occasionally this process may occur suddenly.

Note: It is important to reassure patients and carers that death from choking is rare.

Action
Check all symptom control:
- Pain
- Pressure care
- Dyspnoea
- Dysphagia
- Problematic secretions
- Insomnia
- Anxiety/agitation
- Bowels and bladder
- Mouth.

Reassess emotional, spiritual, religious and practical needs of patient, carer and family.

Medication
The emphasis is on anticipatory prescribing e.g. analgesics, sedatives, antiemetics and anticholinergics.
- Opioid analgesics can control pain, reduce the cough reflex and alleviate the sensation of breathlessness, helping to reduce fear and anxiety
- Anticholinergics such as Hyoscine butylbromide or Glycopyrrolate help reduce and dry up excess secretions, relax smooth muscle, but may cause dry mouth. Hyoscine hydrobromide is a third line option as it can cause acute confusion
- Sedatives such as Lorazepam, Diazepam and Midazolam can reduce distress and anxiety. Lorazepam can be given sublingually, avoiding the difficulty of swallowing for breathless or drowsy patients
- In the terminal stages carefully titrated opioids and benzodiazepines can manage distress associated with excessive respiratory drive. Levomepromazine can be an effective sedative in those who do not respond to the above, although specialist palliative care advice should be sought.

If all other reasonable measures have been taken and the person approaching imminent end of life is still in distress prescribing medications in doses which cause sedation might be necessary.

Administration
If possible, continue to give medication orally. Dysphagia, increasing weakness and nausea or vomiting can indicate a need for parenteral administration. Sublingual, transdermal and per rectal routes are available for some medications.
- Many drugs, including opioids, Midazolam and Hyoscine bromide, can be given subcutaneously (S.C.), bearing in mind the possibility of reduced body mass in the terminal phase. If using “as-required” dosing a S.C. cannula can be placed to avoid repeated injections. In many cases a syringe pump allows continuous delivery of drugs over 24 hours, which can be titrated to effect
- Levomepromazine and Midazolam can be used in conjunction with each other (see above)
- Consider the buccal route as a means of administering fast acting medication such as Midazolam, or sublingual Lorazepam.
Palliative Care

See www.palliativecareguidelines.scot.nhs.uk for information about combinations of medications that can be mixed in a subcutaneous infusion given via a syringe pump, as well as advice on general symptom management. Additional information on administering drugs via feeding tubes is available in Palliative Care Formulary.

www.palliativedrugs.com

A syringe pump, primed every 24 hours may be considered to enable patients to remain at home.

Communicating with the patient
This may become extremely difficult but every attempt should be made to maintain communication even if the patient is unresponsive. Eye movements and single response answers to closed questions may be employed alongside picture/alphabet boards where appropriate or other communication aids.

Care plans and information should be shared by all members of the multidisciplinary care team as appropriate. Notes to effect this can be made in the MND Scotland Patient Information Folder - available from MND Scotland.

Good symptom control is essential in the management of a peaceful and dignified death.

Complementary therapies
Some patients have found these to be of great benefit. These include aromatherapy massage, relaxation techniques and acupuncture.

The family and carers may need ongoing practical, spiritual and emotional support. Consider referral to local specialist palliative care services for additional input if required.

References
Examples of good practice websites and downloads

NHS Scotland Palliative Care Guidelines  www.palliativecareguidelines.scot.nhs.uk

Palliative and End of Life Care  www.gov.scot/Topics/Health/Quality-Improvement-Performance/peolc

Living & Dying Well  www.palliativecarescotland.org.uk/content/living_dying_well


DNACPR - Integrated Adult Policy  www.gov.scot/Topics/Health/Quality-Improvement-Performance/peolc/DNACPR


Health Improvement Scotland: Palliative and end of life care services in Scotland  http://www.healthcareimprovementscotland.org/our_work/patient_experience/palliative_care/palliative_care_indicators.aspx
Appendix A: CARE-MND

Clinical Audit Research Evaluation - MND (CARE-MND)

CARE-MND, the Scottish MND register, is a digital platform which comprehensively captures harmonised clinical information from people with MND across all 14 health boards in NHS Scotland in real time, benchmarked against international clinical care standards including NICE 2016 guidelines.

The platform also acts as:

1) A research interest register supporting various observational and interventional studies. People diagnosed with MND in Scotland are invited to register interest in future research studies providing consent to be contacted by one of the CARE-MND research team members.

2) To accurately study the epidemiology of MND in Scotland (including number of newly diagnosed cases, and people living with MND at any one time).

3) To support regular audit to facilitate improvement in clinical care.

CARE-MND has a dedicated website (www.CARE-MND.org.uk) which has information for people with MND, health professionals and others wishing to learn more of the work currently in progress.

## Appendix B: Commonly Used Equipment

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*This list is not exhaustive; please also note*
- Inclusion in this list is not a guarantee of provision
- Assessors may offer advice even if unable to provide equipment
- Other sources of information include local Independent Living Centres, Disabled Living Foundation, (www.dlf.org.uk) and MND Scotland.

**SCTCI** Scottish Centre of Technology for the Communication Impaired exists to provide a high quality tertiary level of augmentative and alternative communications (AAC) service to people referred to SCTCI from 10 of the 14 NHS Boards in Scotland, Tel: 0141 201 2619, Areas covered, Ayrshire and Arran, Borders, Dumfries and Galloway, Forth Valley, Highland, Grampian, Greater Glasgow and Clyde, Lanarkshire, Tayside and Western Isles.

MND Scotland offers an equipment loan service to bridge the gap between the need for a piece of equipment being identified and eventually supplied by the appropriate service. Contact the MND Clinical Specialist for your area to access this service and ascertain if the equipment is supplied and in stock.
Appendix C: Further Reading

Guidelines and Best Practice

• Motor neurone disease: assessment and management. NICE guideline [NG42] Published date: February 2016 https://www.nice.org.uk/guidance/NG42


• IDDSI (International Dysphagia Diet Standardisation Initiative) https://iddsi.org/ The IDDSI framework 2019

Books

• Amyotrophic Lateral Sclerosis Hiroshi Mitsumoto, Serge Przedborski & Paul H Gordon (Editors) Taylor & Francis 2006, ISBN 9780824729240

• Amyotrophic Lateral Sclerosis– Understanding and Optimizing Quality of Life and Psychological Well-Being. Francesco Pagnini and Zachary Simmons (Editors) Oxford University Press 2018 ISBN 9780198757726


• Palliative Care in Amyotrophic Lateral Sclerosis– From Diagnosis to Bereavement, 3rd edition. David Oliver, Gian Domenico Borasio and Wendy Johnston (Editors) Oxford University Press 2014 ISBN 9780199686025


Review Articles


• Goutman SA. Diagnosis and Clinical Management of Amyotrophic Lateral Sclerosis and other Motor Neuron Disorders. Continuum (Minneap Minn) 2017;23(5):1332-1359.

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Our advocacy service supports people affected by MND to resolve any issues, for example with local authorities.

Benefits Advice
A diagnosis of MND can bring additional expenses and we can help people affected by MND apply for the benefits they are entitled to.

Equipment Loan
We have an equipment loan service which can lend equipment, to help people live more comfortably with the effects of MND.

Grants
We have three different kinds of grant available to people affected by MND, to help with costs associated with MND.

Complementary Therapy
We provide complementary therapy sessions, for example massage or reflexology, to people with MND and/or their family carers.

Communication Aids
Someone with MND may need an aid to help them communicate, whether that’s speech or writing, we may be able to help.

Counselling
Receiving a diagnosis of MND, or having a loved one receive a diagnosis, can be extremely overwhelming. We offer a counselling service to people affected by MND to help them cope with these emotions.

Physiotherapy
Our physiotherapy programme for people with MND can help someone stay independent for longer and improve their quality of life.

Accessible Holiday Accommodation
We have accessible holiday accommodation, near Oban and St Andrews, to provide people with a break from their daily routine.

Support Groups
We host support groups across the country, so people affected by MND can join us in a social setting to exchange information and provide support to one another.

Information
We provide information on a variety of topics to people affected by MND; including carers, family members and health professionals.

Education Service
We provide a free education service to those who wish to learn more about MND and the services we offer.

To find out more about any of our services please get in touch by calling 0141 332 3903, email us at info@mndscotland.org.uk or visit mndscotland.org.uk.

MND Scotland would like to thank the Motor Neurone Disease Association who serve the MND population in England, Wales and Northern Ireland. Their Problem Solving Approach publication has been used with permission as a template to create this document aimed specifically at General Practitioners and Primary Care Professionals in Scotland
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**Scottish MND Clinical Specialists**

The Scottish MND Clinical Specialists visit people affected by MND at home to provide information, assess their needs and pull in the right kind of help at the right time from other agencies. Primarily they liaise with health and social care professionals, including the person’s GP, to help communicate the person’s needs, and offer specialist guidance for particular problems.

To find out who the MND Clinical Specialist is in your area, please call 0141 332 3903 or email info@mndscotland.org.uk or visit our webpage mndscotland.org.uk/care team

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**MND Centres in Scotland**

**North**
- Aberdeen Royal Infirmary

**Tayside**
- Ninewells Hospital

**Central Belt**
- Forth Valley Royal Hospital

**East**
- East • Royal Infirmary, Edinburgh

**West**
- The Queen Elizabeth University Hospital, Glasgow
For further information on our support or services contact us at

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