Motor Neurone Disease

A problem solving approach

for general practitioners and allied health professionals

MND CARE CENTRE – EAST		MND CARE CENTRE - TAYSIDE	
Judy Newton and **Gill Stott** MND Clinical Specialist Department of Clinical Neuroscience Western General Hospital Crewe Road Edinburgh EH4 2XU Tel: 0131 537 2131	Borders East, West and Midlothian Edinburgh City Falkirk South East Fife Stirlingshire Clackmannanshire	**Carolyn Webber & Dee McAleer** MND Clinical Specialist Dept of Neurology – Level 6, South Block Ninewells Hospital Dundee DD1 9SY Tel: 01382 425726	Dundee City North East Fife Perth & Kinross Angus
MND CARE CENTRE – WEST		**MND CARE CENTRE – NORTH**	
Carole Ferguson MND Clinical Specialist Room 4, Ground Floor Neurology Institute of Neurological Sciences Southern General Hospital Glasgow G51 4TF Tel: 0141 201 2380	South Glasgow (Glasgow City) East Renfrewshire South Lanarkshire (G postcodes only) Renfrewshire N, S & E Ayrshire Dumfries & Galloway Western Isles	**Dianne Fraser** Clinical Specialist for MND Autovalet, Room 17, Top Floor Aberdeen Maternity Hospital Cornhill Road Aberdeen AB25 2ZD Tel: 01224 554388	Aberdeen City Aberdeenshire Orkney Shetland
Laura Cunningham MND Clinical Specialist Room 4, Ground Floor Neurology Institute of Neurological Sciences Southern General Hospital Glasgow G51 4TF Tel: 0141 201 2381	North Glasgow (Glasgow City) Renfrewshire Inverclyde East & West Dunbartonshire North Lanarkshire Argyll & Bute Highlands South Lanarkshire	**Andrew Bethell** MND Care Advisor Nairn Town and County Hospital Cawdor Road Nairn IV12 5EE Tel: 07815 507942	Highland Moray

Introduction

Motor Neurone Disease (MND) is probably one of the most challenging diseases 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 1400 patients, is unlikely to see more than one or two MND patients in a professional lifetime.

MND Scotland funds a care team of 4½ full time equivalent staff based at the four Scottish neurology centres. Immediately after diagnosis an MND Scotland Care Team Member serving patients in your area should 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 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 the formulation of advance/ anticipatory care plans.

This booklet aims to outline some of the particular problems encountered by people with MND and to provide signposts to sources of help.

Current models of best practice suggest the following fundamental principles should underpin the development and delivery of optimum care:

- Working with and listening to the needs of people with MND and their caregivers;
- Anticipatory symptomatic intervention and equipment provision;
- Appropriate referral to multidisciplinary teams including local palliative care resources;
- Inclusion on a "Supportive Care Register" (Gold Standards Framework) to enable regular monitoring and intervention when necessary;
- Liaising with the MND Scotland Care Team Member to whom the patient is allocated.

The GP's role is pivotal to the above and will be required to deploy all the skills and art of general practice to enable the person with MND to maintain a dignified quality of life from diagnosis to death.

Quality Improvement Scotland's Clinical Standards: Neurological Health Services

MND Scotland contributed to the development of these clinical standards, in particular standards 11 to 13 dealing with MND. We encourage all health and social care providers to work towards achieving and maintaining these standards and can offer advice and support in doing so. The general standards are listed in Appendix B while the MND Specific standards are listed in Appendix C

It is hoped that this revised edition of "A Problem Solving Approach" will form a useful reference and resource for all members of the multidisciplinary care team looking after someone with MND.

Presentation

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

- Anterior horn cells - resulting in Lower Motor Neurone Lesions (LMN)
- Cortico spinal tract cells - resulting in Upper Motor Neurone Lesions (UMN)
- Cortico bulbar tracts – resulting in UMN lesions in areas controlling the oropharyngeal muscles
- Motor Nuclei in brain stem - resulting in both Upper and Lower Motor Lesions.

Note: Sensory lesions are rare.

- Memory and intellect normally remain intact; however frontotemporal lobar degeneration, although rare, may be present, see section on Cognitive Change.
- Cranial nerves affecting sight and lower sacral segments of the spinal cord affecting continence are usually spared.
- Varied presentation and unpredictable, sometimes very rapid, disease progression.

Initial Presentation

Amyotrophic Lateral Sclerosis (ALS)

Limb onset affects 62%, trunk or respiratory onset affects ~4% of all MND diagnoses.

Both upper and lower motor neurones (UMN & LMN) may be involved
Characterised by: muscle weakness, spasticity, hyperactive reflexes, emotional lability, fasciculations, weight loss. Usually progresses (80% of cases) to include progressive bulbar palsy symptoms.
Average survival 2-5 years from onset of first symptoms.

Progressive Bulbar Palsy (PBP) A form of ALS

Affects ~22% of all MND diagnoses.

UMN & LMN may be involved. Typified by dysarthria and dysphagia.
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.
Average survival 6 months to 3 years from onset of symptoms

Progressive Muscular Atrophy (PMA)

Affects about 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 5 times more commonly than women.
Younger age of onset: average survival five years plus.

Primary Lateral Sclerosis (PLS)

Approximately 2% of all MND diagnoses.

Only upper motor neurones damaged.
Characterised by muscle weakness, stiffness of limbs and increased reflex response. Men are affected twice as often as women.
Onset usually after 50 years of age:
Survival similar to normal life span.

The demarcation between the different clinical groups is frequently blurred. As the disease progresses there may be a considerable overlap resulting in more generalised muscle wasting and weakness.

Onset is 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:

2.5 per 100,000 per annum.

Prevalence:

7 to 8 per 100,000

Male/female ratio:

3 : 2 for ALS,
1 : 1.1 for bulbar onset

Average Age of Onset:

Most common in middle years. Although familial cases tend, on average, to be younger middle aged while sporadic cases tend, on average, to be closer to retiral age, MND can strike in any decade of life.

Differential Diagnosis:

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

Relationship of symptoms to lesions

Medulla	Upper Motor Neurone lesion	Pseudo Bulbar Palsy (other causes - include stroke)	Tongue spastic, no fasciculation Speech spastic and explosive, dysarthria Dysphagia Increased reflexes Emotional lability
Medulla	Upper & Lower Motor Neurone lesions		Dysarthria Dysphagia Wasting of tongue Jaw jerk reflex increased
Medulla	Lower Motor Neurone lesion	Bulbar Palsy	Tongue - shrunken, wrinkled -fasciculation Speech slurred Dysphagia Paralysis of diaphragm
Cortico Spinal Tract	Upper Motor Neurone Lesion		Spastic weakness Stiffness Increased reflexes Extensor plantar responses
Anterior Horn cells	Lower Motor Neurone lesion		Flaccid weakness Muscle wasting Muscle fasciculation

Source: Oliver D (1994) *Motor Neurone Disease*, London, Royal College of General Practitioners. Reproduced with permission

Aetiology & Treatment

Sporadic MND

About 90-95% 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.

Familial MND

Familial MND accounts for around 5 to 10% 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. However, these known genes still account for only half of the familial cases. Research is ongoing to identify the causative genes in the remaining familial cases.

Familial MND almost always displays autosomal dominant patterns of inheritance, although penetrance is quoted as about 80%. Age and site of onset can vary between cases within the same family.

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
- Oxidative stress
- Mitochondrial dysfunction
- Neurotrophic factor dysfunction
- Protein aggregation
- Glial cell dysfunction

Many future research investigations are likely to revolve around the TDP-43 and FUS genes. Deposits of the FUS product were found in spinal cells of all sporadic MND samples tested, except those where their MND was caused by a mutation to the SOD 1 gene. No FUS deposits were found in tissue samples from people not affected by MND.

Treatments Available

Riluzole (Rilutek) is the only drug available 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. There are a number of ongoing studies to test the efficacy of existing and novel drugs in the treatment of MND. More information on these can be obtained from our Information Officer/Librarian or via our website www.mndscotland.org.uk. A link to a current list of clinical trials underway worldwide can be found at www.clinicaltrials.gov.

A baseline respiratory assessment and discussion of future options should be undertaken as soon as possible after diagnosis. Check whether the neurologist has organised this. **Note**: Deterioration can happen quickly, so an urgent referral may be needed.
See *http://guidance.nice.org.uk/CG105* for NICE guidelines on non-invasive ventilation.

Later sections in this booklet discuss many drugs that can be offered in the management of symptoms.

Role of the Primary Health Care Team

The Primary Care Team (PCT) 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:

- Within the PCT by using the Gold Standards Framework approach (GSF)
- With other health and social care professionals through multi-disciplinary meetings, where these take place.

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 PCT 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.

Needs/Symptoms	In addition to management within the PCT, consider referral to:-
Mobility e.g. walking and balancing	Physiotherapist
Swallowing	Speech and Language Therapist
Weight loss, lack of appetite	Dietician
Communication	Speech and Language Therapist
Coughing	Physiotherapist and Speech and Language Therapist
Breathlessness at rest or lying flat, sleep disturbances, daytime sleepiness, morning headaches, tired or lethargic or difficult to rouse.	Respiratory consultant, Respiratory physiotherapist Specialist palliative care services
Fear, anxiety, depression	Psychologist/counsellor/palliative care services
Uncontrolled pain	Specialist palliative care services
Drooling and/or thick viscous mucus	Physiotherapist for advice on positioning or District Nurse/GP for suction unit/cough assist, GP for medication
Difficulties in activities of daily living e.g. washing, dressing, cooking and leisure	Occupational Therapist or Social Worker
Support for individual and family	Social Work, MND Scotland Care Team, Local Hospice Service

Primary Health Care Team (PHCT) Checklist

Question	Yes/ No	If "No" action to take.
Can you identify the person with MND as having a neurological condition on your practice IT system?		Activate the appropriate coding so your system identifies the patient readily.
Is the patient's care regularly discussed within the PHCT, e.g. as part of Gold Standards Framework (GSF) meetings?		Put the patient's name on the Supportive Care Register within your practice.
Is there a member of the PHCT who has been nominated as coordinator for the patient's care?		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.
Are efforts being made to control all unpleasant symptoms currently being experienced by the patient?		Use the sections in this booklet to look at options for management. If symptoms are still uncontrolled, refer onwards.
Is it flagged with the out-of-hours service and NHS 24 that the patient has MND? If required, an electronic palliative care summary submitted?		Notify your out of hours service provider/ complete ePCS and ensure access is given to all relevant facts/wishes.
Do you have a record or copy of any DNACPR order/ADRT/ preferred place of care request in your practice records?		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).
Is the ambulance service aware of any DNACPR order/ADRT/ preferred place of care request?		Notify the ambulance service so it can be flagged in its system to prevent inappropriate care/treatment.
Are you aware of the name of the main carer for this patient?		Record the name of the main carer and their contact details on practice system.
Is the carer registered with your practice, is their record cross-referenced in your system?		Put a flag on the carer's record so you are aware of potential impact on carer's health.
Do you know what the patient's wishes are in relation to end-of-life?		Use active listening skills to clarify these as the opportunity presents.
Have end-of-life wishes been recorded so other members of the PHCT are also aware?		Encourage patients to record their wishes and make other members of their family aware.

Useful resources:

www.scotland.gov.uk/Topics/Health/NHS-Scotland/LivingandDyingWell/ePCS
www.goldstandardsframework.nhs.uk www.adrtnhs.co.uk

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 both the home environment 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 their wishes in relation to end-of-life issues can become very difficult for a person affected by MND. It is therefore of great importance to raise these issues earlier with some MND patients than might be normal.

Alternative and augmentative communication (AAC) devices can help with communication difficulties, but their use is often made difficult and reduced by fatigue in the later stages of the disease.

AAC equipment varies from low-tech to high-tech both for different people and for different stages of the disease.

Commonly used devices include:

- simple pencil and paper,
- a child's "magic slate",
- small dry-wipe boards and markers,
- mobile phones with predictive text functions,
- laptop computers.

In addition to the above, specialist AAC equipment such as:

- "Lightwriters"©,
- touch screen computers and
- eye-gaze control computers,

are also available.

Normally the better high tech solutions have a number of modes of operation, according to the abilities of the user, ranging from straightforward typing, through touch-screen control, external switches in scanning mode or, at the most expensive end of the market, eye-gaze control.

However, the types of high-tech equipment described above are rarely available through the NHS or social work departments due to lack of funding for adult AAC provision.

Anticipatory Care Planning

The GP 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.

Co-ordinating Care

Care requires a truly holistic approach to the needs of the patient 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.

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.

An MND Scotland Care Team Member 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 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 the formulation of advance/anticipatory care plans.

Anticipating possible changes and needs depends on:

- Building and maintaining trust;
- Confidence in a rapid response to requests for help;
- Communication with the care team 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.

Families and Carers

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 means timely intervention and interpretation of what is needed to ensure carers' needs should be fully met.

Evidence based research carried out in 2005 (Mockford *et al*, 2006) showed the satisfaction carers had in supporting a loved one. It also noted:

- The 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 affecting up to 25% of carers;
- Anxiety affecting up to 50% 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.

Coping with the loss of social networks, loss of partner and companion and loss of a future together

Carers' assessments

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 to 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.

Support Organisations

MND Scotland's Welfare Rights and Benefits Officer can review benefit entitlements, handle claims and may be able to access grants from other organisations to help with additional costs associated with caring for someone affected by MND.
Telephone 0141 945 1077

Carers Scotland, Telephone 0141 445 3070
www.carerscotland.org

Princess Royal Trust for Carers
Telephone 0141 221 5066
www.carers.org

Citizens Advice Scotland
www.cas.org.uk

Disability Living Foundation, Telephone 0845 130 9177
www.dlf.org.uk

Young carers
An innovative, online service for Young Carers from The Princess Royal Trust for Carers, which provides a breadth of information to support all young carers.
www.youngcarers.net

Dysarthria

80% of patients are eventually affected by 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:

- Examine the patient’s range of movement in their lips, tongue and palate or will carry out an oromotor examination;
- Advise on communication strategies;
- Arrange for reassessment and provision of communication aids such as Lightwriters and other 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.

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.

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 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.

Dysphagia & Nutrition

Dysphagia eventually affects about two thirds of all patients.

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 may be problems when swallowing liquids resulting in severe coughing and progressing to include:

- Loss of ability to form lip seal, chew, propel food with the tongue and/or form a bolus;
- Poor or absent swallow reflex;
- Weak pharyngeal muscles resulting in uncoordinated swallow;
- Failure to close airway;
- Muscle spasm;
- Acid reflux.

Resulting in

- Drooling;
- Dehydration and weight loss;
- Aspiration and recurrent chest infection;
- Impaired respiratory function.

Monitoring of weight and nutritional intake is necessary for all patients with MND. Equally, involvement of a dietician is essential to assess, monitor and review an individual's nutritional intake and to provide practical dietary advice to enable nutritional needs to be met both orally and non-orally.

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 about how to adapt their diet and may need prescribable food supplements and thickeners.

Advice includes:

- Food fortification to improve food quality;
- Recipe ideas to increase variety;
- Practical solutions to ease food preparation along with physical eating and drinking difficulties by working with occupational therapist;.
- 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.

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).

Treatment

Assess eating and monitor swallow - consult **SLT** who can recommend appropriate thickness for dietary intake.

Assess nutritional intake and weight - consult **dietician.**

Head support and positioning - consult **physiotherapist or OT.**

Note: See section on Co-ordinating Care.

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. See also note on Hyoscine hydrobromide on page 21.
Glycopyrronium I.V./I.M. (Robinul)
(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, re-hydrate, 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 (Carbocysteine)
Propanolol
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.

Notes

Dyspnoea & Choking

A common symptom in later stages but may occur earlier.

Cause of Dyspnoea

Weakened respiratory muscles - intercostal, diaphragm, abdominals and accessory muscles of the neck.

Review current medication and review current fluid/calorie intake (see section on Dysphagia and Nutrition) as 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 CO_2 levels resulting in headaches, particularly on waking.

Treatment

Relieve fear & anxiety - 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 & chest physiotherapy, teach assisted cough technique - consult: physiotherapist**.**

Test blood gases & residual volume in lungs (FVC). Sniff nasal pressure and overnight oximetry.

Consider use of "Insufflator/Exsufflator" (removes lung secretions, helps prevent pneumonia) – consult physiotherapist.

Consider:

- Referral to the palliative care team (recommended);
- Referral to a consultant in respiratory medicine to discuss onset of respiratory problems early on, to explore options before respiratory problems become evident. Involve the family;
- Assisted ventilation;
- Consider annual flu vaccination.

Note: Implications and concerns about end of life decisions, increased dependency and becoming "locked in" (See section on Psychological Support, Palliative Care and Respiratory Management).

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 – increase mucodyne to relieve symptoms – use with caution as patient's cough is weak;
- Midazolam S.C. or buccal;
- Lorazepam;
- Diazepam;
- Liquid morphine;
- Morphine Sulphate modified release tablets e.g. MST, Zomorph;
- Diamorphine.

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 A&E admission and emergency intubation.

For management techniques consult physiotherapist.

The MNDA "Just in Case Kit" is produced by the MND Association, Northampton, but can be obtained free of charge through MND Scotland Care Team Members.

For patients and carers the kit provides tangible evidence that fears have been addressed and practical help is at hand in the form of medication for the relief of anxiety and breathlessness.

For the GP and district nurse it provides guidance on symptom management, medication and storage for the prescribed medications. The GP and DN will need to advise and support the carer in the use of the kit.

How to obtain a Just in Case Kit	
1.	GP agrees with patient & carer that a kit is required
2.	GP requests a kit from care team member for the patient
3.	Kit is supplied to GP free of charge
4.	GP & DN agree on plan to advise & support carer in the use of kit
5.	GP prescribes appropriate medication to be supplied with the kit

Note: The basic kit is an empty box, with instructions, in which to keep the MNDA recommended medications once prescribed by the GP. Many Scottish Health Boards recommend the following for patients dying at home from any illness:

- An opioid injection SC e.g. morphine or diamorphine
- An anxiolytic e.g. midazolam SC or buccal (if special order preparation available)
- An antisecretory SC e.g. hyoscine butylbromide or glycopyrronium.

Some also include low dose haloperidol or levomepromazine as an antiemetic or second line sedative

Laryngeal spasm

May be experienced due to impaired swallow. Patient 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-oesophogeal reflux.

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).

Treatment

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

For stridor try Omeprazole.

Respiratory Management

Respiratory insufficiency affects most people with MND.

Respiratory insufficiency arises in most patients during the course of their disease due to involvement of the diaphragm and accessory muscles and is the usual cause of death in MND.

For some patients, breathlessness, reduced vocal power or poor sleep quality and daytime fatigue may be the initial symptom with which they present to your surgery. Rarely, patients may be admitted to A&E in respiratory distress without any other symptoms at all.

Those with bulbar onset usually experience respiratory problems sooner than those with limb onset MND. A proportion of MND patients have additional "central" brain-stem failure of respiratory drive. These patients can be acutely sensitive to even small doses of benzodiazepines and opioids; oxygen can sometimes lead to rapid coma.

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

Assisted Ventilation

Non-invasive ventilation

National Guidelines on non-invasive ventilation *http://guidance.nice.org.uk/CG105*

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 if mask is used for long periods;
- Inability to tolerate mask or use machine;
- Less effective as MND progresses;
- Increasing dependency over time, which may have inverse impact on quality of life;
- Usually requires assistance of a carer and can increase the carer burden.

Invasive Ventilation

- Not routinely offered in the UK – may be considered once NIV is no longer effective;
- Often a result of a crisis hospital admission;
- Occasionally requested by a person with MND.

Benefits & advantages are as above, plus:

- Complications associated with insertion of and long term use of tracheostomy tubes (with significant rates of early morbidity and mortality);
- The cost and burden of care can be great;
- Total dependency as the MND progresses may be unacceptable.

Discontinuation of assisted ventilation

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. See guidelines at *www.palliativedrugs.com/download/091209_combined_Withdrawing_NIV_Draft_Guidelines_Revised231009.pdf*.

A decision by a patient to discontinue the use of ventilation is legally binding and these wishes should be followed. It should be carefully planned, in consultation with the initiating respiratory physician, 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 support the GP by assisting in the procedure.

The GP role

1. Monitor for signs and symptoms

This is important so that timely interventions can be made to optimise oxygenation and to 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).

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

- Breathlessness on exertion or when lying flat;
- Shallow rapid breathing or use of accessory muscles on inspiration;
- Weak cough/sniff;
- Weak voice;
- Sleep disturbances;
- Unrefreshing sleep;
- Daytime sleepiness;
- Headaches on wakening;
- Poor appetite/easily full;
- Very tired/irritable/confused/drowsy.

2. Make onward referrals

- Refer to the local respiratory consultant for a full respiratory assessment and discussion of options. A baseline respiratory assessment and discussion of future options should be undertaken as soon as possible after diagnosis. 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. There is a critical period with respiratory function beyond which it may be unsafe to operate (FVC<50%).

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 medications (e.g. Just in Case Kit) for the home to prevent a crisis admission? (See sections on Palliative Care and Dyspnoea. 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?

Pain

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

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 medication 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 (Zanoflex) (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.

Joint Stiffness

Physiotherapy:

- 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 thiazide diuretic 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;
- Valproate is now a third line option;
- Phenol block (severs the nerve and is rarely use.)

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.

BuTrans patches are not SMC approved so may not be available in some 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 SC), Oxycodone (oral or SC) or Diamorphine (SC) can be used in titrated doses.
With careful titration, excessive drowsiness and respiratory depression can be avoided. (See section on Palliative Care)
.

- 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 & Nutrition).

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 and 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 day care, respite and complementary therapies may be a good way in.

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 or Gold Standards Framework 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 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 and practical needs of carer and family.

Medication

The emphasis is on anticipatory prescribing in line with the Liverpool Care Pathway guidelines 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 Glycopyrronium 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 can manage distress associated with excessive respiratory drive. Levomepromazine can be an effective sedative.

If all other reasonable measures have been taken and the person is still in distress prescribing benzodiazepines 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 PR routes are available for some medications.

- Many drugs, including opioids, Midazolam and Hyoscine bromide, can be given 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 S.C. 8 – 12 hourly or via syringe pump can also be used as an alternative to Midazolam. Both these drugs can be used in conjunction with each other;
- Consider the sublingual route as a means of administering fast acting medication such as Midazolam.

See also www.palliativecareguidelines.scot.nhs.uk for information about combinations of medications that can be mixed in a subcutaneous infusion given via a syringe pump and "Palliative Care Formulary" (PCF3 Appendix 10) *"Administering drugs via feeding tubes" www.palliativedrugs.com.*

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

Note: The "Just in Case Kit" may be appropriate - see section on Dyspnoea.

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.

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 will need practical and emotional support.

References – examples of good practice websites and downloads

Gold Standards Framework	*www.goldstandardsframework.nhs.uk*
NHS Scotland Palliative Care Guidelines	*www.palliativecareguidelines.scot.nhs.uk*
Liverpool Care Pathway	*www.mcpcil.org.uk/liverpool-care-pathway/index.htm*
Living Well and Dying Well	*www.scotland.gov.uk/Topics/Health/NHS-Scotland/LivingandDyingWell*
Preferred Priorities for Care	*www.endoflifecareforadults.nhs.uk/assets/downloads/ppc.pdf*
Omega (The National Association for End-of-Life Care)	*www.omega.uk.net*
The GMC's ethical guidance on-end-of life care	*www.gmc-uk.org/guidance/ethical_guidance/6858.asp*
Advance decision to refuse treatment	*www.adrtnhs.co.uk*
DNACPR - Integrated Adult Policy	*www.scotland.gov.uk/Topics/Health/NHS-Scotland/LivingandDyingWell/ShortLifeGroups/-DNACPR*

Cognitive Change

Up to 50% of ALS patients can show some degree of cognitive change.

A frontotemporal syndrome occurs in a significant proportion of ALS cases

- Five to fifteen percent of ALS cases meet the criteria for frontotemporal dementia (ALS-FTD) with marked cognitive and behavioural change.
- A further 35% have mild cognitive impairment such as mild aphasia and/or may have behavioural change.

These two conditions are distinct and people with cognitive change should not be labelled as having ALS-FTD or vice versa. The following paragraphs define ALS-FTD and the combination of symptoms that may indicate frontotemporal dementia.

Behaviour changes in ALS-FTD

- Significant personality change;
- Disinhibition and impulsivity (socially inappropriate behaviour;
- Perseveration (continuing to conduct an activity which is no longer appropriate);
- Change in eating behaviour (sweet food preference);
- Loss of emotional understanding (appear egocentric/selfish);
- Withdrawn (apathy/failure to initiate);
- Stereotypical/ritualistic behaviour;
- Behavioural change often occurs **before** cognitive impairment.

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 difficulties (not a primary symptom, but can experience difficulties due to executive dysfunction and “taking in” information rather than the short term memory problems associated with Alzheimer’s for example);
- Visuospatial functions preserved.

Cognitive impairment and/or behavioural changes not classed as ALS-FTD

Greater numbers of patients are affected by cognitive and/or behavioural changes that are not classified as dementia. Cognitive and/or behavioural changes occur in what is thought to be a spectrum ranging from mild to moderate or more severe presentations including a combination of the following symptoms:

- Cognitive impairment may manifest as early deficits on neuropsychological tests of executive functions (verbal fluency) and some people have primary language problems similar to those described above;
- Behavioural changes may partially meet criteria for FTD including disinhibition, loss of insight, lack of volition/apathy, stereotypical and ritualistic behaviour.

Behavioural impairment is a recognised feature of MND and studies suggest that over 60% of patients with MND are apathetic, irritable, inflexible, restless and disinhibited (Grossman et al 2007) and altered sensitivity to social and emotional cues (Lule et al 2005).

It is important to note that cognitively normal patients can have profound behavioural abnormalities (Lomen-Hoerth et al 2003). Further, that cognitive and/or behavioural impairment can co-exist in up to a third of MND patients (Murphy et al 2007).

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 NIPPV);
- Making decisions;
- Planning the future;
- Managing affairs/finances.

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

Behavioural changes (e.g. disinhibition and perseveration) can be difficult for interaction with carers and increases carer burden. 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.

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 care needs to be taken to ensure informed consent during 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. Research is currently underway to explore decision-making and identify factors that influence acceptance of interventions such as gastrostomy. Unfortunately not enough is known to answer important questions such as whether cognitive deficits in MND are progressive.

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 Pick's Disease Support Group provides information and support for carers of people affected by frontotemporal dementia.
www.pdsg.org.uk.

Articles Cited

Grossman AB. Woolley-Levine S. Bradley WG. Miller RG Detecting neurobehavioral changes in amyotrophic lateral sclerosis. **Amyotrophic Lateral Sclerosis 8(1):56-61, Feb 2007**

Lule D. Kurt A. and Jürgens R.*et al.*, Emotional responding in amyotrophic lateral sclerosis, ***J Neurol* 252 (12) (2005), pp. 1517–1524**

Lomen-Hoerth, C. Murphy, J. Langmore, S. Kramer, J H. Olney, R K. Miller. B. Are Amyotrophic Lateral Sclerosis Patients Cognitively Normal? **Neurology. 60(7):1094-7, 2003**

Murphy J. Henry R. Lomen-Hoerth C. Establishing subtypes of the continuum of frontal lobe impairment in amyotrophic lateral sclerosis**.** **Archives of Neurology. 64(3):330-4, 2007**

Psychological Support

People with MND, their families and carers often suffer considerable psychological and emotional distress

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 to people after the diagnosis

Preparation

Find out what the patient knows so far.
Take time to sympathise 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.

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 QIS Neurological Health Services Clinical Standards. 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 an 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 is 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;
- 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:

- Tricyclic Antidepressants e.g. Amitriptyline,
- SSRIs e.g. Fluoxetine, Citalopram, Sertraline

Amitriptyline has a higher incidence of adverse side effects at an antidepressant dose and is not recommended for patients with cardiac disease or the elderly. Fluoxetine and Amitriptyline both need to be used for a few weeks to improve depression. Fluoxetine can be stopped abruptly as it has a long half life, this apart, avoid rapid withdrawal of Tricyclics.

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, Pain and Palliative Care. 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 (p9) and Appendix C regarding the importance of offering a “Needs Assessment” for the carer.

MND forces changes in roles and relationships. Consider:

- Balancing and giving time to needs of other family members;
- 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

Family members, including children, may benefit from ongoing bereavement support. Consider referring to MND Scotland’s counselling service, the local palliative care 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 945 1077

CRUSE
www.crusebereavementcare.org.uk

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

Notes

Appendix A; Scottish Motor Neurone Disease Register

The Scottish Motor Neurone Disease Register is funded by MND Scotland and is part of the Scottish Motor neurone disease Audit, Research and Trials (SMART) unit (www.smart-mnd.org) which is based within Edinburgh University.

SMNDR has four explicit goals:

- To repeat the study of incidence in Scotland to find out if the condition is becoming more common. This will allow us to predict trends in prevalence, and model the demand for services such as non-invasive ventilation.
- To produce a new prognostic model through longitudinal assessments centred on improved phenotyping. This will lead to a well characterised cohort of patients essential for trial recruitment.
- To develop existing biological tissue resources such as the in-house database of four hundred DNA samples, and evaluate new biomarkers of prognosis (in collaboration.)
- To audit the diagnostic phase of care against recently established QIS standards (see appendices A and B) and determine outcome after particular interventions, e.g. gastrostomy and non-invasive ventilation.

All patients whose names are put forward for the register will be contacted by SMART and given the opportunity to receive information about future research or trials in which the unit might have an interest.

Additionally the unit will investigate how centres are meeting the QIS Neurological Standards (The Audit, see appendices B & C) by reviewing health records, with the aim of improving patient care in line with the intentions of the Standards.

Those patients who take part will be given introductory information and will be followed-up annually to assess the progression of their symptoms.

Appendix B; General Neurological Standards

Published by Quality Improvement Scotland as part of the document 'Clinical Standards – October 2009 Neurological Health Services'

Standard 1: General neurological health services provision

Standard statement 1

An effective and comprehensive neurological health service is available and offered across all NHS Boards.

Rationale

Individuals should have access to all aspects of neurological care regardless of where they live.

Patient care is enhanced by the provision of up-to-date and accurate written and verbal information.

Consistent and ongoing data collection allows NHS Boards to monitor and review their services on an ongoing Basis, for the purpose of service improvement. Success will not occur without clinician and management engagement, the active participation of NHS Board IT departments and a long-term approach to system development.

The Neurological Services Data and Audit Group will identify and agree a common dataset for neurological services to ensure consistency across all NHS Boards.

Essential criteria

1.1 The NHS Board makes accurate and current information available about its existing designated services for patients with neurological conditions.

1.2 The NHS Board has a minimum 3-year plan for the provision of neurological health services to its population. This plan is published and subject to annual review.

1.3 The NHS Board works collaboratively with the Neurological Alliance of Scotland, other patient support groups and charities to ensure that patients and their carers are made aware of the resources available nationally and locally through voluntary sector partners.

1.4 The NHS Board provides accurate and current information to patients and their carers about their condition.

1.5 The NHS Board provides resources to ensure collection and analysis of data in relation to neurological services activity and outcomes, as identified by the Neurological Services Data and Audit Group.

1.6 The data collected within the common dataset is used to improve patient care.

Standard 2: Access to neurological health services

Standard statement 2

Patients with suspected neurological conditions are assessed by clinicians who specialise in neurological conditions. Patients are assessed within timescales dictated by their clinical needs.

Rationale

Evidence suggests that neurological conditions are most effectively dealt with by specialist clinicians. Timely access to neurological health services is important in order to achieve good outcomes for patients with some neurological conditions. The Scottish Government Health Directorates have established overall maximum waiting times targets, but some patients will need to be seen within shorter timescales. Providing clear and efficient referral systems will enhance NHS Boards' ability to achieve targets.

Access to neurological health services for patients in remote or rural areas is improved by access to telemedicine.

Essential criteria

2.1 The NHS Board demonstrates that a minimum of 90% of outpatient demand for all neurological health services can be met by substantive resources without resorting to waiting times initiatives, reliance on temporary staffing or other short-term measures.

2.2 Outpatients are referred and triaged electronically. They are allocated to the appropriate waiting list within 5 working days of receipt of the referral at the centre in at least 95% of cases.

2.3a The NHS board ensures that the neurology service has a communication process for discussion of urgent oases with a neurologist at all times.

2.3b Where the neurologist identifies an outpatient referral as urgent, the patient is seen within 10 working days of triage in at least 90% of referrals.

2.4 Initial advice following an urgent request for a neurological opinion for inpatients in non-neurological settings occurs within 24 hours.

2.5 At least 80% of patients with a neurological condition requiring transfer are admitted under the care of a neurologist within 48 hours of acceptance.

2.6 Individuals affected by chronic neurological disease are provided with a contact point within the relevant neurology service to allow for re-entry into the service.

2.7 District general hospitals and regional neurology centres have on-site 24-hour access to telemedicine facilities.

Standard 3: Patient encounters in neurological health services

Standard statement 3

Neurological health services provide a high quality of care that meets the needs of patients, referrers and providers

Rationale

The ability to meet a patient's needs is fundamentally dependent on the quality of the consultation. Other factors such as a good physical environment, timeliness and provision of information are important to enhance the interaction between patient and clinician. Rapid, accurate, legible and accessible communication between all those involved with the patient is also of paramount importance.

Essential criteria

3.1 The professional development and maintenance of standards of all staff working within neurological health services is monitored by the NHS board.

3.2a The NHS board implements systems to collect patient feedback to improve the neurological health services on an ongoing basis.

3.2b The NHS board implements systems to collect feedback from clinicians referring into the service to improve the neurological health services on an ongoing basis.

3.2c The NHS board implements systems to collect neurological service staff feedback on its neurological health services on an ongoing basis.

3.2d The NHS board acts on the patient, referrer, and staff feedback it collects.

3.3 The NHS board provides access to, and demonstrates participation in, communication training for all staff having direct contact with patients with neurological conditions.

3.4 Patient waiting times in clinics are monitored and patients receive an explanation for the delay if they have to wait for more than 30 minutes beyond their appointment time.

3.5 Patients are provided with practical information in advance of their first appointment specific to the appointment and department.

3.6a The outpatient service is conducted in a safe and comfortable environment for patients and is surveyed annually to ensure the quality is maintained.

3.6b Designated private facilities are available on wards for discussions between staff, patients, family and carers.

3.7a New patient encounters are scheduled to allow a minimum of 30 minutes consultation time with a clinician and 30-40 minutes with trainees.

3.7b Return patient encounters are scheduled to allow a minimum of 15 minutes consultation time with a clinician and 15-20 minutes with trainees.

3.7c Clinicians have a facility to schedule additional time where a prolonged consultation is anticipated.

3.8 A minimum of 90% of outpatient letters are electronically dispatched within 5 working days of the consultation.

3.9a All inpatients are discharged with a printed immediate discharge summary.

3.9b Immediate discharge information is sent to the GP electronically in at least 90% of cases.

3.9c Final discharge summaries are dispatched electronically to the GP within 5 working days of discharge in at least 95% of cases.

Standard 4: Management processes in neurological health services

Standard statement 4

Neurological health services have an effective patient management process from the point of first referral.

Rationale

Patients with neurological conditions benefit from the ready availability of a variety of different specialist investigation and management resources. In some cases, complex needs demand access to a wide range of services, including rehabilitation, self-management options and palliative care. A common rate-limiting step in the delivery of care of the acutely ill patient is the ability to access the acute neurology inpatient unit and its associated resources.

One third of all patients attending neurology outpatient departments have neurological symptoms unexplained by disease, but who may benefit from specialist management. In addition, patients with neurological conditions are much more likely to have psychiatric disorders than the general population. Failure to recognise and treat such co-morbid disorders is common and may seriously undermine the patient's neurological management and worsen their prognosis.

Access to clinical neuropsychology is important for the diagnosis of disorders of cognition, the management of patients with neurologically unexplained symptoms and psychological support for patients with other neurological conditions.

Community health partnerships (CHPs), or equivalent, have a role in integrating primary care and specialist services with social care. This role will support services to provide consistent care throughout a patient's journey and discharge from neurological health services.

Essential criteria

4.1 At every consultation, all patients are offered a copy of the GP's letter or a management plan. Any changes to medication are provided in writing immediately to the patient.

4.2a All neuro-imaging procedures are reported by a neuroradiologist, or a general radiologist who has had specialist neuroradiological training.

4.2b All neurophysiology procedures are performed and reported by a neurophysiologist, or medically trained staff with neurophysiological training.

4.3 At the conclusion of the care episode, 80% of patients are transferred back from the neurological unit to the referring unit within 2 working days of transfer request.

4.4a The neurology service has access to an integrated neuropsychology and neuropsychiatry service providing a diagnostic and treatment service for patients with neurological symptoms unexplained by disease, and patients with defined neurological disease that have co-morbid psychiatric disorders.

4.4b At least 80% of patients referred to this service requiring urgent assessment will have initial contact within 24 hours if referred from inpatient consultation, or within 20 working days if referred from outpatient consultation.

4.5 The neurology service has access to specialist neuropsychological assessments and rehabilitation contributing to the diagnosis and management of neurological conditions.

4.6 The NHS board provides designated rehabilitation services specifically for people with neurological symptoms.

4.7 There are multidisciplinary systems in place, with input from a specialist clinical pharmacist for:

- safe use of medicines
- access to formulary medicines
- use of non formulary and unlicensed medicines
- education of non medical and medical prescribers, and
- regular medicines reconciliation and review.

4.8 The neurology service has channels of communication with the individual responsible for long term conditions in the local community health partnerships, or equivalent, to co-ordinate the provision of services, equipment and medication, by the NHS and social services.

4.9 People affected by neurological conditions have ongoing access to self-management options.

4.10 Patients with advanced conditions or complex needs have access to assessment and treatment in their place of residence by a member of the neurology multidisciplinary team where they are unable to access services at hospitals or clinics.

4.11 Patients with neurological conditions have access to equipment for assisting with daily living where that equipment is normally provided by NHS Scotland.

4.12a Palliative care is provided for patients with neurological conditions, their family and carers, as required throughout the course of their illness, and in accordance with the wishes of the patient.

4.12b Specialist palliative care is provided for patients with complex needs.

4.12c Patients with neurological conditions are encouraged to discuss advance care planning, when clinically appropriate.

Appendix C; MND Specific Neurological Standards

Standard 11 Access to specialist motor neurone disease services

Standard Statement 11
An effective and comprehensive motor neurone disease service is available and offered across all NHS boards.

Essential Criteria
11.1a Patients with suspected MND are referred to a neurologist.

11.1b Patients with clinically definite or clinically probable MND are referred to a defined MND service

11.2 The multidisciplinary team consists of, as a minimum, a doctor who specialises in MND, a regional care specialist, the patient and a carer. Additional input from other healthcare professionals with experience and training in neurological conditions is offered from the following services:

- Physiotherapy
- Occupational therapy
- Speech and language therapy
- Dietetics
- Pharmacy services
- Mental health services

11.3 The NHS board provides rapid access to demonstrably effective care pathways covering all aspects of the illness, including links to specialist palliative care and respiratory medicine, gastrostomy services and social services.

Standard 12 Diagnosis of motor neurone disease

Standard Statement 12
Patients with suspected MND have their diagnosis confirmed by a neurologist, with access to appropriate investigation resources and the specialist MND multidisciplinary team.

Essential Criteria
12.1 On request from a neurologist, patients with suspected MND have access to relevant investigation resources including imaging and neurophysiology within 20 working days.

12.2a The diagnosis of MND is confirmed and conveyed to the patient by a neurologist.

12.2b Patient review, after neurophysiology and imaging takes place, is achieved within 15 working days.

12.3a The MND regional care specialist makes contact with the patient within 2 working days of confirmation of the diagnosis.

12.3b Patients with MND and their carers are offered contact details for specialist support services provided by voluntary sector organisations.

Standard 13 Ongoing management of motor neurone disease

Standard Statement 13
Patients with MND and their carers are offered a wide range of support at all stages of their condition.

Essential Criteria
13.1a A needs assessment is offered to patients with MND at all stages of the patient's condition. The assessment is updated as the disease progresses.

13.1b A needs assessment is offered to those caring for people with MND at all stages of the patient's condition. The assessment is updated as the disease progresses.

13.1c At all times an identified individual is allocated responsibility for coordinating the delivery of care relevant to the assessed needs.

13.2a The service assesses and meets the communication needs of patients with MND at all stages of their condition.

13.2b The service assesses and meets the nutritional needs of patients with MND at all stages of their condition.

13.2c The service assesses and meets the respiratory needs of patients with MND at all stages of their condition.

Notes

Appendix D; Commonly Used Equipment		
Item Needed	**Assessor**	**Notes**
Bath Aids	Occupational Therapist (OT)	**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 Care Team Specialist for your area to access this service and ascertain if the equipment is supplied and in stock.
Beds (specialised)	District Nurse (DN)	
Bed Elevators	OT	
Car Seats (rotating)	OT	
Chair, glide about/shower	OT	
Chair, powered riser/recliner	OT , Physiotherapist	
Commode	DN	
Communication Aids	Speech Language Therapist (SLT) or SCTCI**	**** 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
Collars (specialised)	Physiotherapist	
Environmental Controls	Rehab Specialist, OT	
Feeding Aids	OT	
Hoists/Slings	DN	
Lifts/Stairlifts	OT	
Mobile arm supports	OT	
Page Turners	OT	
Pressure Care (Mattresses & cushions)	DN, OT	
Rails, Ramps	OT	
Splints	Physiotherapist, OT	
Suction Units	DN, SLT Physiotherapist	
Wheelchairs	OT, Local wheelchair assessment centre	

***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.

Appendix E; Bibliography & Further Reading

Bibliography

1. McDermott CJ, Shaw PJ, Diagnosis and Management of Motor Neurone Disease **BMJ 336 (March) 658-662**
2. Andersen PM. Borasio GD. Dengler R. Hardiman O. Kollewe K. Leigh PN. Pradat PF. Silani V. Tomik B. EALSC Working Group, Good practice in the management of amyotrophic lateral sclerosis: clinical guidelines. An evidence-based review with good practice points. EALSC Working Group **Amyotrophic Lateral Sclerosis 2007, 8(4):195-213,**
3. Miller. R.G., C.E. Jackson, E.J. Kasarskis, J.D. England, D. Forshew, W. Johnston,., et al "Practice Parameter Update, The care of the patient with ALS multidisciplinary Care," **Neurology 2009;73:1227–1233**
4. Miller. R.G., C.E. Jackson, E.J. Kasarskis, J.D. England, D. Forshew, W. Johnston,., et al "Practice Parameter Update The care of the patient with amyotrophic lateral sclerosis, drug, nutritional, and respiratory therapies (an evidence-based review) **Neurology 2009;73:1218–1226**
5. Bourke S C Tomlinson M, Williams T J, Bullock R E, Shaw P J, Gibson G J Effects of non-invasive ventilation on survival and quality of life in patients with ALS: a randomised control trial **Lancet Neurol 2006; 5: 140–47**
6. Skelton, J. Care of Chronic Conditions. Nursing Role in the Multidisciplinary Management of Motor Neurone Disease **British Journal of Nursing 2005; 14(1) 20-24**
7. Wijesekera LC and Leigh PN Amyotrophic lateral sclerosis **Orphanet Journal of Rare Diseases 2009, 4:3 doi:10.1186/1750-1172-4-3**
8. Oliver, D., Boarasio, G., Walsh, D, Palliative Care in Amyotrophic Lateral Sclerosis. 2nd Edition, Oxford University Press 2006
9. Heffernan C, Jenkinson C et al Nutritional management in MND/ALS patients: an evidence based review **ALS and other motor neuron disorders 2004 5, 72–83**
10. Giordana MT, Ferrero P, Grifoni S, Pellerino A, Naldi A, Montuschi A, Dementia and cognitive impairment in amyotrophic lateral sclerosis: a review **Neurol Sci DOI 10.1007/s10072-010-0439-6**
11. Mockford C., Jenkinson C., & Fitzpatrick R., A Review: Carers, MND and service provision; **Amyotrophic Lateral Sclerosis 2006; 7: 132–141**

Further reading for people with MND, families and carers

12. MND Scotland Patient Information File and Associated Factsheets
13. MOTOR NEURONE DISEASE, The At Your Fingertips Guide. Dr. Stuart Neilson and Dr Frank Clifford Rose. Class Publishing 2003
14. MOTOR NEURONE DISEASE, A Family Affair, 3rd Edition Dr. David Oliver, Sheldon Press 2011

Further reading for professionals:

15. MOTOR NEURONE DISEASE ASSOCIATION MND Resource File: A patient and carer centred .approach for health and social care professionals. 2000
16. BROWN, R. H., MEININGER, V. SWASH, M. Amyotrophic Lateral Sclerosis. Taylor & Francis 1999
17. AMYOTROPHIC: LATERAL SCLEROSIS Hiroshi Mitsumoto, Serge Przedborski & Paul H Gordon (Eds) Taylor & Francis 2006

Other Useful Publications/Clinical Journal Articles.

18. The use of Non-invasive Ventilation in the management of MND *http://guidance.nice.org.uk/CG105*
19. Guidelines for withdrawal of NIV *http://www.palliativedrugs.com/download/091209_combined_Withdrawing_NIV_Draft_Guidelines_Revised231009.pdf*
20. Rio A., Cawadas E., Nutritional advice and treatment by dietitians to patients with amyotrophic lateral sclerosis/motor neurone disease: a survey of current practice in England, Wales, Northern Ireland and Canada **J Hum Nutr Diet, 20, pp. 3–13: 2007**
21. Oliver D, Campbell C, Wright A, Palliative care of patients with motor neurone disease **Progress In Palliative Care 2007, 15(6), 285-293**

MND Scotland

MND Scotland is the only charity providing care, information and funding for research to benefit the Scottish MND community. We have a wide range of services for people with MND, their families, friends and carers.

Services We Provide

Specialist Care and Equipment Loan

Our Care Team and Equipment Loan Services work to ensure people with MND receive the best available support at the right time.

We have an MND Specialist Care Team who 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 client needs and offer specialist guidance for particular problems.

Our equipment loan and delivery service has over 300 specialist items to help people cope with a variety of challenges as their symptoms progress. The service is essential when delays occur in the provision of equipment by the NHS or local authority social work departments.

Small Grants

MND Scotland provides small grants of up to £500 per year to help defray exceptional costs that can come when living with MND. For further information on how to apply ask your MND Care Specialist about this scheme.

Information

Our information officer manages a collection of pamphlets, books, journals and DVDs about MND and related issues. These are available for anyone to borrow, particularly patients, their families and health and social care professionals. The information officer can also answer your queries by researching specialist sources.

Research Funding

MND Scotland provides funding to improve the lives of people with MND and support well conceived research projects.

Education

We run Family Information Evenings across the country to help family and friends find out more about MND. We also organise in-service education days for allied health professionals and study days for professionals who aim to develop specialist knowledge of MND to provide the best possible care.

Counselling

We train and coordinate a special team of volunteer counsellors covering the whole of Scotland. The aim is to help people come to terms with the emotional stress and pressure of living with a terminal condition and its aftermath.

Welfare and Benefits

Our Welfare and Benefits officer is available to offer advice and information about a range of financial and practical entitlements to help and support you.

Local Branches and Support Groups

Offer Support, raise awareness and provide a place for people affected by MND to come together and share support.

Other Services

We also offer other limited services, such as Befriending (Tayside and North Fife only) and some Complementary Therapies in areas where volunteers are available and willing to provide these services. To find out what is available in your area ask your MND Care Specialist or contact MND Scotland directly.

The MND Scotland National Office welcomes all enquiries and invites you to visit our library by appointment. .

.

Disclaimer

This booklet is not intended to be an exhaustive source of information on symptom control, nor is the medication suggested guaranteed to be effective or appropriate in all cases. Professional opinion must be brought to bear, particularly where there are co-morbidities. The decision to prescribe rests with the prescribing doctor or nurse, taking into consideration the needs, wishes, and susceptibility of the patient.

If you have any useful information or comments on this booklet please phone: 0141 945 1077 or email us with your concerns

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

Subject Index

Drug Index

MND Scotland Patient Information File

MND Scotland provides a free information file to every patient diagnosed with MND and in contact with their MND Scotland Care Team Member. The folder comes with factsheets 1, 23, 31 and 32 (underlined below) already inserted; the other available factsheets are listed in the table and all can be downloaded from our website at *www.mndscotland.org.uk/information*. It should be emphasised that not every MND patient will need all of this information. For those without internet access staff at MND Scotland can post out pre-printed factsheets on request.

1 Introductory	
1	What is MND?
1B	What is MND More Detailed
2	Inherited MND
2 Coping with MND	
23	Tools to Help You
27	Coping with MND
29	Telling Children About MND
3 Information and Support	
32	MND Scotland
31	Sources of Support
20	Library and Information
43	Useful Organisations
47	Recommended Reading
4 Caring and Carers	
42	Being a Carer
15A	Care at Home
15	Palliative Care
5 Money Issues	
38	Financial and Legal Issues
4	Benefits in Brief
46	Bereavement Benefits
49	Managing Your Money
6 Managing Symptoms	
33	Managing MND
39	Lithium
22	Riluzole
7 Physical Disability	
5A	Assistive Equipment
5B	Maintaining Mobility
5C	Getting the Best From Physiotherapy
5D	Aids to Maintain Mobility
19	Sex and Sexuality (in development)
21	Specialist Clothing
8 Research, Drugs and Trials	
26	Current Research
18	Clinical Trials
16	Stem Cells
28	Complementary Therapies
10	Creatine
11	Vitamins and MND
	Genetics
9 Genetics	
3	Introducing Genetics
3B	Disease Inheritance
10 Breathing and Bulbar Symptoms	
34	Adapting to Swallowing Problems
6	Feeding Tubes
36	Oral Health
35	Breathing Matters
40	Ventilation in MND
30	Communication Strategies
11 Motoring & Travel	
9	Travelling and Holidays
12	Travel Insurance
17	Motoring and MND
37	Long Distance Travel
12 Death, Dying and Bereavement	
8	Care Planning and End of Life Issues
14	Leaving a Legacy
24	Powers of Attorney
25	Guardianship and Intervention Orders
7	Bereavement and Children
44	Advance Directives
48	Medical Bequests
45	After a Bereavement
13	Coping after Bereavement

MND Scotland

For further information on our support or services contact us at

MND Scotland
Unit 2A, 6 Eagle Street
Craighall Business Park
Glasgow
G4 9XA
Tel 0141 332 3903
Email info@mndscotland.org.uk
Website: www.mndscotland.org.uk
Charity Number SC 002662
Company Number SC 217735