MOTOR NEURONE DISEASE

A PROBLEM SOLVING APPROACH FOR *health professionals*

mnd
Motor Neurone Disease NZ

Motor Neurone Disease New Zealand

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CONTENTS

Here to make time count

Motor Neurone Disease New Zealand (MND NZ) is a charity built on the ethos of helping people affected by motor neurone disease (MND) – and it remains the cornerstone of all that we do today.

We envision a world where no one faces motor neurone disease (MND) alone — where people are supported to live with dignity and hope, and where, one day, MND is treated, cured or prevented.

Until then, our mission is to make time count for people affected by MND by offering personal support; advocacy; education; and supporting research.

For more information visit: www.mnd.org.nz

PRESENTATION

Motor neurone disease

Motor neurone disease (MND) is an uncommon degenerative disorder of motor neurones which leads to progressive paralysis of cranial and skeletal muscles.

The onset is insidious. First symptoms may include stumbling, weakened grip, hoarse voice, difficulty speaking or swallowing, cramp or muscle wasting.

The condition is incurable and leads to death within a few years of diagnosis, generally 1–5 years. Death is most commonly due to respiratory muscle weakness and ventilatory failure.

- **Anterior horn cells – resulting in Lower Motor Neurone Lesions (LMN)**
- **Cortico Spinal tract cells – resulting in Upper Motor Neurone Lesions (UMN)**
- **Motor Nuclei in brain stem –resulting in weakness of 'bulbar' (speech and swallowing) and respiratory muscle function.**

Note

- Sensory lesions are rare.
- Some people with MND may experience cognitive changes, ranging from mild effects to noticeable impairment typical of Frontotemporal Dementia (MND-FTD)
- Cranial nerves affecting sight and lower sacral segments of the spinal cord affecting continence are usually spared, until late in the disease .
- Varied presentation and unpredictable, sometimes very rapid, disease progression.

Relationship of symptoms to lesions

Medulla	Upper Motor Neurone Lesion	Pseudo Bulbar Palsy (other causes including stroke)	• Tongue spasticity, no fasciculation • Spastic dysarthria • Increased jaw jerk • Emotional lability
	Lower Motor Neurone lesion	Bulbar Palsy	• Tongue – shrunken, wrinkled , fasciculating • Flaccid dysarthria – 'nasal speech' • Dysphagia, aspiration • Paralysis of diaphragm
Cortico Spinal Tract	Upper Motor Neurone Lesion		• Spasticity weakness • Increased reflexes, clonus • Extensor plantar responses
Anterior Horn cells	Lower Motor Neurone lesion		• Flaccid weakness • Muscle wasting fasciculation

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

Onset is insidious

Early symptoms may include: stumbling, foot drop, weakened grip, slurred speech, difficulty swallowing, cramp, shortness of breath, muscle wasting and/or tiredness. Some people may present with cognitive features including behavioural changes, emotional lability ("pseudobulbar affect", not related to dementia), or frontotemporal dementia.

Note: Some patients may present with acute respiratory problems.

Incidence 2-3:100,000 per annum
Prevalence 7-9:100,000 (approximately)
Average age of onset 58 years
Male/female ratio 3:2
Differential diagnosis Neurological investigations should normally include EMG, blood tests and investigations that sometimes includes MRI scanning to exclude possibility of other conditions.

Clinical Phenotypes

Amyotrophic Lateral Sclerosis (ALS)

- Involves both UMN and LMN.
- Characterised by: muscle weakness, spasticity, hyperactive reflexes, emotional lability, fasciculation, weight loss. The Bulbar region is usually affected but may not be prominent in everyone with ALS
- Average survival 2–5 years.

Progressive Bulbar Palsy (PBP)

- A small group of people with Bulbar-onset MND have symptoms relatively confined to the bulbar region for several months before it moves to the limbs.
- Both UMN and LMN may be involved.
- Characterised by rapidly progressive dysarthria and dysphagia.
- LMN damage: nasal speech, regurgitation of fluid via nose, tongue atrophy and fasciculation, pharyngeal weakness.
- UMN damage: spastic dysarthria, emotional lability.
- Affects older women predominantly.
- Average survival 6 months to 3 years.

Progressive Muscular Atrophy (PMA)

- Predominantly LMN degeneration.
- Characterised by: muscle weakness and wasting, weight loss, fasciculation.
- 'Flail arm syndrome" falls within this group characterized by slowly progressive, symmetrical and usually proximal upper limb weakness.
- Affects men predominantly.
- Average survival 5 years plus.

Primary Lateral Sclerosis (PLS) - Rare

- Affects UMN only
- Characterised by spasticity/brisk reflexes.
- Balance often impaired
- Longer survival 10 years plus.

Approximately 80-90% have ALS, the remaining 10-20% may have purely UMN or purely LMN or Purely Bulbar involvement. As the disease progresses there may be considerable overlap resulting in more generalised muscle wasting and weakness.

The motor nuclei controlling eye movements and the voluntary pelvic sphincter muscles remain intact.

AETIOLOGY AND TREATMENT

Sporadic

About 90-95% of cases have no evident family history of disease. These "sporadic" cases likely develop due to a combination of genetic susceptibility and environmental factors.

Familial MND

- About 5-10% of cases
- Mutations in several different genes can be detected in about three quarters of people with inherited forms of MND
- 45% of inherited cases are caused by gene expansion mutations in the gene C9ORF72
- Fewer than 20% are caused by mutations in the SOD1 gene
- Changes in the genes TDP43 and FUS are each responsible for 5% of inherited cases

Sporadic and Familial MND are clinically indistinguishable.

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

- Abnormal intracellular protein aggregation
- Glutamate-mediated excitotoxicity
- Oxidative stress
- Mitochondrial dysfunction
- Axonal transport dysfunction
- Neurotrophic factor dysfunction
- Glial cell dysfunction

The Role of the GP is *pivotal* in providing palliative treatments in response to rapidly changing needs

Riluzole is the only medication currently available for use in NZ and requires special authority. Not all patients will meet the criteria for prescription. A recent review suggests that riluzole treatment may extend survival by 6-19 months, which is far longer than that reported in the original clinical trials and points to benefits for both early intervention and prolonged treatment with riluzole.

Normal baseline blood tests, including full blood count and liver function tests, should be taken before prescribing. Full blood count and liver function tests should then be undertaken every month during the first 3 months of treatment, every 3 months during the remainder of the first year, and annually thereafter.

Possibilities for treatment

- There are a number of ongoing studies testing the efficacy of new drugs in the treatment of MND and existing drugs that are currently being used for other conditions
- There are a number of treatment options for symptom management- people should be encouraged to talk about the symptoms they have and the potential treatments available. Symptoms need to be responded to in a timely manner.
- MND demonstrates the need for multidisciplinary care including Respiratory, Gastroenterology and Palliative Care Services
- Unproven treatments – ALS Untangled (www.alsuntangled.com) is an international group of scientists and clinicians who investigate unproven or alternative treatments and conclude with their own recommendations.

DYSARTHRIA

80% patients are eventually affected.

Cause

Weakness and paralysis of the lips, facial muscles, tongue, larynx and pharynx resulting from affected trigeminal, facial, glossopharyngeal, vagus, accessory and hypoglossal cranial nerves.

Impact

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

Treatment

Early referral to speech-language therapist (SLT) who will:

- advise on strategies for communication and swallowing
- arrange for assessment and provision of communication aids

The occupational therapist (OT) and physiotherapist can assist with advice on:

- seating, positioning, wrist supports
- switches, 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" "no" answers; use signals for yes/no.
- Encourage to slow down and over emphasise words.
- Be wary of interruptions or trying to finish sentences.
- Try not to use family/carers to translate for the person with MND unless it is clear that this is what the person wants.

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 because misunderstood or opinion ignored or not sought**
- **increased sadness isolation and frustration felt by patient, carer and family.**

Communication can be aided by:

- writing
- alphabet board
- perspex eye pointing frame (ETRAN BOARD)
- hands-free telephone
- call bells
- personal alarms
- computerised communication aids, e.g iPad, tablet

Check with the **speech-language therapist (SLT)** or **occupational therapist** for up-to-date information and advice on communication aids.

The **TalkLink Trust** (Assistive Communication Solutions) provides specialised assessment and equipment to people with disabilities throughout New Zealand. There are offices in Auckland, Wellington and Christchurch.

DYSPHAGIA AND NUTRITION

Cause

Weakness and paralysis of the bulbar (tongue, facial, and pharyngeal) muscles resulting from affected glossopharyngeal, vagus, accessory and hypoglossal nerves.

Effect

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

Resulting in

- Drooling. In most patients, saliva problems are the result of poor lip seal and/or impaired ability to swallow.
- Dehydration and weight loss.
- Aspiration and recurrent chest infection.

Management

Assessment and monitoring should start at the time of diagnosis. Optimum management of concurrent symptoms is a delicate balance requiring frequent review and fine-tuning.

- Optimise oral hygiene monitoring by nurse
- Assess eating and monitor swallow consult **speech-language therapist (SLT)**
- Assess nutritional intake and weight consult **dietitian**
- Head support and positioning consult **physiotherapist and/or occupational therapist (OT)**
- Assess the person's ability to swallow and protect their airway during eating. Teach safe swallow and assisted cough techniques **speech language therapist and physiotherapist**
- Treat gastro-oesophageal reflux common in MND. May aggravate/confuse secretion management
- Additional support will be required for people with dysphagia and cognitive change who may need reminders about safe swallowing and require supervision.

Saliva problems

Sialorhoea (drooling) is due to difficulty managing the normal flow of saliva and will be exacerbated by a poor lip seal. Advice should be sought from SLT on swallowing, diet, posture, positioning, oral care and careful suctioning where required (over use of suction machines can cause inadvertent salivary gland stimulation).

Optimise non-drug management first.

- correct positioning and head and neck support (physiotherapist)
- -Avoid skin irritation-promote "wipe not dab" (can cause inadvertent salivary gland stimulation) and Consider bibs/ clothing modifications
- oral swabbing and syringing

Reducing the volume

- Consider a trial of anticholinergic medicine as the first-line treatment for sialorrhea. Tailor dose for each patient and monitor closely for side effects. "PRN' dosing may be more effective than regular dosing which may cause tenacious saliva and excessive dryness, and the drying effect can be lost over time. If one anticholinergic medication is ineffective, switching to another anticholinergic medication may be considered. Be aware that anticholinergics can cause confusion in the elderly.
  - Tricyclic antidepressants: **nortriptyline** or amitriptyline at night, starting at lowest doses.
  - **Hyoscine hydrobromide transdermal patch** (NB. central nervous side effects)

Dysphagia affects about two thirds of all patients eventually.

- Hyoscine butylbromide, orally or via PEG/RIG. This has fewer central side-effects.
- Atropine eyedrops can be used as a third-line agent for sialorhoea, but specialist advice should be sought prior to using, as this is an unlicensed indication with potential adverse side effects

- Sialorrhea medication options in advanced disease towards end of life: Glycopyrrolate - can be given via the subcutaneous route. Hyoscine butylbromide - can be given by the subcutaneous route. Both of these medications have less central nervous system penetration and thus are preferable in people with motor neurone disease who have cognitive impairment.
- If first-line treatment for sialorrhea is not effective, not tolerated or contraindicated, consider referral to a specialist service for Botulinum toxin, if available
- The effect of Botox wears off over time. Medication interface may be needed with onset and fading of the impact of Botox
- An alternative to Botox is radiotherapy but the effect is permanent, , and this is only suitable in rare, individual cases.
- Becoming NBM will decrease saliva flow

If a person with motor neurone disease has thick, tenacious saliva:

- Review all current medicines, especially any treatments for sialorrhea

Provide advice:

- Swallowing
- Diet avoid lactose products replace with Almond, soy coconut milk etc
- Posture & positioning; use of tip and tilt chair
- Oral care
- Suctioning and hydration & treatment with humidification, nebulisers and carbocisteine.
- Consider mucolytic agents Pawpaw, papain in E-ZY MEX soda water, dark grape juice, pineapple juice

Vigilant oral cares include the following considerations

- Oral lubricants-Orolube, grapeseed oil and flavouring Biotene moisturising gels (Caution with sprays)
- Low froth tooth pastes, tooth brushes with in build suction or 2 sided bristles
- Dentist appointments (consider inability to flat lie request appropriate positioning perhaps remain in tip and tilt wheelchair) and management of gingivitis
- Consider forward leaning to avoid toothpaste aspiration.
- Impact of medication;eg antibiotics linked to oral and pharyngeal thrush
- Hypo or hyper reflexes (gag reflex supressed or active bite reflex)

Attention to airway protection

- As above
- Safe swallow techniques, breath stacking, Cough Assist Machine use (CAM)
- Surgical intervention - tracheotomy or laryngectomy considered in context of risk. Consideration needs to be given to quality of life and Advance Care planning preferences

Nutrition

Monitoring of weight and nutritional advice is essential for people with MND. Many will need detailed information about adaptation of their diet and may need prescribable food supplements and thickeners. The involvement of a dietitian as part of the multidisciplinary team is therefore very helpful.

Percutaneous Endoscopic Gastrostomy (PEG) should be considered before the effort of eating becomes exhausting, food and fluid intake is inadequate, there is danger of aspiration and/or fear of choking. Recent studies suggest feeding tubes should be placed at approximately 5% of weight loss and before 10% if they are used.

Not every patient will choose this type of intervention, but it is important to offer and discuss the option in good time to achieve optimum benefit. The risks of the

PAIN

procedure increase as respiratory function declines. A combination of oral (for pleasure) and enteral feeding (for fluid and calories) is possible with a PEG in situ.

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 and abdominal muscles. As in other debilitating conditions, constipation may cause symptoms such as pain, nausea and vomiting before it is acknowledged – even by the patient.

Management

Anticipate

- Optimise fluid and fibre intake. **Consult dietitian.**
- **Diet** e.g. prunes, kiwifruit, linseed. (PEG supplements vary in fibre content.)
- **Regular oral laxatives**, usually a combination of bowel stimulant and fecal softener - titrate up cautiously e.g. Codalax or Coloxyl with Senna. Increase dose and frequency as required. Start with Codalax 10 mls or Coloxyl with Senna 2 tablets at night.
- Use of **softeners and stimulants** may help- titrate up cautiously
- Faecal softeners if stool is hard - Docusate sodium (softener),. Macrogols (no longer require a special authority.)
- **Bowel stimulants** if bowel transit is slow eg senna,bisacodyl
- **Liquid versions are available** that can be given via a feeding tube.
- **Rectal laxative** regular suppositories (e.g. bisacodyl and Glycerin) may be required in advanced disease or to suit the practicalities of care at home.

Causes

- loss of muscular control to stabilise large joints and maintain spinal posture
- passive injury to joints when controlling muscles are weak, e.g. shoulder joint damage during assisted transfers
- muscle cramps
- spasticity
- skin pressure
- constipation

Treatment

Physiotherapy assessment and advice is important. By identifying and addressing causes of pain and aggravating movements (active or passive) or postures, the physiotherapist may be able to reduce the need for drugs as well as improving comfort.

Muscle symptoms

Muscle Weakness

- Can affect balance and posture and increase risk of falls
- Mobility and Activities of Daily Living are affected by muscle weakness
- Consider – Physiotherapy – walking aids, splints, active and passive exercises, posture management
- Occupational Therapy – equipment for ADL's, posture management, wheelchair and seating

Fasciculation ("twitching")

This is part of the disease process and almost never intrusive – although often noticed by patients, it cannot easily be treated.

Cramps and spasticity

These are often bothersome and may be disabling in some patients, often earlier in the disease course.

Affects up to three quarters of MND patients at some time. Pain and discomfort may arise from complications of muscle weakness, stiffness, immobility and trauma.

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

- Baclofen can be useful for cramps. Use smaller doses in frail patients.
- If baclofen is ineffective, not tolerated or contraindicated, try gabapentin
- Consider benzodiazepines (e.g. clonazepam) or mexiletine, as a 3rd line treatment
- NB Mexiletine needs monitoring of ECG before and 1 month after reaching stable dose due to potential changes to QT interval.

Patients seldom tolerate more than small doses of benzodiazepines during the day because of unwanted drowsiness and these may be more helpful at night. NB observe for respiratory depression with sedatives in patients with respiratory muscle weakness.

Joint stiffness

Physiotherapy:

- Careful positioning to relieve discomfort
- Passive exercise programme
- Prevention of contractures
- Maintenance of joint mobility.

Start with simple analgesia such as Paracetamol. Non-steroid anti-inflammatory drugs (oral, rectal or topical) are helpful if there is inflammation. Consider single daily slow release preparation. Intra articular injection of steroid will benefit some people.

Skin pressure

Good skin and pressure care is vital. Liaison between nurse and occupational therapist will identify appropriate pressure-relieving management and equipment.

Analgesic drugs

The majority people with MND at some point need medication for relief of pain and/or respiratory symptoms. As in any other situation, dose should be carefully titrated to effect to ensure adequate analgesia and avoid unnecessary side effects, and palliative care specialist is recommended where available. Individual requirements vary widely. Remember to prescribe a laxative. The WHO Analgesic Ladder is applicable:

Step 1 non-opioid Paracetamol

Step 2 mild opioid e.g. codeine, Tramadol (not in elderly)

Step 3 strong opioid Morphine (oral, per gastrostomy, SC, PR) Fentanyl (transdermal, SC – *NB. Opiods can have unwanted side effects and require careful titration as individual requirements vary widely. Due to the complex and individual nature of MND, and risks of respiratory compromise, initiation and titration of opiates should only be done by a physician confident with prescribing opiates in this group of patients. Otherwise consultation with a palliative care physician is recommended.)*

Oedema

May be related to restricted activity and posture, or to DVT or other intercurrent disease.

Treat any specific causes

Attention to posture and seating

Physiotherapy, compression hosiery

- Diuretics are rarely helpful as they can promote urinary urgency and Risk of dehydration & electrolyte disturbance.
- In some areas, referral to the lymphodema service may be an option.

DYSPNOEA AND CHOKING

Shortness of breath is a common, and often frightening, symptom at some stage in the course of MND. Usually, respiratory muscle weakness occurs late in the disease and is the most likely cause of death. However ventilatory failure can develop at any stage and occasionally is the presenting feature of MND.

Choking attacks may be due to aspiration, impaired respiration or muscle spasm. It should be stressed that **death caused by choking attacks is very rare and that the final stages of MND are usually peaceful and dignified.**

Cause of Dyspnoea

Weakened (i.e. denervating) respiratory muscles – intercostals, diaphragm, abdominals.

Aggravated by dysphagia, gastro-oesophageal reflux, weak neck muscles, chest infection, general debility and fear.

Effects

- Early in disease nocturnal hypoventilation may cause Increased CO2 levels resulting in disturbed sleep, nightmares, and headaches, especially on waking.
- Breathlessness on exertion
- Fear, anxiety and panic.
- Orthopnoea
- Fatigue and reduced functional ability.
- Quiet voice; weak cough-Inability to expectorate lung secretions –
- Hypercarbia +/- hypoxia and possible confusion.

Management

Respiratory Specialist assessment and advice	Required if respiratory symptoms or signs of sleep disturbance develop. Respiratory physician may initiate NIV once nocturnal hypoventilation is detected so early referral is essential
Monitoring	Regular clinical history (exertional dyspnoea, orthopnoea, sleep disturbance) and examination (dysphonic voice, weak cough, chest) and lung function testing (every 2-3 months) including sleep studies
Anticipatory care	Discuss treatment options early in disease process. Plan ahead for management of acute breathlessness (e.g. establish out-of-hours medical/nursing contact, explain and prescribe drugs). Refer to palliative care for additional strategies and use of medications for dyspnoea
Relieve fear and anxiety	Explanation, exploration of fears, assurance of ongoing support, +/- medication. Relaxation therapy and complementary therapies
Occupational therapy	Advice on positioning, specialist equipment
Physiotherapy	Breathing exercises, chest physiotherapy, monitoring – ↘ Unassisted breath stacking and /or manual assisted cough ↘ Assisted Breath stacking (using a lung volume recruitment bag with a 1-way valve) for those with bulbar dysfunction or whose cough is ineffective with the unassisted breath stacking.

Use of mechanical insufflation/ exsufflation machine (cough assist)	if assisted breath stacking is not effective and /or during respiratory tract infection
Speech therapy	Speech-Language Therapists are skilled in the assessment and management of dysphagia and may identify a treatable cause of dysphoea during meals.
Ventilatory support	Non-invasive, BIPAP mask used overnight. Cumbersome. Significant symptom relief in selected patients. Tracheostomy and invasive ventilation rarely used in New Zealand. Psychosocial and ethical implications.
Advanced directive	Helpful to document patient's wishes re ventilation for acute event, and end-of-life decisions, although not legally enforceable in New Zealand.

Refer to palliative care for additional strategies and use of medications for dyspnoea.

Oxygen therapy needs to be used with **great caution due to the possibility of CO2 retention leading to reducing LOC and ultimately death.** This should be prescribed by a specialist team.

Choking

- Choking attacks may occur due to aspiration, impaired respiration, muscle spasm or stridor
- Consult physiotherapist for assisted cough techniques and breath-stacking, SLT for secretion control and palliative care for medications that may assist (eg clonazepam, morphine)
- Provide advice to patient, carer & whanau about what to do during a choking attack and when to seek medical advice

Encourage the person to stay calm and wait for the attack to pass

- Encourage the person to focus on slow and steady breathing
- Open a window to give the feeling of air on the face
- Assist the person to the most upright position possible
- If the person has been prescribed medication to help manage choking episodes, use it
- Encourage the person to cough and then swallow their saliva if possible
- Use suction equipment as appropriate, and only if trained to do so

Cognitive Change

There is now increased awareness of cognitive and behavioural changes in MND. People with MND may fall into one of the following groups.

- Those unaffected by cognitive change- about 50%; this decreases to 20% in the final stages of the disease
- Those with mild cognitive change with deficits in executive functions, language, behavioural and / or social cognition – about 35%
- Those with Frontotemporal Dementia either at the same time or after diagnosis of MND, up to 15%

Up to 15% of those presenting with Frontotemporal Dementia go on to develop motor impairments with MND diagnosed after dementia. if there is a concern about cognition or behavior:

- Explore these areas with the person and their family.
- Undertake a formal assessment from a neuropsychologist using ECAS

Documenting cognitive involvement is important. Patients and families should be encouraged to ensure a Power of Attorney is appointed

PSYCHOLOGICAL SUPPORT

People with MND, their families and carers often suffer considerable psychological and emotional distress. **Much can be done to alleviate this 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 the diagnosis

Anxiety can increase as a result of:

- onset of worrying symptoms
- difficulty in identifying cause
- a protracted period of investigation
- the need to be sure before giving the diagnosis.

Telling the diagnosis

Preparation

- Taking time to convey the potential seriousness of diagnosis and prognosis

Time and place

- Quiet, relaxed, private and away from external distractions
- Choice of who will be present – partner/family members

Amount of information

- Initial shock often limits the ability to absorb information
- Need the doctor to give written information, e.g. leaflets from Motor Neurone Disease NZ and contact number for the organisation: 09 624 2148.
- Need opportunities to return for more information and to set own pace
- Need an identified person to provide ongoing support e.g. GP/Motor Neurone Disease NZ support advisor.

Telling the truth

- Honesty is important but avoid leaving the person feeling alone and unsupported
- Prolonged uncertainty can exacerbate fear of the unknown, deny support and the opportunity to come to terms with mortality and make decisions
- Telling relatives in isolation puts strains on relationships and families

Families and carers

MND forces changes in roles and relationships. Consider:

- balancing and giving time to needs of other family members
- meeting the varying information needs of all family members
- counteracting isolation of individuals and promoting awareness of each other's needs
- offering early opportunities for short periods of respite to prevent over dependence on a single carer
- creating opportunities for expressing negative feelings without feeling guilty
- physical exhaustion from the caring role exacerbated by powerlessness to prevent suffering and further deterioration.

Emotional reactions

Fears may include:

- 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 alleviate some of these.

Denial is a coping mechanism that sometimes operates alongside awareness of the condition and its implications.

Anger can occur at any stage of the illness.

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

Depression is not always easy to differentiate from sadness, but treatment is likely to have a positive effect.

Emotional lability (as opposed to low mood) is associated with upper motor neurone involvement and can be very distressing for patients and carers. Amitriptyline usually helps this symptom. Start with 10 mg nocte, warn about side effects such as dry mouth, sedation. Be aware that anticholinergics can cause confusion in the elderly.

Adjustment or coming to terms with the disease is made more difficult by the rate of deterioration and the accompanying changes and losses.

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

Check out your own area which may have a vital local support group.

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.

Some areas have devised specific support manuals which provide compendiums of information with the focus on local help and support available.

PALLIATIVE AND TERMINAL CARE

Palliative care is a critical component of the care of people with MND. It is beneficial to introduce palliative care as part of the MDT at an early stage. As this is a specific area of expertise, consultation with local palliative care services is recommended. MND NZ is pro-active in the involvement of these services and can assist in making the community referral where appropriate.

Coordinating Care

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 carers.

Access to the skills and expertise available from a variety of agencies in providing:

- assessment of needs
- rapid response
- co-ordinated action
- regular review.

The multidisciplinary team

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

A key worker should be identified who will:

- co-ordinate the activities of other team members
- alert team members and initiate effective and timely response to changing needs
- link with other service providers
- become the first point of contact for the family.

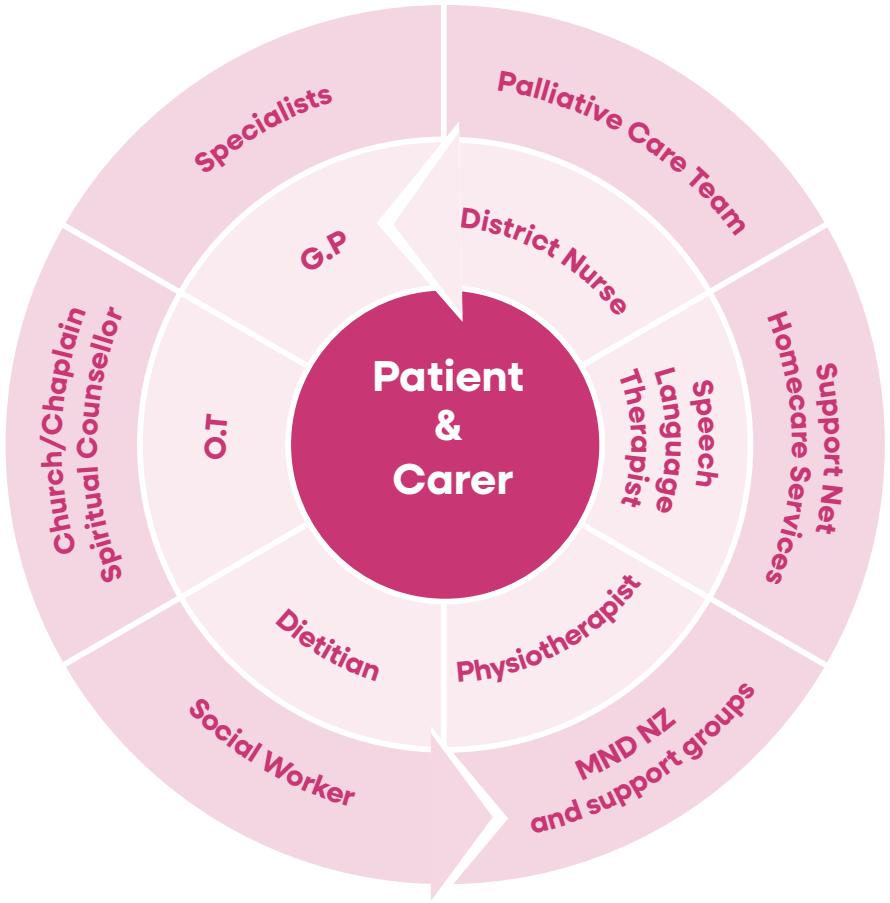
Anticipating possible changes and needs depends on:

- building and maintaining trust
- confidence in a rapid response to requests for help
- communication within the team and effective delegation of responsibility

General Practitioners rarely see a case of MND

It is estimated that a single GP practicing in the community will see, on average. 1 new patient with MND every 10-15 years. Therefore, the role of the allied health care team is critical in providing an experienced multidisciplinary service in caring for people with MND and is a great source of information and advice about the latest equipment and management techniques available. This publication aims to outline some of the particular problems encountered by people with MND and to provide signposts to sources of help.

EQUIPMENT



This concept is patient-centred and relies on effective communication between all participating agencies and individuals. Timely and cohesive team management is vital. Key player focus will change with the progression of the disease.

Equipment can be sourced privately, via the OT/SLT etc, or through the public health service. TalkLink and Enable New Zealand are useful services.

Before decisions are made regarding specific equipment, it is essential that accurate assessment be carried out by an Occupational Therapist, Physiotherapist, Speech-Language Therapist or Nurse, depending on the area of expertise and the availability of professionals in any particular locality.

Accessable administers equipment and modification services in the Auckland and Northland regions (from Meremere north), and Enable New Zealand administers EMS in the rest of the country. Enable NZ provides information on services, equipment and housing modifications for people with disabilities. They are the "who, how, where and when" of funding and services. Phone 0800 171 981 for information, 0800 171 995 for funding. www.enable.co.nz

Equipment which can be helpful for people living with MND includes the following:

Eating utensils

- modified cups, plates and cutlery
- non-slip mats
- cup holders
- collars
- splints

Pressure care

- cushions
- mattress overlays

Grooming aids

- Velcro for assistance with clothing including shoes
- backless blouses/shirts with velcro collars instead of adult bibs

Chairs and beds

- reclining
- adjustable height
- electrically operated for individual comfort
- modification of existing furniture

Mobile arm supports

- wheelchair attached
- free standing or table attached

Aids for carers

- turntables
- hoists
- transfer belts

Environmental controls

- personal alarms
- remote operation of lights/ electrical equipment
- book rest
- page turner
- talking books
- wheeled trolley

Mobility aids

- walking sticks
- walking frame
- rails/ramps
- splints
- wheelchairs – electric or manual, and may be carer or patient operated
- orthotics

Communication

- hands-free telephone
- call bells
- personal alarms
- computerised communication aids – ipad, tablet
- light touch key boards
- voice synthesisers
- 'magic' slates
- amplifiers

Bathroom aids

- grab rails for bathroom and toilet
- chairs
- shower
- over toilet frame
- commode

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RESOURCES

Motor Neurone Disease New Zealand

is a charitable organisation that aims to bring together all people concerned with MND including people with the disease, their carers and professionals. Its objectives are to:

- Co-ordinate and provide up-to-date information
- Provide emotional, social and practical support
- Advocate for the needs of people living with MND
- Develop awareness and understanding of MND
- Encourage research

A number of information booklets, books and videos are available from Motor Neurone Disease NZ. The website www.mnd.org.nz has answers to some of the questions commonly asked by family, friends and carers, and has links to more medically detailed sites. Newsletters are published regularly.

The MND NZ Support Service provides support for people with MND, their family, whānau and the health professionals working with people with MND. There is more information about the service they provide and how to contact them on the MND NZ website: www.mnd.org.nz/support-information

Financial help

GPs and social workers liaise re invalid and disability benefits. Needs assessment is required prior to personal and home help. Clients may be eligible for a community service card and a high-user card for medication. Personal circumstances will dictate eligibility. Information on allowances is provided by WINZ. Tel: 0800 559 009; website: www.winz.govt.nz

Useful websites

Motor Neurone Disease NZ www.mnd.org.nz

MND Australia www.mndaust.asn.au

MND England, Wales and Northern Ireland www.mnda.org

ALS Untangled www.alsuntangled.com

ALS News Today www.alsnewstoday.com

International Alliance www.alsmndalliance.org

World Federation of Neurology www.wfnals.org

Advance Care Planning www.advancecareplanning.org.nz

End of Life Services www.endoflife.services.govt.nz

myTube (nutrition/feeding tubes information) www.mytube.mymnd.org.uk

myBreathing (ventilation information) www.mybreathing.mymnd.org.uk

The MND Registry

The New Zealand MND Registry provides an important picture of MND in New Zealand. This confidential information from patients with MND helps to answer questions about how many people have MND in different areas, how the condition progresses, and how the disease can affect people. The MND Registry also helps MND research grow in New Zealand through access to anonymised patient data and by informing people who wish to be involved in research about research projects that they would be suitable for.

www.mnd.org.nz/research/mnd-registry

Email: mndregistry@otago.ac.nz

Phone: 0800 MND REG (0800 663 734)


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