GUIDELINES FOR
THE PHYSIOTHERAPY MANAGEMENT
OF MOTOR NEURON DISEASE (MND)
September 2014
1ST EDITION
MND GUIDELINE DEVELOPMENT GROUP
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Preface

This is the first edition of the Irish *Guidelines for the physiotherapy management of Motor Neuron Disease (MND)*.

Recent publications provide evidence based guidelines for overall management of MND (Phukan and Hardiman, 2009: Anderson et al., 2012). However, there are no guidelines available, from Ireland or internationally, that specifically inform physiotherapists in their management of this patient group. These guidelines provide a framework of care for the patient with MND, and are intended to be practical and relevant for all physiotherapists encountering a patient with this condition.

The guidelines are the culmination of over two years work by Geraldine O’Callaghan, a senior physiotherapist who received a bursary from the Irish Hospice Foundation, Deirdre Murray, Roisin Vance, and a small number of other clinicians who freely gave of their time to contribute. We hope that the guidelines reflect the commitment and expertise of these clinicians.
Definitions

Motor Neuron Disease (MND) encompasses several different conditions whose common feature is the degeneration of upper and/or lower motor neurons. Nearly 90% of patients with MND have a mixed form of both upper and lower motor neuron degeneration, termed Amyotrophic Lateral Sclerosis (ALS), therefore the terms MND and ALS are used interchangeably, commonly meaning the same thing. As the 10% of cases not falling under the term ALS show specific symptoms, within context of this paper, the term MND is used to reflect the wide array of symptoms observed, and the variety of management approaches required.

Peak Cough Flow (PCF) is the velocity of air being expelled from the lungs during a cough manoeuvre. PCF is measured using a peak cough flow meter.

Non-invasive Ventilation (NIV) is a form of breathing support which does not require intubation or tracheostomy. In MND patients it is usually prescribed in the form of nocturnal non-invasive positive pressure ventilation (NIPPV) which is a bi-level positive pressure form of ventilation and aims to reduce work of breathing and assist with the symptoms of respiratory failure.

A Manually Assisted Cough involves the application of an abdominal thrust or costal lateral compression using various hand placements after an adequate spontaneous inspiration or maximum insufflation.

Maximum Insufflation Capacity (MIC) is the maximum volume of air within the lungs beyond spontaneous vital capacity. MIC is attained when a patient takes a deep breath, holds this breath by closing the glottis, and then performs another inspiration. This is repeated until the maximum lung capacity is achieved.Breathstacking using a bag valve mask, glossopharyngeal breathing or non-invasive ventilation (NIV) may be used to achieve MIC. Where the patient cannot maintain a closed glottis, a one-way valve is used for breathstacking, or alternatively a single insufflation is performed using non invasive ventilation (NIV).

Mechanical In-Exsufflation (MI-E) (Cough Assist) is a unit that applies a positive pressure to the airway achieving maximum insufflation, and then rapidly shifts to a negative pressure.
This rapid shift in pressure produces a high expiratory flow rate from the lungs, simulating a cough.

A **Muscle fasciculation** is a small, involuntary muscle contraction and relaxation, which may be visible under the skin.

**Dysarthria** is a motor speech disorder and is characterised by poor articulation of words.

**Dysphasia** is a partial or complete impairment of receptive or expressive communication.

**Dysphagia** is a term used for describing difficulty swallowing.

**Sialorrhea** defines an excessive production of saliva.
**Key recommendations**

This section summarises the key recommendations presented in Chapter 3.

<table>
<thead>
<tr>
<th>Number</th>
<th>Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>3.1.1A</td>
<td>Patients with MND should be seen as a priority for assessment, and should be offered regular monitoring and review.</td>
</tr>
<tr>
<td>3.2.1A</td>
<td>Effective channels of communication and co-ordination are essential within and between the tertiary, primary and palliative care multidisciplinary team, the patient and the family.</td>
</tr>
<tr>
<td>3.2.1B</td>
<td>Physiotherapists should use all resources available to them for advice and information, and maintain regular contact with each other (phone, e-mail or letter) throughout the course of the patient’s disease.</td>
</tr>
<tr>
<td>3.3.1A</td>
<td>Goals should involve the person and carer, be focused at the level of activity / participation, be timed with short term and long term aims and occur as part of multidisciplinary team planning where appropriate.</td>
</tr>
<tr>
<td>3.4a.1A</td>
<td>Frequency of input should be determined on an individual basis. However, patients should be able to access services urgently when the need arises.</td>
</tr>
<tr>
<td>3.4b.1C</td>
<td>Passive movements (PMs) should be applied to joints that patients cannot move themselves.</td>
</tr>
<tr>
<td>3.4b.1D</td>
<td>Stretching and range of movement exercises form the basis for spasticity management.</td>
</tr>
</tbody>
</table>
3.4b.1G Individualised strengthening exercises during the early stage of MND is probably effective in improving the function of patients and should be recommended.

3.4b.1H Supported treadmill training may be a useful modality in the early stages of MND.

3.4b.1J Active/Passive trainers or unloaded cycling should be considered, ideally for home use, where spasticity is present.

3.4b.1L Timely provision and regular review of aids and appliances is essential in maximising the patient’s function.

3.4b.1O Positioning of the patient in a good alignment while sitting or supine can decrease abnormal muscle tone; prevent or reduce spasticity, contractures, and pain; prevent the development of pressure sores; and minimise the effort required for respiration.

3.4b.1R The physiotherapist should complete a manual handling risk assessment and provide education on handling to carer(s) so that assisted exercises, transfers or mobility are performed at an acceptable level of risk.

3.4c.1S Physiotherapists should monitor closely for signs and symptoms of respiratory insufficiency.

3.4c.1T Peak cough flow should be measured regularly in patients with MND.

3.4c.1V Respiratory adjuncts such as PEP and acapella are not indicated to assist secretion clearance in this patient group, instead manually assisted coughing should be used, and family/carers educated in its technique.

3.4c.1W Facilitating maximum insufflation via breathstacking (using a bag valve mask or a positive pressure device (NIV)), or through the use of a mechanical in-
exsufflator (Cough Assist), can assist in mobilising secretions, maintaining chest wall and lung compliance.

3.4d.1Z Treatment of pain in MND is similar to that for any condition of musculoskeletal origin.

3.4d.1AA Early training on correct manual handling, shoulder care and range of movement exercises can minimise shoulder pain, a common problem in MND.

3.4d.1CC Corticosteroid injections may be indicated to relieve pain where conventional therapy has been unsuccessful. These injections can be accessed through the general practitioner or at the tertiary clinic, should be implemented early, and can be repeated.

3.4e.1FF Physiotherapists should be aware that NIV improves symptoms of hypercapnia in patients with MND, and should refer as appropriate.

3.5.1A The clinician should assure the patient / caregiver of their role as an advocate.

3.6.1A The physiotherapist should avoid providing insufficient information, delivering information callously, taking away or not providing hope.

3.7.1A Clients with MND should not be discharged from tertiary or community physiotherapy services until a transfer of care is made to the palliative care physiotherapist (if appropriate).
Acknowledgments

The Guideline Development Group wishes to acknowledge the funding provided by the Irish Hospice Foundation, which enabled the production of the 1st edition of Guidelines for the physiotherapy management of Motor Neuron Disease.

We would like to thank the following people for reviewing the guidelines and helping to structure the final script:

**Professor Orla Hardiman**, Director of the National Motor Neurone Disease Clinic in Beaumont Hospital, in particular for her input into the sections on background, model of care and medical management.

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**Karen Sayers**, Senior Physiotherapist in Primary Care and Continuing Care, Cashel, Tipperary.

**Angela Ryan**, Senior Physiotherapist in Palliative Care, Milford Hospice, Limerick.

I would also like to acknowledge Antoinette Brady, Physiotherapist, St. James’s University Hospital, Leeds, for performing literature searches.
Conflicts of interests

There were no conflicts of interest declared by any member of the guideline development group.
1. AIMS OF GUIDELINES

1.1 Guideline Objective:

The goal of these guidelines is to provide best practice evidence based recommendations for the physiotherapy management of patients with MND, ensuring that people with MND are provided with the best possible care, thus maximising functional ability and symptom management.

1.2 Applicability:

These guidelines have been developed for the management of Motor Neuron Disease (MND) by physiotherapists, primarily in Ireland, but many of the recommendations will be applicable in other countries and settings.

1.3 Intended Users:

These guidelines target physiotherapists working with MND patients across a range of settings including primary, tertiary, and palliative care settings. A guideline cannot cover every eventuality, and new evidence is published every day so parts of these guidelines will inevitably become outdated. Thus, the recommendations should be taken as statements that inform the clinician, not as rigid rules. The clinician is responsible for interpreting the guidelines, taking into account specific circumstances and considering new evidence that might exist.

The stakeholders who provided input to the guidelines include physiotherapists in tertiary, primary and palliative care services and a consultant neurologist.

1.4 Search Strategy:

Searches were carried out, from July 2011 through February 2013, in the Cochrane Database of Systematic Reviews; Cochrane Central Register of Controlled Trials; MEDLINE (Ovid); PUBMED; CINAHL- EBSCO; EMBASE (Ovid); The Physiotherapy Evidence database
(PEDro); Irish Motor Neurone Disease Association (IMNDA); Motor Neurone Disease Association (MNDA); National Institute for Health and Clinical Excellence (NICE) guidance published; and ALS Association using the following search terms: ‘amyotrophic lateral sclerosis’ or ‘motor neuron(e) disease’ or ‘motoneuron(e) disease’, combined using AND with ‘physiotherapy’ or ‘physical therapy’ or ‘rehabilitation’ or ‘multidisciplinary’ or ‘interdisciplinary’; AND with ‘guidelines’; ‘exercise’; ‘fatigue’; ‘spasticity’; ‘resistance training’; ‘cramps’; ‘pain’; ‘cognitive impairment’; ‘palliative care’; ‘respiratory impairment’; breathing exercises’; ‘cough-assist’; ‘non-invasive ventilation’; ‘NIPPV’; ‘mechanical insufflation/exsufflation’; ‘invasive ventilation’; ‘medical management’ and ‘drug therapy’.
2. INTRODUCTION

2.1 Background:

Motor Neuron Disease (MND) is a rare but fatal syndrome characterised by signs and symptoms of degeneration of upper motor neurons (UMN) and lower motor neurons (LMN) in the cerebral cortex, brainstem and spinal cord, leading to progressive weakness of bulbar, limb, thoracic, abdominal and respiratory muscles. Other brain functions, including oculomotor and sphincter functions, are relatively spared. ALS has an incidence of 0.6 to 2.6 per 100,000 of the population, and a mean onset of 47-52 years in familial cases, and 58 – 63 years in sporadic cases, but can occur at any age. Male sex, increasing age and hereditary disposition are the main risk factors (Anderson et al., 2007; Phukan and Hardiman, 2009).

The European Federation of Neurological Sciences (EFNS) task force on diagnosis and management of ALS (Anderson et al., 2012) recently published updated guidelines on the clinical management of amyotrophic lateral sclerosis. These guidelines, along with a paper outlining the management of amyotrophic lateral sclerosis (Phukan and Hardiman, 2009) are referenced extensively throughout sections 2.1, 2.2 and 2.3.

MND is a heterogenous syndrome that overlaps with a number of other conditions making early diagnosis difficult. As there is no single investigation specific to MND, the diagnosis is based on presentation, a thorough clinical examination, electrodiagnostic tests, neuroimaging and laboratory studies. The revised El Escorial research diagnostic criteria with the Awaji electrodiagnostic algorithm included, summarised in Table 2.1.1, is believed to be sufficiently sensitive to facilitate early diagnosis (Anderson et al., 2012). The clinical features of ALS and its variations, under the umbrella term Motor Neurone Disease (MND), are described in Table 2.1.2 (Phukan and Hardiman, 2009).
Table 2.1.1. The revised El Escorial research diagnostic criteria for ALS with the Awaji electrodiagnostic algorithm (Anderson et al., 2012)

<table>
<thead>
<tr>
<th>Category</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinically definite ALS</td>
<td>UMN and LMN clinical signs or electrophysiological evidence in three regions</td>
</tr>
<tr>
<td>Clinically definite ALS – laboratory supported</td>
<td>UMN and LMN clinical signs or electrophysiological evidence in one region and the patient is a carrier of a pathogenic SOD1- gene mutation</td>
</tr>
<tr>
<td>Clinically probable ALS</td>
<td>UMN and LMN clinical signs or electrophysiological evidence by LMN and UMN signs in two regions with some UMN signs rostral* to the LMN signs</td>
</tr>
<tr>
<td>Clinically possible ALS</td>
<td>UMN and LMN clinical signs or electrophysiological evidence in one region only, or UMN and LMN clinical signs in two regions with no UMN signs rostral to LMN signs. Neuro-imaging and laboratory studies have excluded other diagnoses</td>
</tr>
</tbody>
</table>

*Rostral: situated near the front end of the body
Table 2.1.2. Clinical features of MND (Phukan and Hardiman, 2009)

<table>
<thead>
<tr>
<th>Disease</th>
<th>Clinical features</th>
<th>Other comments</th>
<th>Median survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>ALS</td>
<td>Both upper and lower motor neuron signs in multiple spinal segments</td>
<td>Most common adult-onset form of MND</td>
<td>3-5 years</td>
</tr>
<tr>
<td>Primary Lateral Sclerosis (PLS)</td>
<td>Upper motor neuron (UMN) signs only</td>
<td>Many patients eventually develop clinical or electrophysiological signs of LMN involvement. ALS develops in up to 77% within 3-4 years</td>
<td>For those who remain with the diagnosis of PLS, median survival = 20 years or more</td>
</tr>
<tr>
<td>Progressive Muscular Atrophy</td>
<td>Lower motor neuron (LMN) signs only</td>
<td>Variable evolution to ALS</td>
<td>5 years. A subset survive 20 years or more</td>
</tr>
<tr>
<td>Progressive Bulbar Palsy</td>
<td>Speech and swallowing affected initially due to LMN involvement of CNIX, X, XII</td>
<td>Symptoms include dysarthria, dysphagia, and dysphonia. Aspiration pneumonia is usually the terminal event</td>
<td>2-3 years</td>
</tr>
</tbody>
</table>

Death occurs, on average, 2-4 years after the first symptoms of respiratory dysfunction, but a small group may survive for a decade or more (Anderson et al., 2012). Despite advances in the understanding of the pathophysiology of MND, the cause and pathogenesis remain unknown, with 5.1% familial according to a recent meta-analysis (Byrne et al., 2011).
Skeletal symptoms include progressive muscle weakness and atrophy, muscle fasciculations, cramps and spasticity; while bulbar symptoms include dysarthria, dysphasia, dysphagia and sialorrhea. Patients may also present with fatigue, venous compromise, pseudobulbar emotional lability, depression, anxiety, pain, and respiratory compromise. Impairments present in people with MND limit ability to perform daily activities and to participate in everyday situations, impacting on their quality of life (Morris et al., 2006; Anderson et al; 2012).

Although MND is primarily a motor system degeneration, there is overlap between MND and both frontotemporal dementia and cognitive impairment. The results of a large population based study found cognitive impairment present, predominately but not exclusively, in the form of executive dysfunction in more than 40% of MND patients (Phukan et al., 2012) (Figure 2.1.1). Clinically, cognitive impairment has practical implications for patient management and prognosis, impacting on their ability to engage with recommended treatment strategies, and is associated with a faster rate of functional decline (Elamin et al., 2013).

**Figure 2.1.1. Categorisation of the cohort of incident MND patients (n=160) using the cognitive domain-based categorisation (Phukan et al., 2012).**
Communication difficulties in MND arise mainly from progressive bulbar symptoms such as dysphasia and dysarthria. However, changes in language function can occur, especially in patients with frontal cognitive dysfunction. Language impairment impacts significantly on the quality of life of patients and their carers, makes clinical management of the patient challenging, and is correlated to reduced life expectancy (Kuhnlein et al., 2008).

2.2 Model of Care:

Guidelines for the management of MND recommend that patients attend a multidisciplinary care centre, and be reviewed by the same neurologist on a 2-3 monthly basis (more or less frequently as required). Patients should have access to a specialist team who have effective communication links with the primary healthcare centre, palliative care team and community services (Anderson et al., 2012). Evidence indicates that multidisciplinary care in specialist MND centres results in improved quality of life, reduced hospitalisation and lengths of stay, increased survival rates and reduced disability (Traynor et al., 2003; Chio et al., 2006; Ng et al., 2009; Anderson et al., 2012). Some people with MND cannot attend a specialist multidisciplinary clinic for physical and geographical reasons, so focused interdisciplinary care is essential in the community environment.

2.3 Medical Management:

There are limited pharmacological treatment options in MND. Riluzole is the only treatment shown to slow the course of MND, and is thought to prolong survival by 2-3 months. The drug should be initiated on diagnosis as it may have little effect in the late stages (Miller et al., 2007). Fatigue is reported as a side effect in 26% of patients taking Riluzole (Anderson et al., 2012).

Currently the main focus in MND is on symptomatic, rehabilitative and palliative management to maximise lifespan and optimise quality of life (Miller et al., 2007). Medications are available which may alleviate symptoms of muscle spasms and cramps (quinine sulphate); spasticity (baclofen, tizanadine); sialorrhea (amitriptyline, atropine, botulinum toxin injection, glycopyrronium, hyoscymamine); pain (paracetamol, non-steroidal anti-inflammatories, anti-convulsants, opiates); depression (tricyclic antidepressants or serotonin reuptake inhibitors); and emotional lability (amitriptyline, fluvoxamine, dextromethorphan-quinidine) (Phukan and
Sleep disturbance is medicated with caution to avoid respiratory suppression; while the dose of antispasticity medication is monitored closely for effect on muscle weakness.

Management of respiratory complications and options for ventilatory support should be discussed early. Respiratory muscle weakness is an independent predictor of quality of life in MND, and respiratory failure is the most common cause of death (Bourke et al., 2001). Respiratory muscle dysfunction results in impaired gas exchange, particularly at night, with symptoms of carbon dioxide retention including sleep disturbance, morning headaches and fatigue common; and is associated with dyspnoea, impaired quality of life, and a shortened survival (Senent et al., 2011). Physiotherapy has a significant role to play in monitoring for signs and symptoms of respiratory impairment (Table 2.3.1), which prompt the need for respiratory evaluation; and in management of associated respiratory distress and providing symptomatic relief (Bott et al., 2009; Bento et al., 2010; Senent et al., 2011).

**Table 2.3.1. Signs and symptoms of respiratory impairment in ALS** (Anderson et al., 2007)

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Signs</th>
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<tbody>
<tr>
<td>Dyspnoea on exertion or talking</td>
<td>Tachypnoea</td>
</tr>
<tr>
<td>Orthopnoea</td>
<td>Use of auxillary respiratory muscles</td>
</tr>
<tr>
<td>Frequent nocturnal awakenings</td>
<td>Paradoxical movement of the abdomen</td>
</tr>
<tr>
<td>Excessive day-time sleepiness</td>
<td>Decreased chest movements</td>
</tr>
<tr>
<td>Daytime / morning fatigue</td>
<td>Weak cough</td>
</tr>
<tr>
<td>Difficulty clearing secretions</td>
<td>Sweating</td>
</tr>
<tr>
<td>Morning headache</td>
<td>Tachycardia</td>
</tr>
<tr>
<td>Nocturia</td>
<td>Weight loss</td>
</tr>
<tr>
<td>Depression</td>
<td>Confusion hallucinations, dizziness</td>
</tr>
<tr>
<td>Poor appetite</td>
<td>Papillodema</td>
</tr>
<tr>
<td>Poor concentration or memory</td>
<td>Syncope</td>
</tr>
<tr>
<td></td>
<td>Mouth dryness</td>
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</table>
National Institute for Clinical Excellence (NICE) guidelines (2010) state that respiratory function tests should be performed by healthcare professionals with appropriate competencies every 3 months, more or less often depending on the presence of any signs or symptoms of respiratory impairment, the rate of progression of MND and patient preference. Respiratory function tests may include assessment of sniff nasal inspiratory pressure (SNIP) or maximal inspiratory pressure (MIP); forced vital capacity (FVC); arterial blood gases (ABG); overnight oximetry and/or peak cough flow. SNIP is a convenient measure which correlates well with diaphragm strength and is sensitive to changes in respiratory muscle strength (Anderson et al., 2012).

Nocturnal non-invasive positive-pressure ventilation (NIPPV) has become the standard treatment for MND patients with respiratory insufficiency. The bi-level intermittent positive-pressure ventilator imitates physiological function; it is triggered by the patient’s inspiratory efforts, reduces the work of breathing, and improves gas exchange and sleep quality (Gordon, 2011). NIPPV used in patients with early respiratory muscle involvement, who are compliant and without severe bulbar dysfunction, has been shown to extend survival, improve quality of life and in some cases improve cognitive impairment (Pinto et al., 1995; Bourke et al., 2003; Jackson et al., 2006; Anderson et al., 2012). Indications for initiating NIPPV are found in Table 2.3.2. Humidification can be applied through NIPPV where secretions are retained and are particularly viscous. In general, oxygen is not prescribed so as not to risk inhibition of respiratory drive in the setting of elevated serum carbon dioxide levels.

In the Irish context, NIPPV is usually prescribed by the Consultant Neurologist and/or the Consultant in Respiratory Medicine. Feedback regarding patients’ tolerance of NIPPV from Physiotherapists who are in close contact with the patient is welcomed. Changes in pressure settings may on occasion be made by the Physiotherapist following consultation with the medical team. This can occur via the clinic physiotherapist or the MND advanced nurse practitioner. The change in prescription should be documented in the medical chart. In some cases where NIPPV is becoming less effective, management of the patient’s respiratory distress may be more appropriately managed by palliative care intervention rather than changes to NIPPV. Long term invasive ventilation can prolong survival in MND patients but the decision to mechanically ventilate a patient varies due to concerns regarding quality of life, cultural attitudes, cost, and ethical issues (Anderson et al., 2012) and is not commonly carried out in Ireland.
Table 2.3.2. Proposed criteria for Non-Invasive Ventilation (NIV) in MND patients (Anderson et al., 2007)

<table>
<thead>
<tr>
<th>1. Symptoms related to respiratory muscle weakness. At least one of the following:</th>
</tr>
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<tbody>
<tr>
<td>Dyspnoea</td>
</tr>
<tr>
<td>Orthopnoea</td>
</tr>
<tr>
<td>Disturbed sleep not due to pain</td>
</tr>
<tr>
<td>Morning headache</td>
</tr>
<tr>
<td>Poor concentration</td>
</tr>
<tr>
<td>Loss of appetite</td>
</tr>
<tr>
<td>Excessive daytime sleepiness (ESS &gt;9)</td>
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<tr>
<td>And</td>
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<table>
<thead>
<tr>
<th>2. Signs of respiratory muscle weakness (FVC &lt;80% or SNIP &lt;40cmH2O)</th>
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<td>And</td>
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<table>
<thead>
<tr>
<th>3. Evidence of either:</th>
</tr>
</thead>
<tbody>
<tr>
<td>a) Significant nocturnal desaturation on overnight oximetry, OR</td>
</tr>
<tr>
<td>b) Morning blood gas PCO2 &gt; 6.5Kpa</td>
</tr>
</tbody>
</table>

Malnutrition and weight-loss are predictors of survival, therefore patients with dysphagia or weight loss should be referred urgently to both a dietitian and a speech and language therapist, for nutritional support and techniques to improve swallow (Anderson et al., 2007). Where enteral nutritional support is indicated, early insertion of a percutaneous endoscopic gastrotomy (PEG) or a radiologically inserted gastrostomy (RIG) is recommended, although oral feeding may be maintained while risk of aspiration remains low. The decision to proceed with a PEG / RIG is multifaceted and is made in consultation with the patient, family and neurologist. Patients being considered for a RIG must have adequate respiratory reserve to tolerate the procedure, while patient preference for quality of life may dictate insertion in the first instance (Anderson et al., 2012).
Communication difficulties such as dysphasia and dysarthria require regular speech and language therapy assessment (every three – six months), and timely provision of appropriate communication supports is recommended (Anderson et al., 2012).

Signs of functional decline can be addressed by occupational therapy and physiotherapy through the provision of assistive devices, appropriate seating and home and environmental adaptations. Patients are encouraged to make life style changes (evaluating fitness to drive, assisted living, sharing financial control) and behavioural / environmental modifications.

The MND clinical nurse specialist plays an important role in offering emotional support to the patient and their family; in co-ordination of the multidisciplinary team; and in setting up and supporting families and community teams with home non-invasive ventilation.

Symptoms of cognitive impairment such as personality change, irritability, obsessions, poor insight, and symptoms of executive dysfunction, require early cognitive assessment. Referral to neuropsychology may be recommended, and reversible causes of impaired cognition should be identified and treated. Medications commonly used in Alzheimers disease and selective serotonin reuptake inhibitors (SSRIs) are currently drugs of choice for presentations of overt frontotemporal dementia and aggression or disinhibition (Phukan and Hardiman, 2009).

Increased incidence of major depression in MND patients is controversial, and it is more widely accepted that there is an increase of burden and depression in caregivers (Albert et al, 2005). However, psychological distress is high in MND patients, often running concurrently with caregiver distress. Respite, counselling, support groups, information and hope given to the patient or to the caregiver may alleviate distress (Phukan and Hardiman, 2009).

As MND is a terminal condition, access to palliative care services as well as discussions around end of life wishes should occur early and in advance of the terminal phase. Palliative care in MND contributes to improvements in quality of life, symptom control and caregiver burden. Early involvement is recommended, and effective communication between the multidisciplinary MND team, community services and palliative care team is essential to ensure that distressing end-of-life symptoms that emerge over a short period of time are addressed in a timely fashion (Bede et al., 2013). Patient preferences for life-sustaining treatments should be revisited every six months, and patients and carers should have support when choosing hospice or homecare options; practical help with the legalities regarding advance directives; and respect of the cultural and spiritual issues around treatment choices.
(Anderson et al., 2007; Anderson et al., 2012). In Ireland, there are a number of issues surrounding advance care directives i.e. individualised impact of the disease, difficulty discussing end-of life, legalities and participation in research. This can lead to unplanned interventions, of which mechanical ventilation is one of the most controversial (Corr, 2007).

2.4 Complementary Therapy

Alternative and complementary medicine involves various methods and substances which do not form part of “orthodox” medical treatment. Patients with chronic degenerative or incurable diseases are particularly inclined to seek alternative treatments (Horiwetz, 2007; Hasan et al., 2009; Nabukera et al., 2012). Yoga, relaxation, meditation, massage, and reflexology are among the most commonly used complementary and alternative therapies in a U.K. survey of multiple sclerosis patients (Esmonde and Long., 2008). Subjective assessment of the effects of alternative or complementary medicine used in MND populations are positive overall (Wasner et al., 2001), with massage believed to be a useful adjunctive treatment for spasticity and pain in MND (Blatzheim, 2009). However, further research is needed to investigate the impact of complementary medicine on specific symptom management and quality of life in MND.
3. PHYSIOTHERAPY GUIDELINES

The physiotherapist has a broad range of skills relevant to the support of a patient with MND. Over the course of the disease, they have a significant role in the provision of aids and appliances, prescription of exercise programmes, pain management and respiratory symptom management. Increasingly evidence suggests that exercise is beneficial in maintaining muscle strength, aerobic and pulmonary function, and quality of life in patients with MND (Bohannon, 1983; Gross & Meiner, 1993; Pinto et al., 1999; Drory et al., 2001; Liebetanz et al., 2004; Dal Bello-Haas et al., 2007; Sanjak et al., 2010). Given the often rapid, progressive nature of MND, and the potential “window of opportunity” for treatment, patients with MND referred for physiotherapy should be prioritised and seen urgently.

Literature presented in the guidelines is classified using the system for documenting evidence for intervention studies, as defined by the Scottish Intercollegiate Guidelines Network (Table 3.1). The heterogeneity of MND presents issues with study recruitment, which combined with high drop out rates and the inevitable deterioration associated with the disease, results in a dearth of physiotherapy related research. It is not surprising that most of the evidence for physiotherapy is at the level of clinical opinion rather than randomised controlled clinical trials. However, several studies, which will be discussed later in the paper, provide low level evidence to support physiotherapy in MND, and the strength of this evidence is increasing.

Physiotherapy management evolves through the early, middle and late stages of MND. Specific physiotherapy management through each stage is outlined in Appendix 6.4, and is dependent on the patient’s presentation at that stage. In the early stage the patient can mobilise and manage activities of daily living independently. There may be weakness of specific muscles that limit performance of tasks or endurance. Common deficits noted at this stage may be foot drop or weakness of intrinsic muscles. In the middle stage the patient may be ambulant to some degree but is likely to have severe weakness in certain muscle groups. Spasticity may become an issue, and significant limitations to activity and participation will be emerging. There may be some evidence of respiratory insufficiency and the patient should be taught techniques to support lung expansion and secretion clearance. The late stage is characterised by progressive weakness and deterioration in mobility, dexterity and endurance. At this point the patient is generally wheelchair dependent and / or may present with respiratory compromise. A hoist is likely to be required for transfers, and pain can be a
problem. Patients with progressive bulbar palsy may maintain mobility but present with severe respiratory compromise and should be managed accordingly.

Table 3.1. Levels of evidence for intervention studies

<table>
<thead>
<tr>
<th>Level</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1++</td>
<td>High-quality meta-analyses, systematic review of randomised controlled trials (RCTs), or RCTs with a very low risk of bias</td>
</tr>
<tr>
<td>1+</td>
<td>Well-conducted meta-analyses, systematic review of RCTs, or RCTs with a low risk of bias</td>
</tr>
<tr>
<td>1-</td>
<td>Meta-analyses, systematic review of RCTs, or RCTs with a high risk of bias</td>
</tr>
<tr>
<td>2++</td>
<td>High-quality systematic reviews of case-control or cohort studies. High-quality case-control or cohort studies with a very low risk of confounding, bias or chance and a high probability that the relationship is causal.</td>
</tr>
<tr>
<td>2+</td>
<td>Well-conducted case-control or cohort studies with a low risk of confounding, bias or chance and a moderate probability that the relationship is causal.</td>
</tr>
<tr>
<td>2-</td>
<td>Case-control or cohort studies with a high risk of confounding bias, or chance and a significant risk that the relationship is causal</td>
</tr>
<tr>
<td>3</td>
<td>Non-analytical studies (for example, case reports, case series)</td>
</tr>
<tr>
<td>4</td>
<td>Expert opinion, formal consensus</td>
</tr>
</tbody>
</table>

The roles of the physiotherapist in tertiary, primary and palliative care settings can overlap, in particular the roles of the primary and palliative care therapists as the disease progresses. This will depend on who provides the service in each health service area. In Longford, for example, the primary and palliative care therapist often provide joint care, as the primary care therapist has built up a relationship with the patient and family, and the geography may mean that they can meet some of the patients needs in a more timely fashion. However, with an understanding that we cannot be rigid, an effort has been made to define what is believed to be the core roles for the therapist within each service area (Table 3.2).
Table 3.2. Role of physiotherapist in primary, tertiary and palliative care

| Role of physiotherapist in tertiary care | • First point of contact for assessment  
| | • Deal with immediate mobility or respiratory need  
| | • Send referral to primary care therapist ensuring they have adequate information regarding diagnosis  
| | • Provide more in-depth respiratory management as the disease progresses, often with initial prescription of respiratory adjunct  
| | • Link with primary or palliative services providing expert advice on physiotherapy management in MND  
| Role of physiotherapist in primary care | • Provide more in-depth assessment of need  
| | • Management of symptoms associated with the diagnosis  
| | • Monitor changes in mobility and respiratory function  
| | • Provide aids and appliances in a timely fashion  
| | • Review mobility in the home, work or social environment  
| | • Link with tertiary centre to feedback on current status, in particular if significant changes in respiratory function are observed  
| Role of physiotherapist in palliative care | • Involved in home set-up of respiratory adjuncts and educating family to manage changing respiratory status  
| | • Management of symptoms associated with the diagnosis  

* Role of physiotherapist in palliative care can overlap with that of physiotherapist in primary care
Within these guidelines physiotherapy management for patients with MND is discussed in terms of:

3.1 Assessment and outcome measurement
3.2 Effective communication between interdisciplinary team members and colleagues in primary and tertiary care
3.3 Goal setting
3.4 Treatment including:
   3.4a General Recommendations / Advice
   3.4b Maintaining Mobility and Function
   - Prevention of contractures
   - Exercise prescription for resistance and aerobic training
   - Spasticity management
   - Falls prevention
   - Provision of assistive devices
   - Body positioning
   - Ergonomic assessment
   3.4c Respiratory Symptom Management
   - Monitor for signs and symptoms of respiratory insufficiency
   - Cough and airway secretion management
   - Respiratory muscle training
   3.4d Pain Management
   3.4e Fatigue Management
3.5 Advocacy
3.6 Education of Patients & Carers
3.7 Transfer of Care
3.1 Assessment
Each person with MND is affected in a unique and unpredictable way. The role of the physiotherapist is to provide a detailed ongoing assessment of a patient’s physical needs.

3.1.1 Recommendations
A. Patients with MND should be seen as a priority for assessment, and should be offered regular monitoring and review (4).

B. During the initial meeting the therapist must ensure the environment is and will remain private, with phones and pagers switched off and “Do not disturb signs” in place (4).

C. The physiotherapist should start the initial consultation by asking what the patient already knows or suspects (4).

D. The subjective assessment (Appendix 6.1) should contain details on the referral source, the nature and course of MND to date, any co-morbidities, and the current treatment approach. The patient should be questioned about their symptoms, any functional limitations and any issues that may surround their participation at home, work or in social situations. The expectations of the person with regard to prognosis and physiotherapy input should be discussed, and any mental, personal or external factors taken into account. This discussion should occur with due regard and sensitivity for the patient, and the therapist should avoid overemphasising the potential range of difficulties that may be encountered through the course of disease progression (4).

E. The objective assessment (Appendix 6.1) of the patient should consider impairments (musculoskeletal, respiratory and nervous systems) and functions (balance, mobility, exercise tolerance and global functioning) (4).

F. Outcome measures (Appendix 6.1) may not be appropriate to repeat at all treatment sessions, but should be used at baseline and intermittently at reassessment to monitor changes in disease progression, and to guide appropriate treatment planning (4).
3.2 Effective Communication
The importance of effective communication within and between the multidisciplinary teams, patient, and family continues to be recognised as a fundamental necessity for the successful care and management of patients with MND.

3.2.1 Recommendations

A. Effective channels of communication and co-ordination are essential within and between the tertiary, primary and palliative care multidisciplinary team, the patient and the family (2++).

B. Physiotherapists should use all resources available to them for advice and information, and maintain regular contact with each other (phone, e-mail or letter) throughout the course of the patient’s disease. Outreach support services are also available from the nurse specialists in either the MND clinic, Beaumont, or the Irish Motor Neurone Disease Association (IMNDA), and both can be accessed by the clinician (4).

C. Assessments in primary or palliative care should be completed prior to each multidisciplinary clinic visit. A synopsis of the patient’s level of mobility and function should be forwarded to the relevant physiotherapist in order to provide an accurate picture of the patient’s current status, and should include any specific issues requiring input from the physiotherapist at clinic. If appropriate, a report should then follow from the physiotherapist at clinic detailing any changes to current management, advice, or recommendations for future management. Suggested pro formas for use between tertiary and primary or palliative care settings can be found in (Appendix 6.2). These should be posted, e-mailed, faxed or taken by the patient as agreed by individual therapists or local policies (4).

D. Irish Healthcare professionals in all settings should use the Motor Neuron Disease Patient Passport (Appendix 6.3) held by the patient to assist them in keeping track of all the healthcare staff working with them. It is also a means of improving communication between clinic and community staff (4).

E. Within the team, therapists’ roles can overlap and there needs to be flexibility in deciding who is the most appropriate person to provide advice, education, and therapy for an individual and their care-givers. Decisions must be made around this to avoid
confusing the patient or caregiver, or leave them in any doubt as to whom is managing a particular issue (4).

3.2.2 Evidence
A. A comprehensive literature review of studies describing patient-professional communication interventions for patients with life limiting conditions identified three main themes to effective communication. These include using education to enhance professional communication skills, using communication to improve patient understanding, and using communication skills to facilitate advance care planning. Facilitating a successful intervention might require a healthcare professional to enhance their communication skills through reviewing evidence based research on effective communication tools (Barnes et al., 2012).

3.3 Goal Setting
Goal setting in MND ensures both the therapist and the patient knows what to expect from an intervention. In most patients with MND the goals are likely to be short-term, and aimed at maintaining function and independence rather than gaining anything more. Through setting goals you provide direction and a plan that is open to revision as the disease progresses.

3.3.1 Recommendations
A. Goals should involve the person and carer, be focused at the level of activity/participation, be timed with short term and long term aims and occur as part of multidisciplinary team planning where appropriate (4).

B. The goal in the early stages of MND is to maintain, and optimise, mobility and function. Physiotherapy programmes should be functional, goal orientated and preformed at intensities to avoid fatigue (4).

C. In the middle stage of MND the goal is to continue to maintain functional mobility, manage pain and to carefully monitor and address any symptoms of respiratory compromise (4).

D. Late phase goals are designed to maximise quality of life for the patient (4).
3.4 Treatment Management

Often the goal of physiotherapy in the care of people with MND is not to improve impairment in strength or mobility due to the progressive nature of the condition. Instead, therapy is aimed at assisting the patient to maximise function through provision of aids and appliances, mobility strategies and respiratory management. However, it is important that problems with pain are managed where possible with a restorative approach. As the disability progresses a patient’s care needs may require the application of proper lifting and handling techniques, implemented in ways that ensure the safety of the patient and the carer. Early provision of hoists or manual handling belts can reduce carer burden and prevent secondary musculoskeletal injuries. Multidisciplinary care particularly between physiotherapy and occupational therapy is important in achieving these aims.
The therapist should have a broad range of physiotherapy competencies from the fields of neurology, musculoskeletal and respiratory care. Physiotherapy treatment for patients through the stages of MND progression is presented in Appendix 6.4.

3.4a General

3.4a.1 Recommendations

A. Frequency of input should be determined on an individual basis. However, patients should be able to access services urgently when the need arises (4).

B. Physiotherapy management should be provided, where possible, by a clinician with experience managing MND, who can provide continuity of care in an appropriate environment and with access to the equipment required (Appendix 6.5) (4).

3.4b Maintain Mobility and Function:

Muscle weakness results in limitations in function and mobility in patients with MND. Disuse atrophy and deconditioning, as well as muscle degeneration resulting from the disease process, contribute to the weakness (McDonald, 2002). The development of contractures or the imbalance between opposing muscle groups also impact on functional mobility and fatigue. Spasticity is another common feature of MND, and its effects can be severe, both in general disability and in pain, significantly impacting on mobility and function. Physiotherapists play a significant role in maintaining mobility and function in patients with MND through maintenance of range of movement; prevention of contractures; managing tone; strengthening
and aerobic exercise prescription; the provision of aids and appliances; and education on correct body positioning and manual handling (Appendix 6.6). It should be noted that the intensity of resistance or aerobic exercise should not compromise the reserve required to carry out activities of daily living, and such exercises may not be indicated for the middle to late, or even early phase of ALS.

3.4b.1 Recommendations

C. Passive movements (PM) should be applied to joints that patients cannot move themselves (4).

D. Stretching and range of movement exercises form the basis for spasticity management (4).

E. Education of carers and family members on passive movements and stretches is essential in ensuring PM are carried out regularly (4).

F. The use of standing equipment such as an easy-stand or tilt table to support a stretch in antigravity and weight-bearing muscle groups should be considered (4).

G. Individualised strengthening exercises during the early stage of MND are probably effective in improving the function of patients and should be recommended (1).

H. Supported treadmill training may be a useful modality in the early stages of MND (1).

I. Moderate intensity endurance activity may delay the onset of spasticity (1).

J. Active/Passive trainers or unloaded cycling should be considered, ideally for home use, where spasticity is present (1).

K. Antispasticity medication should be considered for judicious use where spasticity is limiting function. The role of the physiotherapist is to liaise with the medical team regarding limitations associated with increased tone (1+).
L. Timely provision and regular review of aids and appliances is essential in maximising the patient’s function.

M. Many patients will require an ankle foot orthosis (AFO). Off-the-shelf AFOs e.g. Swedish AFO, dictus splint, foot-up splint, will often be sufficient to support gait. However, in more complex patients a custom made device may be indicated. In this instance early orthotic referral is essential as deterioration may be rapid and orthotic wait times long. Liaison with the tertiary clinic regarding prognosis would be important should custom devices be indicated (4).

N. Persons with MND often need a collar as their neck muscles become weaker. An off-the-shelf foam collar can meet the needs of a patient if it is needed for example during transfers, toileting or while in a vehicle. A Headmaster or a Hessinger collar, or other more rigid and supportive collars may be recommended as the disease progresses to maintain head and neck alignment (4). This is provided by the occupational therapist in the tertiary clinic but may be provided by the physiotherapist in primary care.

O. Positioning of the patient in a good alignment while sitting or supine can decrease abnormal muscle tone; prevent or reduce spasticity, contractures, and pain; prevent the development of pressure sores; and minimise the effort required for respiration (4).

P. Adjuncts like riser – recliner chairs, sleep systems, high profile mattresses, profiling beds, or high back wheelchairs may be required as the disease progresses to support and correct body positioning (4). Some equipment which cannot be acquired in a timely manner through the HSE is available through the Irish Motor Neurone Disease Association (IMNDA) (Appendix 6.5).

Q. Close liaisons between the public health nurse, occupational, speech and language and physiotherapists are required to ensure the correct positioning is achieved to maximise comfort and function (4).

R. The physiotherapist should complete a manual handling risk assessment and provide education on handling to carers so that assisted exercises, transfers or mobility are performed at an acceptable level of risk (4).
3.4b.2 Evidence:

C. No randomised controlled trial of stretching has been looked at in patients with MND. Despite this, stretching or PM exercises are widely believed to be beneficial in maintaining joint mobility and managing symptoms of spasticity and related pain in people with spinal cord injury (Harvey et al., 2009), and in unconscious patients in intensive care units (Wiles and Stiller, 2009). Consequently, they are also recommended as a treatment option for patients throughout the course of MND.

F. Bohannon (1983) presented a case study reporting the functional beneficial effects of an upper limb resistance programme in a patient with MND. More recently two randomised studies of resistance exercise in MND patients have suggested that such training is safe and efficacious in reducing the decline in muscle strength (Drory et al., 2001; Dal Bello-Haas et al., 2007) (Appendix 6.6). However, the number of subjects who have been recruited to, and who have completed such studies are small. Endurance exercise may be helpful in reversing or preventing the negative effects of a deconditioning. Positive evidence is emerging to indicate that individualised strengthening exercise programmes during the early stages of MND are probably effective in improving the function of patients and should be recommended.

G. A study by Pinto et al., 1999, investigating whether aerobic exercise affects the functional outcomes in patients with MND concluded that aerobic training of MND patients, even when respiratory insufficiency is present, is not only possible with the use of non-invasive respiratory support, but may have a positive impact on global disease progression and respiratory function. However, details of the activities of the control group during the study are unknown. More recently, Sanjak et al., 2010 reviewed the feasibility, tolerability, safety and exercise treatment effect of a de-weighted (40%) treadmill-training programme for patient with MND. They concluded that treadmill training is feasible, tolerable and safe for this patient group but conceded that the small number of subjects, the lack of control group and non-randomised treatment assignment weakened the argument. More robust research in this field is necessary. However, supported treadmill training may be a useful modality in the early stages of MND. Further details of this evidence can be found in Appendix 6.6.

I. One study (Drory et al., 2001) (Appendix 6.6) has examined the effect of moderate intensity endurance activity on spasticity in 25 patients with MND (Intervention n =14; Control n = 11). In patients who did not exercise, the spasticity increased with time while there was a
net decrease of spasticity in the treatment group. It was concluded that the single trial was too small to determine whether the exercise was beneficial, and that further research is needed.

J. Although the benefits of cycling have not been investigated specifically in MND, a study of unloaded cycling in multiple sclerosis (Sosnoff et al., 2009), found no objective improvement in spasticity but did report a reduction in the perceived level of spasticity. The findings of a study in Iran (Rayegani et al., 2011), despite a number of limitations, suggest that passive rhythmic leg exercise can lead to decreased spasticity, increased passive ROM of lower limbs, and improvements in electrodiagnostic parameters in patients with spinal cord injury (SCI). Patients in both trials expressed sensations of well-being and it could be postulated that unloaded or passive cycling may have the same impact on spasticity in patients with MND.

K. A systematic review of spasticity in MND concluded that judicious use of antispasmodic agents should be considered where spasticity is limiting function, but where muscle strength remains relatively unimpaired. Research should explore whether anti-spasticity medications such as baclofen are really effective in reducing spasticity, or if they may cause harm by increasing muscle weakness and thus further limiting mobility and function (Ashworth et al., 2011).

3.4c Respiratory Symptom Management
Inspiratory, expiratory and / or bulbar muscle weakness in MND patients has a major prognostic impact. Diaphragmatic dysfunction is associated with dyspnoea, impaired quality of life, and a shortened survival; while an ineffective cough results in atelectasis and pneumonia (Senent et al., 2011). The problems can be further complicated by disuse atrophy, shortening and stiffness of chest wall musculature and reduced lung compliance. Effective coughing depends on inspiratory muscles to increase lung volumes; expiratory muscles to produce high thoracoabdominal pressures; and on upper airway muscles to co-ordinate glottis closure and opening. They can all be impaired in MND, resulting in peak cough flows too low for an adequate airway clearance, which can accelerate the development of respiratory failure and death. The focus for physiotherapists in respiratory management is to monitor signs and symptoms of respiratory insufficiency, and to manage secretions.
3.4c.1 Recommendations

S. Physiotherapists should monitor closely for signs and symptoms of respiratory insufficiency (4).

T. Peak cough flow (Appendix 6.7) should be measured regularly in patients with MND (2+).

U. Where increasing respiratory compromise is evident, referral should be made to the tertiary clinic for further assessment (2+) (see page 7).

V. Respiratory adjuncts such as PEP and acapella are not indicated to assist secretion clearance in this patient group (2+), instead manually assisted coughing (Appendix 6.8) should be used, and family/carers educated in its technique (2+).

W. Facilitating maximum insufflation (Appendix 6.9) via breathstacking (using a bag-valve mask or a positive pressure device (NIV)), or through the use of a mechanical in-exsufflator (Cough Assist) (Appendix 6.10), can assist in mobilising secretions, maintaining chest wall mobility and lung compliance (2+).

X. High chest wall oscillations may be useful for secretion mobilisation but access is limited due to the fact that it is cumbersome and can be cost prohibitive (1).

Y. Due to a lack of evidence, no specific recommendations can be made in support of, or refuting, respiratory muscle training in patients with MND (1-).

Z. Oral suctioning using a yanker is useful in patients with upper airway secretions which are difficult to clear. Patients commonly use this form of suctioning at home. Oropharyngeal suctioning is not normally performed in patients with MND as assisted cough techniques are preferred and more comfortable.

3.4c.2 Evidence

T. A peak cough flow of less than 270 l/min indicates that the cough is ineffective for airway clearance, with a value of greater than 160 l/min critical in preventing the onset of respiratory failure during a respiratory tract infection (Bott et al., 2009; Senent et al., 2011).
U. A recent study by Just et al., 2010 found that a score of $\geq 3$ on the modified Borg scale for dyspnoea (sitting and / or supine) (Appendix 6.11), correlates well with a SNIP of $\leq 40\text{cmH}_2\text{O}$ indicating severe inspiratory muscle weakness. A cut off value of 3 on the supine Borg scale provided the best sensitivity (80%) and specificity (78%) to predict a SNIP of $\leq 40\text{cmH}_2\text{O}$, and the tool is recommended for best use in a supine position for this patient group. Clinically, use of this non-invasive assessment tool in the community could assist therapists in early detection of inspiratory muscle weakness and prompt timely referral to tertiary services for further pulmonary function testing.

V. Airway clearance techniques aim to assist with the removal of secretions. Independent airway clearance techniques or other respiratory adjuncts (e.g. PEP or Acapella) are unlikely to be successful in this patient group due to inadequate flow volumes (Bott et al., 2009). Manually assisted coughing significantly increases peak cough flow by a well-timed thrust from an assistant during the expiratory cycle. The upright seated position and an abdominal thrust were found to have the greatest effect in a study of spinal cord injury patients. However, alternative body positions and thrust can be trialled according to patient preference (Bott et al., 2009; Senent et al., 2011).

W. Maximum insufflation or in/exsufflation strategies and assisted coughing techniques have been shown to decrease hospitalisation, and prolong survival, particularly when used in conjunction with non-invasive ventilation at times of respiratory insufficiency (Bott et al., 2009; Senent et al., 2011; Sancho et al., 2013). In patients with poor bulbar function, who have an inability to close the glottis, maximum insufflation can only be achieved through one application or using a one-way valve (Kang and Bach, 2000; Bott et al., 2009). A comparison of assisted coughing techniques in stable MND patients with severe respiratory insufficiency found that mechanical insufflation-exsufflation generated the highest peak cough flow when compared with manually assisted cough alone, or combined with maximal insufflation with either manual breathstacking or the bi-level pressure ventilator. However, subjective reports of comfort were greater using the bi-level pressure ventilator, therefore the choice of techniques should be tailored to the individual (Senent et al., 2011).

X. With high frequency chest wall oscillations positive pressure air pulses are provided to the chest wall via an inflatable vest believed to produce shearing at the air mucus interface and
provide repetitive peak expiratory flows to help expel mucus like mini coughs. Pulses compress the chest wall at frequencies of 5-30 hz. This technique has suggested benefits for secretion mobilisation and airway clearance in MND (Lange et al., 2006). However, the equipment required is expensive and not routinely used clinically.

Y. Cheah et al., 2009, in a randomised controlled study using an inspiratory muscle training device with MND patients over a 12-week period, found a trend towards increased forced vital capacity and vital capacity in the treatment group, and improvements in inspiratory muscle strength in both the treatment and control groups. However, due to the heterogeneity of the available research for patients with MND no specific recommendations can be made.

3.4d Pain Management
Pain is a common problem in MND, with frequency and intensity correlating with a worse functional score and a longer disease duration (Chio et al., 2012). Identification of the precipitants leading to pain is the first priority. Much of this relates to neuromuscular weakness, including the effects of posture and immobility. Emotional distress, muscle spasms, cramps, pressure sores, spasticity, and constipation may all cause pain, while the presence of pre-existing conditions like arthritis may be exacerbated by progressive weakness. Severe pain has been reported in up to 20% of patients with MND (Orrell, 2010). In particular, shoulder pain is a common feature of MND and incidence has been reported at 26% (Newrick and Langton-Hewer., 1985; Brettschneider et al., 2008). Adhesive capsulitis and rotator cuff damage are common and are often attributable to shoulder weakness, spasticity, joint contractures and decreased range of movement and stiffness (Brettschneider et al., 2008). Falls and poor manual handling techniques may also lead to shoulder pain. A Cochrane review by Brettschneider et al. (2008) found no randomized controlled trials of drug therapy for pain in MND and medical management of pain is similar to that of other conditions including non-steroidal anti-inflammatory drugs and opioids. Care with medications which can exacerbate constipation, fatigue or respiratory depression is required.

Physiotherapy strategies such as positioning, stretching, range of motion exercises or other techniques to address pain of musculoskeletal origin, can reduce the need for medications. Corticosteroid joint injections, electrotherapy modalities e.g. TENS, and orthotics like splints and collars are all tools available to the physiotherapist to manage this disabling symptom (Green et al., 2013; Hurlow et al., 2012).
3.4d.1 Recommendations

Z. Treatment of pain in MND is similar to that for any condition of musculoskeletal origin (4).

AA. Early training on correct manual handling, positioning, shoulder care and range of movement exercises may minimise shoulder pain, a common problem in MND (4).

BB. Electrotherapy modalities (e.g. TENS), splints or collars should be considered for use to relieve pain (4).

CC. Corticosteroid injections may be indicated to relieve pain where conventional therapy has been unsuccessful. These injections can be accessed through the general practitioner or at the tertiary clinic, should be implemented early, and can be repeated (4).

DD. Physiotherapists should be aware of the UK Motor Neurone Disease Networking Group paradigm for pain management in Motor Neurone Disease (Appendix 6.12) (4).

3.4e Fatigue Management

Fatigue is a commonly experienced debilitating symptom, and can impact on quality of life for patients with MND (Lou et al., 2003). The level of fatigue experienced is significantly greater than in healthy age/sex matched counterparts (Ramirez et al. 2008). Excessive activity, stress, depression, pain, poor cardiopulmonary function, medication, and impaired sleep patterns can all be considered as contributing factors to fatigue (Mitsumoto et al. 2000, Gordon, 2011), and physiotherapists should therefore be mindful to ask patients about their level of fatigue. Management strategies should be directed at minimising factors known to impact on these contributing factors, referring to other health professionals as necessary.

3.4e.1 Recommendations

FF. Physiotherapists should be aware non-invasive ventilation (NIV) improves symptoms of hypercapnia in patients with MND, and should refer as appropriate (3).
GG. Teaching energy conservation techniques may encourage patients to pace themselves and reduce levels of fatigue (4).

HH. Moderate physical activity can impact on symptoms of MND including weakness, spasticity and depression thereby influencing some of the factors associated with impaired sleep (4). However, the intensity of physical activity should be closely monitored for its positive or negative impact on fatigue.

3.4e.2 Evidence:

FF. Nocturnal hypoventilation is a common symptom of MND and is thought to contribute significantly to impaired sleep. Assistive devices such as NIV have been shown to improve hypercapnic symptoms such as headaches, restlessness, nightmares and poor quality sleep, and therefore improve QOL in patients with MND (Lou et al. 2010).

3.5 Advocacy

3.5.1 Recommendations

A. The clinician should assure the patient / caregiver of their role as advocate (4).

3.6 Education of Patients and Carers:

3.6.1 Recommendations

A. The physiotherapist should avoid providing insufficient information, delivering information callously, taking away or not providing hope (4).

B. The cultural and social background of the patient should be respected: clarifying if the patient wishes to receive information or if he/she wants information communicated to the family (4).

C. Patients and carers should have access to information about ‘physiotherapy and MND’, and get written information when receiving aids and appliances, advice or exercises (4).
D. Clinicians should be able to provide details of the relevant members of the multidisciplinary team in their setting (4).

3.7 Transfer of care

3.7.1 Recommendations
A. Clients with MND should not be discharged from tertiary or community physiotherapy services until a transfer of care is made to the palliative care physiotherapist (if appropriate) (4).

B. When care is transferred to another physiotherapist, written information should accompany the transfer (4).
4. CONCLUSION

Management of patients with MND is challenging, and while evidence indicates that multidisciplinary care in specialist MND centres is recommended for improved quality of life; reduced hospitalisation and lengths of stay; increased survival rates; and reduced disability; a multidisciplinary team approach is also essential in the palliative or primary care settings to augment patient care and outcomes.

Patients with MND should be considered a priority for assessment, and offered regular monitoring and review. While therapy is primarily aimed at assisting the patient to optimise functional mobility and respiratory status, symptoms of pain and fatigue should be addressed with a restorative approach. Physiotherapists therefore require a wide range of neurological, musculoskeletal and respiratory skill, and good communication skills.

These guidelines have endeavored to provide physiotherapists with the knowledge and guidance to provide evidence based and pragmatic care to patients with MND. They make recommendations based on expert opinion, non-analytical studies, and limited but emerging scientific evidence currently available to support physiotherapy input in motor neuron disease. Physiotherapists must use their professional judgment when applying the guidelines to a particular patient’s situation. The guidelines should be revised within the recommended timeframe of 3-5 years, but it is possible international guidelines will be published prior to this.
5. REFERENCES


Bourke, S.C., Shaw, P.J., Gibson, G.J. (2001) Respiratory function versus sleep-disordered breathing as predictors of QoL in ALS. *Neurology*, 57, 2040-2044.


6. APPENDICES

6.1 Assessment and outcome measure tables
6.2 Communication proformas for use between MND clinic and primary/palliative care
6.3 Motor neuron disease patient passport
6.4 Treatment guidelines through the stages of ALS progression
6.5 Environment and equipment
6.6 Evidence supporting physiotherapy in ALS
6.7 Instruction for the use of the peak cough flow meter
6.8 Manually assisted cough
6.9 Maximum insufflation: Breathstacking
6.10 Mechanical In-Exsufflation
6.11 Modified borg scale for dyspnea
6.12 Motor neurone disease pain pathway
Appendix 6.1: Assessment and Outcome Measurement

**Subjective Assessment**

<table>
<thead>
<tr>
<th>Referral source</th>
<th>Consultant details and members of the team involved at tertiary and primary care</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nature and course of ALS</td>
<td>Onset of complaints, time since diagnosis, results of diagnostics, nature of the course</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Current presenting symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Impairments</strong></td>
</tr>
<tr>
<td><strong>Limitations in function and activity</strong></td>
</tr>
<tr>
<td><strong>Participation</strong></td>
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<tr>
<td><strong>Co-morbidities</strong></td>
</tr>
<tr>
<td><strong>Treatment</strong></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Other factors</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Psychological factors</strong></td>
</tr>
<tr>
<td><strong>Personal factors</strong></td>
</tr>
<tr>
<td><strong>External factors</strong></td>
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<tr>
<td><strong>Expectations</strong></td>
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</table>
**Physical Examination and Outcome Measurement**

<table>
<thead>
<tr>
<th>Observation</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Level of consciousness and orientation; posture, fasciculations, asymmetries; communication; sialorrhoea; emotional lability; anxiety</td>
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<table>
<thead>
<tr>
<th>Motor and Sensory Function</th>
<th>Outcome Measure (examples)</th>
</tr>
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<tbody>
<tr>
<td>Joint Mobility</td>
<td>Goniometry</td>
</tr>
<tr>
<td>Muscle Length</td>
<td>Goniometry</td>
</tr>
<tr>
<td>Muscle Strength</td>
<td>Manual muscle testing, Hand grip strength dynamometry</td>
</tr>
<tr>
<td>Spasticity</td>
<td>Modified Ashworth scale</td>
</tr>
<tr>
<td>Sensation</td>
<td>Vibration, Sharp-Blunt</td>
</tr>
<tr>
<td>Co-ordination</td>
<td>Finger-nose test, 9-hole peg test</td>
</tr>
<tr>
<td>Pain</td>
<td>Visual analogue scale, body diagram</td>
</tr>
<tr>
<td>Fatigue</td>
<td>Fatigue severity scale</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Physical Activity</th>
<th>Outcome Measure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Body Posture</td>
<td>Body diagram</td>
</tr>
<tr>
<td>Balance</td>
<td>Berg balance scale</td>
</tr>
<tr>
<td>Mobility</td>
<td>Modified rivermead mobility index, 10m walk test, Timed up and go test</td>
</tr>
<tr>
<td>Exercise tolerance</td>
<td>6m walk test, Modified BORG scale for dyspnoea (Appendix 6.11)</td>
</tr>
<tr>
<td>Global functioning</td>
<td>ALS functional rating scale</td>
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</table>

**Respiratory Examination and Outcome Measurement**

<table>
<thead>
<tr>
<th>Observation</th>
<th></th>
</tr>
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<tr>
<td>Signs and symptoms of respiratory impairment (Table 3.1.4)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Respiratory Impairment</th>
<th>Outcome Measure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sputum retention</td>
<td>Auscultation, Pulse Oximetry</td>
</tr>
<tr>
<td>Effectiveness of cough</td>
<td>Peak cough flow (Appendix 6.7)</td>
</tr>
<tr>
<td>Consider symptoms of nocturnal de-saturation</td>
<td>Modified Borg scale for dyspnoea (Appendix 6.11) Spirometry, Pulse oximetry</td>
</tr>
</tbody>
</table>
Appendix 6.2: Communication: Proformas for use between MND Clinic and primary / palliative care settings

A. Communication Form Beaumont Hospital

![Communication Form Beaumont Hospital](image-url)
B. Suggested Communication Form Primary / Palliative Care

Motor Neurone Disease Clinic
Physiotherapy Report

Name: 
DOB: 

Address: 

Date of Assessment: 
Date for review at MND clinic: 

Subjective:

<table>
<thead>
<tr>
<th>Joint</th>
<th>Right Power</th>
<th>Left Power</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Elbow</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wrist / Hand</td>
<td>HGS</td>
<td>HGS</td>
</tr>
<tr>
<td>Hip</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Knee</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ankle Foot</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neck:</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Tone / Cramps:

Pain:

Respiratory Function:

Balance and Mobility:

Gait: 
Aids/Appliances: 

Physiotherapy Intervention: 

Issues requiring input from Beaumont MDT: 

Physiotherapist: 
Location: 
Contact Details: 

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Appendix 6.3: Motor Neuron Disease patient passport

To the Healthcare Professional

This MND patient management passport aims to help patients keep track of the many healthcare professionals that they may encounter.

It will also assist in improving communication between healthcare professionals and between the hospital and community based teams.

It contains the links to resources on management of MND that may be helpful to healthcare professionals.

Please complete your contact details and initial and date on each contact.

This passport is not a medical record and any specific assessment or treatment information should not be recorded here.
Physiotherapy

The physiotherapist assesses the physical changes that occur in MND. These may include changes in muscle strength, stiffness, cardiovascular fitness, muscle length and joint range of movement.

Treatment focuses on the following areas:
- Providing advice on type & programmes of exercise
- Management of muscle stiffness
- Evaluating the need for walking aids and splints
- Advice on moving and handling skills for the patient, family and carers.
- Management of joint pain.

Beaumont Hospital Physiotherapists:
Name: ____________________________
Tel: ____________________________
Name: ____________________________
Tel: ____________________________
Name: ____________________________
Tel: ____________________________

Appointments or contacts with Beaumont Hospital Physiotherapist

Date | Initial | Comment
-----|---------|--------

Please record appointment dates and initial each contact. This is not a medical record and assessment or treatment information should not be entered.

Primary Care Physiotherapist:
Name: ____________________________
Address: ____________________________
Tel: ____________________________
Name: ____________________________
Address: ____________________________
Tel: ____________________________
Name: ____________________________
Address: ____________________________
Tel: ____________________________

Appointments or contacts with Primary Care Physiotherapist

Date | Initial | Comment
-----|---------|--------

Please record appointment dates and initial each contact. This is not a medical record and assessment or treatment information should not be entered.
## Appendix 6.4: Treatment Guidelines for Early, Middle and Late stage ALS

### Early Stage

<table>
<thead>
<tr>
<th>Goals of Therapy</th>
<th>Strategy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maintain mobility and function, and prevent falls</td>
<td></td>
</tr>
<tr>
<td>Prevention of contractures</td>
<td>Active/Passive Stretching Programme</td>
</tr>
<tr>
<td>Exercise prescription</td>
<td>Active range of movement exercises</td>
</tr>
<tr>
<td></td>
<td>Resistive strengthening exercises, aerobic exercises as appropriate</td>
</tr>
<tr>
<td></td>
<td>Consider use of motomed/ theravital</td>
</tr>
<tr>
<td>Provision of assistive devices</td>
<td>Assistive devices e.g. foot-up, dictus, stick</td>
</tr>
<tr>
<td>Advice</td>
<td>Appropriate footwear, Falls prevention</td>
</tr>
<tr>
<td><strong>Respiratory Symptom Management</strong></td>
<td></td>
</tr>
<tr>
<td>Monitor for respiratory insufficiency</td>
<td>Monitor signs and symptoms of respiratory impairment</td>
</tr>
<tr>
<td>Clear bronchial secretions</td>
<td>Teach supported cough</td>
</tr>
<tr>
<td></td>
<td>Link with SLT</td>
</tr>
<tr>
<td><strong>Fatigue and Anxiety Management</strong></td>
<td></td>
</tr>
<tr>
<td>Advice on energy conservation</td>
<td></td>
</tr>
<tr>
<td>Reassurance</td>
<td></td>
</tr>
<tr>
<td>Teach relaxation techniques</td>
<td></td>
</tr>
<tr>
<td>Psychological support to patient and carer</td>
<td></td>
</tr>
<tr>
<td>Middle Stage</td>
<td></td>
</tr>
<tr>
<td>------------------------------------------------------------------------------</td>
<td></td>
</tr>
<tr>
<td><strong>Goals of Therapy</strong></td>
<td><strong>Strategy</strong></td>
</tr>
</tbody>
</table>
| **Maintain mobility and function, and prevent falls** | Stretching Programme. Educate caregivers  
Consider easy-stand |
| Prevention of contractures | Continue active range of movement exercises, resistive strengthening exercises, aerobic exercises as appropriate  
Initiate active assisted exercises, teaching caregivers to assist patient, Falls prevention |
| Exercise prescription | Consider impact of spasticity on mobility - stretching programme, motomed/theravital  
Link with tertiary clinic to consider antispasticity medication or botulium injections |
| Spasticity Management | Assistive devices e.g. ankle foot orthosis, knee brace, walking frame  
Link with OT regarding seating / wheelchair |
| Provision of assistive devices | Advice on positioning for comfort and to prevent pressure sores (link with Public health nurse), |
| Body Positioning | **Respiratory Symptom Management** |
| Monitor for respiratory insufficiency | Monitor signs and symptoms of respiratory impairment  
Link with tertiary clinic – further assessment may indicate non invasive ventilation |
| Clear bronchial secretions | As for early phase AND,  
Consider breathstacking  
Consider cough assist |
| **Pain Management** | **Fatigue and Anxiety Management** |
| Shoulder Pain / Neck pain / Other | Treat as per pain guidelines according to the pathophysiology of the problem  
Link with GP / Orthopaedics / Tertiary clinic to consider joint injection  
Provide supportive devices (Soft collar / Headmaster collar / sling) |
| Spasticity | Stretching programme, Theravital/motomed  
Link with tertiary clinic to consider antispasticity medication or botulium injections |
| Advice on energy conservation  
Teach relaxation techniques  
Reassurance and psychological support to patient and carer |
<table>
<thead>
<tr>
<th>Late Stage</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Goals of Therapy</strong></td>
<td><strong>Strategy</strong></td>
</tr>
<tr>
<td><strong>Maintain mobility and function</strong></td>
<td></td>
</tr>
<tr>
<td>Prevention of contractures</td>
<td>Stretching Programme. Reinforce with caregivers Consider use of tilt table</td>
</tr>
<tr>
<td>Exercise prescription</td>
<td>Active and passive ROM as able, motomed/theravital</td>
</tr>
<tr>
<td>Provision of assistive devices</td>
<td>Assistive devices e.g. Headmaster collar Link with OT regarding seating / wheelchair Consider sleep system</td>
</tr>
<tr>
<td><strong>Respiratory Symptom Management</strong></td>
<td></td>
</tr>
<tr>
<td>Monitor for respiratory insufficiency</td>
<td>As for middle phase management with increased liaison between tertiary support services (clinical nurse specialist) and community services</td>
</tr>
<tr>
<td>Clear bronchial secretions</td>
<td>As for middle phase management Consider oral suction</td>
</tr>
<tr>
<td><strong>Pain Management</strong></td>
<td></td>
</tr>
<tr>
<td>Shoulder Pain / Neck pain/ Other</td>
<td>As per middle phase management</td>
</tr>
<tr>
<td>Spasticity</td>
<td>As per middle phase management</td>
</tr>
<tr>
<td><strong>Fatigue and Anxiety Management</strong></td>
<td></td>
</tr>
<tr>
<td>Advice on energy conservation Reinforce relaxation techniques Reassurance and psychological support to patient and carer</td>
<td></td>
</tr>
</tbody>
</table>
Appendix 6.5: Environment and Equipment

Patients with ALS should ideally have access to an experienced physiotherapist with a good knowledge of the disease process and its management; an appropriate environment in which to be treated; and access to the basic equipment required to facilitate effective evidence based management. Physiotherapists must anticipate patient and family needs, and meet those needs in a clean, safe, private environment. Given the rapidly progressive nature of ALS, it is advisable to have certain items in stock that can be provided in a timely manner.

The Primary Care Setting
Assistance towards the costs of the purchase of medical and surgical aids and appliances, such as wheelchairs, walking aids etc, may be provided directly by the HSE to Medical Card holders.

Physiotherapist should be aware that some items of equipment are available to clients from the Irish Motor Neuron Disease Association (IMNDA), and can be accessed through their equipment officers at 1800403403.

Such equipment must be re-useable and may include:

- Motorised wheelchair, transit wheelchair
- Hoist (not slings)
- Riser recliner chair and other seating equipment
- Commode
- Headmaster collar
- Suction machine

A proposal can be made to the IMNDA if the patient has a definite equipment need that cannot be met through established sources within the physiotherapy service.
### Appendix 6.6: Evidence supporting physiotherapy in ALS: Review of exercise studies

<table>
<thead>
<tr>
<th>Types of exercise</th>
<th>Studies</th>
<th>Type/Intensity/Frequency</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stretching</td>
<td>No RCT</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Strengthening</td>
<td>Bohannon et al., 1983 Case study</td>
<td>Patterned exercises to most upper limb muscle groups Two sets of 10 repetitions – 2 days per week by physiotherapist, 2 days per week by family, 2 days per week self with theraband</td>
<td>Improved isometric strength in 14 upper limb muscle groups Decreased isometric strength in 4 upper limb muscle groups Subjective improvement in functions</td>
</tr>
<tr>
<td></td>
<td>Drory et al., 2001 RCT</td>
<td>Exercise group prescribed muscle endurance work with UL, LL and trunk major muscle groups working against modest loads but with significant muscle length changes 15 mins, twice daily at home Control group instructed not to perform physical activity outside normal daily activities</td>
<td>10 and 8 intervention group subjects reviewed at 3 and 6 months respectively Significantly less decreased function (ALSFRS) and spasticity (ASS) in the intervention group at 3 months No significant difference in MMT, fatigue or QOL, but more marked decline in control group.</td>
</tr>
<tr>
<td></td>
<td>Dal Bello-Haas et al., 2007 RCT</td>
<td>Stretching exercise programme for UL and LL, to be carried out once daily at home for both the exercise and control group Exercise group also prescribed an individualised moderate-load and moderate-intensity resistance exercises (only if specific muscle group was Oxford grade 3 or greater) to be carried out three times weekly</td>
<td>Significantly less decreased function (ALSFRS-R) and decline in QOL (in a subscore of the QOL SF36) in the intervention group at 6 months Fatigue severity score (FSS) did not differ between groups at 3 or 6 months Trend towards less decline in maximal voluntary isometric contraction (MVIC) in 3 UL and 8 LL muscle groups in the intervention group versus the control</td>
</tr>
</tbody>
</table>
| Aerobic | **Pinto et al., 1999**  
Prospective, single blind controlled trial  
- 20 patients (8 intervention group) | The intervention group performed a treadmill exercise programme using ramp protocol (Bruce or Norris) with assistance of Bipap as required to control peripheral and muscle oxidation. Exercise was applied with goal of attaining an anaerobic threshold in 10-15 mins and ceased with fatigue. The control measure is unknown. | 20 patients completed the study at 1 year.  
Significant difference between groups in the FIM scale, but not on the Barthel.  
Significantly slower clinical course (Spinal Norris) and difference in slope in respiratory function tests observed in the intervention group. |
| --- | --- | --- | --- |
| Sanjak et al., 2010  
Interventional with repeated measures  
- 9 patients  
- Exclusion: Unable to walk | Treadmill exercise programme using up to 40% body weight support 60 minute session with 6 x 5mins exercise interspersed with 6 x 5mins rest, 3 days per week for 8 weeks. Intensity did not exceed level 12-13 on the Borg perceived exertion scale. | 6 subjects reviewed at 8 weeks.  
Statistically significant improvements in walking distance and speed on treadmill 6MWT, and in distance on the ground 6MWT at 4 and 8 weeks.  
Trend towards improvements in walking speed (25FWT) and results showing higher than baseline for upper and lower extremity MVIC at 8 weeks.  
Improvements observed in function (ALSFRS-R), rate of perceived exertion (RPE) and fatigue (FSS) at 8 weeks showing good tolerability to treatment. |
Appendix 6.7: Instructions for use of Peak Flow Meter to record peak cough flow

Indications
A peak cough flow of less than 270 l/min indicates there is ineffective cough generation, and as such should be measured regularly to identify a need for augmented respiratory support and a need for reassessment at tertiary clinic.

Equipment Required
- Peak flow meter modified using a face mask for cough measurement

Technique
- Ensure pointer is set at zero
- Hold peak flow meter so that fingers are clear of scale and slot
- Place mask securely over the patients mouth
- Instruct the patient to inhale deeply and cough as strongly as possible

Return the pointer to zero and repeat 3-5 times. Record the highest score.
Appendix 6.8: Manually Assisted Cough

Indications
Assisted coughing is a longstanding technique used to produce an increase in velocity of expiratory flow sufficient to mobilise secretions from the airways in instances where a patient, due to weakened respiratory muscles, is no longer able to generate this force. As the patient coughs (or is instructed to cough) the therapist applies pressure to increase the strength and effectiveness of the cough.

Technique(s) (in sitting or supine)

Technique 1:
Place the heels of your hands underneath the ribs as illustrated.
As the patient attempts to cough push inwards and upwards.
Technique 2:
Place one forearm across the upper abdomen of the patient with your hand curved around the opposite side of the chest. Your other hand is placed on the near side of the chest. As the patient attempts to cough, push simultaneously inwards and upwards with your forearm, squeezing and stabilising with the other hand.
Technique 3:
Spread your hands anteriorly around the lower rib cage and upper abdomen. With your elbows extended push inwards and upwards with both arms as the patient attempts to cough.

Contraindications / Precautions
Assisted coughing would be contraindicated if medical advice were not sought in the presence of the following:
- Unstable angina or arrhythmia
- Excessive chest trauma, rib fractures/ flail segment, osteoporosis

In these situations assisted coughing **may not be contraindicated** but modifications to technique may need to be applied:

- Poor skin integrity
- Pain
- Spasticity
- Clotting disorders
- Surgical emphysema
- Compromise of spinal alignment
- Chest drain
- Bronchospasm
- Abdominal injury / abdominal wounds
Appendix 6.9: Maximum insufflation - Breathstacking

Performing maximal insufflation prior to a cough will increase inspiratory volume and consequently the expiratory flow and cough efficiency. A form of maximal insufflation should be used to improve effectiveness of a cough in patients who have a vital capacity of less than 1500 ml, or a peak cough flow of less than 270 l/min, as there is ineffective cough generation below these levels. Maximal insufflation also provides a full range of movement to the chest wall therefore consideration should be given to a role in maintaining mobility for function, as well as in secretion clearance (Kang and Bach, 2000).

Indications
Breath stacking enables a deep inspiration thus facilitating higher peak expiratory flow and peak cough flow.

Breathstacking: Using a Bag-Valve-Mask

Equipment Required
- Bag
- One-way valve
- Face mask
Bag-Valve-Mask Breathstacking Technique

- Ask the patient to take a deep breath in and simultaneously squeeze the bag.
- The patient should hold the breath in (the 1-way valve will help this).
- The patient should try to take another breath in on top of their deep breath while you simultaneously squeeze the bag.
- This can be repeated 3-5 times until the patient gets a full deep satisfactory breath in.
- The patient can then exhale or cough
- This can be considered one cycle of breath stacking

The number of cycles to be completed at a time and frequency per day will be advised by the prescribing physiotherapist. This could be between 3-5 cycles, 2-3 times a day.

If the patient has a weak cough, breath stacking can be accompanied by an assisted cough.

Position 1
**Breathstacking: Via Non-invasive Ventilation (NIV)**

**Equipment Required**
A positive pressure device such as the non-invasive positive pressure ventilation (NIPPV) machine as pictured. This will have been prescribed to patients, after specialist respiratory assessment, to manage severe dyspnoea in the later stage of the disease.

**Non-invasive Ventilation Breathstacking Technique**
Instructions for use will depend on the type of machine used, and will be guided by the MND nurse specialist from the tertiary clinic. When using NIV to breathstack, inspiratory pressures can be increased to achieve a maximum insufflation capacity in cycles similar to that of the bag-valve-mask technique, and are often followed by an assisted cough.

**Contraindications / Precautions**
There are no additional contraindications/precautions aside from those that would prevent prescription of non-invasive ventilation in the first instance.
**Breathstacking: Via Glossopharyngeal Breathing**

Glossopharyngeal breathing, also known as “gulping”, involves a series of gulps using the lips, tongue, pharynx and larynx to push air into the lungs when normal inspiratory muscles are not functioning. Glossopharyngeal breathing can augment maximum inspiration in patients and has been reported to increase vital capacity, functional residual capacity and peak cough flow assisting in airway clearance. Limitations in the technique have been reported in patients with oropharyngeal weakness. The technique can be considered difficult and time-consuming to learn and teach but, although the studies discussed in Bott et al, 2009 are small and uncontrolled, all report positive findings, and combined with positive feedback from patients, make it a useful technique to consider.

**Glossopharyngeal Breathstacking Technique**

- A mouthful of air is taken, depressing the tongue, jaw and larynx to get maximum volume.
- The lips are closed and the soft palate raised to trap the air.
- The jaw, floor of the mouth and the larynx are raised. This together with the progressive motion of the tongue, forces the air through the open larynx.
- After as much air as possible has been forced through the larynx it is closed and the air is retained in the lungs until the next cycle is initiated.
- Exhalation occurs when the glottis is opens and the inflated lungs deflate passively due to elastic recoil.
Appendix 6.10: Mechanical In-Exsufflation

Indications
Mechanical in-exsufflation is commonly accompanied by manually assisted coughing, and should be considered in patients with bulbar muscle involvement or any patient who is unable to increase peak cough flow to effective levels with other strategies. It should be considered in patients where secretions are causing discomfort or in those with increasing incidences of chest infections (Sancho et al., 2004; Bott et al., 2009; Bento et al., 2010).

Equipment Required
- Cough Assist Machine
- Tubing
- Large Bacteria Trap
- Cough Assist Face Mask

Cough assist can also be used with a mouth piece, lip seal, tracheostomy and endotrachael tube

Mechanical In-Exsufflation Technique
Instructions for use depend on the type of machine used – The supplier and physiotherapist should be present when the patient receives the device. Pressure cycles of between +60 cmH2O and -60 cmH2O are titrated to suit the individual to optimise the insufflation and
exsufflation required to achieve an effective cough, but should be started low, and increased gradually.

**Contraindications or Precautions**

- Pneumothorax or pneumo-mediastinum
- Facial trauma – interface should be considered
- Tracheoesophageal fistula
- Recent or existing barotraumas
- Bullous emphysema
- Spinal Instability
- High/Low blood pressure – monitor
- Acute pulmonary oedema
- Acute lung injury

**Consumables and cleaning**

- The cough assist facemask should be washed regularly and dried thoroughly prior to use to ensure the filter does not become damp. The facemask should be replaced once worn.
- The bacterial filter should be replaced weekly, with average use (3 times per day).
- If the bacterial filter is at the mask end of the circuit, the tubing can be replaced every 3-4 weeks as the filter prevents contamination of the tubing.
- A change of all components is recommended following a respiratory tract infection
### Appendix 6.11: Modified Borg Breathlessness Scale for Dyspnea

<table>
<thead>
<tr>
<th>Number</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>NOTHING AT ALL</td>
</tr>
<tr>
<td>0.5</td>
<td>VERY VERY SLIGHT JUST NOTICEABLE</td>
</tr>
<tr>
<td>1</td>
<td>VERY SLIGHT</td>
</tr>
<tr>
<td>2</td>
<td>SLIGHT</td>
</tr>
<tr>
<td>3</td>
<td>MODERATE</td>
</tr>
<tr>
<td>4</td>
<td>SOMEWHAT SEVERE</td>
</tr>
<tr>
<td>5</td>
<td>SEVERE</td>
</tr>
<tr>
<td>6</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>VERY SEVERE</td>
</tr>
<tr>
<td>8</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>VERY VERY SEVERE</td>
</tr>
<tr>
<td>10</td>
<td>MAXIMAL</td>
</tr>
</tbody>
</table>
Appendix 6.12: Motor Neurone Disease Pain Pathway (Motor Neurone Disease Networking Group, 2001)

The purpose of the pathway was to reduce variations in treatment; enable the identification and assessment of patient needs; provide guidelines for decision-making and care interventions; provide a structure for patient care and enable improved collaboration and efficiency in the provision of care.

![Motor Neurone Disease Pain Pathway](image-url)