# Neurological Diseases and Palliative Care

St Clare Hospice
Hand in hand we care

## LEARNING TOGETHER STUDY DAY

## 25th June 2024

# Dr Ros Marvin

## Palliative care and symptom management of neurological diseases (including secretions and fatigue)

# Palliative care and symptom management of neurological diseases


**Katy Felgate**

Senior Occupational Therapist

**Dr Ros Marvin**

Consultant in Palliative Medicine

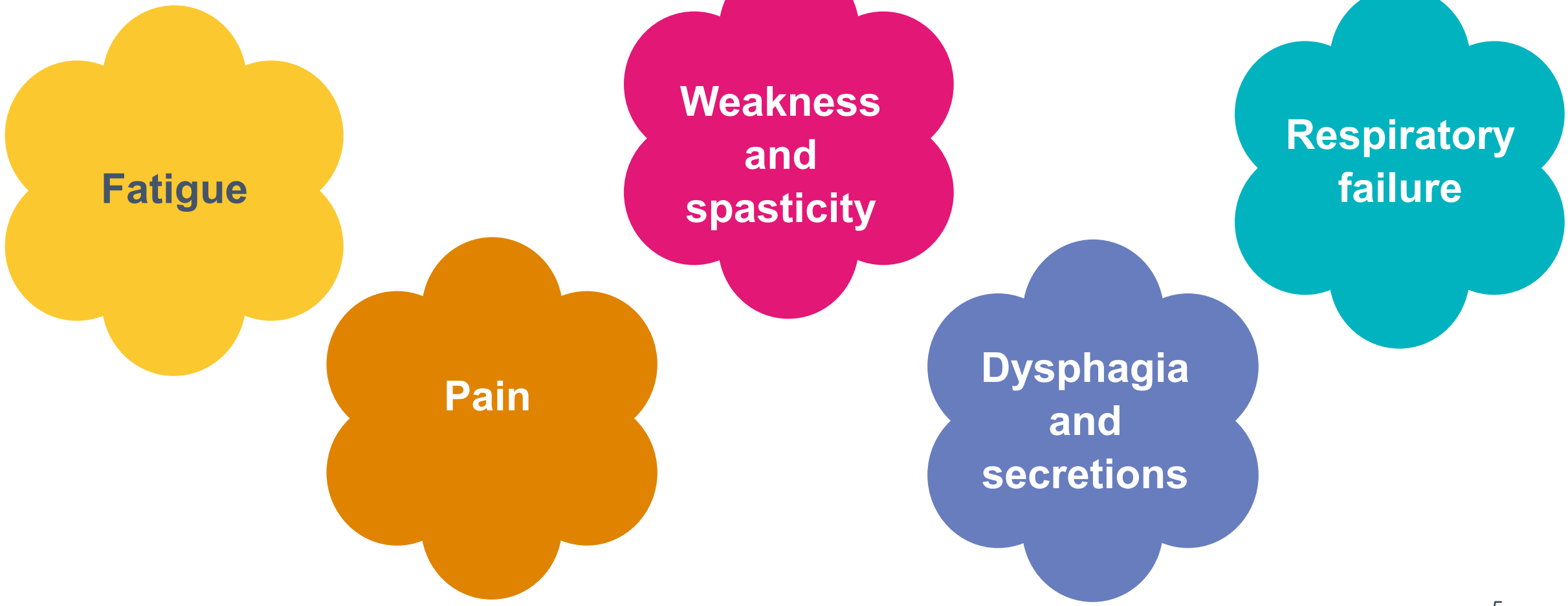

St Clare Hospice

Hand in hand we care

# Aims

- Common symptoms in neurological diseases
- Palliative management – pharmacological and otherwise

# Common symptoms



# What is Fatigue?

Persistent, overwhelming sense of tiredness or exhaustion.

Mental, physical or both.

Disproportionate to the activity level and not relieved by sleep or rest.

# Prevalence

Fatigue impacts up to 45% of patients with a neurological condition (Kluger et al 2013)

Disease specific – 36.5% to 78% of patients with MS (MS Society)

# Non-pharmacological Management of Fatigue

Lack of reliable evidence for neurological conditions (Khan and Amalya, 2018).

**Importance of Addressing Fatigue:** Improves quality of life, emotional well-being, and overall function.

# Causes of Fatigue in Neurological Conditions

**Impaired Nerve Function: Reduced Signal Transmission, Muscle Weakness.**

**Neurotransmitter Imbalances: Chemical Disruptions.**

**Central Nervous System Overload: Increased Cognitive Load, Sleep Disturbances.**

**Immune System Activation: Inflammation.**

**Energy Production Deficits: Mitochondrial Dysfunction.**

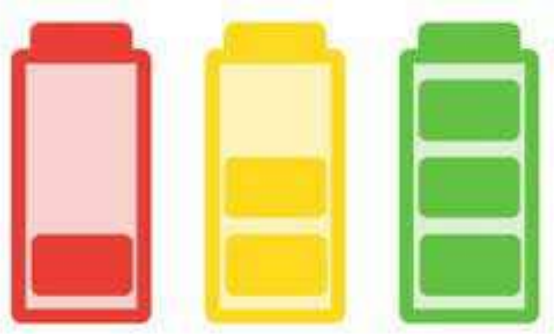
**Psychological Factors: Depression and Anxiety.**

**Medications and Treatments: Side effects.**

# Energy Conservation Strategies

| Prioritising | Planning | Pacing | Positioning | Permission |
| --- | --- | --- | --- | --- |
| What is most important? | The activity or task | Rest before exhaustion | Of self or the activity | To say no or do things differently |
| Ditch, Delay Delegate | Timing, Distance | 20% rule | Equipment | Education |

# Manage Energy to Enable Activity



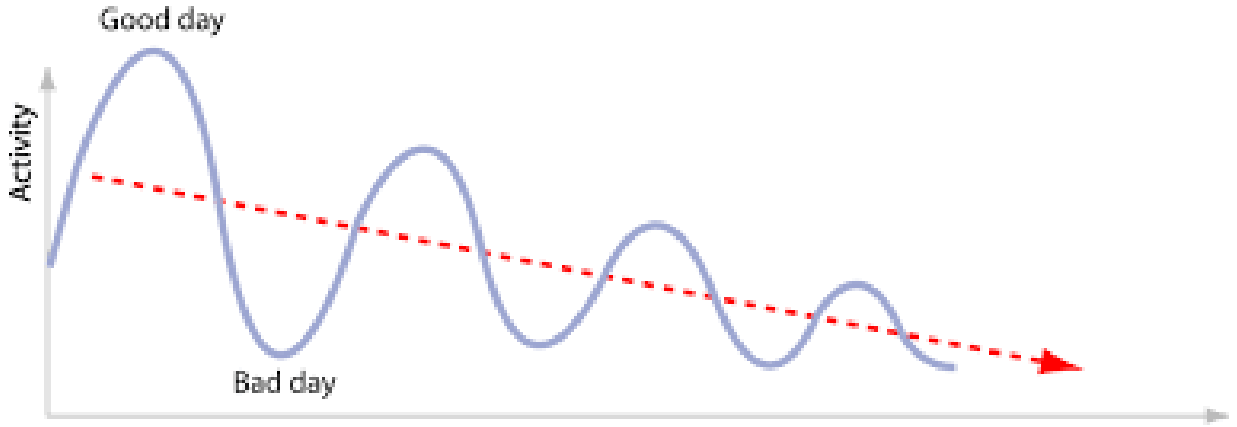
Avoid boom and bust

Management of energy levels – activity diary

Analogies

Spend energy on activities most important

Value based goal setting



# Management of Fatigue

Sleep hygiene advice
Relaxation and mindfulness – anxiety and rest
Nutrition and appetite – reduce caffeine
Adaptations and equipment
Coping with feelings

# Benefits of Exercise for Fatigue

## Spending some energy on an intentional ‘movement’ can help by:

- Building or maintaining cardiovascular fitness (the efficiency of the heart and lungs)
- Building or maintaining your muscle mass (your strength and the body’s energy stores)
- Due to this, daily tasks and chores feel easier
- Helping you sleep better at night
- Improve your balance and your confidence
- Building self-esteem and improving mood
- Bonding with others

# Managing Mental Exhaustion

- Set Boundaries
- Reduce Distraction
- Compensate
- Relaxation
- Seated Leisure Activity

# Education

Provided in a group setting where possible – St Clare Wellbeing Hub

Peer support

Self-management of symptoms

Help patients feel less isolated and misunderstood – invisible symptom

Can improve mood and confidence

Increase social interaction

Increased understanding

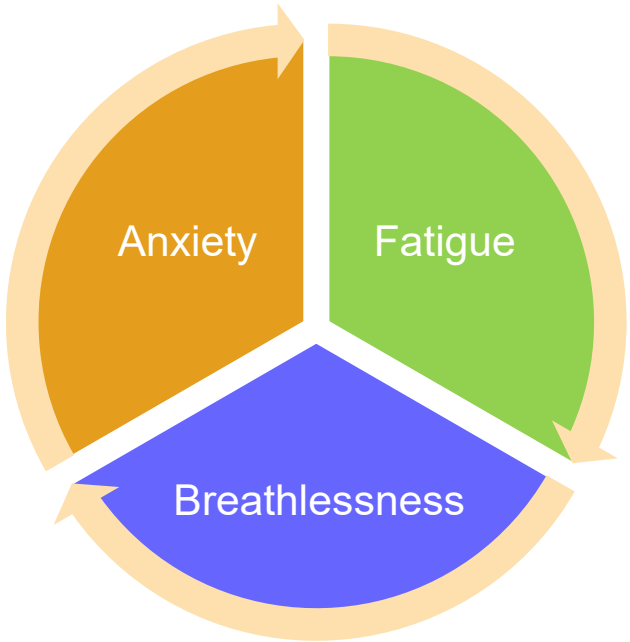
Can be provided 121

# Fatigue and Effect on other Symptoms

Reducing breathlessness to help manage fatigue

Anxiety management

Balance between activity to build strength and conserving energy to build efficiency.

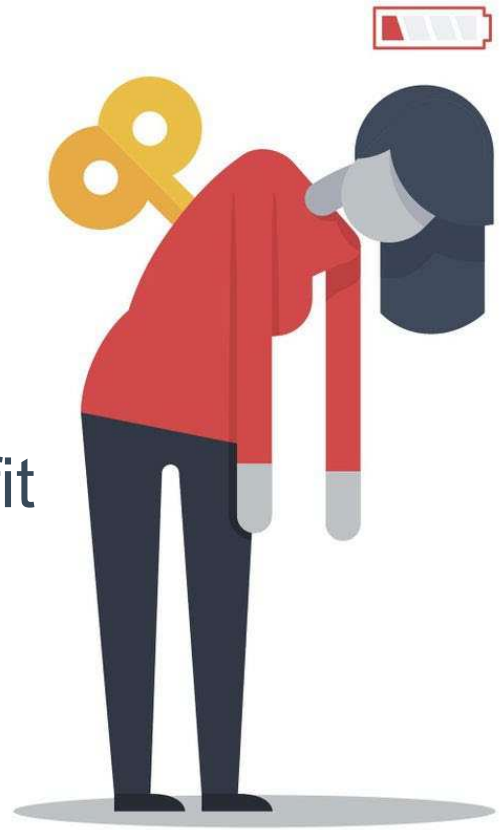


# Fatigue

## Use non-pharmacological strategies first!

Dexamethasone, amantadine, methylphenidate and modafinil have all been used as therapies for fatigue

Unfortunately, the evidence for their use shows little benefit over placebo, and they are not used routinely



# Pain

## Consider underlying cause

- Nerve damage
- Muscle spasm/spasticity
- Immobility causing joint stiffness or pressure sores

Optimise therapy for the underlying disease

Assess pain regularly, using pain scales

Reassess following intervention, to ensure it has been effective

# Pain

## Analgesics:

- **Mild to moderate pain** can often be managed with **non-opioid analgesics** like paracetamol or NSAIDs
- **Opioids**: For severe pain, opioids like morphine or fentanyl may be prescribed, with careful monitoring for side effects. Be aware of the risk of dependency in patients with a longer prognosis
- **Antidepressants and Anticonvulsants**: Neuropathic pain may be treated with medications such as amitriptyline, gabapentin, or pregabalin

# Pain

## Other interventions:

- **Muscle Relaxants**: Spasticity-related pain may be alleviated with baclofen or tizanidine. Benzodiazepines may also be helpful
- **Botulinum toxin**: Botox can be used for focal dystonia which may be painful
- **Nerve blocks or corticosteroid injections** can provide significant relief for localized pain
- **Acupuncture and TENS**: Mixed evidence, but some patients benefit

# Pain

## Non-pharmacological interventions

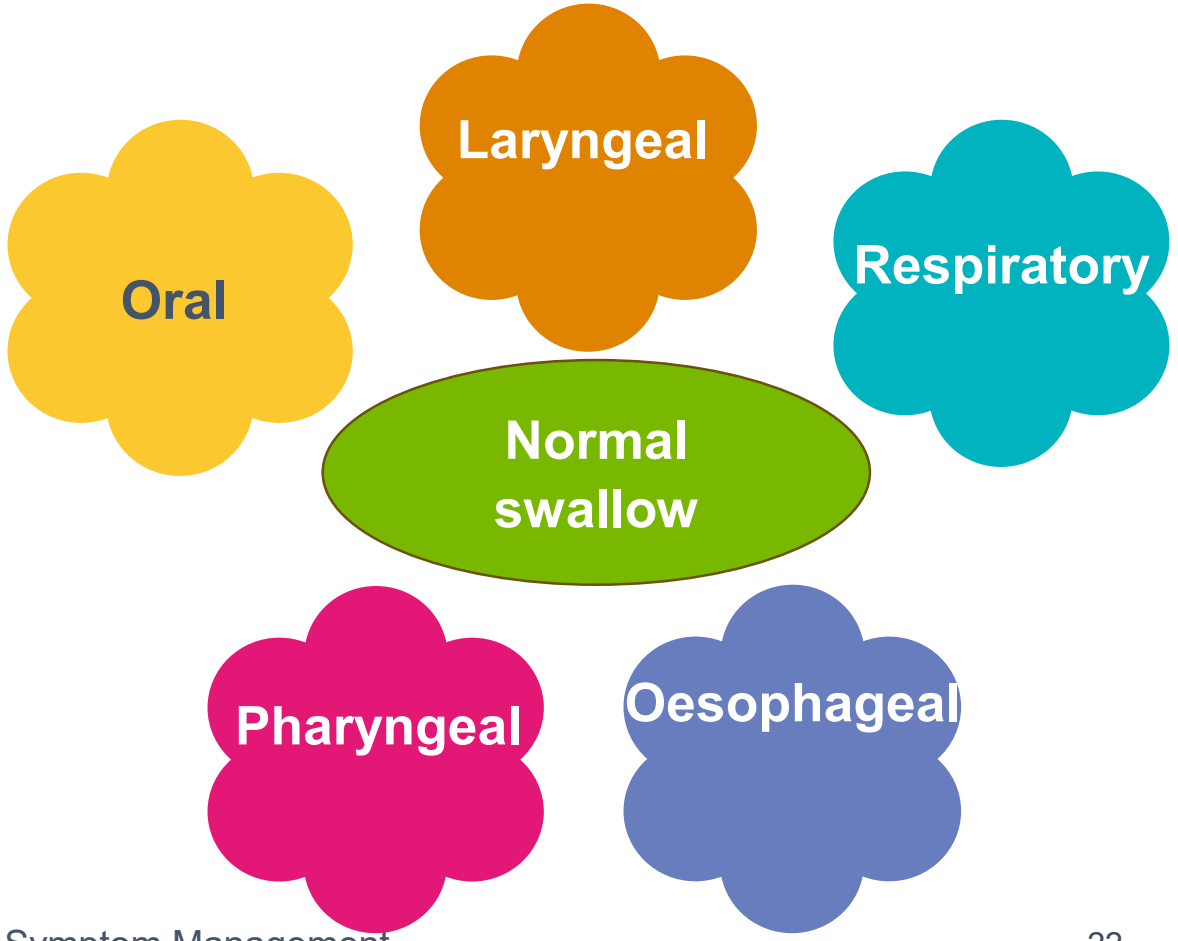
- **Physiotherapy**: Stretching, range-of-motion exercises, and physical therapy can help manage pain from muscle stiffness and spasms
- **Occupational Therapy**: Adaptive devices and techniques to assist with daily activities and reduce strain
- **Massage Therapy**: Gentle massage can alleviate muscle pain and improve circulation
- **Heat and Cold Therapy**: Applying heat or cold packs can reduce muscle pain and inflammation

# Dysphagia (difficulty swallowing)

Multiple anatomical structures need to function in synchrony

Dependent upon the motor and sensory nervous system being intact

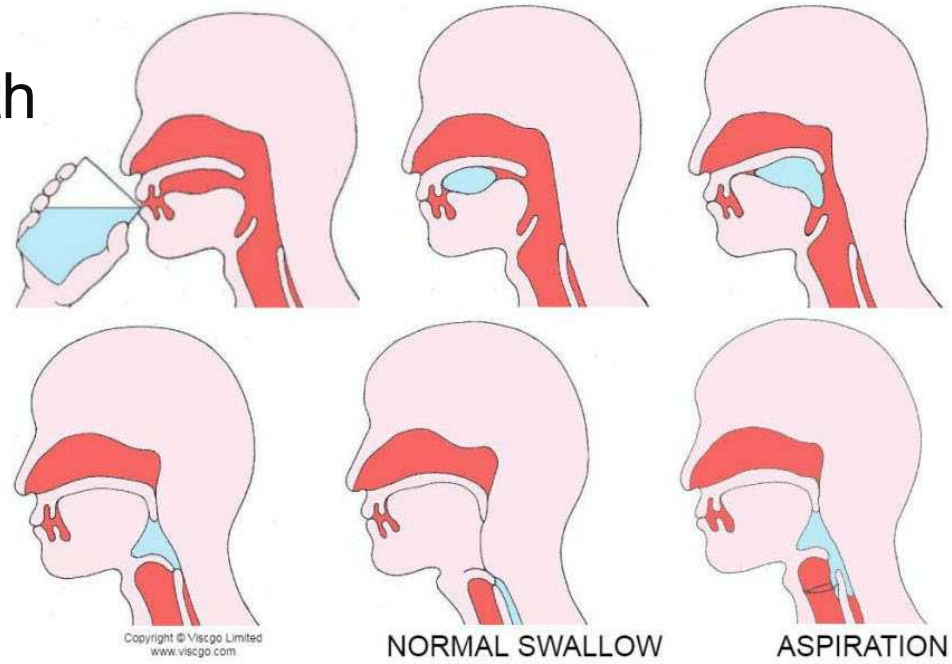
Dysphagia is common in neurological diseases



# Dysphagia

## Increased risk of

- Aspiration pneumonia
- Choking
- Weight loss
- Malnutrition
- Dehydration
- Poor oral health




Therefore increased morbidity and mortality, and reduced quality of life

# Dysphagia

## Symptoms include

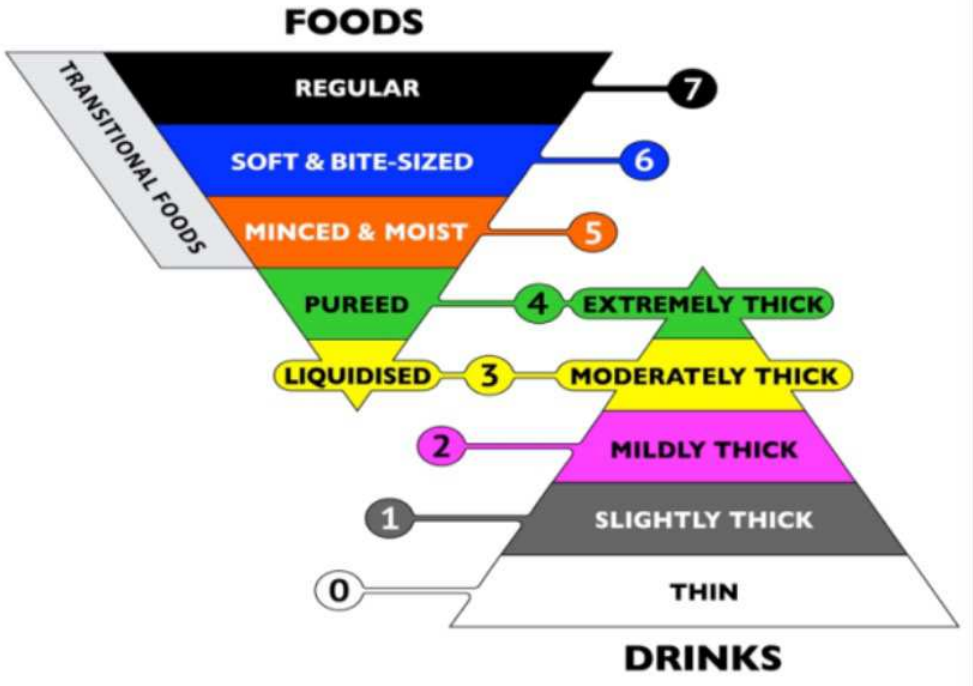
- Food spillage from lips
- Wet voice
- Dry mouth
- Nasal regurgitation
- Regurgitation
- Poor chewing ability
- Drooling
- Coughing and choking

# Dysphagia

## Refer to Speech and Language Therapy when appropriate

Strategies may include:

- Modifying food and liquid consistencies
- Positioning techniques
- Swallowing therapy
- Enteral tube feeding




The International Dysphagia Diet Standardisation Initiative

# Dysphagia

## Is PEG or NG feeding appropriate?

- Decisions are medically and ethically complex
- Weigh up the risks of increasing morbidity and prolonging suffering against the potential benefits
- Patients may lack capacity
- Advance Care Planning
- Multidisciplinary decision – e.g. Addenbrooke's Feeding Issues MDT

# Sialorrhoea (drooling)

## Common in MND

Same amount of saliva, but increasing weakness of muscles in mouth, tongue and throat causes difficulty managing saliva, in the mouth and during swallowing

Worse if the person has a poor lip seal, neck weakness

or swallowing difficulties (dysphagia)

Non-pharmacological:

Posture, positioning, oral care and consider suctioning

# Sialorrhoea (drooling)

## Pharmacological management

First line: Anti-muscarinic medications

- Glycopyrronium bromide, hyoscine butylbromide, hyoscine hydrobromide, atropine, amitriptyline, clonidine
- Choice depends on route (oral, PEG, patch, SC) and side effects (e.g. sedation)

# Sialorrhoea (drooling)

## Further management options

Botox: Botulinum toxin A injected into salivary glands can reduce production for weeks/months (may worsen dysphagia)

Single dose radiotherapy: part of the salivary gland destroyed permanently. Effect occurs gradually (weeks).

# Thick saliva, mucus and phlegm

## Also common in MND

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

- dehydration
- mouth breathing or open mouth posture, which can lead to evaporation of saliva
- non-invasive ventilation (NIV) drying out their airways.

Accumulates due to dysphagia and muscle weakness leading to ineffective cough

Airway occlusion

Distressing for patient and carers

# Thick saliva, mucus and phlegm

## Review medications, especially antimuscarinics

Non-pharmacological:

- Advice on swallowing, diet, posture, positioning
- Increase fluid intake, oral hygiene
- Avoid mouth breathing if possible
- Pineapple or papaya juice – enzymes break down protein in mucous (also available as tablets)
- Trial of reducing dairy intake

# Thick saliva, mucus and phlegm

## Pharmacological management

- Nebulised saline to loosen secretions
- Mucolytics e.g. carbocisteine (break down mucus structure)
- Expectorant e.g. guaifenesin (thins secretions, over the counter cough mixture)
- Beta blockers (need to monitor for hypotension/bradycardia, limited evidence)

- **Difficult balance between too thick and too thin**

# Respiratory Failure

**Weakness of respiratory muscles may lead to respiratory failure – type 1 or 2**

Symptoms and signs:

- Dyspnoea (breathlessness)
- Orthopnoea (SOB when lying down)
- Use of accessory muscles
- Reduced FVC (forced vital capacity) and other pulmonary function tests
- $O_2$ sats
- Arterial blood gases (low oxygen, high $CO_2$)

# Respiratory Failure

## Treating the Underlying Neurological Condition

**Immunosuppressive Therapy**: For conditions like myasthenia gravis (e.g., corticosteroids, IVIG, plasmapheresis).

**Disease-Modifying Treatments**: For multiple sclerosis, such as interferons or monoclonal antibodies.

## Respiratory failure/pneumonia is the commonest cause of death in MND

# Respiratory Failure

## Symptomatic Treatment

**Mucolytics and Expectorants**: To facilitate airway clearance.

•Nebulised saline can enable patients to cough

**Antibiotics**: To treat respiratory infections promptly.

# Respiratory Failure

## Cough Assist

**Mechanical insufflation: exsufflation (MI:E) machine** simulates a cough to aid secretion clearance

## Non-Invasive Ventilation (NIV)

**BiPAP**: Commonly used for patients with MND and muscular dystrophy.

**CPAP**: Used primarily for obstructive sleep apnea, which can coexist with neurological conditions.

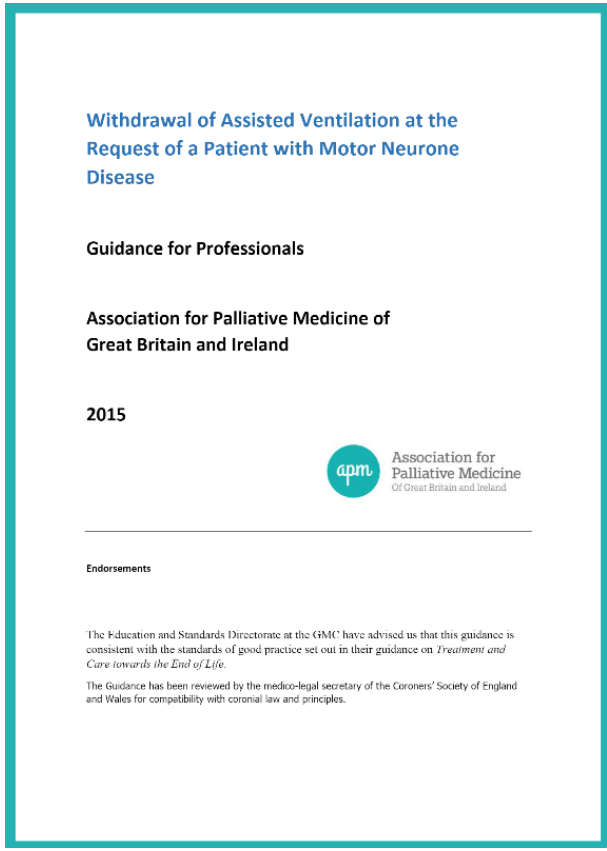
## Invasive Mechanical Ventilation

- **Via tracheostomy**: Not routinely offered, may happen if a person first presents with respiratory failure acutely without planning

# NIV withdrawal

## Patients may choose to discontinue when quality of life worsens

- If a patient is dependent on NIV, withdrawing it will hasten death
- If they have capacity, it is their decision to make
- We can predict that withdrawal will lead to significant symptoms prior to death, therefore sedative medication is used pre-emptively to reduce breathlessness and distress
- Withdrawal ideally should be planned, with palliative care support



Withdrawal of Assisted Ventilation at the Request of a Patient with Motor Neurone Disease

Guidance for Professionals

Association for Palliative Medicine of Great Britain and Ireland

2015

Association for Palliative Medicine Of Great Britain and Ireland

Endorsements

The Education and Standards Directorate at the GMC have advised us that this guidance is consistent with the standards of good practice set out in their guidance on *Treatment and Care towards the End of Life*.

The Guidance has been reviewed by the medico-legal secretary of the Coroners' Society of England and Wales for compatibility with coronial law and principles.

# Last days of life

## Manage holistically

Symptoms managed as for other palliative conditions:

- Anticipatory medications prescribed for pain, breathlessness, nausea/vomiting, secretions, agitation
- Individualised to patient
- Syringe driver when patient unable to swallow

# Final thoughts

**Neurological conditions need multidisciplinary management**

**Don’t forget non-pharmacological management**

**Advance care planning vital, especially discussions regarding nutrition and NIV**

**Symptom management is complex, and may involve trial and error to get the right balance**

**Signpost to hospice services early – including psychological support**


ROB BURROW
‘WE MUST STILL DARE TO DREAM’
DAILY EXPRESS

# References


Fatigue and fatigability in neurologic illnesses - PMC (nih.gov)

Prevalence and burden of multiple sclerosis-related fatigue: a systematic literature review | BMC Neurology | Full Text (biomedcentral.com)

management_of_fatigue_in_neurological_disorders_.4.pdf

**Welbank T, Kurien M. To PEG or not to PEG that is the question. *Proceedings of the Nutrition Society*. 2021;80(1):1-8. doi:10.1017/S002966512000703X** https://www.cambridge.org/core/journals/proceedings-of-the-nutrition-society/article/to-peg-or-not-to-peg-that-is-the-question/2D684AD92D0B3ED1E5AF1EA560A2F970

**Managing saliva problems in motor neurone disease** https://www.mndassociation.org/media/145

**Managing respiratory symptoms in motor neurone disease** https://www.mndassociation.org/sites/default/files/2022-11/P6%20Respiratory%20symptoms.pdf

**Association for Palliative Medicine Guidance on Withdrawal of Assisted Ventilation** https://apmonline.org/apm-professional-guidelines-2/