Symptom management in motor neuron disease

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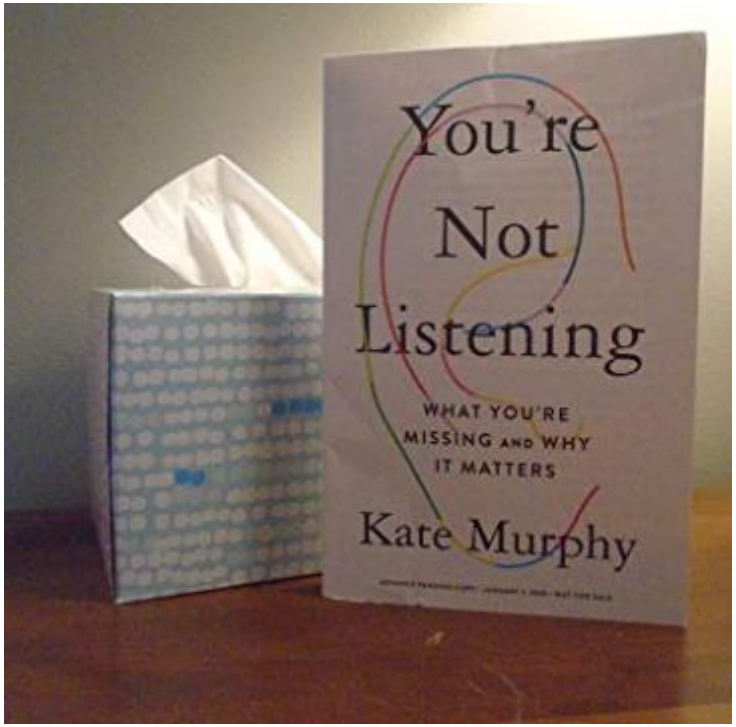

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UMN

- motor cortex in the frontal lobes to activate the LMNs.
- **Spastic muscles**
- **Exaggerated** deep tendon **reflexes**
- upgoing plantars

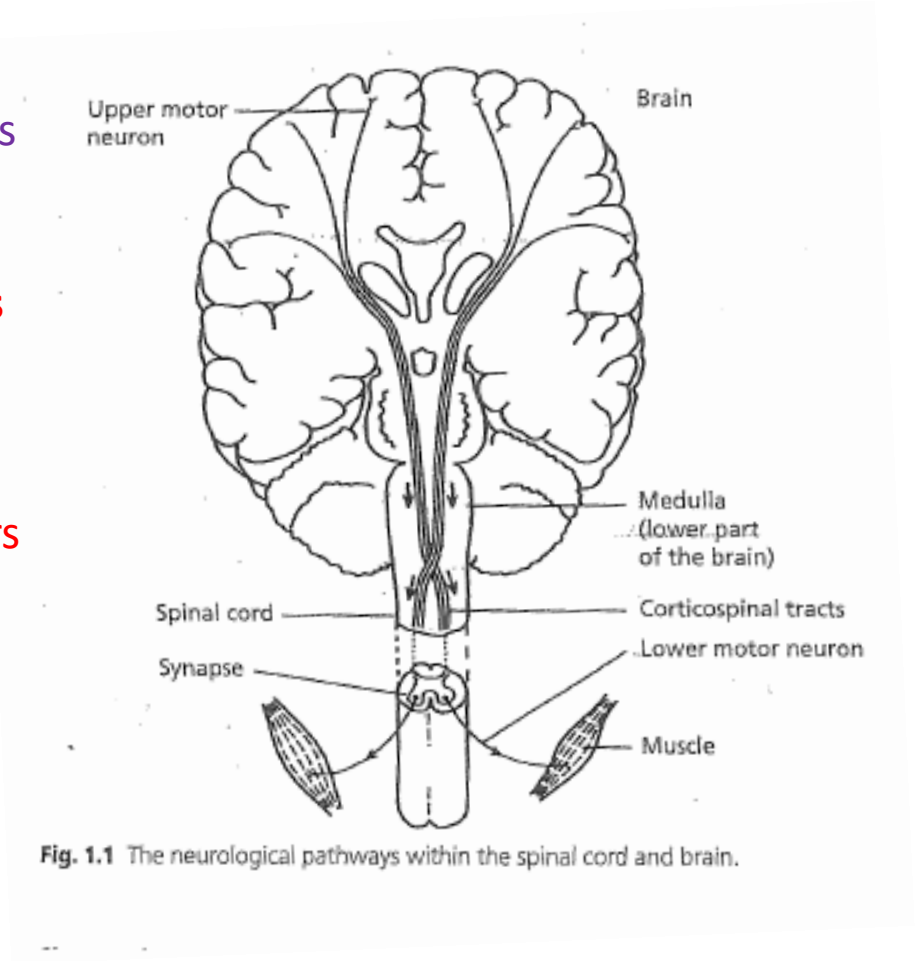

Fig. 1.1 The neurological pathways within the spinal cord and brain.

LMN

- Spinal cord and brainstem
- project out in peripheral nerves & makes direct contact & activates muscle fibres.
- **Weak , wasted muscles** which **fasciculate** (twitch).

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Symptoms due to MND/ALS

Symptoms directly attributed to ALS/MND	Motor	Muscle weakness and atrophy, fasciculations, cramps, spasticity (including laryngospasm, trismus and tongue biting), dyspnoea, dysphagia, dysarthria
	Cognitive	Apathy, behavioural disturbances, impaired decision-making, cognitive impairment, dementia
	Pseudobulbar affect	Pathological laughing and crying
Symptoms indirectly caused by ALS/MND	Pain	Immobility, injury, weak unsupported joints, skin pressure and breakdown, headache, limb pain including dependent oedema, exacerbation of pre-MND conditions
	Secretions	Sialorrhoea (drooling), thick tenacious secretion, nasal congestion, choking
	Urinary	Urinary frequency, incontinence, retention
	Gastrointestinal	GORD, bowel management (including constipation) , weight loss
	Psychological	Fatigue, insomnia, depression and anxiety
	Respiration	Symptoms of chronic hypoventilation , coughing

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Palliative care

- Mainstay of care from diagnosis is palliative
- No cure
- Earlier referral - intact communication & cognition

- *“In the absence of a cure or any medical intervention which might stop the progression of MND , the focus is on symptomatic, rehabilitative and palliative therapy with an overall aim of optimizing QOL “ (Cochrane systematic review 10th January 2017)*

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- Disease starts years/decades before 1st symptoms
- 50% motor neurons degenerate before symptoms
- Disease of progressive loss and increasing disability
- Trajectory variable and uncertain

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Principles of palliative care

- Uses MD team approach to address and integrate physical ,psychological , social and spiritual aspects of care of the patient
- Provides relief from pain and other distressing symptoms
- Offers support system to help family cope during illness and in bereavement period
- Patient – centered care

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Principles of palliative care

- Affirms life.
- Enhances QOL and positively influences course of illness
- Dying is a normal process
- Neither hastens or postpones death

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Total care

Cochrane systematic review of symptomatic treatments in ALS/MND

- Robust evidence lacking
- Not true lack of efficacy
- Study design
- “Highly unlikely that there will ever be classically designed placebo-controlled RCT in this field ”

January 2017

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Dyspnoea

- Diaphragm - inspiratory
- Abdominal - expiratory/cough
- Accessory muscles
- Initially day-time ventilation preserved
- Nocturnal hypoventilation
  - Supine – diaphragm less efficient
  - During REM sleep accessory muscles less active

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Progressive dyspnea

- Orthopnoea
- Exertional dyspnoea
- Dyspnoea at rest
- Use of accessory muscles
- Decreased/absent chest wall movement
- Paradoxical movement of abdomen

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Nocturnal hypoventilation – ask these questions

- Restless disturbed sleep, non-refreshing sleep
- Vivid dreams/nightmares
- Daytime sleepiness
- Mood and personality changes and LOA
- Dry mouth
- Morning headache

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Management of dyspnoea

- Manage secretions
- Manage cough
- NIPPV/invasive ventilation
- Pharmacological/medical
- Manage anxiety
- Terminal hypercapnic coma

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Tenacious secretions

- Hydration
- Mucolytics > acetylcysteine 200 - 400mg tds
- Propranolol/metoprolol
- Nebulise with saline/anticholinergics
- Humidifier, steam inhalation
- Suction

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Tenacious secretions

- Sip fruit juices - red grape juice, pineapple , apple
- Papaya enzymes
- Sugar-free citrus lozenge
- Grape seed oil
- Reduce alcohol, caffeine and dairy

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Develop effective cough strategy

- PCF of > 160L/min to clear airway
- Weak cough > recurrent chest infections
- Cough augmentation :
  - Manual assisted cough
  - Unassisted breath stacking
  - Assisted breath stacking
  - Mechanical cough assist machine

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- Manually assisted cough

https://www.youtube.com/watch?v=KTmELt49TUE

- Breath stacking

https://www.youtube.com/watch?v=JlgeRoI5vCw

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NIPPV

- Increases survival
- Improves QOL = TV
- 4hrs > 24hrs
- Mild-moderate bulbar dysfunction – greatest benefit
- Severe BD – QOL benefit only
- Discuss option to discontinue when initiated

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Common complaints from patients on NIPPV

- Nasal discomfort
- Rhinorrhoea
- Nasal congestion
- Abdominal bloating
- Leaks
- Increasing dyspnoea

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Oxygen / vaccinations

- Hypoxic
- Co-morbid lung disease e.g. COPD
- Intolerant of NIPPV or terminal dyspnoea
- Can reduce respiratory drive
- Severe dry mouth
- Influenza and pneumonia vaccinations

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Medical management

- Semi-recumbent
- Airflow
- Morphine
  - does not worsen respiratory failure
  - reduces tachypnoea
- Benzodiazepines

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Diaphragmatic pacing for chronic resp failure

- QOL and survival benefit
- Electrodes condition and strengthen diaphragm
- Delays need for mechanical ventilation
- Improves survival

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Dysphagia

Choking
Drooling
Aspiration
Malnutrition (16-55%)
Dehydration
LOW
Reduced function
Increasing disability

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Early dysphagia – adaptive strategies

- Positioning – upright
- Postural changes of head
- Dietary modifications
- Safety strategies
- Fatigue - several small meals

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Advanced dysphagia – augmented feeding techniques

- NGT
- Gastrostomy/jejunostomy
- PEG
- RIG
- Jejunostomy
- Palliative surgery rare

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- 41% - Hypermetabolism
- + reduced intake >> LOW
- Food presentation important
- Exercises
- EVOO ??

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Choking

- Poor swallow, weak cough, muscle spasm
- Reduced tongue force
- Delayed triggering of swallowing reflex
- Weakness of laryngeal adduction
- Food spills towards larynx - aspiration vs penetration
- Thin liquids, dry crumbly foods
- Breath stacking/assisted cough/medication
- Death rare

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Dry mouth

- Dry mouth on waking; drooling later in day
- Sip water
- oral lubricants
- Toothpaste and mouth wash – alcohol and SLS free
- Grapeseed oil/peppermint oil
- Bicarb/salt in water

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Sialorrhoea (drooling)

- 50%
- Social isolation
- Not hypersalivation
- Weak facial muscle/reduced swallowing
- Lip closure

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Sialorrhoea

- Glycopyrrolate or buscopan
- Scopaderm patch
- Amitriptyline
- Atropine
- Botilinum toxin type B --**EBT
- Radiotherapy to salivary glands
- Surgery to sever nerve to parotid gland

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GORD

- Weak diaphragm - affects LOS
- PEG overfeeding
- Positioning - avoid meals 2-3 hrs before lying down
- PPI
- Prokinetics

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Pain management – MDT

- Flaccid limbs
  - Careful positioning and support
  - Splints, braces, physiotherapy
- Painful joints
  - Regular passive exercises – ROM
  - Intra-articular injection of LA/steroids
- Paracetamol
- NSAIDS – ibuprofen, naproxen
- Opioids

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Fasciculations

- Fasciculation
- Visible to eye
- Can precede weakness
- Rarely need treatment
- Anecdotally baclofen, gabapentin helpful

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Cramps

- Fasciculations can lead to painful muscle cramps
- Abdominal and paraspinal muscles
- Levitiracetam
- Quinine Europe not USFDA
- Magnesium
- Baclofen
- Diazepam
- Cochrane review Gabapentin, Vit E and riluzole - no benefit

Spasticity

- Assists mobility
- Cochrane review 2017. no benefit -baclofen, gabapentin, BZD, Vit E, riluzole
- Commonly used: baclofen, BZD, dantrolene, gabapentin
- Sativex (THC/cannabidiol) > spasticity in MS. Nabilone
- Physiotherapy - passive movements. RCT x 1; moderate exercise improved outcome at 3 mths
- Severe refractory spasticity: intrathecal baclofen; IM botulinum toxin; IV dantrolene

Weakness > poor mobility

- Cardinal symptom – poor mobility
- Neck weakness/head drop
- Provision of equipment, home adaptation, aids, assistive technology – symbolism
- Physiotherapy
- Limited pharmacological mgt
  - acetylcholinesterase inhibitors (pyridostigmine)
  - Creatine monohydrate

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Nasal congestion

- Due to weakness of nasopharyngeal muscles
- Use nasal tape to elevate nasal bridge at night
- Use nasal decongestants
- MND can exacerbate chronic/seasonal rhinitis

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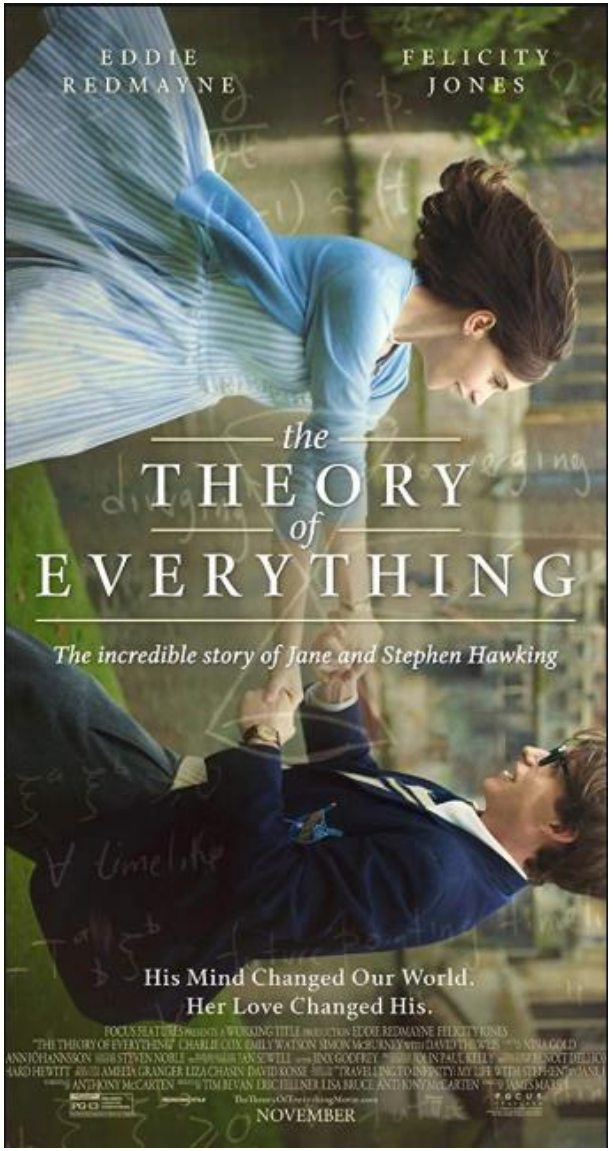
Laryngospasm

- Sudden reflexive closure of vocal cords
- Sensation of choking
- Provoked by diff stimuli
- Self-resolves in seconds. Repeated swallowing while breathing through the nose helps to resolve it
- Trismus, jaw spasm, clenching – precipitated by cold, anxiety, pain
- Benzodiazepines



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Sexual bowel bladder function

- Sexuality is affected NOT sexual function
- Bladder control - weakness of perineal muscles
- Spasticity of bladder - urgency & frequency – oxybutynin, amitriptyline
- Sphincters spared
- Constipation: Power pudding, hydration, laxatives, rotate to fentanyl

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Mood affect

- Reactive depression after diagnosis
- High risk of suicide in 1st year after diagnosis
- Stress and depression in caregiver
- Anxiety
- Denial/anger
- Hopelessness
- Counselling, SSRI, Amitriptyline

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Sleep

- Inability to change position, mood, cramps, fasciculations, dyspnoea, dysphagia, aspiration
- Sedatives impair muscle strength. Use with caution- zopiclone, mirtazapine
- Sleep hygiene

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Pseudobulbar affect – DARWIN

- Involuntary emotional expression disorder
- Abnormal display of affect - Uncontrollable laughter & crying
- NOT a mood or psychiatric disorder
- 50% of patients – socially disturbing
- Found in other CNS diseases
- Amitriptyline; SSRI
- RCT x 2: 30mg dextromethorphan/30mg quinidine BD - approved in US and Europe

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Fatigue

- Fatigue of remaining nerves and muscles
- Reduced intake, LOW
- Reduced ventilation
- Depression
- Modafinil 300mg daily

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Ulcers, oedema ,clots

- Pressure ulcers
- Dependent oedema
  - reduced muscle pump activity
  - Elevation
  - physiotherapy
  - compression hose
  - diuretics
- DVT: higher risk. Untreated high risk for PE

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Cognitive and behavioural dysfunction

- Non-compliance and shortened survival

- Range of phenotypes
  - ALSci – cognitive impairment
  - ALSbi - behavioural impairment
  - ALS-FTD – frontotemporal dementia- 15%

- All can precede physical signs

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Cognitive deficits in MND

- Word-thought generation
- Executive function
- Cognitive flexibility
- Inattention
- Impulsivity
- Memory
- Language difficulties
- Social cognition

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Behavioural changes

- Self-centered
- Apathy, blunted emotions
- Disinhibition/lack of embarrassment
- Irritability, aggression
- Repetitive behaviours, rituals, compulsions
- Lack of interest in hygiene
- Altered sensory behaviour – to heat/cold/pain

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Cognitive dysfunction

- Bulbar vs limb onset: no diff in prevalence
- Bulbar show greater deterioration over time
- CD often improves after NIPPV
- Exclude depression
- Psychotrophic medication - BZD, TCA, riluzole
- CD does not occur in tandem with motor decline

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Interventions for cognitive dysfunction

- Simplify environment and communication
  - Short phrases
  - Stop/think
  - Reduce distractions
  - Memos
- Educate caregivers
  - biological basis for behavioural change
  - Frontal lobe pathology > aggression, impulsivity , irritability
  - Certain behaviours not amenable to change e.g. change in affection

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Interventions for CD

- No medication to improve cognitive outcome
- Disruptive behaviours
  - Anti-psychotics
  - behavioural intervention
    - Distraction
    - Diversion

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ALS-FRS

ALS Functional Rating Scale Revised (ALS-FRS-R)

Date:.....**Name patient:**.....**Date of Birth:**.....

Patient's number.....**Right-/left-handed**

Item 1: SPEECH

- 4 ☐ Normal speech process
- 3 ☐ Detectable speech disturbance
- 2 ☐ Intelligible with repeating
- 1 ☐ Speech combined with non-vocal communication
- 0 ☐ Loss of useful speech

Item 2: SALIVATION

- 4 ☐ Normal
- 3 ☐ Slight but definite excess of saliva in mouth; may have nighttime drooling
- 2 ☐ Moderately excessive saliva; may have minimal drooling (during the day)
- 1 ☐ Marked excess of saliva with some drooling
- 0 ☐ Marked drooling; requires constant tissue or handkerchief

Item 3: SWALLOWING

- 4 ☐ Normal eating habits
- 3 ☐ Early eating problems – occasional choking
- 2 ☐ Dietary consistency changes
- 1 ☐ Needs supplement tube feeding
- 0 ☐ NPO (exclusively parenteral or enteral feeding)

Item 4: HANDWRITING

- 4 ☐ Normal
- 3 ☐ Slow or sloppy: all words are legible
- 2 ☐ Not all words are legible
- 1 ☐ Able to grip pen, but unable to write
- 0 ☐ Unable to grip pen

Item 5a: CUTTING FOOD AND HANDLING UTENSILS
Patients <u>without</u> gastrostomy → Use 5b if >50% is through g-tube

- 4 ☐ Normal
- 3 ☐ Somewhat slow and clumsy, but no help needed
- 2 ☐ Can cut most foods (>50%), although slow and clumsy; some help needed
- 1 ☐ Food must be cut by someone, but can still feed slowly
- 0 ☐ Needs to be fed

Item 5b: CUTTING FOOD AND HANDLING UTENSILS
Patients <u>with</u> gastrostomy → 5b option is used if the patient has a gastrostomy and only if it is the primary method (more than 50%) of eating .

- 4 ☐ Normal
- 3 ☐ Clumsy, but able to perform all manipulations independently
- 2 ☐ Some help needed with closures and fasteners
- 1 ☐ Provides minimal assistance to caregiver
- 0 ☐ Unable to perform any aspect of task

Item 6: DRESSING AND HYGIENE

- 4 ☐ Normal function
- 3 ☐ Independent and complete self-care with effort or decreased efficiency
- 2 ☐ Intermittent assistance or substitute methods
- 1 ☐ Needs attendant for self-care
- 0 ☐ Total dependence

Item 7: TURNING IN BED AND ADJUSTING BED CLOTHES

- 4 ☐ Normal function
- 3 ☐ Somewhat slow and clumsy, but no help needed
- 2 ☐ Can turn alone, or adjust sheets, but with great difficulty
- 1 ☐ Can initiate, but not turn or adjust sheets alone
- 0 ☐ Helpless

Item 8: WALKING

- 4 ☐ Normal
- 3 ☐ Early ambulation difficulties
- 2 ☐ Walks with assistance
- 1 ☐ Non-ambulatory functional movement
- 0 ☐ No purposeful leg movement

Item 9: CLIMBING STAIRS

- 4 ☐ Normal
- 3 ☐ Slow
- 2 ☐ Mild unsteadiness or fatigue
- 1 ☐ Needs assistance
- 0 ☐ Cannot do

Item 10: DYSPNEA

- 4 ☐ None
- 3 ☐ Occurs when walking
- 2 ☐ Occurs with one or more of the following: eating, bathing, dressing (ADL)
- 1 ☐ Occurs at rest: difficulty breathing when either sitting or lying
- 0 ☐ Significant difficulty: considering using mechanical respiratory support

Item 11: ORTHOPNEA

- 4 ☐ None
- 3 ☐ Some difficulty sleeping at night due to shortness of breath, does not routinely use more than two pillows
- 2 ☐ Needs extra pillows in order to sleep (more than two)
- 1 ☐ Can only sleep sitting up
- 0 ☐ Unable to sleep without mechanical assistance

Item 12: RESPIRATORY INSUFFICIENCY

- 4 ☐ None
- 3 ☐ Intermittent use of BiPAP
- 2 ☐ Continuous use of BiPAP during the night
- 1 ☐ Continuous use of BiPAP during day & night
- 0 ☐ Invasive mechanical ventilation by intubation or tracheostomy

Interviewer's name.....

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Poor prognostic factors

- Bulbar presentation – speech/swallowing problems
- Weight loss
- Poor respiratory function
- Older age
- Lower ALSFRS score
- Shorter time from first symptom to time of diagnosis

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End of life

- Difficult to recognize
- Individual variation
- Gradual insidious
- Specific triggers for MND
  - Respiratory failure
  - Declining mobility
  - Dysphagia
  - Repeated aspiration pneumonia
  - Weight loss
  - Marked general decline

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EOL medications

- Subcutaneous infusion
- Opioids for pain/sob
- Midazolam for stiffness/agitation
- Anticholinergics for secretions i.e. buscopan
- Haloperidol for agitation/nausea

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“I don't have much positive to say about motor neuron disease, but it taught me not to pity myself because others were worse off, and to get on with what I still could do. I'm happier now than before I developed the condition.”

“My advice to other disabled people would be, concentrate on things your disability doesn't prevent you doing well, and don't regret the things it interferes with. Don't be disabled in spirit as well as physically.”

Stephen Hawking

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