Caring for people with Motor Neurone Disease (MND)

National Clinical Specialist Team
2017
Outline

• What is MND?
  – MND in Scotland
  – Types
  – Symptoms
  – Treatments

• MND infrastructure in Scotland
  – CARE-MND
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Motor Neurone Disease: Key Points

- A neurodegenerative disease
- Rapidly progressive
- No cure
MND in Scotland

- Median survival 3 years after symptom onset
- Approximately 200 incident cases/year in Scotland
- Prevalence of approximately 450
Motor Neurone Disease

- Amyotrophic Lateral Sclerosis
- Primary Lateral Sclerosis
- Progressive Muscular Atrophy
- Progressive Bulbar Palsy

COGNITIVE IMPAIRMENT
Amyotrophic Lateral Sclerosis (ALS)

- Approx. 85% of patients have ALS
- ‘Spinal’ = limb onset or ‘Bulbar’ = speech/swallow onset
- Typically progresses to involve all limbs, bulbar muscles and respiratory muscles

Swinnen & Robberecht (2014)
Pathophysiology

• Anatomy: Motor neurones

• Pathology: Motor neurone degeneration/death

• Pathophysiology: Genetic – familial and sporadic; emerging field

• Investigations: Electrophysiology
Diagnosis of MND

• Clinical diagnosis defined by progression
  – Average time from symptom onset to diagnosis is about 12 months

• Patients are allocated a clinical specialist at time of diagnosis
Clinical Assessment

• MND management requires regular rapid ongoing assessments

• Anticipatory care should commence at point of diagnosis
Treatment Options in MND

Symptoms?

Drugs?

Interventions?
MND by Symptoms

• Bulbar Dysfunction
• Upper and Lower Limb Dysfunction
• Respiratory Dysfunction
• Emotional lability and Cognition
Bulbar Dysfunction

- Affects tongue muscles, facial muscles and pharyngeal muscles
Bulbar Dysfunction

- Speech (dysarthria)
- Communication
- Communication Aids
- Swallowing (dysphagia)
- Hydration
- Nutrition
- Saliva
- Gastrostomy
Communication: Strategy

• Face your patient and be at eye level so you can hear and see articulation of words

• Turn off distractive devices

• Allow for extra time

• Don’t answer for the patient and if you do not understand them ask them to repeat themselves or write it down
Augmentative & Alternative Communication (AAC)

- **Low-tech:** Paper and pen, Word Boards – very common, especially if patient is not familiar with technology

- **High-tech:** iPads, Lightwriter, Eye-Gaze
Nutrition and Hydration

• Metabolic rate in MND is doubled, weight loss is expected
• A weight loss of >10% at diagnosis is a poor survival indicator
• Patients require dietetic advice from diagnosis and reviewed throughout the disease
  1. Give small high energy supplements regularly
  2. Consideration of upper limb (hand to mouth action) is important, carers may need to help
  3. Early insertion of gastrostomy tubes should be clinical priority
Gastrostomy

- PEG, RIG, PIGG or NG are used
- Some family members/carers are involved in the use of these
Symptomatic Treatments

**Sialorrhoea**
- Hyoscine/Buscopan
- Glycopyrronium (especially if cognitive impairment)
- Botox
- Suction/Humidification/Carbocisteine

**Nutrition**
- Supplements/Thickeners
- Liquid drug preparations
Spinal Dysfunction: Upper and Lower Limbs

- Variable
- Muscle wasting
- Loss of tone or contractures
- Consider: washing, dressing, feeding, turning in bed
- Mobility aids/hoists often necessary
Symptomatic Treatments

**Muscle cramps**
- Quinine
- Baclofen

**Muscle spasms**
- Baclofen
- Tizanidine
- Dantrolene
- Gabapentin
Respiratory Dysfunction

• Weakness of respiratory muscles is one of the main causes of death in MND

• Respiratory management from diagnosis is vital for quality of life and survival

• Assess using MND red flag signs

• Non–invasive ventilation is offered to support Type 2 respiratory failure
Red flag signs of respiratory failure

- Breathlessness
- Orthopnea
- Recurrent chest infection
- Disturbed sleep
- Non-refreshed sleep
- Nightmares
- Daytime sleepiness
- Poor concentration
Non-invasive Ventilation

- BiPAP mask with adjustable settings
- Commenced at night initially and gradually increased depending on need
Symptom Control for Breathlessness

- NIV is not for everyone
- Symptom control should be commenced
- Patients wishes should be noted
- Advanced care planning up to date
Symptomatic Treatments

- Shortness of breath/
  Anxiety: Lorazepam

- Coughing: Breath stacking, Cough assist
Withdrawal of NIV

- Needs to be supported by the patient, family and healthcare professionals

- Ensure someone familiar with the machine is involved

- Ensure up to date DNACPR, AWI, POA
Emotional Lability

• Inappropriate crying or laughing

• Part of the disease (an Upper Motor Neurone sign)

• Patience and understanding not pity

• Sometimes treated with antidepressants
Cognitive Impairment

• 50% of patients may have cognitive impairment – spectrum from mild to dementia

• Associated with Frontotemporal Dementia

• Apathy, disinhibition, poor planning/decision making

• Be aware of capacity if severely affected
Palliative Care

• Underpins *all* of these interventions

• May be referred for specialist input for:
  – Current or anticipated complex needs:
    • Psychological distress
    • Social distress
    • Troublesome or rapidly progressing symptoms
  – Complex future care planning needs

*NICE 2016*
Key Challenges

- Communication and capacity
- Young patients
- Complex and rapidly progressing needs
- ?
Carer burden

- Respite care / Day services
- Offer support and education to family/carers
- Bereavement care
- ~10% of MND is familial
CARE-MND Team

Person with MND

- MND Clinical Specialist
- Psychology
- Neuro
- Palliative Care
- Resp Team
- SLT/Physio/OT
- Research
- Dietetics/Gastro
We Aim To Engage Every Person Living With MND In Scotland In Shaping Clinical Care And Research
Research

- Scottish MND Register
- Genetic research
- Clinical trials
- Introduced by Clinical Specialist, Nurse Consultant, Researchers
Recommended reading and websites

2016 NICE guidelines
https://www.nice.org.uk/guidance/ng42

MND Scotland
www.mndscotland.org.uk/

MNDA
https://www.mndassociation.org

RCN
www.mnd.rcnlearning.org.uk

Communicating with patients
https://vimeo.com/179918383

CARE-MND Website:
www.care-mnd.org.uk

MyNIV

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