MOTOR NEURONE DISEASE

Caroline Davis
MND Clinical Nurse Specialist
The name of a group of related diseases that attack the motor neurones
MOTOR NEURONES

Healthy motor neurone

Motor neurone affected by MND

- Cell body (receives message to move)
- Axon (transports messages)
- Muscle

- Affected nerve cell
- Muscle after wasting

MOTOR NEURONES
**LOWER MOTOR NEURONE DYSFUNCTION**

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Signs</th>
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<tbody>
<tr>
<td>Fatigue</td>
<td>Weakness</td>
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<tr>
<td>Weakness</td>
<td>Atrophy</td>
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<tr>
<td>Cramps</td>
<td>Fasiculation</td>
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<tr>
<td>Muscle twitching</td>
<td>Supressed reflexes</td>
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<tr>
<td>In-coordination</td>
<td>Hypotonia (low muscle tone)</td>
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</table>
UPPER MOTOR NEURONE DYSFUNCTION

Symptoms
• Weakness
• In-coordination
• Stiffness
• Slowing of movement

Signs
• Spasicity
• Brisk reflexes
• Babinski sign
• Weakness
• Emotional Lability
Four Main Types of MND:

• Amyotrophic Lateral Sclerosis
• Progressive Bulbar Palsy
• Progressive Muscular Atrophy
• Primary Lateral Sclerosis
AMYOTROPHIC LATERAL SCLEROSIS

- Most common form
- Involves upper and lower motor neurones
- Weakness, wasting in limbs
- Muscle stiffness and cramps
- Life expectancy is 2-5 years from onset symptoms
PROGRESSIVE BULBAR PALSY

- Approx. quarter of patients
- Involves upper and lower motor neurones
- Early stages characterised by involvement of bulbar muscles
- Slurring of speech, difficulty swallowing
- Average life expectancy 6 months to 3 years from onset of symptoms
PROGRESSIVE MUSCULAR ATROPHY

- Affects 5-7% of people with MND
- Mainly damage to lower motor neurone
- Characterised by progressive muscle weakness, fasiculations, shrinkage in muscle bulk and weight loss.
- Average life expectancy is 5-10 years
PRIMARY LATERAL SCLEROSIS

• Affects a small proportion of people with MND
• Damages only the upper motor neurones
• Spasticity and weakness
• Speech and swallowing difficulties
• Average life expectancy may be more than 10 years
DIAGNOSIS

- Blood Tests
- Electromyography (EMG)
- Nerve Conduction Tests
- Magnetic Resonance Imaging (MRI) scans
- Neurological Examination
- El Escorial Criteria for ALS
WHO GETS MND?

• Most people diagnosed are over 40
• Highest incidence between 50-70 years of age
• Men are twice as more likely then women to be affected
• Incidence of people who will develop MND per year is approx 2 in every 100,000 people
• Prevalence of people living with MND at any one time is approx 7 in 100,000 people
CAUSES

GENETIC FACTORS

Usual

10% of patients
TREATMENT

• Riluzole
  Anti-glutamate
  Blood tests needed
  Through QE (unless GP says yes!)

• The care cure
• Symptom Control
SYMPTOMS

- Vary between patients greatly
- Every experience of MND is unique
MUSCLE WASTING

- Immobility
- Uncomfortable
- Care needs
- Activities of everyday living
- Communication
- Eating Drinking
WHAT CAN HELP?

- Physiotherapy assessment
- Occupational Therapy Assessment
- Mobility aids
- Care provision
- Aids
- Adaptations to house - minor and major
- Environmental controls
MUSCLE STIFFNESS, FASCICULATION AND CRAMPING

- Annoying
- Uncomfortable
- Painful
- Interrupts sleep

- Medications e.g Baclofen, quinine
- Fasciculations - can reduce over time
SWALLOWING - DYSPHAGIA

- Reduced nutritional intake
- Fatigue when eating
- Embarrassment
- Discomfort
- Dehydration/ malnutrition
- Aspiration pneumonia
WHAT CAN HELP?

- SALT / Dietician assessment
- Thickened fluids
- Softer diet
- Positional
- Swallow techniques
- Eating aids- one way valve straws
- Taking time
- Smaller meals
- Food supplements
- PEG/RIG
- NG Tube
Patients all have a lung function test at QE on diagnosis.
If no respiratory issues: PEG at QE (2-3 weeks)
If resp issues: PEG at Stoke Hospital under NIV (8 week+)
All patients should be referred to Birmingham Nutrition team.

Concerns:
- Not quick enough? NG in community?
- Transport issues!
- Family visiting
- NIV/suction/ cough assist at same time?
- Winter pressures!
- RIG better?
May affect up to 50% of patients with MND

Approx 2-3 pints of saliva produced a day

Bulbar muscle weakness can cause pooling of saliva / difficulty managing saliva in the mouth (dysphagia)

Thick and sticky or thin and pools

Thick mucus in throat

Causes choking; drooling; increases risk of aspiration; embarrassment; discomfort
Detailed discussion with patient to determine:

- thick, sticky and tenacious?
- thin and watery – drooling (sialorrhoea)
- Mixture of the 2
- How much saliva? Time of the day?
- Thick mucus?

Speech and Language Therapy Assessment

- Assessing swallow and lip seal etc
- Oral care routine

Frontal Temporal Dementia/ cognitive impairment

Respiratory Assessment

Occupational Therapy Assessment
INTERVENTIONS

- Reducing production of saliva
- Changing character of saliva
- Aiding clearance
THIN SALIVA MANAGEMENT

- Speech and language Therapist
  - Swallow techniques
  - Barrier cream to protect skin
  - Risk of aspiration

- Occupational Therapist:
  - Good posture to improve flow of saliva
  - Neck collars
  - Rise and recline chair/ wheelchaire to improve posture
  - Positioning at night
THIN SALIVA MANAGEMENT

- Respiratory Team Assessment
  - Suction machine provision
  - Subtle clothing protectors
  - Medications prescribed by GP,
  - Consultant Neurologist, Palliative
  - Care Consultant or Nurse Prescriber
Anti-muscarinic medication as first line treatment (NICE guidelines 2016)

- Hyoscine Hydrobromide (Scopoderm)
- Hyoscine Butylbromide (Buscopan)
- Atropine eyedrops
- Glycopyrronium Bromide (glycopyrrolate)
- Tricyclic Antidepressant (Amitriptyline)
  - Side effects: Constipation, confusion in elderly, skin dryness, urinary retention or urgency (see BNF for further)

In cognitive impairment Glycopyrronium first line treatment
If medications and other intervention fail then Botox injection into the salivary glands

- Done by Specialist
- Botox Toxin A first line
- Botox Toxin B if natural immunity
- Can last up to 3 months
- PEG in situ is required
- Dr Soryal in QE

Radiotherapy of saliva glands (x-rays to destroy saliva glands)
THICK SALIVA AND MUCUS MANAGEMENT

- Speech and Language Therapy Assessment
  - Swallow assessment
  - Oral hygiene advice (District Nurses)
  - Mouth breathing?
  - Review of current meds (overdrying with anti-muscarinics?)
  - Hydration? PEG needed urgently
  - Lollies, jelly, frozen mousses, sucking ice
  - Pineapple juice and papaya juice before meals
  - Wiped around mouth with pink sponge
  - Sucking on sugarless sweets (choking risk though)
THICK SALIVA AND MUCUS MANAGEMENT

Respiratory Team Assessment
- NIV drying mouth?
- Humidification through NIV
- Saline nebs prescribed
- Weak cough for mucus:
- Unassisted breath stacking
- Assisted breath stacking
- Cough Assist
- Suction Unit

Dr Mustfa, Respiratory Consultant and CNS team: Royal Stoke Hospital
THICK SALIVA AND MUCUS MEDICATIONS

- Mucolytics (carbocisteine)
  - Caution: peptic ulcer
- Saline – through nebuliser
DRY MOUTH

- Artificial saliva
- Oral hygiene
- Fluid increase
- Checking for oral thrush
- Medication review
GOOD SALIVA AND MUCUS MANAGEMENT

- Improvement of quality of life
- Reduction in aspiration pneumonia
- Reduction in hospital admission
Managing Saliva Problems in Motor Neurone Disease – MNDA information booklet P3 for professionals.

NICE Guidelines for Motor Neurone Disease, February 2016. Saliva Management

• Very distressing
• Frustrating
• Advance Care Planning
Communication aids

Low-tech
- Alphabet sheets
- Communication books with pictures

High-tech
- Light-writer
- Tablets with apps on
- Augmentative and Alternative Communication (eye-gaze)
MND doesn’t affect the sensory nerves

However:

- muscle cramps
- Spasticity
- stiff joints
- muscle spasms
- skin pressure
- constipation
Up to 35% of people with MND affected with some cognitive change
- Other causes e.g. dehydration, fatigue, rise in CO2
- Frontal Temporal Dementia - very small percentage
- Psychological services
- Improving causes
- Tested by ECAS
Edinburgh Cognitive and Behavioural ALS Screen
20 minutes
Completed by any health professional
Looks at cognition specific to ALS
Behaviour issues in FTD
Forward referral
Capacity issues

BEHAVIOUR SCREEN – Carer Interview

- Please ask the carer about the following possible behaviours. Symptoms should have occurred repeatedly and not just on one instance, and may have occurred prior to the development of any motor signs. Tick 'Yes', 'No' or 'Don't Know'. If 'Yes', please provide a brief written description. Give one mark for every 'Yes' response (maximum = 10).

### A Behavioural disinhibition

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<tbody>
<tr>
<td>1</td>
<td>Socially inappropriate behaviour, e.g. inappropriate behaviour with strangers, criminal behaviour</td>
<td>Y</td>
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<tr>
<td>2</td>
<td>Loss of manners or decorum, e.g. crude or sexually explicit remarks, jokes or opinions that may be offensive to others, lack of response to social cues</td>
<td>Y</td>
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<tr>
<td>3</td>
<td>Impulsive, rash or careless actions, e.g. new onset gambling, buying or selling property without regard for consequences, giving out personal information inappropriately, e.g. credit card numbers</td>
<td>Y</td>
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### B Apathy or inertia

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<td>4</td>
<td>Loss of interest, drive or motivation, e.g. passivity and lack of spontaneity, needs prompting to initiate or continue routine activities</td>
<td>Y</td>
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### C Loss of sympathy or empathy

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<td>5</td>
<td>Diminished response to other people’s needs and feelings. Positive rating on this feature should be based on specific examples that reflect a lack of understanding or indifference to other people’s feelings, e.g. hurtful comments, disregard for others’ pain or distress</td>
<td>Y</td>
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https://ecas.psy.ed.ac.uk/
EMOTIONAL LABILITY

- Uncontrolled crying or laughing
- Inappropriate
- Embarrassing
- Socially isolating
- People feel they are ‘going mad’

- Medication such as anti-depressants can help
PSYCHOLOGICAL EFFECTS

• Changes in relationships
• Coming to terms with terminal illness
• Disability
• Loss of independence
• Future plans
• Distressing symptoms
PSYCHOLOGICAL EFFECTS

Used with permission of the patient
**RESPIRATORY MUSCLE WEAKNESS**

Muscle groups involved in deep breathing (neck, intercostals, diaphragm)

- windpipe (trachea)
- bronchus
- lung
- alveoli
- diaphragm

_Muscles involved in breathing_
<table>
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<tr>
<th>Symptoms</th>
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<tbody>
<tr>
<td>• Breathlessness</td>
<td>• Increased respiratory rate</td>
</tr>
<tr>
<td>• Orthopnoea</td>
<td>• Shallow breathing</td>
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<tr>
<td>• Recurrent chest infections</td>
<td>• Weak cough[a]</td>
</tr>
<tr>
<td>• Disturbed sleep</td>
<td>• Weak sniff</td>
</tr>
<tr>
<td>• Non-refreshing sleep</td>
<td>• Abdominal paradox (inward movement of the abdomen during inspiration)</td>
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<tr>
<td>• Nightmares</td>
<td>• Use of accessory muscles of respiration</td>
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<tr>
<td>• Daytime sleepiness</td>
<td>• Reduced chest expansion on maximal inspiration</td>
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<tr>
<td>• Poor concentration and/or memory</td>
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<tr>
<td>• Confusion</td>
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<tr>
<td>• Hallucinations</td>
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<tr>
<td>• Morning headaches</td>
<td></td>
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<tr>
<td>• Fatigue</td>
<td></td>
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<td>• Poor appetite</td>
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[a] Weak cough could be assessed by measuring cough peak flow.
NON-INVASIVE VENTILATION (NIV)

- Symptom control
- Relief from respiratory distress
- Portable
- Can be used on/off
- Improved sleep
- Improved concentration/less fatigue/appetite
- Care Issues
- End-of-life concerns
END OF LIFE ISSUES

• Patient wishes known
  - Advance Decision to Refuse Treatment
  - Advance Care Planning
  - Discussions with family
  - Distinguishing between NIV and invasive ventilation

• Communication issues

• Speed of progression
  - Referring into MDT early
  - Hospice/ Palliative Care Involvement

• Withdrawal of Non-Invasive Ventilation
“While legally a competent patient is entitled to make such a decision, it may cause considerable concern for some members of the patient’s family or healthcare professionals and managers, as the patient’s choice may be viewed by some as tantamount to assisted suicide. However, what is taking place is the patient (or their proxy) taking an active decision to refuse a medical treatment or have it withdrawn, which from a medical ethics and legal perspective is not assisted suicide. Continuing with a treatment that the patient does not want is not legal and could be constituted as assault.”

Guidelines for Withdrawing Non-Invasive Ventilation (NIV) in Patients with MND Reviewed January 2014. Leicestershire and Rutland MND Supportive and Palliative Care Group

WITHDRAWAL OF NIV

- MDT involved
- Keyworker appointed
- Appropriate setting
- Discussions with patient and family – all issues addressed
- Appropriate medications in place i.e. sedation
- Awareness of the machine settings etc
- Family and patient prepared for death to possibly occur shortly afterwards
- Bereavement support after death for family
- Debriefing for staff
WHAT IS NOT AFFECTED

• Senses – hearing, sight, smell, touch
• Continence
• Sexual function
• Eye muscles
• Heart Muscles

These things should therefore be investigated
Queen Elizabeth Hospital Birmingham

One of the 22 MND Care Centres funded by the Motor Neurone Disease Association (new Stoke one!)

2 Consultant clinics every Wednesday afternoon, attended by Specialist Nurses

Monthly MDT clinic attended by the whole MND MDT team (you can request last minute slots for pts onto this if needed)
Referrals: Other consultant Neurologists; GPs; other hospitals; ENT; Neurosurgery etc.

Referral can be to Consultant Neurologists or only to CNS team

If you hear of a patient happy to chase referral!

If pt told diagnosis by a Consultant Neurologist (no matter where) – we can see (if consultant okays this!)
THE MND CARE CENTRE TEAM

- Specialist Nurses: Caroline Davis – Care Coordinator and Nicola Ryder – Specialist Nurse
- Consultants Neurologists: Dr Hardev Pall and Dr Venkat Srinivasan
- Respiratory Consultant: Dr Mustfa, Royal Stoke Hospital
- Palliative Care Consultant: Dr Jon Tomas
- MNDA, Regional Care Development Advisors: Alison Noakes and Jackie Dornford-May
- Physio- Claire Mobberley
- Occupational Therapist: Russell Wigley
- Speech and Language Therapist; Sally Brakjovich
- Dietician: Erin Forker
Every patient offered a home visit within 2 weeks of diagnosis, or referral to us

Home visit:
- Offer support
- Answer questions
- Discuss community team
- Referral into community team and local pathway
- Discussion of PEG if bulbar
- Benefit discussions
- MNDA referral consent
ROLE OF THE MND CNS

- Ongoing support in clinic, or home if required (dependent on community team support)
- Attends local MDT to support community team
- See patients in hospital for advice
- Joint visits with other health professionals
- Phoneline/ email / text available for questions etc. Triaging to services/ clinics etc
AREAS COVERED BY CARE CENTRE

- 310 patients on active caseload
- 13 MDT teams
- Birmingham
- Black Country
- Solihull
- Herefordshire
- Worcestershire
- South Warwickshire
- Parts of Staffs and Shropshire
The needs of people with MND

Before diagnosis
- Rapid and accurate diagnosis through:
  - The GP's early recognition of symptoms which might suggest the diagnosis
  - Earliest possible assessment by a neurologist

At diagnosis
- Sensitive communication of the diagnosis, ensuring:
  - Appropriate emotional/psychological support
  - Appropriate information made available, in a timely manner about:
    - the condition and its implications
    - sources of help and support
    - the MND Association and MND Connect
  - Comprehensive information is sent to the GP
  - Follow up clinical appointment within two weeks of diagnosis
  - Direct referral to the MND Association
  - Information regarding availability of local services.

A proactive approach to managing the disease is key.

Following diagnosis
- Immediate identification of a single point of contact (key worker/case manager) to ensure:
  - Access to information and service provision
  - Planning and co-ordination of support and care

Individual needs will vary but the following elements are essential:
- Access to information, service provision, and benefits tailored to current needs of the person with MND
- Intermittent holistic assessments
- Flexibility and priority of response to ensure speed in service delivery
- Access to appropriate expertise and services at the appropriate time, including:
  - Timely referral to specialist palliative care and respite care
  - Speech and language therapy
  - Occupational therapy
  - Physiotherapy
  - Diazines
  - Social care
  - GP/primary health care team
  - Neurology and neuro-rehabilitation
  - Respiratory specialist
  - Psychologist
  - Access to pharmaceutical and other relevant treatments
  - Regular communication between disciplines
  - Regular monitoring and review
  - Continuing offer of Association support
  - Access to advice regarding advanced care planning.

Key principles that underpin care of people living with MND
- Provided in partnership between the statutory services and the MND Association

Provide information to enable people living with MND to make informed choices to ensure:
- Management of the disease determined by the needs and wishes of the person with MND, their families, and carers

Provide services proactively to ensure:
- Flexibility and speed in response to referral

Provide support to people with MND, including regular monitoring and review to ensure:
- Timely access to clinical intervention
- Continuity of care throughout the progression of the disease
- Co-ordination and co-operation between service providers
- Dignity in death
THE FUTURE

- A Cure
- Improved medications
- Technological developments
- Earlier diagnosis
The Motor Neurone Disease Association  
http://www.mndassociation.org/

National Institute of Neurological Disorders and Stroke  
http://www.ninds.nih.gov/

NICE guidelines  

MND Nurses  
Caroline.davis@uhb.nhs.uk  
Nicola.ryder2@uhb.nhs.uk  
Mobile: 07771 624712