

Caring for people
with
Motor Neurone
Disease (MND)



National Clinical Specialist Team
2017

Outline



- What is MND?
 - MND in Scotland
 - Types
 - Symptoms
 - Treatments

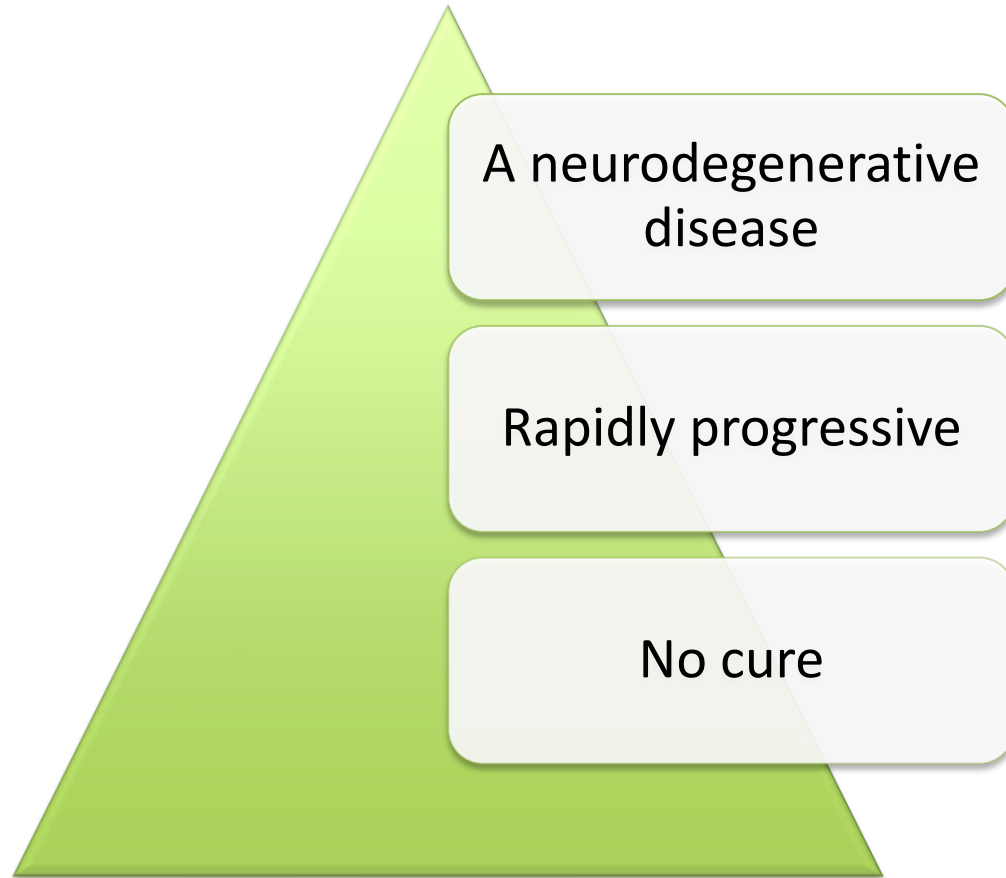
- MND infrastructure in Scotland
 - CARE-MND



The West Team

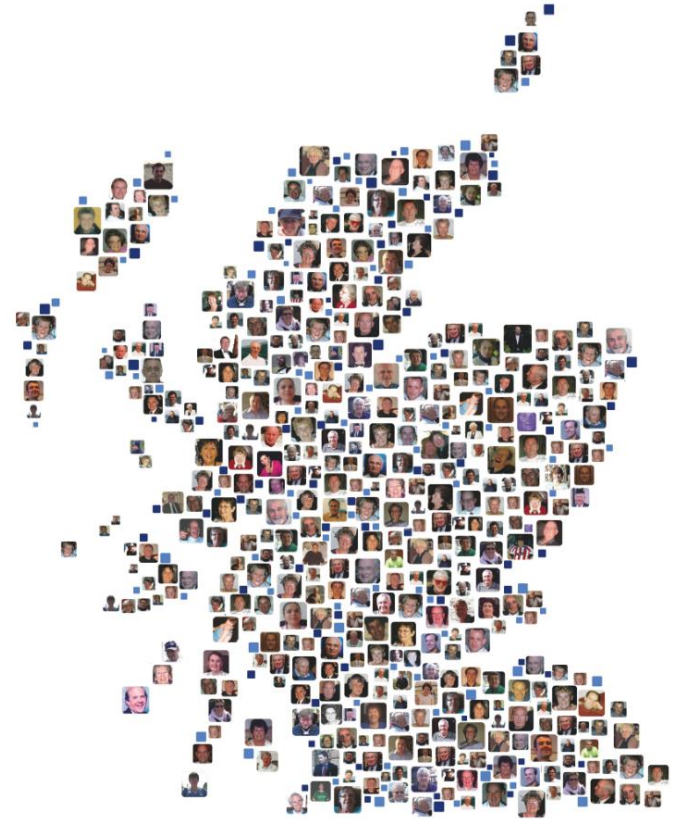
- Ann.silver@ggc.scot.nhs.uk- NHS GGC- S, Lanarkshire (G postcodes), Western Isles
- Laura.cunningham@ggc.scot.nhs.uk- NHS GGC- NE, Lanarkshire (ML postcodes)
- Janice.hatrack@ggc.scot.nhs.uk- NHS GGC- Inverclyde, East Ren, Renf, E and W Dun
- Helen.Lennox@ggc.scot.nhs.uk- NHS Ayshire and Arran, Dumfries and Galloway

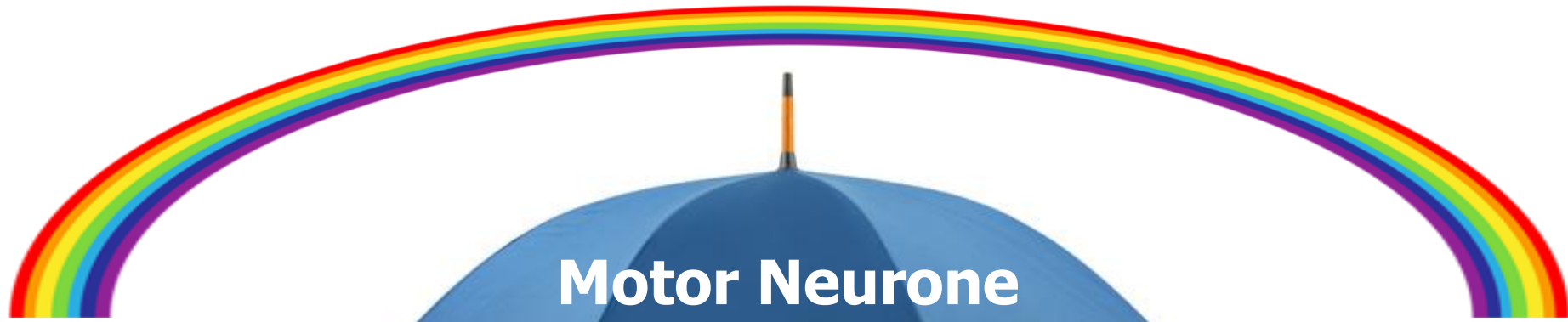
Motor Neurone Disease: Key Points



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

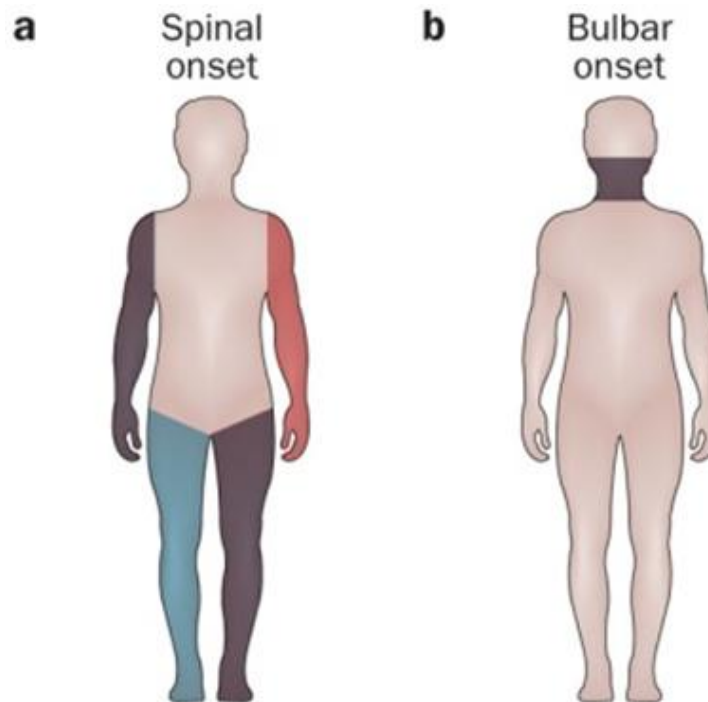
Progressive
Muscular
Atrophy

Progressive
Bulbar Palsy

COGNITIVE IMPAIRMENT

Amyotrophic Lateral Sclerosis (ALS)

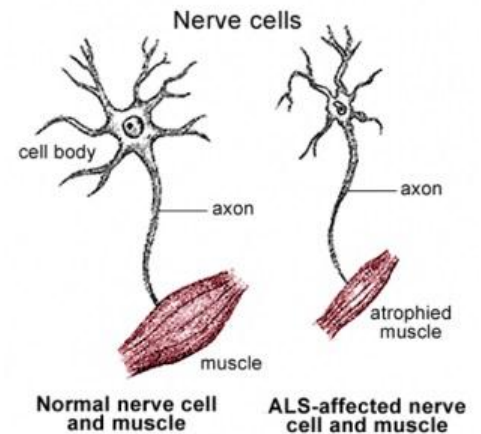
- Approx. 85% of patients have ALS
- ‘Spinal’ = limb onset or ‘Bulbar’ = speech/swallow onset
- Typically progresses to involve all limbs, bulbar muscles and respiratory muscles



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

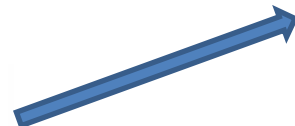
- MND management requires regular rapid ongoing assessments
- Anticipatory care should commence at point of diagnosis

Treatment Options in MND

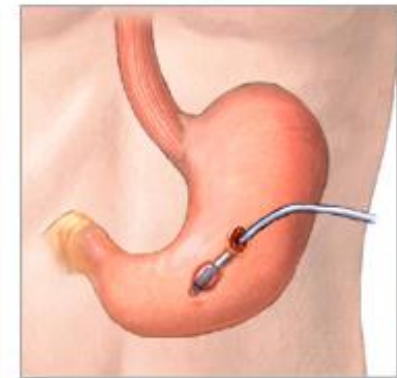
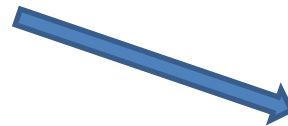
Symptoms?



Drugs?



Interventions?



MND by Symptoms

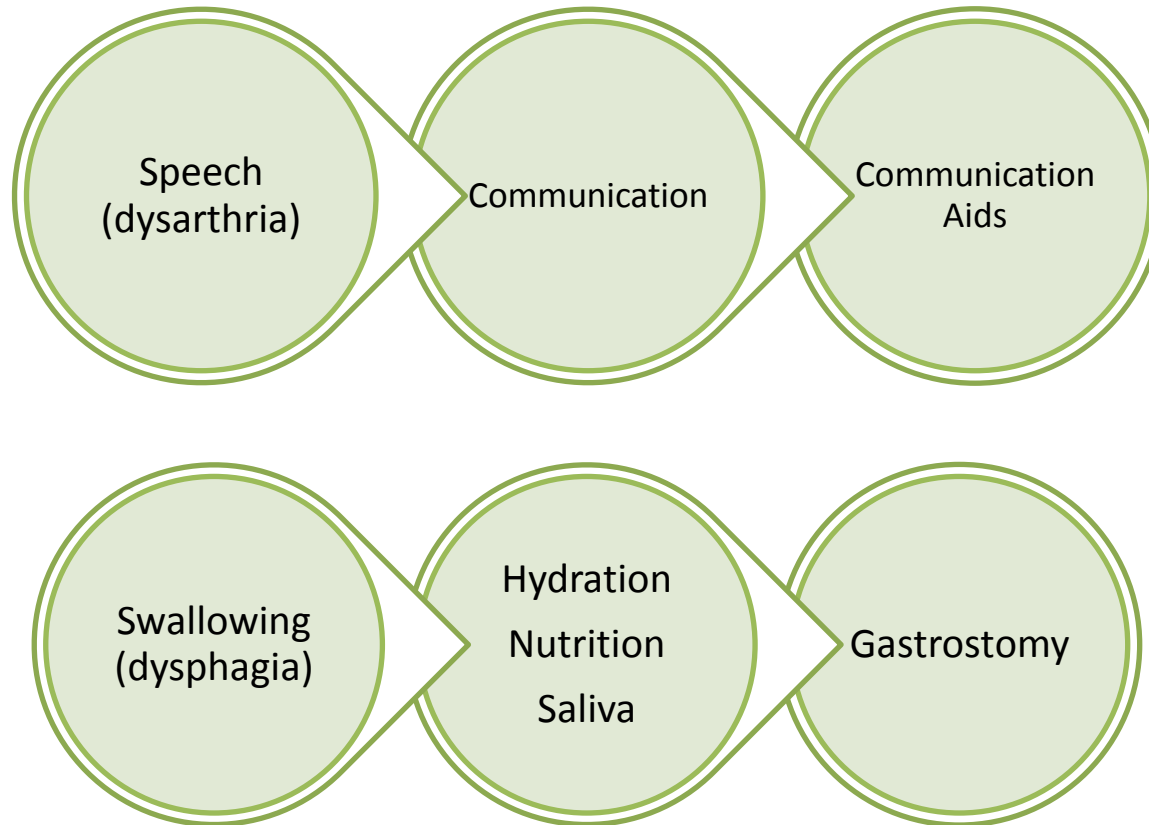
- Bulbar Dysfunction
- Upper and Lower Limb Dysfunction
- Respiratory Dysfunction
- Emotional lability and Cognition

Bulbar Dysfunction



- Affects tongue muscles, facial muscles and pharyngeal muscles

Bulbar Dysfunction

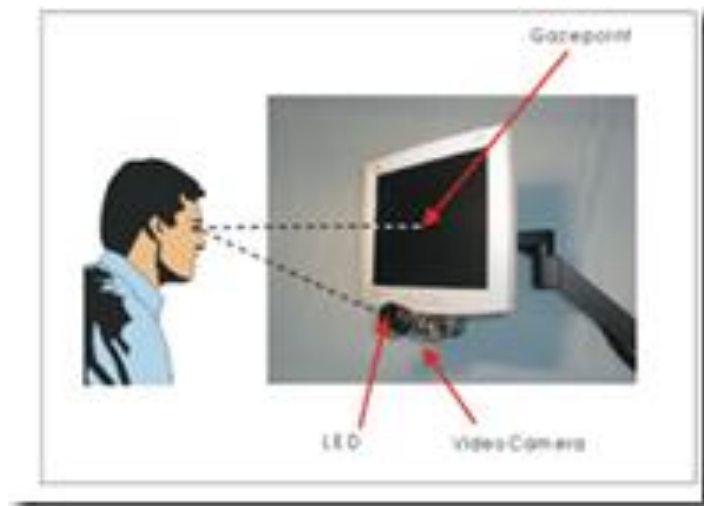


Communication: Strategy

- 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

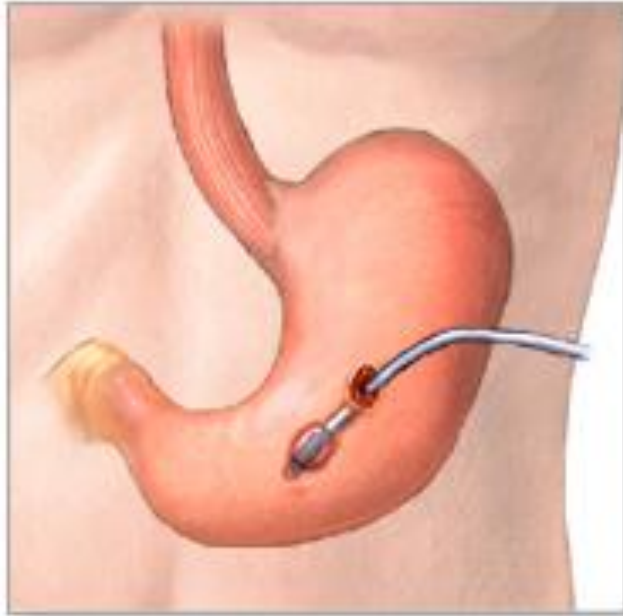


Nutrition and Hydration



- 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 through out 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

EDINBURGH COGNITIVE AND BEHAVIOURAL ALS SCREEN – ECAS English Version (2013)		
Date of testing:	Name:	
Age at leaving full-time education:	Date of Birth:	
Occupation:	Hospital No. or Address:	
Handedness:	
LANGUAGE - Naming		
⇒ Ask: Say or write down the names of these pictures:		
		Score 0-8 <input type="checkbox"/>
		
		
		
LANGUAGE - Comprehension		
⇒ Ask: point to the one which is:		Score 0-8 <input type="checkbox"/>
1. Something you can fly in	2. Something with webbed feet	
3. An animal that climbs trees	4. Something used for chopping	
5. A means of transport	6. Something with a sharp edge	
7. Something with a sting	8. Something with a diet of nuts and seeds	

S. Abrahams & T. H. Bak 1

Palliative Care

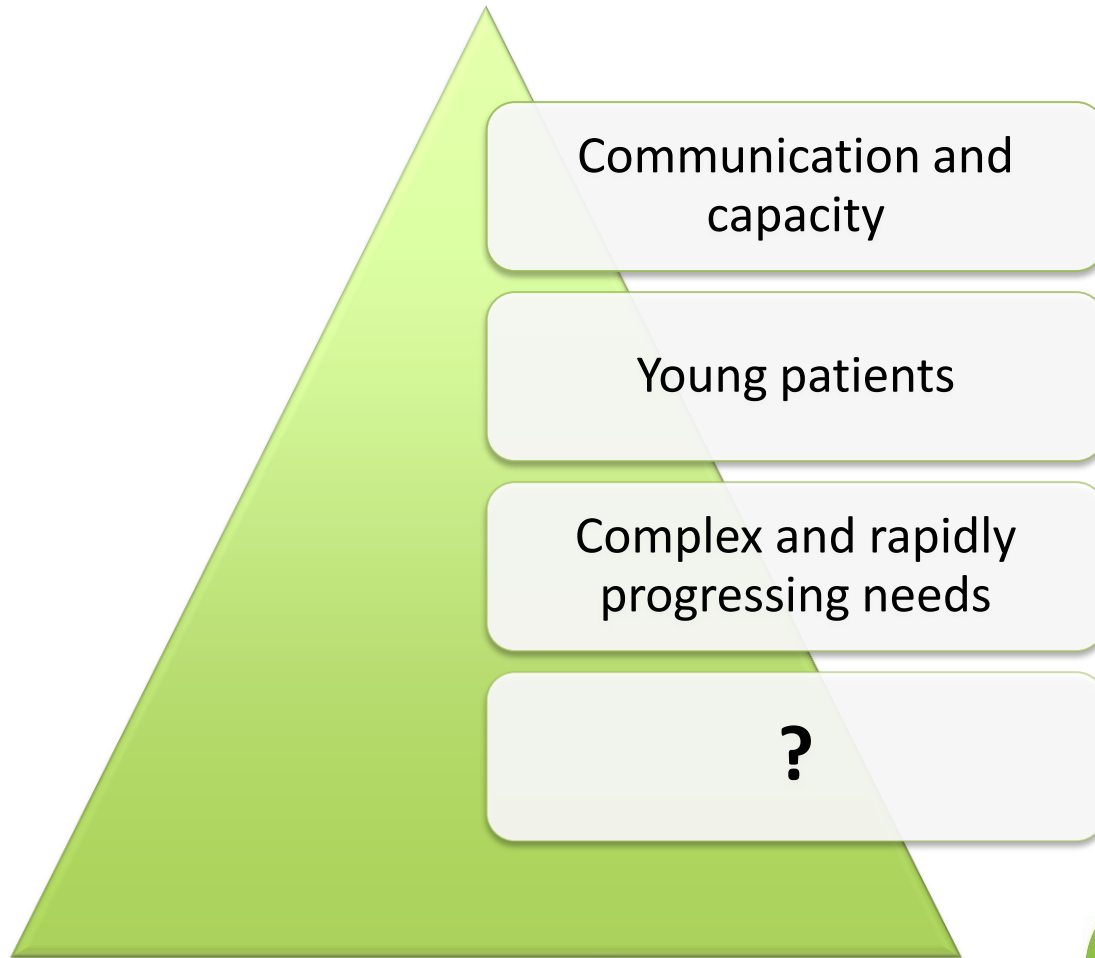
- Underpins *all* of these interventions
- May be referred for specialist input for:
 - Current or anticipated complex needs:
 - Psychological distress
 - Social distress
 - Troublesome or rapidly progressing symptoms
 - Complex future care planning needs

NICE 2016

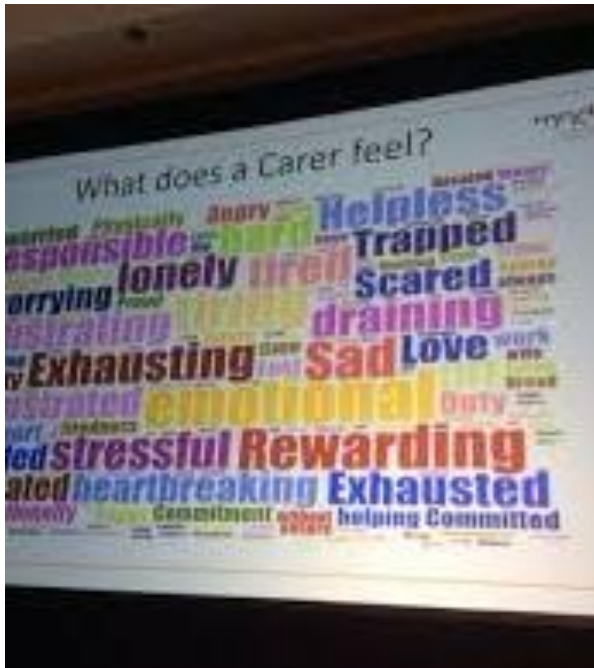


Clinical Audit Research Evaluation

Key Challenges

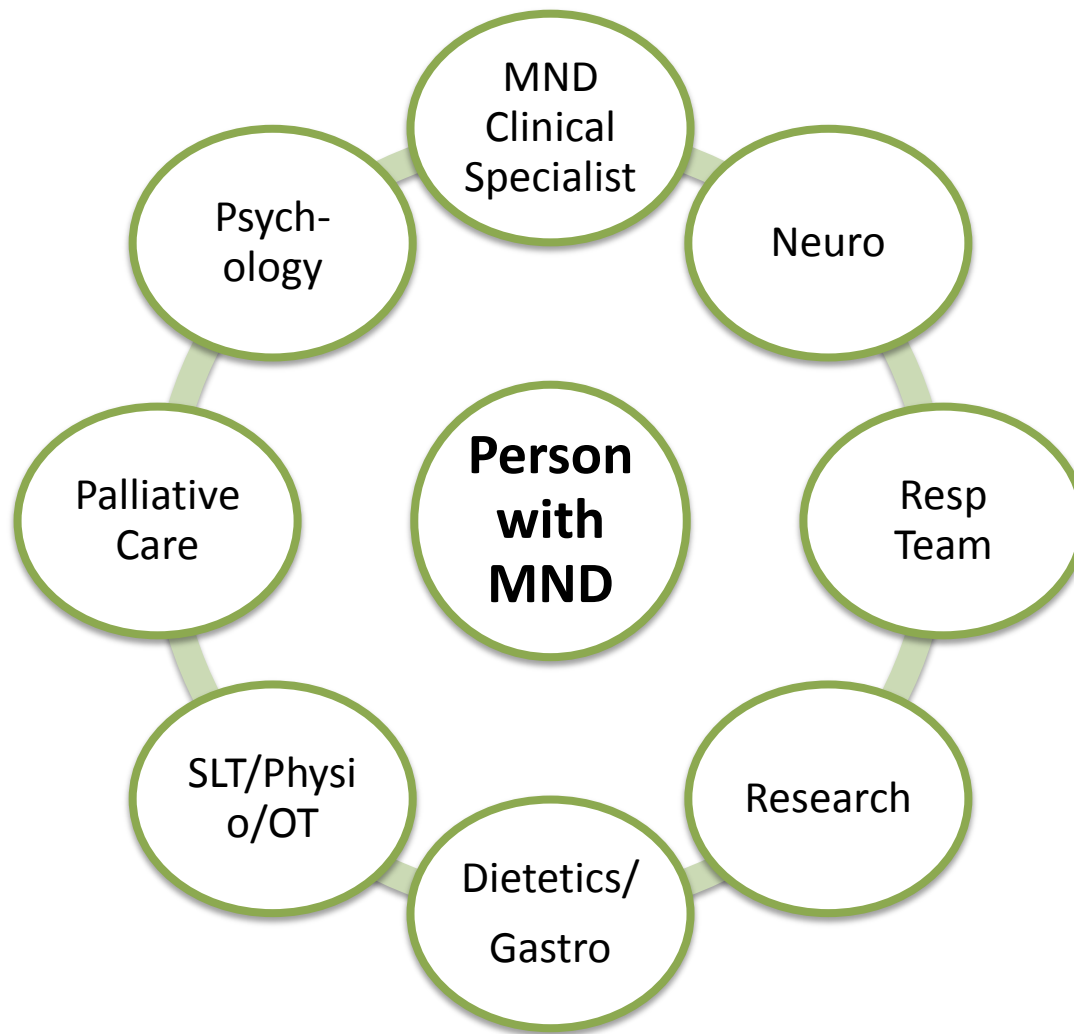


Carer burden



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

CARE-MND Team





We Aim To Engage Every Person Living With MND In Scotland In Shaping Clinical Care And Research

REGISTER YOUR INTEREST

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

Any questions just e mail on:

Judith.newton@ed.ac.uk (MND
Nurse Consultant)



Clinical Audit Research Evaluation