**Palliative Care in MND**

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**Palliative Care – WHO Definition**

> "The active total care of patients whose disease is not responsive to curative treatment. Control of pain, of other symptoms, and psychological, social and spiritual problems is paramount. The goal of palliative care is the achievement of the best quality of life for patients and their families... palliative care...affirms life and regards dying as a normal process..."

**Who Receives Palliative Care**

Any patient with a life threatening, non-curable disease

- Malignant  
  - Cancer
- Non Malignant  
  - Heart Failure, Renal Failure  
  - HIV  
  - Chronic Obstructive Pulmonary Disease  
  - Motor Neurone Disease  
  - Parkinsons Disease

**Motor Neurone Disease**

- Motor Neurone Disease is a relentlessly progressive neurodegenerative condition
- No cure, at best Riluzole slows progression
- 1 person diagnosed with MND every 4 days in Ireland
- 1 person with MND dies every 4 days in Ireland
- Palliative care treatment of the MND patient should ideally commence at the time of diagnosis and care throughout the disease trajectory is crucial for later stages
- Approximately 70% of MND patients referred to palliative care services
- Patient autonomy, dignity and quality of life

**Multidisciplinary Team**

- Speech and Language Therapist  
- Dietitian  
- Specialist Nurses  
- Care Workers  
- Palliative Care Team  
- Counsellor  
- Occupational Therapist  
- Respiratory Physiotherapist  
- Social Worker  
- General Practitioner

**Palliative Care**

- Palliative care is a type of approach which looks after physical, psychological, social needs and spiritual well being
- Achieve best of quality of life for patients and their families – identify personal goals and preferred place of care
- Amelioration of symptoms, for example pain management
- Involve the patient in decision making about various care options including respiratory and nutrition
- Promote and maintain function and independence for as long as possible
- Discuss end of life issues, concerns, fears, wishes
Stigma attached to including Palliative Care

Patients feel it’s the “end” if referred to Palliative Care

Unpredictable course of MND makes it hard to define point at which referral to palliative care should be made

European guidelines advise early referral in the disease process is appropriate

Forced vital capacity (FVC) 60% predicted
Clinical signs of respiratory insufficiency or
Respiratory weakness requiring non-invasive positive pressure ventilation or
Nutritional decline requiring enteral feeding or
Severe pain or psychosocial distress requiring intensive palliative care interventions (including opioid medication) or
Rapidly progressive paralysis (over 2-3 months) in two body regions or

Palliative Care Services

Palliative Care Hospital Consultant
Hospice
Community Palliative Team
Day Hospice Care
Respite Care

Ireland

No established national care pathway
No defined triggers for activation of palliative care services
Single multidisciplinary clinic in Beaumont Hospital incorporating palliative care provides services for up to 80% of the MND population
Coordinated by neurologist to community based palliative care services
Later stages between hospital specialist and community care
GP pivotal within palliative care community service

Suggested Triggers

Progressive Muscle Weakness and Wasting
Fasciulations, cramps and spasticity
Weight Loss
Saliva and mucus problems
Dysarthria and slurred speech

Symptom Effects of MND

Respiratory Muscle weakness
Emotional Lability
Dysphagia – poor swallow
Cognitive changes

Bede et al, J Neurol Neurosurg Psychiatry 2011; 82:413-418
**Muscle Cramps and Spams**

- Common
- Early MDT involvement – OT/PT
- Assist Devices:
  - Head rest, braces, wheelchair etc
- Drug Treatment: Cramps
  - Quinine Sulphate
- Drug Treatment: Spascity
  - Baclofen

**Sialorrhea**

- Overall decrease in saliva production
- Treatment includes:
  - Robinol
  - Amitriptyline
  - Transdermal scopolamine one or two patches every three days
  - Botulium toxin
  - Manually assisted cough technique
  - Suction machine

**Dysphagia**

- Decreased oral intake
- Requires early dietician referral
- Modification of liquid and food consistency

**PEG/RIG**

- Indications:
  - Hunger
  - Choking
  - Decreased calorific intake
  - Poor Quality of life
  - FVC >50%
- RIG insertion prolongs life by average 6 months

**Pseudobulbar Effect**

- Emotional Lability
  - Uncontrolled crying, laughing/giggling
- Affects approximately 50% of patients
- Usually treated with Amitriptyline

**Dysarthria**

- Speech Therapy
- Communication devices
  - White board
  - Computer technology such as iGaze, iPAD
  - Light writer
**Respiratory Care**
- SVC <50% is an indicator of respiratory symptoms and consideration of NIV
- Decrease in SNIP (40% cm per H2O)
- Night-time Pulse Oximetry
- Clinical Symptoms include:
  - Dyspnoea
  - Daytime fatigue
  - Poor concentration
  - Headache
  - Disturbed sleep and nightmares
  - Nervousness, depression, anxiety
  - Phonation difficulties

**NIV**
- Delivery of mechanical ventilation to the lungs
- Does not require intubation
- Treatment of acute respiratory failure
- Improves survival (200 days)
- Patients can remain on NIV in the final stages

**Disadvantages of NIV**
- Slower correction of gas exchange abnormalities
- Gastric distension
- Mask
  - Irritation
  - Air leakage
  - Facial skin necrosis (most common problem)
- Claustrophobia
- Compliance

**Pain**
- Muscle contractures
- Joint stiffness
- Pressure ulcers due to lack of immobility

**Sleep Issues**
- Anxiety
- Often derive from other symptoms of the disease
- Identify cause and treat
- Sedation should be used with caution
Psychosocial

- Loss of control, self esteem, independence and confidence
- Anxiety, fear
- Communication difficulties
- Knowledge of impending death
- Bereavement and grief counselling for patients and families
  - Beneficial from beginning
  - Continued support for patients and families

Other Symptoms

- Urinary frequency/urgency
  - In the absence of UTI, often due to spasticity
- Peripheral edema
  - Often dependent: elevation, massage, compression stockings
- Laryngospasm
  - Sudden reflex closure of vocal cords due to variety of stimuli, usually resolves spontaneously

End of Life Decisions

- Advanced Care Planning
- Advanced Statement of wishes and preferences
- Preferred Priorities of Care (PPC)
- Withdrawal of treatments
- Tissue donations/Organ Donation
  - Brain bank
  - Organ donation not suitable

Triggers for End of Life discussions

- The patient opens the discussion
- The presence of severe psychological, social or spiritual distress or suffering
- The presence of pain requiring high dosages of analgesic medications
- The presence of dysphagia requiring feeding tube
- The presence of dyspnoea, symptoms of hypoventilation, FVC <50%
- Loss of function in two body regions

Conclusion

- Palliative care shifts the emphasis from disease modification to symptom management
- Appropriate timing of palliative intervention is essential
- Outcome measurements are primarily centred around quality of life rather than disease progression

- Requirement for a national framework and patient pathway
- Formal cognitive testing prior to advance directions
- MDT care – incorporation both hospital and community palliative services
- Structured caregiver support
- Need for a development of international consensus guidelines for palliative management of MND

Bede et al. J Neurol Neurosurg Psychiatry 2011; 82:413–418
Thank you